American Academy of Pediatrics

Developmental and Behavioral Pediatrics

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Library of Congress Control Number: 2010906842
ISBN-13: 978-1-58110-274-1
MA0414

9-273/0910

1 2 3 4 5 6 7 8 9 10

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Chapter 1

Introduction
Child Development: The Basic Science of Pediatrics

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Over 40 years ago, Dr Julius Richmond characterized child development as the basic science of pediatrics.[1] The processes of child development and behavior fundamentally differentiate pediatrics from all other areas of medicine and impact all pediatricians and other primary pediatric health care professionals* and pediatric subspecialists. Thus clinical competence in child development and behavioral/mental health is vital to the success of all pediatric health care encounters.

Although the Accreditation Council of Graduate Medical Education requires all pediatric residents to experience one block month in developmental-behavioral pediatrics during their residency training,[2] surveys of pediatricians in practice continue to indicate that pediatricians feel ill-prepared in this basic science of pediatric practice.[3,4] In addition, family medicine residents and family and pediatric nurse practitioners, who will provide medical homes for at least one-third of all US children,[5] generally receive limited, if any, training in developmental-behavioral pediatrics. As illustrated in Table 1.1, and even more concerning in this setting of limited preparation and training, developmental disorders are the most prevalent chronic medical conditions encountered in primary pediatric care, and psychosocial and behavioral issues are even more ubiquitous in day-to-day pediatric practice.[6–9]

Currently the American Board of Pediatrics reports only 520 board-certified specialists in developmental-behavioral pediatrics and only 253 with board certification in neurodevelopmental disabilities.[10] This does not represent 773 different subspecialists, since a significant minority of individuals have both certifications. This critical

*Throughout this manual, the term "primary pediatric health care professionals" is intended to encompass pediatricians, family physicians, nurse practitioners, and physician assistants who provide primary care to infants, children, and adolescents.

lack of subspecialists to whom to refer results in unacceptably long waiting lists at most tertiary care child development and behavioral health centers. Thus, given both the prevalence and chronic nature of developmental-behavioral disorders and lack of subspecialists in the field, it is essential that primary pediatric health care professionals establish medical homes for children and youth with developmental and behavioral concerns from which care is initiated, coordinated, and longitudinally monitored and with which families can form a reliable alliance for information, support, and advocacy from the time of diagnosis through the transition to adulthood.[11]

This manual represents a cooperative effort of the American Academy of Pediatrics Section on Developmental and Behavioral Pediatrics and Council on Children With Disabilities. Given the crucial need to enhance the education of all primary pediatric health care professionals in this basic science of pediatrics, this manual attempts to blend the overlapping perspectives of both neurodevelopmental disabilities and developmental-behavioral pediatrics with the goal of improving care for all children.

This manual is neither intended to be an exhaustive reference geared for the subspecialist nor a cursory introductory list of developmental and behavioral pediatric topics. Instead, this manual aims to be a resource providing the essentials of what all primary pediatric health care professionals need to know to successfully care for children with developmental and behavioral concerns in their practices and to identify those who may require subspecialty referral. It is hoped primary pediatric health care professionals will use this manual to improve confidence in managing children with developmental and behavioral concerns and provide evidence-based developmental-behavioral pediatric care within the medical home.

Table 1.1. Prevalence of Developmental-Behavioral Disorders and Other Chronic Medical Conditions in Children[6-9]

Condition	Cases per 1,000
Asthma	135
Learning disabilities	90
ADHD	78
Anxiety	64
Depression	60
Developmental delay	32
Mental retardation	12
Congenital heart disorders	9
Autism	6.6
Epilepsy	6.5
Cerebral palsy	3.1
Diabetes	2.2
Juvenile rheumatic diseases	1.5
Spina bifida	0.4
Cystic fibrosis	0.3
Cancer	0.2
Inflammatory bowel disease	0.07
Chronic renal disease	0.07

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Chapter 2

Nature, Nurture, and Their Interactions in Child Development and Behavior

Paul P. Wang, MD, FAAP

Nature and nurture have long been regarded as rival influences on child development and behavior. One school of thought has contended that a child's fate is determined by nature, consisting essentially of a child's biological endowment, while a rival school of thought has argued that nurture, which consists of a child's environment and experiences, is dominant in determining adult outcomes. In truth, intuitive parents and pediatricians have long known that both sets of factors—the innate and the experiential—are important in the complex processes of child development and behavior. Over the last several decades, science has amassed substantial evidence to document the importance of both nature and nurture.[1] Moreover, current research is elucidating the complex interactions of nature and nurture that occur throughout the childhood years.[2]

This chapter attempts to provide a framework in which to consider how nature, nurture, and their interactions shape children's lives. Many examples are provided of both innate and experiential factors that influence children's development and behavior, and of the mechanisms through which those factors are believed to act. Throughout the chapter, the reader is asked to hold 2 overarching concepts in mind: *individual variability* and *developmental plasticity.* Because of individual variability, children differ in the extent to which any factor may shape their development and behavior, regardless of whether that factor is biological or environmental. As research is beginning to show, and as illustrated further below, much of this variability may be rooted in the interaction of nature and nurture. Because of developmental plasticity, the effects of both innate and experiential factors can be either augmented or ameliorated by other factors over time. No developmental influence, whether biological or environmental, should be regarded as rigidly deterministic, always consigning a child to a specific and certain fate. Rather, the processes of development continue throughout childhood, adolescence, and even adulthood, allowing biological and behavioral interventions to influence ultimate outcomes.

Biological Factors in Child Development and Behavior

Nature is variably construed in either a more narrow or more broad manner. From the narrow perspective, a child's nature is defined as her innate biological endowment, her heredity and, thus, consists primarily of her genes. From the broader perspective, all factors that are believed to operate through a direct biological mechanism are construed as being within the category of nature.

Genetic Sequence

Scientists long believed that full sequencing of the human genome would show that our species, *Homo sapiens,* had up to 100,000 genes, but the Human Genome Project revealed that the total number of human genes is only 20,000 to 25,000.[3] This is only slightly more than the number of genes found in simple creatures, such as worms. Furthermore, there is surprisingly little difference between the genomes of *Homo sapiens* and *Pan troglodytes* (the common chimpanzee), though controversy surrounds the oft-quoted figure of a 5% difference. Science has yet to fully elucidate how quantitatively small differences in genetic sequence can give rise to seemingly dramatic phenotypic differences, but it is believed that the answer may be found in at least 3 areas: (1) the very common human phenomenon of alternative splicing, by which mRNA is spliced into multiple different versions, each being translated into a different protein product; (2) the role of "non-coding" regions of human DNA, which help to regulate the time and location in the body where the coding regions are actually transcribed into RNA; and (3) the enormously complicated interaction of human genes with one another, which is the focus of the field of genomics.

Genetic factors can have either large or smaller, more subtle influences on child development and behavior. Examples of the former include genetic disorders associated with intellectual disabilities (mental retardation) or severe behavioral abnormalities. These disorders can result from single gene mutations (eg, fragile X syndrome, Lesch-Nyhan disorder, Rett syndrome) or from anomalies in multiple genes (eg, Williams syndrome; velocardiofacial syndrome; Down syndrome; and other chromosomal aneuploidies, segmental chromosomal deletions, or duplications). In some of these disorders, the exact pathogenic mechanism is not fully understood. For example, which genes on the triploid chromosome 21 are implicated in the neurobiological differences associated with Down syndrome? In other disorders, including fragile X syndrome[4] and many metabolic conditions, such as Lesch-Nyhan, the pathogenesis is understood to a somewhat greater extent.

Researchers are avidly seeking genetic factors with smaller effects on child development and behavior. Several genes have been tentatively identified that may increase the risk of reading disability and language delay, but replication of these results has been problematic.[5] In the case of autism spectrum disorders, some cases may be associated with a single gene mutation (eg, fragile X syndrome), but many hypothesize that most cases of autism result from the effects of 15 to 20 genes, or perhaps

more, acting in concert.[6] If only one or a few of these genes are present in their pathologic form, symptoms may be mild, but if several are present, they may act synergistically to result in a full case of autism. *CNTNAP2,* for instance, is associated with both autism and language disorders.[7,8] Perhaps its influence is manifested as an autistic disorder only when other pathologic factors (genes or otherwise) add to its negative influence on communication skills. General cognitive abilities are likely affected by an even larger number of genes, though again, single gene differences can result in significant cognitive impairment. The identification of the many genes influencing traits such as cognitive ability and autistic symptoms, and the complex interactions of these genes with each other, is likely to require many more years of investigation.

The interaction between nature and nurture is apparent even for the classic, severe genetic disorders, which were long thought to result in preordained, minimally changeable developmental outcomes. For many metabolic disorders, with phenylketonuria serving as a particularly well-known example, early diagnosis and careful nutritional management can greatly enhance outcome. Even in multigene, chromosomal disorders such as Down syndrome, the past decades have yielded powerful evidence for developmental plasticity that depends on environmental factors. When Down syndrome was first recognized in the medical literature, virtually all affected individuals were confined to state-run institutions, and their level of cognitive function was moderately or severely impaired in almost all cases. Today, children with Down syndrome typically grow up with their own family, they receive extensive early intervention and special educational support, and their cognitive and functional capacities are far greater than they were a generation ago.[9]

Epigenetics

Even from the most narrow perspective of what nature consists of, it is now recognized that there are epigenetic factors that are part of a child's biological endowment. These are innate factors that are not dependent on the sequence of base pairs in a person's DNA. The best understood example of this is imprinting, by which a child's DNA is somehow marked as having come from either the mother or the father. For example, the genetic syndromes of Prader-Willi and Angelman both can result from a deletion at chromosome 15q11-q13. If this deletion occurs on the chromosome 15 that was inherited from the father, then the child is affected by Prader-Willi syndrome, but if the deletion occurs on the maternally inherited chromosome 15, then the child will have Angelman syndrome.

Another example of an epigenetic phenomenon is found in fragile X syndrome, which is caused by a mutation (a triplet repeat expansion) in a non-coding region of the gene for fragile X mental retardation protein (FMRP). In almost all cases, this mutation is accompanied by methylation of the FMRP gene, which blocks the gene from being expressed, causing the affected child to fully manifest fragile X syndrome.

In rare cases, however, the mutated gene is not methylated, allowing some FMRP to be produced, thereby resulting in less severe symptoms.

A third example of an epigenetic modification is that of social skills in Turner syndrome. According to one report, which still requires replication,[10] girls with Turner syndrome who have inherited their only X chromosome from their father, as a group, show social skills that are superior to girls whose only X chromosome is inherited from their mother. The investigators hypothesize that there may be a gene or genes on the X chromosome that are active in males (who have only one X chromosome) but may be methylated and turned off on one of the female X chromosomes. If the methylated X chromosome is passed on to the girl with Turner syndrome, then her social skills may be disadvantaged.

Intriguing research has shown that monozygotic twins, who by definition have identical genetic sequences, can have differences in their epigenetic states.[11] As a corollary, the twins can have differences in the extent to which the epigenetically regulated genes are expressed, which would lead to differences in their phenotypes. The data show that twins' epigenetic differences increase with age, particularly if the twins live apart from each other. The tentative conclusion is that the different environments that the twins experience are the cause of their epigenetic divergence and, thus, of the phenotypic differences that arise between identical twins. This conclusion is supported by several findings in animal research: that an individual's epigenetic state can change during life as a consequence of specific environmental experiences.

Biological Experiences

The broader perspective on nature includes not only genetic and epigenetic factors, but also other biological influences that are experienced after birth and that do not necessarily have a genetic basis. Examples include traumatic brain injury, hypoxic-ischemic encephalopathy, meningoencephalitis, congenital brain malformations, and prenatal drug exposures. Because these factors have a well-defined biological mechanism, they are generally thought to have a strongly constraining influence on developmental-behavioral outcomes. The fallacy in such reasoning can be found in other biological conditions, such as malnutrition, epilepsy, in utero infections, and chronic illness. These conditions are among the most common biological insults experienced by children worldwide, with protein malnutrition and deficiencies of iron and iodine as prime examples. Each of these conditions also has a clear biological mechanism, but each is associated with a range of developmental-behavioral outcomes. It is widely appreciated that the outcome in this second set of conditions depends on the success of interventions directed either to the underlying conditions or to their developmental-behavioral consequences. We are now learning that the same is true for genetic differences and for biological conditions that were previously regarded more fatalistically. It seems very likely that future research will point the way to interventions that may help to treat the biological conditions that have been

thought largely resistant to treatment. As an example, constraint therapy, a possible therapy for focal brain injury and cerebral palsy, is discussed below.

Environmental Factors in Child Development and Behavior

The array of environmental, or experiential, factors affecting development and behavior is wide. As is evident from even a short listing of these factors—from birth order, maternal educational level, and family dynamics to poverty, child abuse, and strength of religious affiliation—experiential factors can be either beneficial or deleterious in their effects, and can be either direct or indirect in their mechanisms. In almost all cases, the mechanisms by which these factors exert their effects are still unknown, but it is an unstated assumption that the action of experiential factors depends somehow on brain plasticity. That is, the brain has the potential to be molded by environmental factors (nurture), in either a beneficial, functionally positive direction, or in a deleterious, functionally negative direction. Stated in yet another way, it can be expected that experiential factors ultimately act through neurobiological mechanisms to shape the status and the function of children's brains, just as Freud hypothesized that the action and effects of his psychotherapy were made manifest through neurobiological processes.

Neurobiological Effects of Environmental Enrichment

A classic, early demonstration of the neurobiological effect of environmental enrichment is found in the work of Volkmar and Greenough[12] on rats. This research built on earlier work that showed superior problem-solving abilities in rats raised as pets compared with rats raised without such extensive human interaction. The neurobiological studies of Volkmar and Greenough[12] showed that environmental enrichment is associated with greater cerebral volumes, a larger number of synapses, and increased complexity of dendritic branching. Thus differences in environmental variables were translated into changes in neurobiological characteristics.

Maternal Education, SES, and Poverty

In this light, it is no surprise that maternal education level (or analogous variables such as maternal socioeconomic status [SES] or maternal IQ) has repeatedly been shown to be one of the most powerful predictors of developmental outcome, regardless of the primary risk factor being studied (eg, prematurity, meningitis, adoption after institutionalization, etc). The specific mechanisms through which maternal educational level exerts its action on the child is not known, but it may take the form of an enriched environment, whether that enrichment is provided directly by the mother or indirectly through other interventions that the mother obtains for the child. More recently, many studies have begun to look beyond demographic variables such as SES to make a detailed assessment of observable environmental variables, such as the presence of books in the home, hours of electronic media exposure, or types of conversation in which the child is engaged.[13] These studies suggest that the

demographic variables that are associated with child development may be mediated at least partially through these more "proximal," concrete factors.

Other environmental factors that influence child development, such as poverty and institutionalization, may operate at least partly through similar mechanisms, though there has been little study to date of the detailed mechanisms of these environmental factors. Some recent data suggest that poverty is associated with impairments in executive functions, such as attention and inhibition, and that these impairments are reflected in differential patterns of brain activation during tasks that require these skills.[14] These data provide further evidence that environmental factors are associated with changes in neurobiological processes in children's brains.

The mechanisms by which environmental factors influence behavioral and mental health outcomes are even less well understood. Factors such as child abuse or neglect, familial dysfunction and divorce, and exposure to violence obviously are associated with increased stress, and may operate through the biological pathways associated with stress, while resilience factors may mitigate such stress.[15] Alternatively or additionally, some such factors may shape children's behavior by providing a behavioral model that, unfortunately, is assimilated and later recapitulated.

Sensitive Periods

The extremely rare cases of "wild" or "feral" children have contributed greatly to the concept of "sensitive" or "critical" periods of child development. In these tragic cases, children are deprived of normal human interactions, and they grow up to be pervasively impaired.[16] In addition to these exceptionally unusual cases, there are also commonly encountered conditions that give insight on sensitive periods and illustrate the importance of environmental influences and their interactions with biological factors. The example that is best understood on a neurobiological level is strabismus. Neural inputs from both eyes are known to converge in the primary visual cortex of the occipital lobe. In children with strabismus, the connections from 1 of the 2 eyes to specific cortical cells are lost, and the capacity for binocular vision is lost with them. The pathogenic mechanism for amblyopia secondary to refractive errors is analogous. Related animal research has studied other unusual constraints on visual input, such as exposing young animals to an environment that has only vertical stripes. Such animals lose much of their capacity to perceive visual features other than stripes. As is the case for strabismus and amblyopia in humans, this research demonstrates how normal brain development requires appropriate environmental inputs, and how that input must occur within a certain age range.

Language development provides another common example of a sensitive period for environmental stimulation in child development. The cases of feral children again provide an extreme example, in which it is argued that language cannot be acquired if there is an absence of appropriate language stimulation in the first few years of life. More prosaically, it is widely observed that children tend to acquire a

second language much more easily and proficiently than adults do. While there are exceptions to this rule, there is likely to be a combination of biological and environmental explanations for children's advantage over adults in second language learning. Besides having brains that may be more receptive to a second language, children typically learn a second language in a different social and psychological environment than adults do—they tend to be immersed with other children who provide plentiful, possibly simpler and more concrete language input, and children may be less embarrassed than adult learners to practice their emerging language skills.

More generally than language development, pediatricians, psychologists, parents, educators, and policy makers understand that the first few years of life constitute a very important period for child development. This belief is rooted in the rapid biological development of the brain in those years. Recent research on children adopted internationally illustrates this principle clearly.[17] Whether they are from Romania, Russia, China, or elsewhere, children who remained in institutions past 24 months of age show more severe and more enduring developmental impairments than those adopted before age 2 years. Ongoing research suggests that the prognosis of all institutionalized orphans improves when they are placed in foster care or its equivalent, even if they remain in their country of origin.[18]

While the term *critical period* was used frequently in the older literature, the term *sensitive period* now is used more commonly to reflect the fact that most developmental processes are not completely limited to a specific window in time. Rather, while specific developmental processes might proceed most quickly and efficiently during their sensitive period, they can still move forward after the sensitive period has passed. Thus the phenomenon of stereovision requiring early correction of strabismus seems to be an exception rather than the rule.

Education as Nurture

Most of the innate and experiential factors discussed above are inadvertent (that is, the affected child and her parents did not seek out those influences) and have negative influences, but there are, of course, many environmental factors that are positive in their influence and that are actively sought by children and their caretakers. Education can and should be thought of as a prime example of a positive environmental experience that is deliberately sought, with early intervention and Head Start being 2 specific examples within the larger category of educational experiences. Very few parents would deny the importance of formal education, and many go so far as to choose where they live so that they can optimize their children's education. Others pay thousands of dollars of private tuition for the same purpose, despite relatively limited data supporting the superiority of one particular school over another for a particular child. By contrast, the data supporting the general benefits of early intervention and Head Start are now unassailable (see Chapter 5).

Education researchers have long been engaged in the development of new curricula that would be more effective than previously used curricula. Recently these efforts have turned their focus beyond just academic skills to include the teaching of fundamental cognitive skills. In one example, a new preschool curriculum was created with the goal of promoting better executive functions (such as working memory, inhibition, and attention) in children.[19] Data from a small number of sites suggest that the new curriculum is effective.

"Extraordinary" Manipulations of the Environment

Therapeutic environmental experiences can also take very unusual approaches in their efforts to tailor interventions to specific populations of children. To return to the example of amblyopia, monocular occlusion (patching) of the better-sighted eye seems as extreme as the manipulations imposed on research animals in the study of vision, as discussed above, but its value is proven. A relatively unknown and still incompletely proven therapy for cerebral palsy takes a related approach. In constraint therapy, individuals with a hemiplegia have their better-functioning limb restrained or immobilized, forcing them to use their affected limb more intensely, just as eye patching forces the use of the impaired eye. It is hypothesized that the more intense use of the impaired limb results in faster and more thorough remapping of brain circuits, culminating in better recovery of function. Data on the benefits of constraint therapy are accumulating both in adult and child patients.[20]

The discrete trial/applied behavioral analytic (ABA) approach to autism intervention might also be considered an unusual, experimental approach to a severe developmental disturbance. Certainly the rigid programming and narrow focus of the therapeutic environment that ABA uses bears little resemblance to the typical environment that children encounter; however, the encouraging data on its efficacy suggest that such marked alterations in the learning environment can be beneficial for such children (see Chapter 13).

Other experimenters are using computer-based methods to tailor educational interventions so that the difficulty level of the instruction fits each student as well as possible. While the promise of these methods has not been sufficiently well proven, early reports are encouraging. In one example, healthy adult subjects trained intensively on a working memory task whose difficulty was adjusted so that it challenged but did not overwhelm the subjects. The subjects not only improved their performance on a different working memory task but also increased performance on measures of fluid intelligence.[21] The results demand broad replication, but they suggest that even traits such as intelligence, that are generally thought to be highly genetically determined and impervious to educational interventions, especially in adulthood, may actually be sensitive to behavioral (environmental) intervention when a carefully crafted approach is taken.[22] Similar programs have been created for children with working memory deficits, and the results are also encouraging.[23]

Environmental Factors and Behavioral Development

While much of the above discussion has focused on children's cognitive development, it is well known that environmental factors can have enormous effects on children's behavioral development. Many factors discussed in relation to cognitive development also have very important influences on child behavior. A dramatic example is that of institutionalization and its effects on attachment-related behavior. Early and severe deprivation from typical bonding and attachment experiences has enduring effects on later behavior in this domain.[18] Thus there seems to be a very important sensitive period for infants and toddlers to experience normative formation of close interpersonal relationships.

Children in North America more commonly encounter other environmental factors that are less dramatic but that can also have important influences on behavioral development. Birth order, parental age, and parents' marital status (especially having a teenaged, unmarried mother) are among the demographic variables known to be associated with child behavioral outcome. Examples of other variables that are associated with behavioral outcome, and that are more proximal to children's day-to-day lives, are parenting style; parents' mental health; parental substance abuse; and exposure to violence in the home, in the media, or in the community. A substantial body of research also has identified positive environmental factors that support resilience in behavioral and developmental outcomes.[15] Findings from an early but still influential study of resilience, the Kauai Longitudinal Study,[24] indicate that both internal (eg, temperament and academic competence) and external (eg, membership in a religious community and a close relationship with a supportive adult) factors modulate outcomes in the face of negative risk factors.

Nature and Nurture: Research Issues

Influences on child development and behavior, both innate and experiential, rarely if ever work in isolation. This section of the chapter reviews an assortment of theoretical concepts that illustrate this point.

Heritability

Many behavioral genetic studies are designed to estimate the relative contributions of genetics versus environmental factors on various outcomes or diagnoses (Figure 2.1).[25] The numerical parameter that is used to quantify genetic influences is known as heritability, which has a numerical range of 0 to 1.00 and is symbolized as h^2. For example, studies of attention-deficit/hyperactivity disorder estimate its heritability to be between 0.60 and 0.90, while IQ studies estimate its heritability from 0.50 to 0.85, with values tending higher with increasing age (that is, genetic factors have a larger influence in older ages).[22] It should be understood that heritability is an abstract mathematical parameter that is not known to have a tangible reality. That is, if the heritability of reading disability is 0.75, it does not imply that 75% of all cases of dyslexia have an exclusively genetic etiology, or that the child of a person with

dyslexia has a 75% chance of having dyslexia, or any such implication. Heritability merely describes the proportion of the statistical variance in a trait that was attributable to genetics in a particular research study.

Many behavioral genetic studies, such as those described in Figure 2.1, yield estimates of genetic heritability that are greater than 0.50 for many developmental-behavioral diagnoses and traits,[26] leading some commentators to claim that biology is more important than environment. Such a claim is misleading at best. First, it is simply not clear that the adjective "more" is a useful descriptor. Indeed, it cannot be concluded that any particular case of a disease is "more" caused by genetic or environmental factors, or that more cases of that disease are caused by genetics than environment. Since no study of any trait has shown complete genetic heritability (ie, heritability always has been found to be <1.00), and since even identical twins do not show 100% concordance for any diagnosis or trait (eg, autism, schizophrenia, or reading skills, etc), it implies that environmental factors can make a clinically significant difference even when 2 individuals are genetically identical. It seems much more likely that all or almost all cases of a disease have both genetic and environmental influences in their pathogenesis. Robert Plomin, one of the most prominent researchers in the field, has commented that behavioral genetic studies can be regarded as providing some of the best evidence of the importance of environmental factors in shaping health and disease.[27]

A further argument against the over-interpretation of heritability parameters is that they are dependent on the amounts of genetic and environmental variation in the populations under study. For example, if all the subjects in a study had identical environments, the study would show misleadingly high estimates of genetic heritability and low estimates of environmental influence. In fact, it is well known that extreme environmental manipulation can have enormous effects on behavior and development. Whether those extreme manipulations are rare (eg, children who are abusively deprived of human interaction) or historically common (eg, the practice of foot binding in late imperial China and its effect on foot length), it is clear that environmental factors can have dramatic influences. Common, real-life variability in the environment also affects estimates of heritability, as Turkheimer and colleagues[28] demonstrated. They found that estimates of the heritability of IQ are much higher in populations with higher SES, while heritability is near zero in populations of lower SES. Explanations for this finding are only speculative, but it seems possible that families with higher SES provide a more consistently beneficial environment to their children, thus minimizing the environmental differences between them and thereby making genetics a larger source of variance. In populations with lower SES, on the other hand, some children may encounter more beneficial environments (eg, a particularly nurturing teacher) while others do not, which results in higher estimates of environmental variance in IQ and minimal genetic heritability.

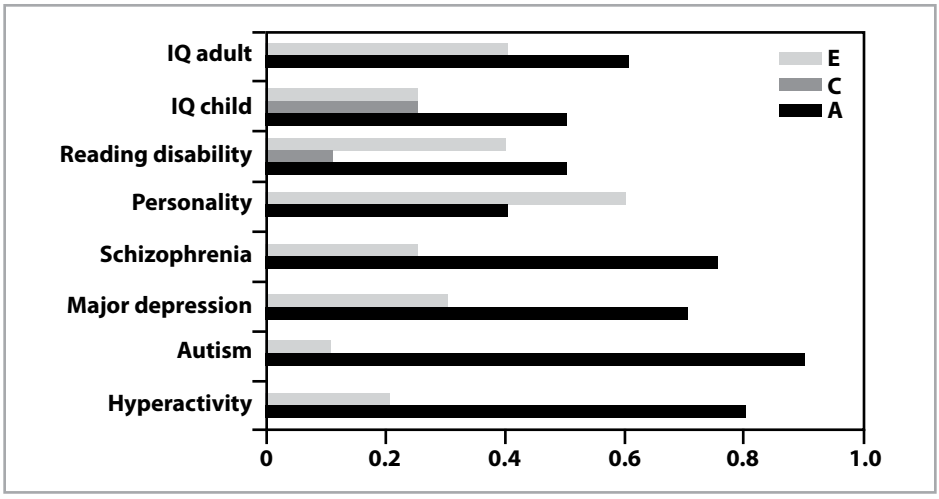

Figure 2.1. Heritability of behavioral traits. Behavioral genetic studies have provided estimates of the amount of variance in several behavioral traits and diagnoses that are accounted for by genetics (labeled "A"), environmental factors that are shared by both twins (C), and nonshared environment (E). From: McGuffin et al[25] with permission from AAAS.

Experience-Dependence Versus Experience-Expectancy

The interplay between nature and nurture, and the limitations inherent in experimental estimates of heritability, are perhaps most evident in those developmental processes that show sensitive or critical periods. As discussed previously, if children are deprived of language input during the first few years of life, they are unlikely ever to develop normal language. Neurobiologists now use the term *experience-expectant* to describe neurobiological processes like language development, in which the brain expects, and needs, appropriate inputs during an early sensitive period for development. Similarly, the visual cortex is experience-expectant for binocular input during the first few years of life, otherwise stereovision will be permanently impaired (as with untreated strabismus). A contrasting term, *experience-dependence,* is used to describe processes in which the brain and developmental skills are shaped by experience, but which do not result in fundamental functional impairment if the environmental stimuli are never encountered. Experience-dependent processes are not associated with sharply defined sensitive periods, in contrast to experience-expectant processes.[29] For example, when children have the opportunity to learn soccer or piano playing or geometry, the brain adapts as it acquires the new skills and knowledge, but lack of exposure to this type of stimulation does not result in a fundamental adaptive impairment.

The Interaction of Nature and Nurture

As the landmark publication *From Neurons to Neighborhoods* concluded,[1] "the longstanding nature versus nurture debate is over-simplistic and scientifically obsolete." Instead, the book's authors went on to explain, "…the question is…how early

experiences and genetic predispositions interact…." Researchers have only recently acquired the tools and underlying knowledge needed to answer this question, but their early efforts are beginning to elucidate how nature and nurture interact at a fundamental level.

Much of the research on the interaction between genetic and environmental factors is particularly relevant to pediatrics. For example, one pioneering study investigated the relationship between childhood experiences of maltreatment and violent or antisocial behavior in adulthood. By performing genotyping studies on a very large cohort of children who had been victims of abuse in childhood, Caspi et al[30] found an interaction between the *MAOA* gene and child maltreatment (Figure 2.2). Boys who had one version of the gene (the version associated with lower enzyme activity) had a much higher risk of developing antisocial behavior themselves. In a group of control subjects with no history of childhood abuse, the *MAOA* gene was not associated with a higher risk of adult antisocial behavior. Thus a synergistic interaction between gene and environment was critical in determining outcomes; neither factor alone showed nearly the effect as did the 2 risk factors in combination.

Another study on gene-environment interactions examined the relationship of breastfeeding to measures of IQ. Breastfeeding is associated with many positive

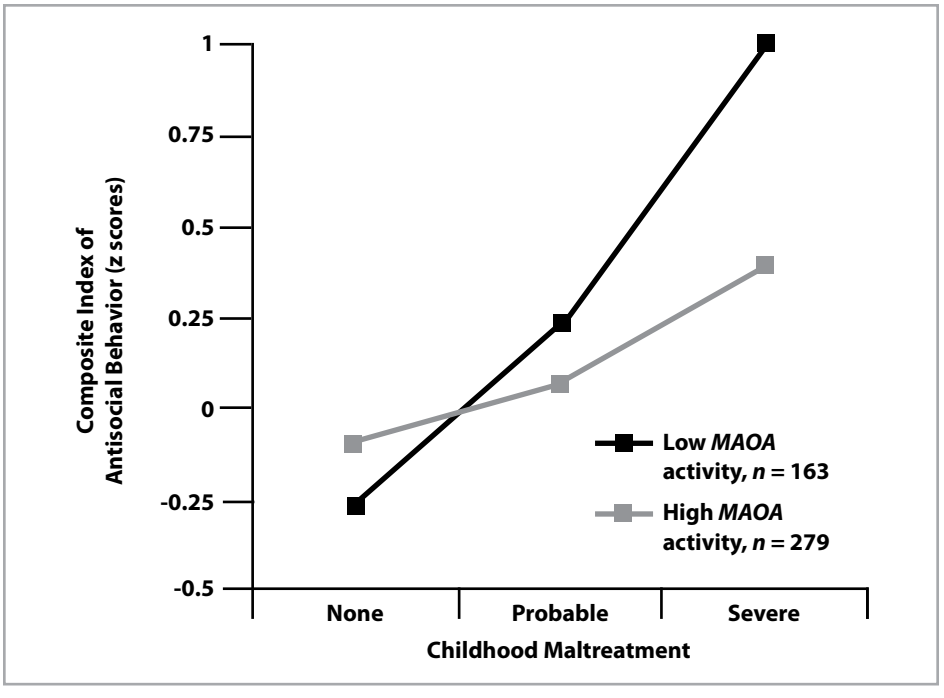

Figure 2.2. Interaction of *MAOA* genotype and exposure to maltreatment. Exposure to maltreatment increases the severity of antisocial behavior in boys, but this effect is much larger in boys with the low activity version of the gene for *MAOA,* than for boys with the high activity version of this gene. From: Caspi et al[30] with permission from AAAS.

benefits, and higher IQ is one of these. Suspecting that some of this advantage may be related to the unique fatty acid composition of breast milk, investigators found that a gene involved in fatty acid metabolism *(FADS2)* may be critical to the IQ benefit associated with breastfeeding.[31] In children with one particular allele of *FADS2,* IQ scores were about 7 points higher if they were breastfed, but children with the other allele of *FADS2* did not show a significant effect of breastfeeding on IQ.

Lead intoxication provides another example of gene-environment interaction. Even at levels that are below the generally accepted limit of 10 μg/dL, population studies show that lead exposure is associated with lower IQ scores, after accounting for maternal IQ, various home environment factors, and other confounding factors. However, it may also be true that some individuals are more severely affected than others at any given level of lead exposure. This thesis has been articulated by many, perhaps most elegantly by Ruff,[32] but it is only recently that researchers have begun to explain why this may be so. A study of the effects of lead on executive functions (which are a diverse set of frontal lobe–dependent functions that include attention, inhibition, cognitive flexibility, and more) suggests that for some of these functions, the deleterious effects of lead exposure only appear in children with a specific allele of a gene for a dopamine neurotransmitter receptor.[33] These results require replication, but they again point to the intricate interplay between environmental and genetic factors in child development and behavior.

A final example of gene-environment interaction is widely appreciated in clinical medicine, but perhaps not usually thought of in this context: pharmacogenomics. While drug therapy operates through a biological mechanism, it can also be regarded as an environmental influence on child behavior. Physicians understand that individual patients often respond differently to any given drug therapy, and these differences may often be rooted in genetic differences. Individual differences in hepatic metabolism are the mechanism through which genetics interact with the effects of drug therapy. For patients treated with atomoxetine, for example, those who are "poor metabolizers" through the *CYP2D6* pathway have a higher incidence of side effects than patients who are "extensive metabolizers," and they also show a trend toward greater efficacy.[34] Genetic differences in neurotransmitter receptors or neurotransmitter metabolism may be associated with differences in the efficacy and tolerability of other drugs as well.

Yet another line of current research examines the interaction of environmental and epigenetic factors. Though this work has not yet been extended to humans, the data in rats suggest that maternal grooming and nurturing can lead to changes in gene methylation. This, in turn, results in changes in gene expression, which subsequently increase grooming and nurturing in the offspring when it matures.[35] The cycle thus perpetuates itself in an echo of human behavioral patterns.

Fragile X: An Example of Nature, Nurture, and Their Interactions

Fragile X syndrome is the most common inherited cause of intellectual disability, with Down syndrome being the most common non-inherited genetic etiology. Fragile X syndrome is the subject of very active research on multiple levels, from the molecular to the behavioral, and data generated from this research provide examples of the effects on child development and behavior of nature, nurture, and their interactions.[36]

The single gene mutation that defines fragile X syndrome causes intellectual disabilities and behavioral problems in most affected individuals. As with essentially all genetic conditions causing intellectual disability, however, there is a significant range of cognitive and behavioral function within the fragile X syndrome population. The biological and environmental factors that account for this variability are starting to be illuminated. First, while all affected individuals share the so-called full mutation in the FMRP gene, the epigenetic factor of gene methylation is absent in a small percentage of individuals. In those patients, the FMRP gene is still partially expressed, some protein product is made, and the cognitive impairment is moderated. Second, as is the case for many other genetic conditions, genetic mosaicism occurs in some individuals, such that some cells carry the fragile X syndrome mutation, but others do not. A closely related phenomenon, random X inactivation, also known as lyonization, also is associated with variability in symptom severity. As a result of random X inactivation, females who are heterozygous for the fragile X syndrome mutation can have either an uncommonly high or low percentage of cells in which the active X chromosome has the fragile X syndrome mutation.

Investigators recently examined the role of other genes, besides the FMRP gene itself, on behavioral symptoms in the fragile X syndrome population. One finding was that variations in a gene for a serotonin transporter were associated with different levels of aggressive and also of stereotypic behavior in male patients with fragile X syndrome.[37] These results need replication before they can be regarded as definitive, but they show the direction of current research and its potential to elucidate how genes interact with each other to affect child development and behavior.

Environmental factors are also of great importance in shaping children with fragile X syndrome. As many had suspected might be the case, the effect of having a mother who also has fragile X syndrome is very important. This phenomenon also has been demonstrated in mice that have the equivalent of fragile X syndrome.[38] More generally, it has been found that home environment, as assessed by a multidimensional, direct-observation instrument, has a significant influence on cognitive and adaptive outcomes in children with fragile X syndrome. The IQ of males with fragile X syndrome was most strongly predicted by the home environment, which included such dimensions as parental responsiveness to the child, presence of learning materials in the home, and parental efforts to provide developmental enrichment.[39] In fact, the association of home environment with IQ was numerically larger than for any other

variable, including the child's levels of FMRP and mean parental IQ (which was not a statistically significant predictor). For adaptive behavior, the association between home environment and total score on the Vineland Adaptive Behavior Scale was numerically larger than for any other predictor, including the child's own IQ.[40]

Conclusion

Nature and nurture both have profound effects on child development and behavior. Genetic differences can be associated with either subtle or dramatic effects, and the field of genomics is starting to unravel the complex interactions among the many genes involved in brain development and behavior. Environmental factors also can have either subtle or profound effects, and in contrast to genetic factors, many environmental influences are typically thought of in the context of their beneficial or protective effects. Education and other behavioral interventions are rightly regarded as environmental influences on child development and behavior. Newer approaches to intervention may be expanding the range of influence of educational efforts.

While many behavioral genetic studies have attempted to quantify the relative influence of genetic versus environmental influences, this endeavor is afflicted by various interpretative limitations and is of little relevance to individual patient cases. Of greater research and clinical interest is the burgeoning understanding of gene-environment interactions. This research is beginning to show that the variability of genetic and environmental influences may often arise because of their interaction. In the future, clinicians may need to avail themselves of genetic diagnostics to determine how individual patients will be affected by either positive or negative environmental influences.

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Chapter 3

Interviewing and Counseling Children and Families

Prachi E. Shah, MD

Don't walk in front of me; I may not follow.
Don't walk behind me; I may not lead.
Just walk beside me and be my friend

~Albert Camus (1913–1960)

The Pediatric Visit: An Opportunity for a Goal-Directed Partnership

There is much wisdom from the French novelist, essayist, playwright, and Nobel Prize Laureate Albert Camus about what is needed to successfully interview and counsel families in the context of a pediatric visit. The provider need not dictate the agenda of the visit and "lead" the family according to a predetermined agenda. Nor should the provider limit the interview to the initial concerns that are expressed without inquiring about and exploring other potential areas salient to child health and development that the family may not explicitly mention. Successful interviewing and counseling of families requires that the provider "walk with" a family and develop a therapeutic alliance with the child and caregivers based on mutual respect and the shared goal of optimizing the child's health and developmental-behavioral outcomes. This holistic, family-centered approach to patient care is most likely to be adopted into practice when this perspective is introduced and integrated into pediatric residency training and incorporates the core concepts described below.

A mutual, goal-directed partnership between the primary pediatric health care professional and family is best achieved when pediatric care is continuous, comprehensive, family-centered, coordinated, compassionate, and culturally effective. This model is most successful when the provider is known to the child and family, and when there is a partnership of mutual responsibility and trust between them.[1] These characteristics of the ideal physician-family alliance are best captured in the model of the medical home. The medical home is a vision for how all individuals who are

involved in the delivery of health care services can partner with their patients and their families to help them achieve their maximum potential. This vision posits that optimal care is provided in a system that fosters collaboration and cooperation among all members of the community in which the child and family live.[2] In this model of the medical home, there is a need to learn to engage families and a responsibility to inquire about risk factors that can affect a child's growth and developmental trajectory.[3,4]

The goal of the pediatric health supervision visit, according to the American Academy of Pediatrics, is to promote children's optimal growth and development.[5] As a result of his or her regular and ongoing contact over time with infants, toddlers, and their families, the primary pediatric health care professional is well positioned to monitor and support early child development and behavior, and optimize child health outcomes.[6] However, evidence suggests that the pediatric visit is an often underutilized opportunity to identify developmental and behavioral concerns and to provide anticipatory guidance to families. To better address developmental and behavioral concerns in the context of the primary care visit, the pediatric health supervision visit must be adapted to address this unmet need. The pediatric visit must provide an opportunity for the parent and provider to communicate about the issues that are most salient to childhood health behavior and development. To understand how to use this opportunity during the clinical visit, let us explore the content and meaning of dialogues between a family and primary pediatric health care professional in the context of the clinical interview.

Capturing Missed Opportunities: Optimizing the Pediatric Encounter

Creating a "Holding Environment" in the Context of the Pediatric Visit

The opportunity to identify problems begins when the provider first inquires about positive developments within and between the child and family. Potential problems are further elucidated by discussing any concerns about their child's development and behavior. The clinical interview provides an ideal opportunity for providers to enhance their relationships with patients and families, assess the emotional states of patients, and uncover clues that might point to psychosocial distress or disturbance within the family context. The clinical interview can serve as a tool for gathering information, provide an opportunity to form a therapeutic alliance with the family, and serve as a means for influencing behavior. Successful interviewing is maximized by using a developmental approach and some principles of family engagement and assessment.[7]

Employing some general principles regarding interviewing and assessment is helpful to create an environment where the family can feel that their concerns are heard and respected. This involves building a therapeutic alliance with both the caregiver and pediatric patient and viewing developmental and behavioral concerns through a

culturally sensitive lens. This process is best achieved when the primary pediatric health care professional can provide a psychological "holding environment," in which parents feel safe to articulate their fears, vulnerabilities, challenges, or concerns as parents.[8] The concept of the holding environment was first described by Donald Winnicott, a British pediatrician, psychiatrist, and psychoanalyst, as a means of articulating what infants need from their caregivers to feel safe and secure.[9] In Winnicott's holding environment, the primary caregiver provides an environment of physical and psychological support in which the infant can feel safe and develop his sense of self. In much the same way, the primary pediatric health care professional can create a supportive holding environment for parents in which they feel a sense of safety, nurturance, support, and trust and in which the provider can explore issues with the family that are most relevant to the child's and family's well-being: The health and well-being of children are intimately related to the parent's physical, emotional, and social health and social circumstances.[10]

Based on trust built of an ongoing caring relationship, the physician can comfortably ask important personal questions that shed light on the key resources for achieving each child's developmental potential. Relevant areas of inquiry with parents include their mood, family ties, work issues, perceived social support, social affiliations, child care needs, health status, and financial security. These conditions influence family dynamics, support a child's health and growth, and are essential domains for the pediatrician to explore.[11]

A helpful framework to address parents' concerns and provide anticipatory guidance in the context of the pediatric encounter can be conceptualized by the mnemonic SHARE.[12]

- **S:** *Set* the tone: Create a holding environment in the context of the pediatric visit.
  Support parent and child: Build a therapeutic alliance.
- **H:** *Hear* the parent's concerns about their child's behavior and development and the effects on family functioning through the use of some guided questions.
- **A:** *Address* specific risk factors for child development and family functioning.
  Allow parents to reflect how cultural traditions contribute to their expectations of child behavior and development.
- **R:** *Reflect* with parents their experience of their child.
  Reframe child behavior and development in terms of the child's developmental level.
  Revisit the therapeutic goals set.
- **E:** *Empower* the parent and child by formulating an action plan to address the concerns voiced during the visit.

The Art of Interviewing

S: Setting the Tone/Supporting the Parents and Child in the Pediatric Visit: Developing a Therapeutic Alliance

Creating a safe space and open environment to share the details that are most salient to child development and family functioning requires special attention to the nuances of the initial family encounter. Nurturing emotional development in children must begin with support and nurturing for the parents in their caregiving role. As the family is the primary vehicle for children's early development, then the family is the pediatric patient.[11] This can begin by creating the time and space for parents and children to feel that they can voice their concerns. Some general principles regarding interviewing and counseling should be considered. Efficient communication requires unbroken attention. Privacy will increase the information shared during an interview and is especially important when sensitive psychosocial issues are being discussed. Equally important is explicitly addressing issues of confidentiality with patients and parents prior to clinical assessment and having adequate time to address the family's concerns.[7]

A successful therapeutic alliance between a physician and patient is thought to incorporate 3 main components: (1) agreement on goals that are the desired outcomes of the therapeutic process; (2) agreement on tasks that are the steps undertaken to achieve the goals; and (3) a bond between the physician and patient built on shared values, such as trust, respect, genuineness, positive regard, and empathy.[13] Although important factors for achieving a therapeutic alliance include listening to patients sensitively, being empathetic, and building trust and respect, perhaps the most important factor for building a therapeutic alliance between a physician and family is the ability to collaboratively identify mutually agreed on goals for the physician-family encounter.[14]

The pediatrician has the added challenge of developing a therapeutic alliance with both the parent and the pediatric patient. This can be facilitated if an alliance can be forged with the child in a developmentally sensitive manner.

Infancy (0–1 years): In the period of infancy, the most primary developmental process is the development of a sense of trust in his caregiver and the world around him. The young infant is dependent on his caregiver for a sense of safety, security, and to help him control and regulate his emotions.[15] In the first year of life the "interview" of the child, or more accurately put, the assessment of the child, should take place in the presence of the caregiver, preferably in the caregiver's arms if possible. A soft tone of voice and gentle handling of the infant are important means of helping him develop a sense of trust and comfort with the primary pediatric health care professional. Narrating to the infant what will happen

in the context of the visit can help both the infant and the caregiver feel more comfortable. A therapeutic alliance can be forged with both the infant and the caregiver when the caregiver's concerns are articulated and directed toward the infant during the assessment, "Your mommy wonders why you are so fussy at night. Is there something in your ears…in your tummy…" etc.

Toddler Years (1–3 years): The hallmark of the toddler years is the desire for autonomy, stranger wariness, separation, and individuation. In the context of the health supervision visit, the toddler may seek to have more control and active participation. At the end of the first year of life, the infant has developed a framework of attachment to his primary caregiver based on his history of early experiences.[16] In this attachment relationship, the caregiver serves as both a "secure base" from whom the toddler can explore an unfamiliar environment and a "safe haven" to whom he can return when he is distressed.[17] Rapport can be built with the toddler by allowing and encouraging exploration, being sensitive to the toddler's needs for "emotional refueling" from the caregiver, and indulging the toddler's desire for autonomy and control, "First I will listen to your heart, then you can listen…."

Preschool Years (3–6 years): Preschoolers are developmentally in Piaget's "preoperational thinking" stage. This developmental stage is characterized by egocentric and magical thinking. The preschool child has greater verbal and cognitive capacities than the infant and toddler but often views the world in a very concrete and self-oriented way.[18] The preschooler may view illness as a "punishment" for certain behavior (eg, "My stomach hurts because I did not eat my vegetables last night.") or may view illness as something that was caused by "magic." To engage the preschool child, it is often helpful to reassure the child that illness is not their fault or a result of bad behavior, and it can be helpful to probe with the child their understanding of why they don't feel well. The provider can further build rapport with the preoperational child by engaging them in the solution, "What do you think we should do to help you feel better? Maybe we can give you some 'magic medicine' to take away your earache."

School-aged Child (7–12 years): The school-aged child is at a period of advancing verbal and cognitive development. His thinking is more logical, organized, and concrete, and he is better able to understand cause and effect. Rapport with the school-aged child can be facilitated by inquiring about school, hobbies, and friends. The school-aged child can be more actively engaged in the clinical interview and can be directly queried about how he feels, what his concerns are, and what his goals are for the visit. The school-aged child can also be invited to assume a greater responsibility in the treatment process, "Now that you are such a big boy/girl, I need you to help your mom remember that you will need to take your medicine every day until it is all gone."

Adolescence (13–21 years): Developmentally, the adolescent patient is in the process of gaining autonomy from his parents. This emerging autonomy can be respected by structuring the pediatric visit to include both time with the parent and adolescent together and time with the adolescent alone. Confidentiality and its limitations should be addressed and acknowledged before the parent leaves the room.[7] Building an alliance with the adolescent and caregiver can be facilitated if mutually agreed-on therapeutic goals can be identified in the context of the visit, and if both the adolescent and caregiver are committed to achieving the desired change by working together toward the therapeutic goal.[19]

A successful therapeutic alliance between the primary pediatric health care professional, caregiver, and pediatric patient is best achieved when a partnership can be forged between the parents and the professionals caring for their children. This family-oriented approach incorporates the view that parents play an important role in the health and well-being of their children and is based on the following assumptions: (1) the parents know the children best and want the best for them, (2) each family is different and unique, and (3) a child's well-being is affected by the stress and coping of other family members.[20] The pediatric encounter should be structured such that the parent's desire for the well-being of their child is acknowledged and addressed and that the parent can feel free to share the issues that are serving as a stress to the relationship. This can be facilitated through the careful use of guided, open-ended questions.

Facilitating a Dialogue

H: Hear the Parent's Concerns About Their Child's Behavior and Development, and the Effects on Family Functioning

Parents often present to the pediatric visit with concerns about their child's health, behavior, or development. It has been suggested that nearly half of parents have concerns about their young child's behavior, speech, or social development,[21] but some parents are reluctant to share their concerns about behavioral and developmental issues with their pediatrician.[22,23] For some parents, societal and cultural beliefs influence what child behavior concerns they feel comfortable divulging to their pediatrician and what concerns remain unshared.[22,24] To ensure that the parents' deepest concerns and needs are expressed and addressed, the pediatric health supervision visit must be adapted to address this aspect of the child's health. The pediatric health supervision visit must provide the opportunity for parents to "tell their story," to express their perceptions about the strengths and vulnerabilities of their children and families.[25]

This can more readily occur when a therapeutic alliance built on trust and mutual respect has been established between the provider and family. Problems, concerns, and beliefs that a family may present may differ radically from the attitudes and beliefs of the provider. To ensure that a parent feels free to express their concerns

honestly and openly, special attention must be placed on creating an interview environment of openness and acceptance. In an environment of "radical acceptance," the provider actively welcomes all comments from parents and children in a nonjudgmental fashion and communicates a personal and professional commitment to openness during the interview, assessment, and treatment process.[26] In the spirit of this "bidirectional openness," the parents and child may articulate deep underlying beliefs that may play an important role in a child's health and behavioral outcomes.

The interview can begin with very open-ended, general questions such as, "What is going on in your child's life? How has he been lately? Is there something specific that you would like us to focus on today?" This sets the agenda and the "therapeutic goal" toward which the visit can be directed. As is developmentally appropriate, this question can be addressed to the child, and his input on the goals for the visit can be incorporated as well. In addition to addressing the concerns that were explicitly articulated in the pediatric visit, the provider must also gently probe for areas of concern related to a child's development and behavior. Developmental and behavioral problems have been described as the "new morbidity" in pediatrics[27,28]; psychosocial concerns are more prevalent in pediatric primary care[29]; and the need to manage developmental, behavioral, and emotional concerns is increasing.[30] To probe more deeply about a parent's developmental or behavioral concerns about their child, the parent can be asked, "Do you have any questions or concerns about how your child is learning, behaving, or developing?" Exploration of a parent's opinions and concerns about their child's behavior and development has been shown to be especially predictive of their child's developmental and behavioral status and is a key to identifying children at risk for developmental or behavioral problems[31,32] If a screening questionnaire was given prior to the encounter, often responses can guide the conversation and alert the provider to potential areas of concern.

When developmental or behavioral concerns are identified, it is often helpful to explore how the parent sees and views the child in light of these developmental and behavioral differences. What are the parent's perceptions of the child? What is the parent's understanding of the child's behavior and development? What are the parent's hopes and fears for the child? Exploration of these underlying issues often plays an important role in formulating a successful therapeutic intervention. It is also important to quantify how impairing the child's behavior is. Because there is often a reluctance to label a child's behavior as pathological, significant behavioral problems might be minimized or overlooked. A useful framework to help parents understand their child's behavior is to inquire how the child's behavior is affecting the family system and family functioning. If a child's behavior interferes with a parent's ability to maintain employment, maintain family routines, go out in public, or complete household chores, further evaluation may be warranted.[24] In addition, the parent's perception of their child should be explored as this perception influences parental

behavior toward the child and affects the child's behavior toward the parent.[33] These parental perceptions of the child can provide valuable clues to the dynamics of the parent-child relationship and can provide clues about the origin of behavioral disturbances.[34] These issues regarding a parent's perception of the child's behavior, and the impact of behavior on family functioning, can be explored with the caregiver using select "trigger questions" to help elicit information that may be difficult for parents to talk about.[8]

- Does your child's behavior interfere with your ability to maintain family routines (eg, eating dinner together at home)?
- Does your child's behavior interfere with your ability to go out in public (eg, eating out at a restaurant, going to the grocery store)?
- Does your child's behavior keep you from getting things done at home (eg, doing chores at home, talking on the phone)?
- Has your child's behavior affected your ability to maintain employment (eg, because of a difficulty in maintaining child care)?
- How has your child's behavior affected your relationship with your spouse?
- Has your child's behavior affected his functioning at home or school or with his friends?
- Tell me, what is most difficult about your child's behavior?
- Why do you think he does it?
- How does his behavior make you feel?
- Who does his behavior remind you of?
- What are your hopes for your child?
- What are your fears for your child?
- If you could pick 3 words to describe your child's personality, what would they be and why?

Understanding the parent's perceptions of their child's behavior and hearing the parent's hopes and fears for their child are important pieces of information that can guide the therapeutic process. When a parent's deepest hopes and fears are freely expressed, the interventions can be targeted to actualize the parent's hopes and mitigate their fears.

Probing More Deeply

A: Address Specific Risk Factors for Child Development and Family Functioning

Allow Parents to Reflect How Cultural Traditions Contribute to Their Expectations of Child Behavior and Development

In addition to addressing the concerns that were brought up by the family in the context of the pediatric visit, it is important to inquire about specific risk factors for child developmental and behavioral problems. The health and well-being of children are intimately related to their parents' physical, emotional, and social health and social circumstances.[10] Divorce, marital discord, domestic violence, substance

abuse, poverty, stress, mental illness, financial stresses, and a lack of social support are just a few of the struggles that affect today's families and impact the early parent-child relationship. Within the context of the pediatric visit, the primary pediatric health care professional can explore issues with the family that are most relevant to their well-being.

Based on trust built of an ongoing caring relationship, the provider can comfortably ask important personal questions that shed light on the key resources for achieving each child's developmental potential. Relevant areas of inquiry with parents include their mood, family ties, work issues, perceived social support, social affiliations, child care needs, health status, and financial security. These conditions influence the dynamic of the family and supporting a child's health and growth. As such, they are therefore within the pediatrician's rightful domain to explore.[11]

Nurturing emotional development in children must begin with support and nurturing for the parents in their caregiving role. Because the family is the primary vehicle for children's early development, the family is the pediatric patient.[11] Risk factors that affect the stability of the caregiving environment will undoubtedly manifest as child health and behavioral concerns[35,36] and can affect parenting practices.[37] The pediatric health supervision visit is a natural opportunity to inquire about the presence of risk factors that can challenge caregiving, including maternal depression, domestic violence, poverty, community violence, parental discord, and the presence of social and community support.

In addition, special attention must be placed on the role of culture on parenting and child development and behavior. Cultural sensitivity implies an awareness of the influence of multiple factors that can shape the priorities and perspectives of individuals and families in society.[38] Culture can influence a parent's understanding and interpretation of child behavior and development[39] and can influence parenting practices.[40] The role of culture in families should be explored to better understand the issues that affect the health care of patients and their families.[41] For the pediatric patient, this can include, but is not limited to, parenting philosophies, the influence of the American culture, parenting practices/discipline, religion and spirituality, and behavioral expectations. Some targeted questions to explore the role of culture in parenting are as follows.[40]

- Who do you live with?
- Who do you trust to take care of your child?
- What are some important values that you want to teach your child?
- Do you want your child to keep your cultural traditions? In what way?
- Does spirituality/religion play a role in raising your children? How?
- How do you teach your child right from wrong?
- How do you discipline your child?
- Do you use any cultural or home remedies?
- What are some hopes and dreams that you have for your child?

Understanding the cultural lens through which the parents view child behavior and development can serve as an important framework in providing anticipatory guidance, feedback, and formulating a therapeutic plan with the family.

Providing Anticipatory Guidance and Feedback

R: Reflect With Parents Their Experience of Their Child/Reframe Child Behavior and Development in Terms of the Child's Developmental Level/ Revisit Therapeutic Goals Set

Once the family's concerns are identified in the pediatric interview and the appropriate assessment has been undertaken, it is important to provide feedback and anticipatory guidance to families. Anticipatory guidance is the provision of information to parents and or children with an expected outcome being a change in parent and/or patient attitude or knowledge,[42] which can serve as the mechanism for strengthening a child's developmental potential.[43] The pediatrician has the opportunity to support and encourage parents in the parenting role and help foster a sense of parental competence in the midst of challenging caregiving experiences.[8] In the context of the supportive pediatric holding environment, the primary pediatric health care professional can help the parent experience a sense of safety, nurturance, and support and can address, acknowledge, and validate the parent's fears, anxieties, and experiences. In reflecting back to the parent the dialogue they have shared (eg, "What I hear you saying is…"; "It sounds like you are most concerned about…"), sends a very powerful message to the parent that, "I have heard your concerns, and I am with you in this process." When a parent is assured that their fears and concerns are heard and recognized, it then becomes easier to offer another perspective of the child's behavior, perhaps informed by the child's developmental level: "I wonder if when your child hits you because he is frustrated it is because he doesn't have the words to tell you how he is feeling, so he communicates his feelings to you the only way he knows how… with his body instead of with his words."

Undoubtedly, there will be times when the primary pediatric health care professional and the parent have differences in philosophies, perspectives, and even in management strategies. At times, parents may even be resistant and difficult to engage. This is when it is helpful to revisit the initial therapeutic goals that were mutually agreed on at the beginning of the visit. Often the parent, child, and the provider can find common ground and a therapeutic alliance by joining together to actualize the expressed hopes (eg, "I want us to have a better relationship.") and prevent the spoken fears (eg, "I don't want him to get into drugs or a bad group of friends.") When there are areas of ideological difference, the parent and provider can connect at the level of their shared desire: to optimize the health and development of the pediatric patient. In an open and supportive bidirectional communication, the provider can engage in an open and nonthreatening dialogue with the parent about what can be done to foster and optimize the child's health, developmental, and behavioral outcomes.

Steps for the Future

E: Empower the Parent and Child: Formulating an "Action Plan" to Address Concerns

One of the most powerful strategies a physician can offer a family experiencing challenges is to highlight the family's strengths and identify their capacity to achieve the therapeutic goals set. In the context of this family-centered approach, the parents and child are viewed as partners in the treatment process in hopes of facilitating family empowerment.[44] This family-centered approach to addressing developmental and behavioral concerns should address the needs of the whole family and should address the needs of the child in the context of the caregiving environment. Child behavior problems cannot be divorced from the experiences of the immediate caregiving environment,[45] and interventions to address child behavior concerns should address the needs of the family unit as a whole. These interventions should include such strategies as expanding social supports, using family strengths, individualizing resources, and delivering services consistent with the family's cultural values and beliefs.[44] However, it has been posited that the most important element for treatment success is related to a family's ability to feel a sense of empowerment and locus of control over the presenting problem. By empowering families to develop possible solutions to problems or needs, the pediatrician is not only helping with the current situation but is also helping the family to develop skills to solve future problems independently.[44] In this approach, the pediatrician involves the family as an active collaborator in finding a solution to the problem presented. The pediatrician, through thoughtful and empathetic interviewing, seeks out the treatment goals of the family as they present for evaluation and then actively seeks their input and alliance in formulating a treatment goal.[14] In creating this physician-family alliance, the family members become active participants in identifying and implementing a solution to the identified concerns. Family empowerment has been demonstrated to be an important factor in child behavioral outcomes. When children and families were included in the decision-making process and were provided services that were sensitive to their unique needs, values, and strengths, parents reported improvement in their child's behavior and reported a greater confidence in their ability to handle behavioral concerns in the future.[44]

To foster the parent's feeling of empowerment and competence in the parenting role, it is important that the parents experience a sense of support and connectedness in their caregiving role. This can occur through connecting the family with other supports and services available in the community. Supporting the needs of the child and family as part of a seamless continuum has been captured by the concept of "contextual pediatrics," or the idea that developmental support and guidance to families cannot be provided in isolation: Pediatric health care must be integrated within the framework of other community services.[46] The changing face of pediatric health care calls for a greater emphasis on collaborating with community partners, such as teachers, child care providers, early intervention specialists, etc, to optimize family

functioning and child health.[10] The importance of the pediatrician's collaboration with community partners was highlighted in a recent report by the Commonwealth Fund on the research from the ABCD-II Consortium.[47] This report suggested that to optimize identification of early social-emotional and behavioral concerns, the primary pediatric health care professional should provide formal screening for social-emotional concerns and collaborate with community partners to provide follow-up services. Once the pediatrician identifies behavioral concerns, collaboration with community partners ensures that families get the support and services they need. Effective support for social-emotional development can begin with greater attention to developmental and behavioral concerns during office visits and with the collaboration of partners in the community. An interdisciplinary, cross-agency commitment to fostering healthy emotional development is necessary to provide the supportive, responsive care children need in all of the environments in which they live, grow, and develop.

Conclusion

Given the frequent contact a primary pediatric health care professional has with families, there is a natural opportunity to talk about development, behavior, and psychosocial issues. The provider has the opportunity to optimize developmental, behavioral, and social-emotional development by being attentive to the quality of the family environment in which the child lives, by being attuned to risk factors in the caregiving environment, and by providing support and intervention and empowering families when vulnerabilities are identified. By partnering with other childhood resources in the community, the primary pediatric health care professional can foster the health and well-being of the family and optimize the health and development of children across the life span.

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Chapter 4

Basics of Child Behavior and Primary Care Management of Common Behavioral Problems

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Introduction

Behavioral and emotional difficulties are among the most common reasons that children are seen by primary pediatric health care professionals. Over 12% of children meet criteria for a behavioral or emotional disorder.[1] More often parents will raise concerns and seek intervention for behaviors that are not indicative of a specific disorder but are distressing to the child or family. To help these children and families, practitioners must assess the magnitude of the problem and identify contributing factors. Then they must opt to provide reassurance, recommend a specific intervention, or refer the child to a specialist. In this chapter we provide a brief overview of why behavioral and emotional problems occur, how to assess the magnitude of the problem, and factors to consider in deciding to intervene or refer. We then offer principles of behavior management to pediatric providers so that they can effectively help guide parents in the promotion of appropriate behaviors in their children. Finally, we review interventions for some common behavioral syndromes.

Assessing Behavioral Difficulties

Screening for Behavior Problems

At well-child visits primary pediatric health care professionals are expected to accomplish a lot in a brief period. Some parents will directly raise their concerns about their child's behavior, but others will do so only if specifically asked. Parents may hesitate to raise concerns about their child's behavior because of the perceived implications, including parental blame and uncertainty regarding the appropriate setting to discuss behavioral concerns. Medical providers may hesitate to ask or

screen for behavioral concerns if they lack knowledge of interventions or community resources to which to refer.[2] Thus not infrequently parents report unmet concerns about their child's behavior or emotions following well-child visits.[1] The use of standardized behavioral screening tools is one strategy to efficiently improve detection of behavioral and emotional problems in primary care.

The Pediatric Symptom Checklist (PSC) is a 35-item parent-report scale that has been demonstrated to improve detection of behavioral and emotional problems in children ages 4 to 15 years.[3] It is available online at http://www2.massgeneral.org/allpsych/pediatricsymptomchecklist/psc_home.htm. Parents can complete the PSC in 5 to 10 minutes, and it can be scored in less than 5 minutes. Recently, an abbreviated 17-item version (PSC-17) has been reported to be an even more efficient behavioral screening measure, although it may miss children with anxiety disorders and it is not as well studied.[4] For younger children, the Brief Infant-Toddler Social and Emotional Assessment (BITSEA)[5] or Ages and Stages Social-Emotional (ASQ:SE)[6] are brief behavioral screening tools appropriate for use in primary care.

Defining the Problem

Once a behavioral concern is identified, the practitioner needs to obtain a description of what the child is doing; the settings in which the problem behavior occurs; and a description of the frequency, intensity, and duration of the behavior. Understanding these aspects of behavior is necessary to distinguish typical child behaviors from those that are more problematic. For example, a 3-year-old child described as having temper tantrums would not seem unusual, but tantrums that occur many times a day, last 30 to 60 minutes, and are associated with aggression at home and preschool are much more concerning. Behaviors that occur at high frequency, high intensity, or in the presence of different caretakers or in multiple environments are more suggestive of a behavioral or emotional disorder.[7] In these situations, intervention in primary care settings will be more difficult and referral to a specialist is generally indicated.

Understanding the Problem

After defining the behavior, the practitioner must develop an understanding of why the problem behavior is occurring. This involves identifying contributing factors in the child and in the environment, as well as external factors that precede and follow the problem. Child factors to consider include the child's age and developmental level; the child's temperament or personality; and the possibility of behavioral, developmental, emotional, or physical disorders. One must interpret the behavior with regard to the child's age and developmental level. A 2-year-old child who often says "no" and cries if she does not get her way may be asserting her autonomy in a developmentally expected manner, but this same behavior would be more concerning in a 4-year-old. When evaluating a 4-year-old child with these behaviors, one needs to consider the possibility that the child has developmental delays. Considering the

possibility of a developmental disorder is also important because children with language delays, intellectual disabilities, and autism spectrum disorders have an increased frequency of behavioral and emotional disorders.

Children vary in how they approach and respond to different situations. Some children are upset by a sudden change in plans, while others are more flexible. Some children get tired at predictable times, while others get tired at more variable times. Some children are very active, while others are more sedate. These differences in behavioral tendencies are often referred to as variations in temperament. The different dimensions along which one sees variability in these behavioral tendencies is the subject of ongoing debate, but one frequently used model describes 9 dimensions along which differences in behavioral tendencies can be observed: adaptability, regularity, activity, intensity, persistence, approach/withdrawal (to new situations), sensitivity to sensory stimuli, mood, and distractability.[8] A moderate portion of the variation in temperament seen in children is due to genetic factors.[9] Thus children living in very similar environments can exhibit very different behaviors. Certain temperament characteristics tend to be associated with an increased likelihood of difficult behaviors. These characteristics include low adaptability, high intensity, low regularity, withdrawal in new situations, and a frequently negative mood. However, the child's specific temperament is less important than the relationship between temperament and environmental expectations or demands.

When there is a mismatch between a child's temperament and the expectations or demands placed on the child, it is likely that behavioral problems will occur. For example, a very active child being raised by very active parents, who are thrilled by the fact that the child is "always on the go," will not be seen by the parents as having a behavior problem. In contrast, less active parents who expected a toddler who would sit on their lap and engage in more sedate play may view the child's behavior as problematic. If parents do not understand the child's temperament and insist on the child engaging in more sedate activities, the child's resistance may be seen as an oppositional behavior, and tantrums or other disruptive behaviors may develop. In these situations, helping parents to appreciate their child's temperament and adjust their approach to better match the child's temperament can minimize behavior problems and decrease parental tendency to feel like they are to blame for their child's behaviors.[10]

In addition to understanding the child's temperament and parent (or other caretakers') expectations, a thorough understanding of the behavioral difficulties will require understanding how certain situations increase the likelihood of problematic behaviors occurring and understanding how the responses to these behaviors will make it more or less likely that these behaviors will occur again in the future. Factors occurring prior to a behavior that increase the likelihood of the behavior occurring are referred to as the antecedents of that behavior. Antecedents include examples of similar behavior that the child then imitates. Some antecedents may be characteristics of the timing of or setting in which the problem behaviors occur. For

example, problem behaviors are more likely when a child is tired, hungry, or not feeling well or when the child is in under- or overstimulating environments. Other antecedents may be specific demands (or triggers) placed on the child, such as to perform a task or entertain oneself for a time. Demands and timing may interact with each other. A child is likely to respond differently to the instruction to get ready for bed if it is given at the end of his favorite TV show than if it is given in the middle of the show. Because antecedents occur prior to the problem behavior, understanding antecedents and modifying them (when possible) is critical in preventing problem behaviors.

Before intervention, the practitioner must know how the parents understand the behavior, how the parents have been responding to the behavior, and what attempts they have made to change the behavior. The parents' understanding of the behavior may be influenced by family, community, or cultural factors, all of which the practitioner will need to consider when counseling the family. When a problem behavior is occurring repeatedly, it is very likely that the response to the behavior is reinforcing it. Any response to a behavior (regardless of the intent of the response) that maintains or increases the frequency of the behavior is referred to as a reinforcer. A common scenario in many homes is when a child who wants parental attention while the parent is on the phone engages in disruptive behavior. If the parent gets off the phone and yells at the child their intent may be to punish the child by yelling, but it is likely that they are reinforcing the disruptive behavior by providing the child attention. Positive reinforcers increase the likelihood of a behavior by providing the child something the child wants, often adult attention or a desired item. Negative reinforcers increase the likelihood of a behavior by removing a demand. A teacher who tells students that if they complete homework on Monday through Thursday they will not be given homework on Friday is using negative reinforcement to increase the likelihood that students complete homework during the week. Punishment is a response to a behavior that decreases the likelihood of the behavior occurring again in the future.

When assessing parents' past attempts to change problem behavior, it is very important to have parents describe what happens and not just name the procedure they used. Parents may have tried to ignore a specific behavior, but have only done this inconsistently or for brief periods. Other parents may have tried to reward appropriate behavior, but the child rarely, if ever, earned the rewards. These parents believe that they have tried ignoring or rewarding as behavioral change strategies, but as we will describe later in this chapter, they have not followed these strategies effectively. In contrast, if a number of well-executed behavioral change strategies have not been successful, it is much more likely that a behavioral or emotional disorder is present.

After developing an understanding of the behavior, the practitioner must decide whether to recommend interventions to the family or refer for intervention. At least in part, this decision will be made on the basis of the time available to the

practitioner and the practitioner's interest in and comfort with managing behavior problems. Factors that make it more difficult to successfully help families improve their child's behavior are summarized in Box 4.1. When one or more of these factors are present, practitioners should strongly consider referral.

Box 4.1. Factors That Make It More Difficult to Change Behavior

- Problem behaviors are pervasive across time, persons, and settings.
- Problem behaviors cause severe disruption at home, school, or community.
- Problem behaviors threaten the safety of the child or others.
- Previous attempts to change the behavior have failed (particularly if previous attempts were well executed).
- Problem behaviors occur in the context of multiple psychosocial stressors.
- Parents do not agree on behavior management strategies.

Principles of Behavior Management

Behavior management starts with gaining an understanding of the behavior, which then guides the next step: changing behavior. The focus of behavioral change should be on encouraging the behavior we want to see, as well as discouraging the undesired behavior. Behavioral intervention occurs on several different levels, depending on the particular problem and situation (Table 4.1). Antecedent modification refers to changing the factors that trigger the problem in order to prevent the problem behavior from occurring. Giving instructions communicates how we want the child to behave. Finally, consequence modification refers to changing how the parents (or others) respond to both problem and desired behaviors.

Modifying antecedents

One method for modifying antecedents is to change the child's physical environment. Childproofing, for example, removes access to hazards, decreasing the likelihood that the child might play with hazardous materials. Tantrums in a toy store can be reduced by avoiding the store. Aggressive behavior noted after the child plays fighting video games can be reduced by removing the child's access to such games. Placing a high latch on a door decreases the likelihood of a young child escaping.

Antecedent modification may include teaching skills needed for appropriate behavior. Misbehaviors that occur because of frustration often require these interventions. Teaching effective communication skills, for example, would be part of the behavioral plan for a child who gets frustrated because of difficulty with language. Similarly, obtaining occupational therapy for graphomotor computer keyboard skills might be a useful intervention for a child whose misbehavior is related to frustration with handwriting.

Table 4.1. Examples of Antecedent and Consequence Modification for Different Triggers

Trigger	Antecedent Modification	Consequence Modification
Restricted access (being told "no," "you can't," "stop," "don't," etc)	Remove forbidden items from environment (eg, childproofing). Clearly state rules for access. Distract the child.	Consistently enforce rules. Allow access for appropriate behavior. Remove child from situation. Ignore misbehavior. Punish aggression associated with tantrum.
Need or desire for attention	Plan activities with child. Have adult available to engage child when parent is busy.	Increase frequency of attention for appropriate behavior. Ignore misbehavior. Institute time-out for misbehavior.
Difficulty with task demands or communication	Alter task demand if difficulty is too high, provide breaks. Help child before frustration occurs. Model the behavior desired. Show the child in pictures the sequence of tasks expected. Augment communication with sign language, pictures, etc.	Reward successful task completion. Follow non-preferred tasks with preferred tasks. Insist on task completion despite child's protest behavior. Provide help when request for help is made appropriately.
Transitions, change in routine, or change in what child expects	Create visual schedules. Prepare the child for changes and transitions. Establish routines. Teaching skills related to flexibility.	Calmly persist with change while ignoring inappropriate behavior. Praise successful transitions. Place preferred activities before non-preferred activities.
Provocation by sibling or peer	Monitor interactions. Teach appropriate interacting. Find activities both enjoy.	Punish provocation and retaliation Pay attention when siblings are interacting well.
Fear or aversive sensory stimulus	Avoid fearful or adverse stimulus. Expose gradually.	Reward exposure. Respond to appropriate requests for breaks or escape.

Another means of antecedent control is to ensure that appropriate behavior is modeled by the child's peers and other adults. One of the most important ways that children learn behavior is through imitation.[11] By the time a child is 2 years old, they have taken on many of the mannerisms of their family and culture. They speak with similar words and inflection, have similar gestures and facial expressions, incorporate their observations into their pretend play, and begin to interact with each other the way others interact with them. In order to use modeling to change behavior, parents need to be mindful of the behavior they want their child to display and to display such behavior themselves. For example, for a child who gets explosively angry, the appropriate behavior to model would be a calm response when upset, using words to relay the problem rather than behavior. Getting angry and yelling back at the child models the undesired behavior. For a child who is very anxious, a parent needs to model calm, confident reassurance.

Children who are exposed to inappropriate behavior often imitate that behavior. Most children are quick learners; that is, within their ability, they can model new behavior even after only a brief exposure: a parent's curse word is repeated by the child, or a wrestling hero's move is tried on a peer. When children display inappropriate adult behavior, such as sexualized behavior or threats of violence, one must also explore where such behavior has been witnessed. Such behavior may have been directly experienced in the home environment; at a babysitter's or child care; or indirectly through TV, videos, or the Internet.

Parents can modify antecedents by changing their expectations of their child's behavior based on their understanding of the child's developmental abilities, temperament, learning differences, and physiologic and mental health status. For example, parents of a very active child may give the child "movement breaks" throughout a meal instead of expecting the child to sit quietly for the duration of the whole meal. Parents may minimize demands when a child is tired or stressed. Parents of a 13-year-old child may give the child more responsibility and allow more participation in family decision-making.

Giving Instructions

A child's behavior can be shaped through the instructions the child is given. Helping parents give instructions more effectively is an important part of behavioral counseling.[12] Effective instruction starts with gaining the child's attention. This sounds obvious, but too often parents try to engage their child when the child is doing something else. If a parent yells "It's time for dinner!" from the kitchen when the child is absorbed in a video game in the basement, it's likely that the child is selectively attending to the video game and not the parents' instruction. Both the parent and child get rapidly exasperated if the parent sees the child as making an active decision to disobey. Such exasperation can be avoided if the parent first makes eye contact or receives verbal assurance that the child is listening before giving the instruction.

Instructions to alter or engage in a specific behavior need to be given in simple language that fits the comprehension abilities of the child. In general, the length of the instruction should match the typical length of the child's sentences. This doesn't mean that parents should always talk to the child in short sentences, but simplifying language when directing the child's behavior helps the child understand what is expected. Instructions should also be given as firm statements and not as a question, unless the instruction is a choice ("It's time to clean up" vs "Can we clean up now?"). A "firm" voice (or "sergeant" voice) helps the child differentiate a parent's directive from a choice.

Instructions that specify the desired behavior are more likely to be followed than general commands or "don't" instructions. "Behave" is less likely to be successful than "come sit by mommy." "Walk" is better than "stop running." Minimizing "don't"

instructions often requires parents to ignore minor misbehavior and redirect the child to what they want them to do.

Finally, successful instruction requires that the parent follow through with the instruction. Figure 4.1 illustrates a protocol to help parents follow through consistently with instructions to a child. After getting the child's attention, the parent issues the instruction or directive followed by 5 to 10 seconds of silence. The period of silence is to allow the child to process the instruction. If the child listens the parent should notice, and sometimes provide a reinforcer (next section). If the child does not listen the parent should reissue the instruction with a warning ("If you don't put your shoes on now, the TV is getting turned off."). If the child listens, the parent notices. If the child does not comply, the parent carries out the warning, and if the instruction was to complete a task, the parent reissues the directive. Children who do not comply unless the parent repeats the instruction multiple times or threatens them are likely playing the odds that a parent will drop the demand. From experience they have learned that the likelihood a parent will follow through on a demand increases once a threat is issued, or once a parent's voice reaches a certain angry pitch. Once a child learns that a parent is more consistent with follow through, warnings will not be needed as often in the future.

If a child does not follow an instruction because they do not understand the instruction, a different approach is needed. In this case, parents can reissue the instruction with physical guidance, with a prompt, or by modeling the behavior.

If a child is chronically oppositional, parents may need to practice this sequence with only a few instructions a day, and start with instructions with which the child is likely to comply. Success with these instructions will help bolster the parent's confidence and teach the child that compliance will be recognized with praise and adult attention. This will facilitate success with harder directives later.

Manipulating Consequences

Behavioral change can be augmented by altering the consequences of behavior, that is, changing the rewards or punishments that occur as a consequence of the behavior. Rewards are more effective behavioral interventions than are punishments. Therefore, a child needs to succeed at meeting parents' expectations most of the time. Initially this may require adults to change or lower expectations (an antecedent modification) to more easily achievable goals, which then can be altered as the child and parent become more successful.

To be effective, reinforcing consequences need to have a positive value for the child and punishing consequences a negative value. They also need to be acceptable to the parents. Attention has a positive value for most children. Reinforcer preference is otherwise quite individual and will vary over time. Rewards that a child earns frequently may lose their efficacy over time, as they become less valued by the child.

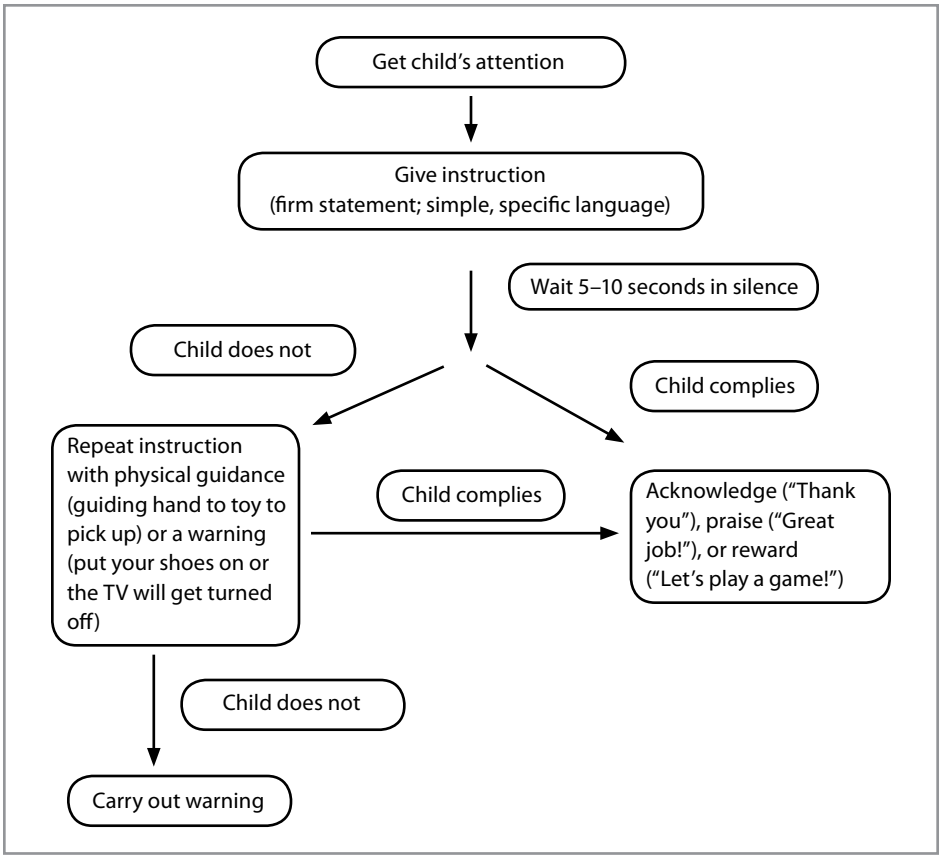

Figure 4.1. Giving instructions.

To avoid this satiation of the child with a specific reward, rewards should be varied to keep the child's interest (Table 4.2). For parents, the acceptability of the reward means both that the parent is willing to provide the reward each time the child engages in the desired behavior and that the parent is willing to restrict access to the reward if the child does not engage in the desired behavior. If a child is working toward earning a trip to a recently released movie, will the parents be willing to say "no" if the child has not earned the trip and his best friend's family invites him to the movie? If the parents are not willing to say "no" in this situation, then the trip to the movie is not acceptable as a reward for the behavioral plan.

Behavioral consequences are most effective if they immediately follow the behavior and specify the behavior that is being rewarded or punished. For example, telling a child "good job" without reference to what "good job" refers is unlikely to be effective at increasing any specific behavior. However, if a teacher says "good job on your handwriting, you're really working hard to stay in the lines," the praise is much more likely to be effective in increasing that behavior in the future. Most older children can

Table 4.2. Types of Reinforcers

Reinforcer	Examples
Social attention	Praise, comments, reprimands (negative attention is still attention), playing games, pat on the back, hair tousling
Tangibles	Treats, stickers, and toys
Privileges	Computer or television time, later bedtime or curfew, greater independence
Tokens (that can be exchanged for tangible reinforcers or privileges)	Money, stickers, checkmarks, as well as list of backup reinforcers, eg, 3 stickers = 30 minutes of a favorite game or activity
Negative reinforcement	Breaks from chores, break from homework

have delayed rewards and punishment, but there has to be a clear link between the behavior and the consequence. For younger children, or for those with developmental disabilities, a delayed consequence may inadvertently reinforce or punish an unintended behavior. For example, if a child says, "mommy, mommy" to get her mother's attention, then kicks her, and the mother responds, "what do you want, sweetie?" she is rewarding the kicking behavior, not the verbal behavior ("mommy"). Once a child's behavior is responding consistently to a reward, the frequency of the reward can be decreased and the behavior can be reinforced intermittently.

Many behavioral expectations of children are not single event behaviors but require compliance across time (eg, sitting still, staying by the cart in the grocery store, or keeping one's seatbelt buckled). To reinforce these behaviors, parents should reinforce periodically throughout the period of the expected behavior. A parent may be advised to bring special treats and small rewards (eg, stickers) to use at regular intervals during these types of activities.

If a child engages in an undesired behavior, a parent can ignore or punish the undesired behavior. Ignoring the undesired behavior may seem counterintuitive to parents. However, undesired behavior is often being reinforced because it results in parental attention, albeit negative attention. Negative attention can be as powerful as positive attention, given the child's goal is for parental attention of any type. When the undesired behavior is no longer reinforced by parental attention, the frequency of the behavior decreases over time. This process is termed extinction. If a behavior has been reinforced by allowing the child to escape from a demand, the use of extinction would require the parent to persist in getting the child to perform the task despite the inappropriate behavior. Practitioners counseling parents to use extinction as a behavior change strategy should warn families that often the behavior may worsen before it improves. If a child has learned that tantrums are an effective means of getting a candy bar, the first few times the parent refuses to give the child a candy bar, the child's tantrum may be longer or louder than it ever was before. Only after this fails on a few occasions will the child learn that the tantrums will no longer be reinforced, and the frequency of the tantrums will decrease.

Punishment does not teach desired behaviors but may be a necessary component of behavior management plans for undesired behaviors that cannot be ignored. Similar to rewards, punishment must be contingent on the behavior, specify the behavior being punished, and be acceptable enough to use each time the behavior occurs. If punishment is used every other time a specified misbehavior occurs, the child gets away with the behavior every other time. Corporal punishment is not recommended. Most parents do not find it acceptable to use corporal punishment each time a specified behavior occurs. In addition, corporal punishment has negative effects on parent-child relationships and child and adult mental health across cultures. Corporal punishment models behavior that would be inappropriate for the child to demonstrate with peers or adults. Although most people in the United States would agree that not all forms of corporal punishment are child abuse, most child abuse starts as corporal punishment.[13] For these reasons, practitioners should discourage parents from using corporal punishment.

Alternative punishment strategies are listed in Table 4.3. One of the most frequently recommended punishment strategies for children is time-out from positive reinforcement (referred to as time-out from here on). Time-out is usually implemented by having a child sit in a chair, stand in the corner, or go to their room for a brief period (1–5 minutes). Time-out is a well-studied and effective punishment procedure, but many families have difficulty administering it correctly. Pitfalls of time-out include not specifying the behavior being punished. In order to be consistent across time and caregivers, all those working with the child need to know what behaviors will result in time-out. Another problem is that parents pay attention to children while they are in time-out, or make time-out in an area of the house where they have access to attention and activities. Time-out facing a corner means the child has limited access to those around her. Finally, some children do not stay in time-out. This may occur if the amount of time one expects the child to sit is too long. A time-out of 30 seconds may be appropriate for a child who never sits still for longer than a few minutes. Until a child is aware of how time passes, 2 minutes would be the maximum time needed. Once a child knows the difference between a minute and 5 minutes (around 5 years), longer periods may be appropriate. If the child does leave time-out before time is up, they should be escorted back with minimal attention from the parent. No one punishment works for all children and families, and if time-out has not worked even when used properly, other options can be explored.

Behavior management counseling can be a very rewarding experience when the advice provided targets what the parent and child need. However, it requires time to fully understand the child factors, settings, expectations, and consequences that are influencing the behavior. One-size-fits-all advice is generally neither rewarding nor successful.

Table 4.3. Types of Punishment

Punishment	Description
Time-out from positive reinforcement	Contingent withdrawal of social attention and activities for a brief period.
Verbal reprimand	Brief instruction to change behavior.
Privilege withdrawal	Not allowing the child to engage in a fun activity for a brief period.
Response cost	Usually used in conjunction with a reward system in which the child can earn tokens for appropriate behavior and lose tokens for inappropriate behavior. If the child is losing more tokens than they are earning, it will not be effective.
Grounding	This is a specific type of privilege withdrawal in which the child is required to stay at home and not interact with friends for a specified period.
Job grounding	The child is grounded as defined above until they complete a specific task or chore. The length of the grounding is determined by when the child completes the task.
Natural consequences	Allowing the child to experience the consequences of a poor choice as opposed to fighting with the child about it. Works well for activities such as getting cold when not wearing a coat, but not for behaviors with more serious potential consequences.

Common Behavioral Syndromes

Infant Crying/Colic

The time infants spend crying progressively increases to a mean of approximately 2.5 hours per day during the second month of life and decreases progressively thereafter. When otherwise healthy infants cry intensely for an excessive duration, they are often referred to as having colic. The amount of crying required for a diagnosis of colic is not agreed on, but in research studies, more than 3 hours a day for more than 3 days per week is a frequently used definition.

The diagnosis of colic requires that the child be otherwise healthy and feeding well. Thus physical problems that can cause excessive crying must be excluded. Acute disorders that should be considered in a crying infant include infections, corneal abrasion, glaucoma, skull or long-bone fracture, incarcerated hernia, supraventricular tachycardia, intussusception, midgut volvulus, and a hair tourniquet on a digit.[14] A number of chronic conditions have been proposed to be the cause of infant colic, including cow's milk allergy, lactose intolerance, constipation, and gastroesophageal reflux. Although in any one case these problems are potential causes of crying, no well-designed study has suggested that these are common causes of excessive crying, and controlled studies using interventions targeting these problems have not been found to be effective for most infants.[15]

Infants with excessive crying have been found to have differences in temperament from those who cry less. Perhaps, not surprisingly, parents tend to rate these infants

as more intense and more difficult to soothe (less easily distracted). However, these temperament characteristics of infants with excessive crying are also supported by independent observations. For example, independent observers of infants undergoing a physical examination rated infants with colic as crying more intensely and being more difficult to console.[16] Infants with more persistent crying differ from those with less crying in that they have a higher crying-to-fussing ratio (suggesting greater intensity), and infants with colic were found to be less likely to soothe in response to an orally administered sucrose solution than were infants without colic.[17]

Management of colic involves reassuring parents that their child is healthy and helping parents to understand their infant's temperament traits contributing to the increased crying. Parents should be counseled that crying is the infant's way of communicating that there is something that the infant wants but it is not necessarily a sign of pain or illness. Parents usually think of some things that an infant might want, such as to be played with, fed, or have their diaper changed. However, parents may not think of the crying as a sign of the need to be quietly held, the need for nonnutritive sucking, or the need to be left alone to sleep.[18] Furthermore, parents need to understand that these infants are more difficult to soothe and, thus, even when the parent is providing what the infant wants, it may take many minutes before the infant stops crying. If parents rapidly change from one activity to another in futile attempts to get the infant to soothe, they may stop providing the infant what he or she wants before the infant communicates to the parent that it is what he or she wants by stopping crying.

Bedtime Resistance/Night Wakings

Sleep occurs in cycles that typically last about 90 minutes in older children and adults but are often shorter in younger children. During the cycles, the child goes from light sleep (described as Stage 1 and Stage 2 sleep) to deep non-REM sleep (Stage 3 and Stage 4 sleep) or into REM sleep. Most deep non-REM sleep occurs in the first third of the night, and most REM sleep occurs in the second half of the night.[19] Individuals wake briefly between sleep cycles but tend not to be aware of this as they rapidly go into the next sleep cycle.

Falling asleep is facilitated by a calming and familiar environment. For toddlers or preschoolers, a specific blanket, stuffed animal, pillow, or other item may be part of the familiar environment that helps the child to fall asleep. (Note that these items are not indicated for infants, given an increased risk of sudden infant death syndrome.[20]) These types of items are referred to as sleep associations, and when these items are not present, initiating sleep may be difficult. In most children with night wakings, the problem is not actually the waking, but rather trouble falling back asleep without the involvement of the parent, because the parent is part of the child's sleep associations.[21] For example, a parent and child cosleeping in the same bed is typical in many cultures and does not contribute to night waking because the parent is present throughout the night. However, in the United States, where most parents

want the child to sleep in their own bed, a parent lying down next to the child to get them to fall asleep is problematic, because the parent is not present when the child wakes between sleep cycles during the night. Other common problematic sleep associations include having an infant fall asleep while being rocked or nursed. Sometimes the parent is not part of the sleep association, but may be needed to help with a sleep association. For example, an infant may fall asleep sucking on a pacifier, but if the pacifier falls out of the child's mouth, the child may need the parent to put the pacifier back in their mouth during the night. Night wakings can be managed by teaching the child to fall asleep without the problematic sleep association. This often results in bedtime resistance that should be managed as described below.

If a child is having trouble falling asleep, practitioners should assess whether principles of good sleep hygiene are being maintained (Box 4.2). A regular morning wake-up time, consistent nap length, and a positive calming bedtime routine followed by a consistent bedtime are particularly important. When unclear, it is helpful for parents to complete a 1- to 2-week sleep diary focusing on whether the expected time of sleep is consistent with the child's physiological needs or tendencies. Trying to get a child to sleep when they are not tired or sleep past the time their biological clock is waking them is not likely to be successful. However, even at an appropriate bedtime, a child that is being required to learn to fall asleep in the absence of an established sleep association is likely to resist going to bed. In this situation, parents often have to let the child cry until they fall asleep.[22] After a few nights, most infants and toddlers will learn to fall asleep on their own, but the protests the first couple of nights can be dramatic and prolonged. Ignoring the tantrums is difficult for many parents, but if parents are going to check on the child it needs to be very brief, and the parent must not reintroduce the sleep association (eg, lie down with the child), as this would just teach the child that tantrums are effective in getting what the child wants.

Breath-holding Spells

Breath-holding spells are involuntary (reflexive) events. Typically they occur in response to an event that causes anger, frustration, fear, or minor injury. The child cries, becomes apneic at the end of exhaling, and then becomes pale or cyanotic. The child may lose consciousness and have a brief convulsion. Breath-holding spells usually begin between 3 and 18 months of age and may occur infrequently or multiple times a day. It would be rare for breath-holding spells to persist beyond 7 years of age.[23] Breath-holding spells are thought to be caused by dysregulation of the autonomic nervous system.[24]

Children with breath-holding spells should have hemoglobin and iron studies performed, as these spells have been associated with anemia and iron deficiency. If the history is typical, no further medical evaluation is needed. When the history is not clear, an electroencephalogram may be helpful in distinguishing breath-holding spells from seizures. If a child is pale and loses consciousness, an electrocardiogram

Box 4.2. Good Sleep Hygiene

Environment
- Dark (no more than a night-light)
- Quiet
- Comfortably cool

Schedule
- Regular AM wake-up time
- Consistent nap length
- Regular bedtime

Position—on back or side in infancy

Activities Around Bedtime
- Child is put into bed drowsy, but still awake
- No frightening TV or stories before bed
- No vigorous physical exercise in the hour before bed
- Consistent and soothing bedtime routine
- Avoid meals or hunger around bedtime

should be obtained to distinguish the spells from conditions associated with cardiac arrhythmias, such as long QT syndrome. In infants, one should consider the possibility of gastroesophageal reflux resulting in apnea. In very rare cases, breath-holding spells have been associated with brain stem dysfunction caused by tumors or Arnold-Chiari malformations.

In most cases, treatment primarily involves demystification and reassurance that even if the child loses consciousness, the child will start to breath again without intervention. Parents should be cautioned to avoid allowing breath-holding spells to prevent them from setting limits, even if the limits provoke the spells. If children learn that emotional outbursts prevent parents from setting firm limits, the frequency of the outburst and the associated breath-holding spells are likely to increase.

If the child is anemic, he or she should be treated with iron, as this will decrease the frequency of breath-holding spells in many children. Although much less well investigated, treatment with iron has also been reported to decrease the frequency of breath-holding spells in children who are not anemic.[25] Thus if the breath-holding spells are severe, treatment with iron could be considered.

Enuresis

Nocturnal enuresis is defined as nighttime urinary incontinence that occurs at least 2 times per week for at least 3 months after the age of 5 years. Approximately 10% of school-aged children have nocturnal enuresis, the incidence becoming smaller as children get older. After the age of 5, 16% of children will spontaneously remit each year.[26] Parents should be reassured that bed-wetting in children 5 years and younger is considered in the range of normal, and pull-ups at night are very acceptable and

appropriate. After the age of 5 years, when to treat depends on the amount of distress bed-wetting is causing to the child or family. Daytime (diurnal) enuresis is much less common in children once toilet trained, and is more commonly associated with physical or functional abnormalities of the urinary tract, requiring urological consultation. Less than 10% of children with nighttime enuresis have similar abnormalities. Both diurnal and nocturnal enuresis can be associated with chronic constipation.

Interventions for nocturnal enuresis include biobehavioral treatments or medications. The most successful biobehavioral treatment uses a bed-wetting alarm, a small moisture detector that attaches to the pajamas and alarms when the child begins to void. This wakes up the parents and child, so the child can go to the bathroom, finish voiding, clean up, and go back to bed. Sometimes the alarm does not awaken the child and, in these cases, the parents need to awaken the child while the alarm is going off so that the child learns to awaken to the alarm. Occasionally the alarm can be scary for children, particularly if it has not been demonstrated to the child during the daytime before it goes off in the middle of the night. The success rate of bed-wetting alarms is on average 78%, and treatment generally goes more smoothly if the child is motivated to stop the bed-wetting. Additional procedures sometimes used with the alarm include retention control training (having the child drink a large glass of water and, once they feel the need to urinate, holding it as long as possible with rewards for successively longer periods, up to 45 minutes) and cleanliness training (having the child remake the bed before going back to bed after the alarm). Overlearning (after the child is dry for 2 weeks on the alarm, giving the child fluid before bed, and continuing the alarm) has been used to decrease the relapse rate.[27] Other biobehavioral treatments include scheduled wakenings and toileting through the night, hypnotherapy, and cognitive-behavioral approaches. These treatments have shown some success in limited studies but do not have the robust results of those studies using bed-wetting alarms.[28]

Medications used for bed-wetting include desmopressin (ddAVP) tablets[29] (intranasal formulations of ddAVP are no longer indicated for the treatment of primary nocturnal enuresis in children due to an increased risk of hyponatremia) and imipramine.[30] Using either medication, approximately 20% of children become consistently dry at night, but effects are not generally sustained once medication is discontinued. Although ddAVP is generally safe, water intoxication leading to hyponatremia is an infrequent but serious adverse side effect. Identifiable and preventable risk factors contributing to hyponatremia are inappropriately high fluid intake, administration of a larger than recommended dose, young age (<6 years), and concomitant administration of another medication. Thus, when ddAVP is prescribed, parents should be instructed to avoid letting the child drink more than 8 ounces of fluid in the evening before bed; not take a higher than recommended dose; and promptly discontinue the medication and seek medical care if headache, nausea, or vomiting develops. Imipramine is rarely used because of the higher risk of side effects and the life-threatening consequences of an overdose.[31]

Encopresis

Encopresis is usually caused by chronic constipation with stool impaction and overflow incontinence. Incontinence can involve either solid or liquid stool in the underwear. Parents may misinterpret the leaking of stool as diarrhea. Occasionally the child may have large bowel movements that can block the toilet. Often parents think the child is deliberately soiling the underwear and set up elaborate toileting schedules, punishments for accidents, and rewards for no accidents. These interventions are not effective because the encopresis is a physiological problem, not a behavioral problem. The severe constipation disrupts functioning of the anal sphincters, so the overflow incontinence is beyond the child's control.

Treatment involves explaining the physiology of the disorder to the child and parents so that the demands of treatment make sense, and the parents and child are willing to comply with treatment. When a child is chronically impacted with stool, the muscle of the bowel wall is overstretched and stool evacuation is compromised, further exacerbating the problem. Due to the chronic stretching of the bowel wall, the child does not get a strong urge to defecate as the stool accumulates. Some children lose the urge to defecate altogether or only get the urge to defecate when the stool is at the verge of the external sphincter.

Treatment for encopresis requires first that the colon be disimpacted, followed by maintenance treatment. The safest, most effective way to clear the colon of stool is with daily or every other day Fleet enemas. Catharsis via orally administered agents may cause abdominal cramping and diarrhea at unpredictable times. If a child has failed previous treatment attempts, an abdominal x-ray may be needed to confirm that the catharsis has been successful, as one of the common problems encountered in treatment is inadequate bowel emptying.

Once the child is disimpacted, the maintenance phase of treatment consists of

- Daily treatment with laxatives at a dose that results in a daily soft, easy to pass bowel movement. One may need to experiment with different laxatives (polyethylene glycol 3350, magnesium hydroxide, lactulose, bisacodyl, senna) depending on the child's ability to take medication as well as effects and side effects. Polyethylene glycol 3350 is often the easiest to administer, as it is tasteless and colorless, so it can be easily ingested.
- Twice-daily toileting times should be incorporated into the family's schedule (best done after meals, usually breakfast and dinner). The child should sit for about 10 minutes, pushing periodically. Their feet should be flat on a solid surface with knees slightly higher than hips, as often even school-aged children may need support for their feet. Some children need to learn to push again, and valsalva practice is helpful.

- Toileting times should be followed by a preferred activity or treat, such as TV time, games, or dessert.
- If a child does not have a bowel movement for 48 hours, a stimulant laxative (bisacodyl or senna), an enema, or suppository should be used.

Once a child achieves consistent stool continence for several months (generally 4–6), laxatives can be slowly weaned. If the child has only been constipated for a short time, the laxative can be weaned more quickly.

Fearful/Anxious Behaviors

Fear and anxiety are both part of the typical human experience. Each is necessary to alert us to real dangers. However, for some, the severity of the fear response, and the inability to regulate emotions once aroused, can lead to an inordinate amount of time and energy spent upset or worrying. Formal treatment is suggested when fears and anxiety interfere with sleep, daily activities, social functioning, or academics. Initial treatment consists of behavioral approaches. Medication is used when behavioral approaches are not feasible or are unsuccessful. Studies of behavioral therapy for anxiety in children have shown it to be effective in greater that 50% of cases.[32]

Behavior therapy consists of gradual exposure to a fearful stimulus, usually using a stepwise approach from the least frightening element of a stimulus or situation to the entire stimulus or situation. Relaxation techniques are often taught to help regulate emotional arousal. A schedule for daily practice is set up, and rewards may be used for successfully tolerating the exposure.[33] For example, if a child is afraid of cats, the child might first be shown pictures of cats, followed by being in the room with a cat, then petting a cat someone else is holding, and eventually holding and petting the cat. At about 8 to 9 years of age, a therapist can begin to work directly with the child using cognitive behavioral approaches.

Medications for anxiety are considered when behavior therapy is not possible or effective. Studies of pharmacologic treatments for anxiety in children are limited, but selective serotonin reuptake inhibitors (SSRIs) are the most frequently used medications.[34] Very small doses of SSRIs can be effective, and the dose should be increased very slowly to minimize side effects. Children on SSRIs should be monitored for suicidal ideation, which may occur in a small percentage of children on these medications. Benzodiazepines may be prescribed for children with anxiety related to a medical procedure, but are not usually used on a regular basis because of their potential negative effects on cognition.

Repetitive Behaviors and Habits

Repetitive behaviors are part of normal child development. Rocking, banging, or digit sucking occur in most infants during the first year of life.[35] Usually these behaviors decrease in frequency during the toddler and preschool years, but occasionally they persist even into adulthood. Repetitive behaviors may help children modulate

their level of arousal. They can serve a self-calming function during anxiety-provoking situations and a self-stimulating function during periods of low arousal. Repetitive behaviors may be viewed as problematic when they cause tissue damage or subjective distress for the child. Distress for the parent alone would not usually be a reason to treat repetitive behaviors.

Repetitive behaviors represent a diverse group of behaviors that include rocking, digit-sucking, head-banging, nail-biting, hair-pulling (trichotillomania), tics, and Tourette disorder. Of these, thumb-sucking is the behavior most frequently addressed by pediatricians and is the behavior that will be focused on in this section. Thumb-sucking is usually harmless in infants and young children, but it can cause problems when it persists at high frequency after 4 to 6 years of age. Some of the most frequent sequelae involve dental problems, such as an anterior open bite, decreased alveolar bone growth, mucosal trauma, and even altered growth of the facial bones. Thumb- or digit-sucking is a common cause of paronychia in children, and it may be associated with an increased incidence of accidental ingestion. Rarely, deformities of the fingers and thumb may occur. Thumb-sucking is associated with stigma among both adults and peers. Peers view children who thumb-suck as less desirable playmates.[36]

Treatment of thumb-sucking should be considered in children older than 4 to 6 years. However, if the thumb-sucking is occurring infrequently (eg, only at night) or only as a temporary response to a stressor, treatment is not usually indicated. Treatment is indicated if thumb-sucking causes dental problems, digital malformations, or distress to the child. If thumb-sucking has resulted in significant negative reactions from the parents, a moratorium on parental comments on the thumb-sucking should precede any other treatment. This reduces tension between the parent and child and may decrease the sucking if it had been reinforced by parental attention. If other sources of stress or anxiety are thought to be related to the thumb-sucking, a plan to manage these stressors should be developed before the sucking is treated. Treatments are most effective when the child is a willing partner in the process.

Praise for not thumb-sucking in combination with use of a device to remind the child not to suck is the most common treatment. A variety of devices have been used from Band-Aids on the thumb to a thumb splint. Aversive taste treatments use a spicy hot commercially available substance applied to the thumb nail to remind children not to suck their thumb. Intraoral appliances inserted by dentists that block the child from thumb-sucking can be used. All of these treatments can be problematic if the child is not motivated to stop the thumb-sucking. The digital appliances can be removed by the child, and the intraoral devices can be bent or broken by children who want to continue to thumb-suck.

Conclusion

Primary pediatric health care professionals will encounter many parents who are concerned about their child's behavior. Often, an understanding of child physiologic and temperament factors contributing to the behavior, along with an assessment of the settings in which the behavior occurs, events preceding the behavior, and consequences of the behavior, will allow practitioners to provide advice to parents that will facilitate successful management of their child's behavior. Occasionally this assessment will reveal that the behaviors are severely disruptive, dangerous, pervasive, or occurring in the context of multiple psychosocial stressors. In these situations referral to a mental health professional should be considered.

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Chapter 5

Early Intervention

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One of the critical roles of primary pediatric health care professionals is to encourage and facilitate the optimal development of children. An active and engaged relationship between the medical home and local early intervention programs (EIPs) is essential. The American Academy of Pediatrics (AAP) Council on Children With Disabilities provides guidance for primary pediatric health care professionals through the policy statement, "The Role of the Medical Home in Family-Centered Early Intervention Services."[1] This policy statement reviews that various federal and state statutes mandate that community-based, coordinated, multidisciplinary, family-centered EIPs be established, and primary pediatric health care professionals, in close collaboration with families and the early intervention team, should assume a proactive role in ensuring that at-risk children receive appropriate clinical and developmental early intervention services.[1]

Many studies have documented improved outcomes for children who are engaged in early intervention; however, primary pediatric health care professionals sometimes delay referring eligible children and their families for early intervention services.[2–10] It is always uncomfortable to broach bad news or medical concerns with a family yet, in the case of developmental delays, EIPs are available in every community in the United States to provide an evaluation at no cost to the family and provide and coordinate appropriate services if eligible. Additionally, most research has shown that families are highly satisfied with their involvement in EIPs, which should boost primary care providers' confidence in making referrals for early intervention services.[4,5,8,11–16]

What Is Early Intervention?

Early intervention is a federally mandated program available in every community in the United States. It is authorized under the Individuals with Disabilities Education Act (IDEA). Its mission is to identify and coordinate special services for eligible infants and toddlers and their families. Under a series of federal laws beginning in 1975, both eligibility and definitions of special education services, including early

childhood special education/early intervention services, have been expanded over time (Box 5.1). Currently, early intervention services are mandated to meet the needs of children from birth to 3 years of age for any child who has delays in one or more of the following areas:

- Physical development
- Cognitive development
- Communication development
- Social or emotional development
- Adaptive development

The IDEA provides states and territories with specific requirements for providing early intervention services to infants and toddlers with special needs. In turn, each state and territory develops its own policies for carrying out the IDEA requirements. States must offer early intervention services to children with delayed development or those with an established diagnosis with a risk for disability. Services can be mandated for children who have a diagnosed condition that has a high probability of resulting in delayed development, such as Down syndrome (trisomy 21) or other chromosomal/genetic disorders, extreme prematurity or low birth weight, or structural brain abnormalities. The at-risk conditions and diagnoses are established by each state and can include family and/or maternal factors (eg, teenaged mother in

Box 5-1. A Legislative History of Early Intervention

1975 PL 94-142 Education for All Handicapped Children Act
Free and appropriate public education (FAPE) for children 5–18 years old. Services for children 3–5 years old was optional.

1986 PL 99-457 Education of the Handicapped Amendment of 1986
Development of programs for children birth to 3 years of age with disabilities or delayed development. FAPE included ages 3–5 years old by 1990–1991 school year. Required coordinated, multidisciplinary service for children and families.

1990 PL 101-476 Individuals with Disabilities Education Act (IDEA)
Part H—the Program for Infants and Toddlers with Disabilities. Mandated early identification and provision of services to infants and toddlers at risk of developmental delays. No cost comprehensive, multidisciplinary assessment by a trained team to assist in determining which services were needed. Care coordination is mandated. Subsequently becomes "Part C" of IDEA.

1997 PL 105-17 Amendment to IDEA
Encouraged tracking/monitoring of at-risk children to facilitate initiating services when/if indicated.

2004 PL 108-446 IDEA Improvement Act
Mandated referral of all children with substantiated cases of neglect or abuse, affected by substance abuse and/or exposed to family violence, as well as children who are homeless or in state custody. Expanded rights of families to choose to have their children at early intervention sites until eligible for kindergarten.

poverty). For children at risk for developmental delays, states are required to maintain a tracking system to identify any child who begins to manifest delays. Such longitudinal surveillance is also an important role of primary pediatric health care professionals and the medical home.[17]

The latest revision to the IDEA in 2004 went into effect in January 2006 (PL 108-446). The IDEA mandates the establishment of community-based, coordinated, multidisciplinary, family-centered programs that are accessible to children and families. The primary pediatric health care professional and the medical home should be an important component of this process, providing identification, referral, medical information, and prescription for appropriate therapies.[1,17,18]
By federal statute, available services must include[1]

- Early identification, screening, and assessment services
- Care coordination services
- Medical services (only for diagnostic or evaluation purposes)
- Family training, counseling, and home visits
- Special instruction (provided by early intervention specialists)
- Speech-language pathology
- Audiology services
- Occupational and physical therapy
- Psychological services
- Health services that are necessary to enable the infant or toddler to benefit from other early intervention services
- Social work services
- Vision services
- Assistive technology devices and services
- Transportation, interpretation services, and other related costs that are necessary to enable a child and family to receive the other services

The type and amount of services provided are determined through multidisciplinary evaluation and the development, with the parents, of an Individualized Family Service Plan (IFSP).

What Is an IFSP?

The intervention plan required for EIPs is an IFSP, which is analogous to the Individualized Education Program (IEP) for children older than 3 years with special education needs. As suggested by this name, the family is a critical member of the early intervention team. The type and extent of services required for each child and family are delineated in the IFSP through a formal multidisciplinary evaluation process. The IFSP is a legal contract under federal law. By statute, the family is an integral component of the IFSP process and should play a lead role in the development of the program through identification of concerns, family needs, and goals for the child.

This is accomplished in collaboration with an EIP service coordinator, who is assigned when the family initiates a request for services.[1,17,19]

Following an assessment, an IFSP is created to include the following:

- The child's present developmental attainments
- Family strengths
- How to enhance the development of the child
- Major outcomes expected and timelines to achieve goals
- Specific services the child and family will receive
- Projected dates for initiating services and their duration
- Name of the service coordinator responsible for coordinating and helping the family implement the plan
- Steps to help the child and family with the transition to school services

The primary pediatric health care professional serves to ensure that his or her patients have a medical home in which surveillance, screening, evaluation, and diagnosis of developmental disorders are made; referrals to early intervention services are provided; IFSPs are reviewed; and parents receive counseling, advice, and advocacy for continued services as needed.[1] The primary pediatric health care professional also needs to consider referrals for pediatric medical specialist evaluations (eg, genetics, developmental pediatrics, etc).

Benefits of Early Intervention

Early intervention has been shown to be quantitatively most successful, in terms of altering developmental trajectory, in children with mild versus severe delays, in children at environmental versus biological risk for developmental delay, and in children at risk for developmental disability versus in children with known disability.[19] Neurocognitive research has shown that there are optimal periods of brain development in which learning is most efficient.[1] Intervention that is well planned and provided as early as possible can improve the functional outcome and the quality of life of children at risk for developmental delays.[11–14] Intervention services provided during the preschool years have the potential to provide the greatest benefit to children and their families and provide an opportunity to minimize developmental and behavioral problems in children at risk and maximize potential in children with developmental disabilities.[11–14]

The early identification and referral of children with developmental delays, as well as those children at increased biological or environmental risk for developmental delays, is necessary in order to provide early intervention services. Primary pediatric health care professionals are in a unique position to identify children at risk because they have regular longitudinal contact and ongoing relationships with young children and their families. Through the use of evidence-based surveillance and screening,

primary pediatric health care professionals can best identify children who have developmental delays or are at risk for developmental delays in a timely fashion, so that community resources for intervention can be used.[18,20]

It is estimated that 18% of children have a developmental and/or behavioral disability[21,22]; however, only 30% of these are identified before school entrance.[23,24] Approximately 11% of school-aged children (6–17 years) receive special educational interventions through the school system for academic or behavioral concerns. However, only 5% of preschool-aged children (3–5 years) and 1.8% of infants and toddlers (birth–2 years) receive early intervention services.[4,16] The National Early Intervention Longitudinal Study[25] found that the age of first concern was often later for children with developmental delays when compared with children with diagnosed conditions, such as Down syndrome, or children at high risk, such as premature infants. However, it is important to note that only those children with more severe delays can be reliably identified at the earliest ages. Children with severe cerebral palsy or intellectual disability should be identified by 1 year of age, while those with higher prevalence but lower severity problems, such as learning disabilities or attention-deficit/hyperactivity disorder, who make up the majority of the 18% of children with developmental or behavioral disabilities, are not reliably identified until nearer school age. In an effort to improve the early identification of children with developmental and behavioral concerns, the AAP recommends that developmental surveillance be performed at every well-child visit, that formal standardized developmental screening tools be used at specified ages (9 months, 18 months, and 24 or 30 months), and that primary pediatric health care professionals attend to all developmental concerns raised by parents using developmental surveillance (see Figure 6.1 in Chapter 6).[18] This underscores the need for improved standardized developmental and behavioral screening tools with strong psychometric properties to best identify mild delays, which are extremely difficult to identify at young ages and which may often be overlooked even by the most experienced pediatric clinicians.

There have been several national long-term studies that have revealed the effectiveness of early intervention.[8,11–15,17,26,27] In terms of children at risk for developmental delay, it has been shown that children with low birth weight who receive comprehensive, multidisciplinary early intervention services score higher on cognitive assessments at 3 years of age when compared with children who receive health services alone.[27] In this same group at 8 years of age, school outcomes and family development were improved for those children and families who received intervention than for those who did not.[2] Early intervention services that are able to combine child-focused educational activities with strengthening of the parent-child relationship are most effective.[7] Families of children with disabilities face significant challenges, and family support is an essential component of early intervention for these families. Early intervention has been shown to improve parent-child interactions, help parents modify their behavior in response to their child's needs, provide

support for families, and help families learn strategies for effectively advocating for their children.[28]

The presence of psychosocial risk factors can place a child at an increased risk for developmental and behavioral disorders. The presence of 4 or more of the following psychosocial risk factors has been shown to negatively impact cognitive abilities[29,30]:

1. Less than a high school education in parent
2. Parental unemployment
3. Parental marital status (single)
4. Three or more children in the home
5. Parental mental health problems
6. Parental history of abuse as a child
7. Domestic violence
8. Frequency of household moves

These psychosocial risk factors can be objectively measured through the use of a screening measure, such as the Family Psychosocial Screener (FPS).[31] The FPS can be downloaded for free at www.brightfutures.org/mentalhealth/pdf/professionals/ped_intake_form.pdf. Mediation of psychosocial risk factors may involve parent training in how to promote child development, early prevention programs for children, and efforts to reduce family stressors, such as social work services for housing and food and income provisions for at-risk families. Children at high environmental risk for developmental and/or behavioral problems based on adverse psychosocial circumstances who receive early intervention have been shown to score higher on cognitive and achievement tests, complete more years of education, are more likely to attend a 4-year college, and are older when their first child is born.[8]

When to Refer to Early Intervention

There are clearly windows of opportunity when the human brain is more amenable to targeted intervention and, thus, the earlier intervention begins, the more likely it is to be effective in improving functional outcomes.[11–15] Thus it is critical for primary pediatric health care professionals and the medical home to be proactive in identifying children with delays, and those with possible delays, and facilitating referral for evaluation by local early intervention providers, a service that is free of charge to families.[17] For children with mild developmental delays or for those at risk for developmental delays, intervention at a younger age has been shown to result in improved long-term developmental trajectory.[8] For children with significant developmental disabilities, early intervention can have a positive impact on functional outcomes, as well as help with long-term family dynamics and acceptance, even if the interventions provided do not significantly alter the developmental trajectory.[18,26,32–34] When a developmental delay is even suspected, a referral for evaluation should be initiated by the primary pediatric health care professional, although the

determination of eligibility for services is the responsibility of the EIP. The AAP Developmental Surveillance and Screening Policy Implementation Project recently reported that a diverse sample of practices could successfully implement developmental screening as recommended by the AAP.[35] However, these practices reported referring only 61% of children who failed screens, and many practices struggled to track their referrals. Those that did track found that many families did not follow through with recommended referrals. Thus it is critical for primary care practices to take a more active role in the referral process, by either calling in or sending in the referral directly to the EIP, rather than just providing information about the local EIP for families to contact. When the family is directly referred by their primary pediatric health care professional, the early intervention evaluation team can contact the family directly and arrange for the evaluation to determine eligibility in the family's home or the child's child care setting.

Early intervention services are optimally provided in the "most natural environment," meaning either the child's home or child care program, obviating the need for the family to travel to center-based services. The in-home services of an early intervention specialist (an early childhood special education teacher or developmental therapist that works with the family on family-centered home-based activities) can be provided, along with direct speech and language, occupational, and physical therapies or services for the visually or hearing impaired, as indicated by the IFSP. Early intervention programs facilitate evaluations for transition to school-based services, once the child reaches 3 years of age, and assist with coordinating the IEP, if the child is eligible for continued early childhood special education services. If the family moves, the IFSP will transfer as written to any county in the United States, and the EIP service coordinator will coordinate transfer of services to the new site for the family.

Primary pediatric health care professionals are often called on to prescribe the frequency of specific habilitative therapies (speech, occupational, physical) that supplement the services provided by early childhood special educators.[36] Unfortunately, there is not a strong evidence base in the medical literature to guide health care professionals in this endeavor. While speech, occupational, and physical therapies clearly improve developmental trajectory when employed in rehabilitative settings (such as in children who need to relearn previously acquired skills following a head injury), evidence to support the ability of these therapeutic modalities to clearly alter developmental trajectories in habilitative settings is less clear, although many studies have focused on scores on standardized developmental measures rather than on functional outcomes.[37,38] The most important concept for therapeutic intervention is that therapeutic goals need to be worked on throughout daily activities across settings, rather than being limited to the short therapy session. In this model, it is the parent or primary caregiver who becomes the true therapist and who generalizes the specific therapy across settings. This model requires a buy-in from both parents and therapists. Simplistically, a child may not need a speech therapist to start to talk or

point; however, parents may require the services of the therapist to learn to engage their child in regular activities throughout the day and to establish a milieu to encourage the child's generalization of the specific goals of therapy. This model of parents as primary therapists is supported by results of the Early Intervention Collaborative Study, which showed that mother-child interaction was a key predictor of change in both child developmental outcomes and parent well-being beyond the type of disability experienced by the child.[3,39] In this study, family relations also predicted change in children's social skills.[39] Unfortunately, this consultative, family-centered model is not the historical approach to therapy; rather the direct service model—therapist with child—is the model most therapists are trained in and parents expect. Primary pediatric health care professionals have an educational role with the family in explaining the importance of generalizing therapeutic goals and activities across settings. The results of the Early Intervention Collaborative Study showed that, despite the great variability of child and family function and of the types and extent of services offered, most young children in EIPs improved in many domains of functioning.[1,39]

Conclusion

As soon as a primary pediatric health care professional identifies a child with developmental delays or behavioral concerns through the processes of developmental and behavioral surveillance and screening, that child should be immediately referred to their local federally mandated EIP, which is available in every community in the United States. Every EIP provides diagnostic developmental evaluations, free of charge to families, to confirm the concerns raised by surveillance and screening. The local EIP will then provide an IFSP of early intervention services to eligible children and their families primarily in the natural environment of the home or child care setting. Such services are most effective when provided at the earliest possible age. Early intervention services are quantitatively most effective for children at psychosocial risk for developmental delays. However, even in the setting of severe developmental disability caused by known biological risk, early intervention has been shown to foster a more comfortable and developmentally appropriate interaction between parents and their children with disabilities; to enhance the ability of parents to care for their children with disabilities within the home; to prevent secondary behavioral handicaps; to enhance long-term social, emotional, and adaptive functioning; and to improve long-term quality of life for both children with disabilities and their families.

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Chapter 6

Developmental and Behavioral Surveillance and Screening Within the Medical Home

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Developmental disabilities and behavioral disorders are among the most common health conditions present in children, with current prevalence estimates suggesting a frequency of approximately 12 or 13 affected per 100 children in the United States.[1,2] Intellectual disability, the most common of the severe developmental disorders, affects 1 in 83 children,[3] while autism spectrum disorders (ASDs) have been most recently identified in 1 in 110 children.[4] Cerebral palsy (CP), the most common severe motor disability, affects 1 in 278 children.[5] The more common and lower severity disorders, such as attention-deficit/hyperactivity disorder (ADHD)[6] or the speech and language disorders,[7] can each affect as many as 1 in 10 children. Given the frequency of these disorders and the benefits of early intervention, the first task for primary pediatric health care professionals in the management of affected children is early and appropriate identification of these disorders through developmental surveillance and screening. Given the chronicity of these disorders, primary pediatric health care professionals need to establish medical homes for affected children and youth from which care is initiated, coordinated, and monitored and with which families can form a reliable alliance for information, support, and advocacy from the time of diagnosis through the transition to adulthood.[8]

Developmental surveillance and screening are critical functions of a family-centered medical home. The child who fails developmental screening and who is identified with a developmental disorder (such as CP, intellectual disability, or autism) on developmental evaluation should be designated as a child with special health care needs within their medical home.[9–11] It is now clear that a child with special health care needs who receives care in a family-centered medical home can experience improvements in the receipt of evidence-based care; minimized delays in receipt of appropriate health care; and improved health status and family, social, and community functioning.[12]

Box 6.1. American Academy of Pediatrics Key Recommendations on Developmental Surveillance and Screening[14,40]

- Perform developmental surveillance on every child at every well-child visit.
- Perform developmental screening on every child using a standardized screening tool at 9, 18, 24, 30, and 48 months.
- Perform developmental screening using a standardized screening tool when a concern regarding development is expressed.
- Perform autism-specific screening on every child at 18 and 24 months.
- If screening results are concerning, refer to developmental and medical evaluations and early intervention services.
- Follow up on referrals made.
- Continually track the child's developmental status.

The surveillance and screening for developmental and behavioral disorders follows the same principles used in other health conditions managed within the medical home. Pediatric health surveillance occurs at every well-child visit through routine performance of the health history and physical examination by the physician or pediatric nurse practitioner. In contrast, screening involves the administration of a low-cost, brief, standardized laboratory test by health assistants at an age-determined visit, with interpretation of the results of screening and treatment initiation performed by the medical staff. In the example of a case of anemia, signs or symptoms such as tachycardia, pallor, or fatigue may be noted by surveillance at a routine visit. Screening for anemia, on the other hand, is performed routinely by laboratory testing at the newborn visit (sickle cell anemia), 12 months of age (iron deficiency, hereditary anemias), and adolescence (iron deficiency anemia in menstruating females).[13] When a concern for anemia is identified by surveillance or screening, the health care professional will pursue further evaluation. Such methods are similarly incorporated into developmental surveillance and screening, as recommended in the American Academy of Pediatrics (AAP) policy statement on these practices (Box 6.1).[14]

As currently defined in this statement, *developmental screening* is the administration of a brief standardized tool for the identification of children at risk of a developmental disorder. It is administered at specific ages, based on known patterns of development. In contrast, *developmental surveillance* is defined as a flexible, longitudinal, continuous, and cumulative process aimed at identifying children who may have developmental problems and is performed at every well-child visit. Five key components are grounded in the history and the observation of the child: (1) eliciting and attending to the parents' concerns about their child's development; (2) documenting and maintaining a developmental history; (3) making accurate observations of the child; (4) identifying risk and protective factors; and (5) maintaining an accurate record of documenting the process and findings. As in the example of anemia,

when a child is identified by screening or surveillance as high risk for a developmental disorder, the health care professional performs or refers the child for more detailed evaluation. The *developmental evaluation,* like the evaluation of the child with anemia, is aimed at identifying the specific developmental disorder or disorders affecting the child. When coupled with further medical evaluation of the child, both a specific developmental disorder and related medical etiology and/or associated medical conditions can be identified (see Chapter 7), and a program of treatment and care management, such as referral for early intervention services (see Chapter 5), can be initiated.

Developmental Surveillance

Pediatric practice dedicated to the identification of developmental problems has been rooted over past decades in the practice now defined as developmental surveillance. Traditionally, primary pediatric health care professionals have performed informal developmental monitoring through a review of developmental milestones with the parent at well-child preventive care visits. The health care professional has typically coupled this history with the child's medical history of known risk factors for developmental problems and observations of the child's skills and interactions at the visit. While often referred to in the past as screening, such practice now is referred to as developmental surveillance, in concert with other concepts of health surveillance. This practice, coined by Paul Dworkin in 1988, remains a mainstay in the early identification of children affected by developmental disorders.[15] As now defined in the AAP policy statement, it is distinct from developmental screening.[14] Developmental surveillance is performed at each preventive care visit. It is composed of several key historical and observational components: (1) inquiry about developmental concerns, (2) developmental history, (3) identification of historical risk and protective factors identification, and (4) observation of a child's development during the physical examination and visit (Box 6.2).

Box 6.2. Components of Developmental Surveillance

- History
  - Parental developmental concerns
  - Developmental history: milestone achievement, with identification of abnormal patterns
    - Delay
    - Dissociation
    - Deviancy or deviation
    - Regression
  - Medical and family historical risk factor identification
  - Protective factor identification
- Developmental observation
  - Gross and fine motor skills
  - Speech, language, and social engagement
  - Related neurologic function on physical examination

Medical History

The history obtained from the family is a powerful tool in the process of developmental surveillance. Parental concerns about their child's development can be a powerful predictor of developmental problems and are therefore a key area of inquiry during the preventive care visit.[16–18] The history obtained from the parent should also consider significant family biological and psychosocial risk factors, such as other family members with genetic, developmental, or behavioral disorders. The family history may reveal a pedigree of developmental disorders, like that seen in X-linked disorders, such as fragile X syndrome or Duchenne muscular dystrophy. Patterns of language disorders can be seen in families of children with ASDs. The milder learning disorders and ADHD are also known to have strong familial components, often noted by academic underachievement in family members. A family history of substance abuse is also of note, due to concerns of a child's prenatal exposure, parental underachievement, and social and environmental risks posed. Similarly, the child's medical history may contain known risk factors for increased risk for developmental disability. The child born preterm or with perinatal complications is at known risk for a wide range of developmental disabilities, from CP and sensory impairments to learning, attention, and intellectual disorders. A history of neurologic problems, such as seizures or traumatic brain injury, also place a child at higher risk for associated neurodevelopmental disorders. A child with multiple congenital anomalies may also have underlying neurodevelopmental disorders identifiable through surveillance. Abnormal growth patterns, including failure to thrive, overgrowth, macrocephaly, or microcephaly, also suggest higher risk. Finally, the history should include a thorough social history to identify psychosocial and socioeconomic factors within the family and community that may place a child at increased risk for developmental delay.

Developmental History

The developmental history is classically obtained through the tracking of a child's attainment of key developmental markers, commonly referred to as milestones, throughout childhood. The milestones are classified into 5 skill areas: gross motor, visual-motor problem-solving, speech and language, social, and adaptive skills. Milestone tracking and review may reveal known patterns of timing, order, or sequence seen in developmental disorders.[19,20] *Delay* is the most widely known pattern of atypical development. With developmental delay, the child acquires skills in the typical sequence but at a slower rate. It may occur within a single stream or across several developmental streams. *Dissociation* is noted when a child's development is delayed in one stream and not another. Such patterns can help distinguish developmental disabilities. For example, a child with a communication disorder would have delay in speaking skills, while acquiring walking at a normal age. *Deviancy* or *deviation* occurs when a child achieves milestones out of the usual sequence within a stream of development, such as when a child crawls before sitting, as seen in some types

of CP, or uses words before their meaning is understood, a pattern commonly seen in autism. Echolalia, or repeating of words or phrases, may be seen briefly in typical development. However, it represents deviant development when it occurs in place of interactive speech and language or when it occurs for a prolonged period. *Regression* is the least common but most concerning pattern. It typically presents with a child losing milestones, but can more subtly be recognized when a child stops acquiring new developmental skills or has a slowing in rate of developmental progress over time.

While obtaining a history of developmental milestone acquisition, primary pediatric health care professionals can calculate a child's rate of developmental progress with use of the developmental quotient (or DQ). The DQ is calculated by dividing the child's developmental or best milestone age (DA) by the child's chronologic age (CA) (DQ = DA/CA x 100). A DQ of 100 represents the mean or average rate of development, while a DQ below 70 is approximately 2 standard deviations below the mean and suggests a significant delay that requires further evaluation. For example, a child with a motor DQ of 70 or less may have CP or other motor disability. A DQ below 70 for language milestones strongly implies a communication or intellectual disorder.

Developmental Observation

Observation during the course of the preventive care visit may also demonstrate the child as having an abnormal pattern of development. Problems in movement or posture may be seen while the child sits with the parent or when placed on the examination table. Associated neurologic abnormalities, such as increased or decreased muscle tone, should be considered when this is observed. Difficulties with interpersonal engagement or eye contact with the parent may also be noted during the course of the visit. The young child is often reticent during the visit, limiting one's ability to observe a child's speech and language skills. However, a speaking child's pattern of communication may provide insight into the child's development in this area.

Implementation of each of these components of developmental surveillance at each preventive care visit within the medical home can identify the child with developmental concerns and a possible developmental disorder. When the primary pediatric health care professional identifies a concern, these findings should be documented in the paper or electronic medical record for ongoing tracking of these issues. In addition, formal developmental screening or referral for developmental evaluation should be completed to verify these concerns. Such screening or evaluation may need to be completed at a separate visit based on time demands at the preventive care visit. When a typical pattern of development and no concerns are identified during surveillance, the primary pediatric health care professional can make recommendations for specific developmental stimulation activities for the child and parent. Whether the child is noted to have concerns and risks or has typical development,

child-parent activities should be promoted based on simple age-specific developmental goals. Working in family-professional partnerships within the medical home, developmental care plans can be developed to encourage optimal developmental stimulation as a component of family-centered care. The parent of the sitting child can be encouraged to provide opportunities for standing-based play. The parents of the child with limited word use or recognition can be instructed in the benefits of reading-based picture vocabulary development. The timing of subsequent visits with ongoing surveillance should be determined at the conclusion of the preventive care visit. When a child has a typical pattern of development, the usual periodicity schedule can be followed. If the child has concerns, subsequent visits for surveillance or screening should be planned earlier. At all of these follow-up visits, the child's attainment of the promoted skills should be reassessed.

Behavioral Surveillance

As in the AAP statement on developmental surveillance and screening, the universal health care concepts of surveillance and screening that are critical components of the medical home can also be incorporated into the early identification of behavior disorders.

Medical History

Parental concern about their child's behavior and social skills has been shown equally powerful to concern about development for the identification of significant problems in the child, particularly in children older than 4 years.[21] The family and social histories may also reveal areas of concern. A child's behavior issues may be tied to known familial mental health problems, such as anxiety disorders, mood disorders, or ADHD. Family psychosocial risk factors, such as substance abuse or family disorganization, should also be identified.

Behavior History

The behavior history obtained during surveillance is more typically symptom based, rather than using the developmental milestone model. The primary pediatric health care professional should inquire about the child's relationships and engagement with parents, siblings and other familiar persons, other children (particularly age peers), and unfamiliar children and adults. Inquiry should be made about the child's behavior during daily living activities, including eating, sleeping, and playing. Concern about problems with compliance, tantrums, attention, activity level, impulsivity, and aggression should be elicited. A history of unusual patterns of behavior may also be reported. Examples include repetitive speech or play, excessive preoccupation with objects or specific ideas, hand flapping, unusual visual gaze, or potentially self-injurious hand biting or face slapping.

Behavior Observation

During the preventive care visit, the health care professional should observe the child's engagement and communication with the parent in the office. Impulsivity, decreased attention span, or increased activity level suggest attention problems. Observed tantrums and oppositionality should be considered in the context of the child's age and the history provided by the family. The unusual behavior problems described above should elicit further evaluation when directly observed in the medical office setting. As with developmental surveillance, the primary pediatric health care professional should pursue screening or further evaluation when concerns about behavior are identified through behavioral surveillance. In addition, counseling around behavior management and discipline is also a critical component of a family-centered care plan derived from the family professional partnership contained within the medical home (see Chapter 4). If the patterns of behavior are age typical, counseling on typical behavior patterns and their management should be offered. The parents of a child with stranger or separation anxiety can learn techniques of anticipation and consolation. Discipline methods for management of mild tantrums or oppositionality, such as time-out techniques, can also be included in a behavioral care plan. Promotion of child-parent activities and child play should be promoted.[22,23] As with developmental surveillance, subsequent visits should be arranged based on behavior issues noted. Early follow-up should be arranged when there are specific concerns identified.

Developmental Screening

In the pursuit of the early identification of developmental disorders, the primary pediatric health care professional is charged with developmental screening periodically during early childhood. Such screening involves the administration of a brief and standardized test in the medical home and is similar to screening of other health conditions. It differs from developmental surveillance in several key features. First, developmental screening involves use of a formal, standardized test with known reliability, validity, sensitivity, and specificity (Boxes 6.3 and 6.4). Second, given its implicit time demand and cost, screening is not employed at every preventive care visit, as is surveillance. Instead, it is administered at ages based on key times for identification of developmental disorders. Third, as in other health care screening, the developmental screening test is typically ordered and interpreted by the clinician, but may be administered by associated health care staff, while surveillance is usually performed directly by the primary pediatric health care professional in the course of the preventive care visit. Like surveillance, screening identifies children at high risk of a developmental disorder. Scoring of screening tests typically is categorical, with assignment of a child into a risk category, such as no risk, suspect, or high risk, rather than a numeric score. Those with scores in the suspect category are typically followed by additional surveillance or screening, while those at highest risk are in need of more detailed developmental evaluation and medical testing for the determination of a diagnosis and treatment needs.

Box 6.3. Characteristics of Developmental Screening Tests Used in Test Selection

Test Properties

Reliability—the ability of a test to produce consistent results

Predictive validity—the accuracy of a test to predict later test performance or development

Sensitivity—test accuracy in the identification of delayed development or disability

Specificity—test accuracy in the identification of children who are not delayed

Standardization sample—the group of children whose test performance comprises the test norms; used for comparison to later individual child performance on the same test

General screening test—test that evaluates multiple areas of development

Domain-specific screening test—test that evaluates one area or domain of development (eg, motor, language)

Disorder-specific screening test—test aimed at identifying a specific developmental disorder (eg, autism)

Implementation Properties

Parent-completed or clinician-administered

Age range

Administration time

Languages available

Test cost

Box 6.4. Common Current Developmental Screening Tests[14]

- General (Broad-Band) Screening
  - Parent Report
    - Ages and Stages Questionnaires (ASQ)
    - Child Development Review-Parent Questionnaire (CDR-PQ)
    - Parents Evaluation of Developmental Status (PEDS)
  - Clinician-Administered
    - Battelle Developmental Inventory Screening Tool (BDI-ST)
    - Bayley Infant Neurodevelopmental Screen (BINS)
    - Brigance Screens-II
- Domain-Specific Screening
  - Language
    - Capute Scales (also includes problem-solving, cognition)
    - Communication and Symbolic Behavior Scales-Developmental Profile (CSBS-DP)
  - Motor
    - Early Motor Pattern Profile (EMPP)
    - Motor Quotient (MQ)
- Disorder-Specific Screening
  - Autism spectrum disorders
    - Modified Checklist for Autism in Toddlers (M-CHAT)
    - Pervasive Developmental Disorders Screening Test-II, Stage 1 Primary Care Screener (PDDST-II)
    - Screening Tool for Autism in Two-Year-Olds (STAT)
    - Social Communication Questionnaire (SCQ)

The AAP currently recommends formal developmental screening at 9 months, 18 months, and 30 months of age during the first 3 years of life, as well as at any time that the parent or primary pediatric health care professional has concerns about appropriate development[14] (Table 6.1). Of note, when a 30-month visit is unavailable or it is anticipated that a child will not be seen at that age, screening should be performed at 24 months. In addition, screening for school readiness is useful at the 4-year preventive care visit. Screening specifically for ASDs is also recommended at the 18-month visit and again at the 24-month visit.[24,25]

The ages for screening were selected based on times when key developmental problems can be identified and time availability at each preventive care visit. Screening at the 9-month visit is aimed at identification of hearing impairment not identified in newborn screening, visual disorders, and delays in motor development resulting from neuromotor disorders such as CP. At the 18-month visit, the clinician can identify milder motor problems, such as abnormalities of gait or coordination seen in mild CP. In addition, this visit is critical for the early identification of communication or intellectual disabilities, including ASDs. The next screening visit is recommended at 30 months of age and is now included in the AAP periodicity schedule and the Bright Futures guidelines for health supervision.[13,26] However, when such a visit is not available, screening should be performed at 24 months. This visit is also centered on the development of communication or intellectual skills and may also identify ASDs, intellectual disorders, or milder speech-language disorders. Performance of screening at these early ages enables referral of those children who fail screening to local early intervention agencies for further evaluation and initiation of early intervention services[24] (see Chapter 5). Such early screening also allows early identification of related medical conditions, with associated early initiation of related medical treatments. Screening at ages 4 to 5 years may also be useful for

Table 6.1. Developmental Screening Visits in the AAP Periodicity Schedule[13,14,26]

Age of Visit, months	Critical Developmental Streams Screened	Common Developmental Disorders Identified
9	Vision, hearing, gross motor, fine motor, receptive language	Visual impairment, hearing impairment, cerebral palsy, and other neuromotor disorders
18	Gross motor, fine motor, language (expressive and receptive), social	Cerebral palsy (mild to moderate), mild neuromotor disorders, autism spectrum disorders, specific language impairments, intellectual disability
30[a]	Language (expressive and receptive), social	Specific language impairments, intellectual and other learning disabilities, attention disorders

[a] When a 30-month visit is not available or there is concern about a child returning for this visit, screening should be performed at 24 months.

school readiness determination in order to identify children at increased risk for academic problems.

Screening Tests

In performing developmental screening at these key visits, the primary pediatric health care professional must choose a screening test based on multiple factors (Box 6.3).

First, a screening test should be normed and standardized on a large sample of children and should meet appropriate standards in reliability, validity, sensitivity, and specificity. These test standards ensure that the test can accurately identify those children about whom there is a developmental concern as well as distinguish them from those without. A good screening test should provide consistent results if administered by multiple testers (reliability). It should strike an appropriate balance in identification, minimizing both overidentification and referral of those without developmental problems and under-identification and referral of those with a true developmental disorder (predictive validity, sensitivity, specificity).[27,28] The predictive validity of a test is generally most useful for the clinician. If the test result is positive for concern, positive predictive validity provides insight into what percentage of children with such a result are truly affected by a developmental disorder. Sensitivity measures the percentage of affected children identifiable with a test, while specificity reports the percentage of unaffected children testing negative. As such, these measures are most useful in public health and test development, frequently reported in the test manuals and in studies examining their validity. Sensitivity and specificity rates of 70% to 80% or a sum of both greater than 150 are commonly recommended for developmental screening tests.[28,29] Caution must be exercised, however, in interpreting these rates in developmental screening. While some measures are validated in the general pediatric setting, others have been performed on a known high-risk group, such as premature infants, or outside of a health care environment, such as in child care or school. The measured outcome typically is another general broad test of development, rather than presence of a specific developmental disorder. The clinical utility of these measures has also been questioned due to their instability, outcome verification bias, small sample sizes, test score bias with young children, and errors in selection and use of diagnostic tests.[30]

Other characteristics of the available screening tests should be considered by the clinician when selecting one for use in practice. The most common format for developmental screening tests is *general,* also referred to as broad-band, where multiple developmental domains are screened at each test administration, modeled after the first widely used screening test, the Denver Developmental Screening Test (DDST).[31,32] These tests also typically cover a broad range of ages, typically infancy through school age. This format offers the clinician economy of time and cost in the use of a single test for all children. Such screening tests are most often validated

against other developmental tests. Commonly used tests at the present time include the Parents Evaluation of Developmental Status (PEDS)[33] and the Ages and Stages Questionnaires (ASQ).[34] Alternatively, screening tests can be *domain-specific,* where the test screens a specific area of development, such as motor or language, for specific disorders, such as CP (motor)[35,36] or communication disorders (language).[37,38] While less widely used and validated, such screening tests may be useful for the identification of specific developmental problems or disorders. Use of such tests requires that the clinician select a test appropriate to the specific age of screening. For example, at the 18-month visit, a test for motor and another for language should be chosen. Finally, a screening test can be *disorder-specific,* screening for a known developmental disorder. This model is most similar to screening for other medical conditions, with the positive result measured against a defined developmental disorder. Currently, screening for ASDs follows this model, with screening test results measured against criteria for autism or other pervasive developmental disorders.[39] Diagnosis-specific screening also requires appropriate timing for the screening test administration. In the case of autism screening, the AAP policy statement initially recommended such screening at 18 months.[14] However, soon after, based on new research, this recommendation was expanded to also include 24 months of age, illustrating the challenge of such methodology, requiring ongoing reconsideration of the screening recommendation.[40] The Modified Checklist for Autism in Toddlers (M-CHAT) is widely used currently for this purpose and is available in the AAP autism toolkit.[41–43] At present, it is unclear which of these methodologies have the best potential for early identification of the developmental disorders, particularly given the wide array of screening tests available. However, current efforts in both general screening and autism screening may provide better knowledge of the best pathway to be taken in future screening.

Practical aspects of implementation may also guide the clinician in choice of instrument. The time required for test administration is important, as the primary pediatric health care professional fits the test into the preventive care visit. The AAP recommendations have specifically taken this concern into account, considering time availability at each preventive care visit, typically recommending tests taking less than 15 minutes to administer. Screening tests may be parent-completed or directly administered by the primary pediatric health care professional. Parent-completed developmental questionnaires allow screening to be performed in part outside of the clinic examination room, in the home, or in the medical office waiting area. They can then be scored by office staff or by the clinician, who can interpret the results, review them with the family, and make appropriate recommendations. Tests directly administered by the primary pediatric health care professional are typically incorporated into the preventive care visit and offer the clinician a direct view of the child's achievement, opening up a conversation with the parent about the child's development, with related recommendations. Such methodology may require more clinician time than a parent-report measure. Finally, in choosing a

test, consideration must also be given to languages available if a practice serves multilingual populations.

Many screening tests are now available for use in the pediatric health care setting, making the selection challenging for the clinician. When choosing, the clinician should consider the test's style and properties, as well as the population being screened and the office characteristics. The AAP has provided a table of available and acceptable screening tests in the policy statement on developmental screening.[14] Other practical guides are available on the Internet[44-46] and in the literature[47,48] for selecting developmental screening tests.

Implementation of Developmental Surveillance and Screening

In the AAP Periodic Survey of 2002, pediatricians reported high rates of developmental surveillance (up to 71%) but low rates of formal developmental screening using a standardized instrument (23%), voicing concerns regarding time limitations, lack of staff, and inadequate reimbursement for performance of screening.[49] In response, the 2006 AAP recommendations on developmental surveillance and screening aimed to create a practical guideline for implementation. A clinical algorithm was created with the intent of clarifying the recommendations with a visual practice tool with electronic health record compatibility. As illustrated in Figure 6.1, children identified with a developmental concern through developmental surveillance at any pediatric visit should undergo formal developmental screening for confirmation of the concern. When developmental screening is performed, either as part of preventive health care or in response to concerns identified through surveillance, a child will be found to either have a normal pattern of development or a pattern of concern for a developmental disorder. When typical development is seen, the child should be followed through the routine schedule of developmental surveillance and screening or through more frequent surveillance if concerns remain. When a concern is identified, further management strategies should be initiated by the primary pediatric health care professional (see Management of the Child With Developmental Concerns on page 83).

Insights gained from the experiences of surveillance and screening implementation projects provide guidance for its practical implementation.[44,50,51] Successful implementation of developmental surveillance first requires that the medical home identify a clinician practice champion, who can lead the office effort for creation of a system for surveillance and screening. The first task for the clinical staff will be selecting an appropriate screening test for use at the 9-, 18-, 30-, and 48-month preventive care visits. If current office practice does not allow the 30-month visit, planning for screening children at 24 months must be put in place. In addition, for specific screening for ASDs, an autism-specific screening test must be chosen for implementation at both the 18- and 24-month visits. Implementation projects have shown that developing an office system approach is critical, with medical office staff

playing key roles in scheduling, test distribution and scoring, and referral and tracking of children identified in need of further evaluation and treatment. Refer to Box 6.5 for additional details on key factors for successful implementation of developmental surveillance and screening. The AAP Developmental Surveillance and Screening Policy Implementation Project recently reported that a diverse sample of practices could successfully implement developmental screening as recommended by the AAP.[52]

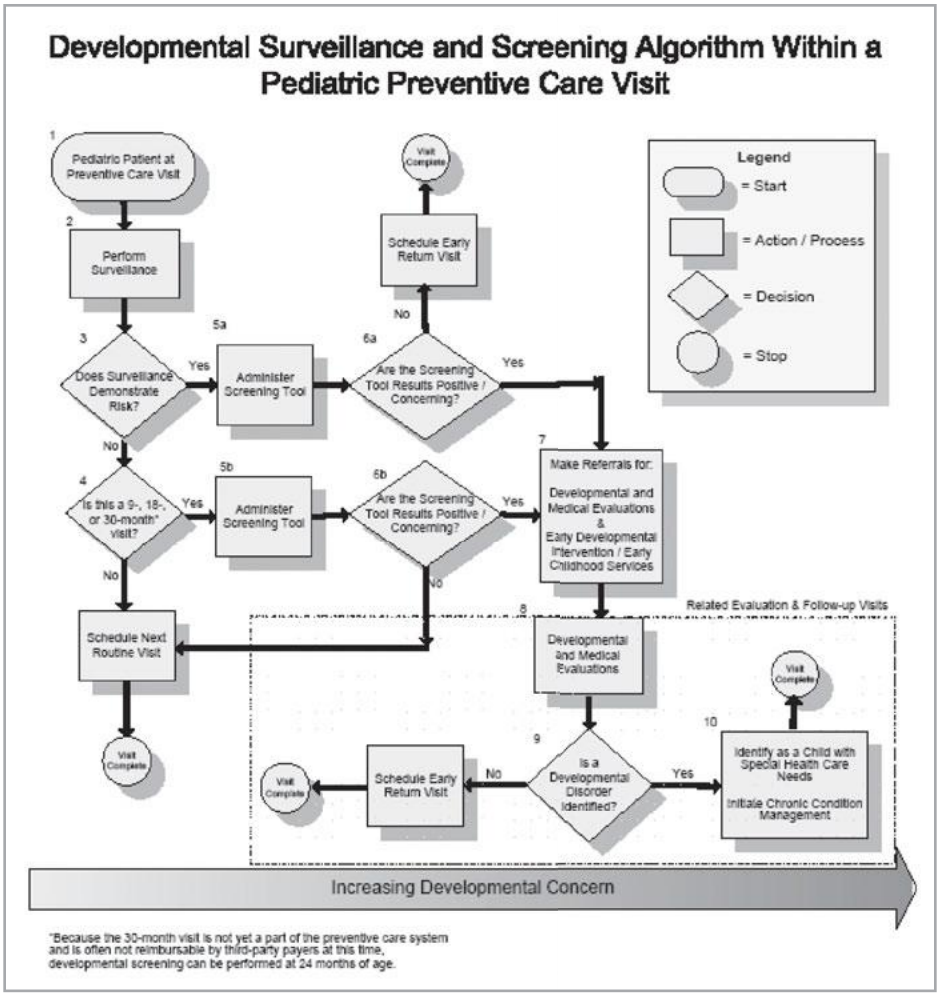

Figure 6.1. American Academy of Pediatrics Algorithm for Implementation of Developmental Surveillance and Screening.[14]

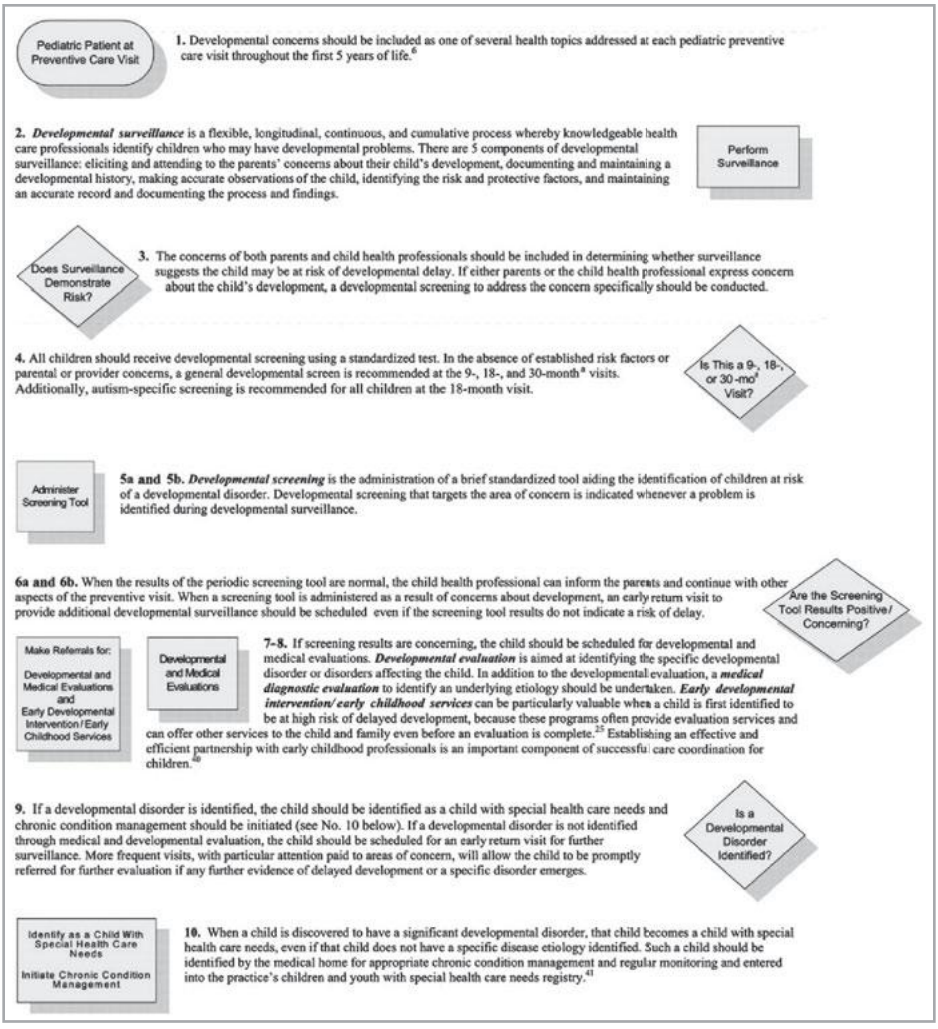

Figure 6.1 continued. American Academy of Pediatrics Algorithm for Implementation of Developmental Surveillance and Screening.[14]

Box 6.5. Key Strategies in Implementation of Developmental Surveillance and Screening[14,24]

Screening Procedures

- Choose a screening tool.
  - Population served (parental education level, languages spoken)
  - Office staff skill level
  - Medical staff skill level
  - Test time and cost
- Train staff in patient visit flow changes and in screening tests used.
- Plan test distribution in advance.
- Consider need for readers and translators.
- Develop office and medical staff reminder systems for screening at targeted ages (9, 18, 24 and/or 30, and 48 months).
- Train staff and use coding and billing practices appropriate to screening.

Referral Procedures

- Develop office system for referral of children to early intervention and community therapy programs.
  - AAP referral form available[19]
- Identify network of medical consultants for medical evaluations.
- Identify resources and referral systems for further developmental evaluation.

Medical Home Strategies

- Create a practice registry and office system for chronic condition management and planning of children with developmental disorders and special health care needs.
- Ensure ongoing close developmental surveillance.
- Create tracking system for referrals.
- Develop a chronic condition management/care plan.
- Deliver family-centered care, develop partnerships with families, and get regular feedback.
- Identify community resources for support services.
  - Parent-to-parent organizations
  - Government agencies
  - Advocacy groups
  - Respite systems

Management of the Child With Developmental Concerns

When screening confirms a developmental concern in a child, the primary pediatric health care professional should initiate a series of actions aimed at the identification and treatment of the child with a developmental disorder: (1) full developmental evaluation for diagnosis of a specific developmental disorder; (2) medical evaluation for determination of etiology of the developmental concerns; (3) referral for therapeutic intervention to the local early intervention program (EIP), community therapy, or early childhood education providers; and (4) initiation of a medical home program and chronic condition management for the child with special health care needs.

The developmental evaluation of a child who has been identified through screening is aimed at identifying a specific developmental disorder and can provide prognostic

information and specific direction for therapeutic interventions. Pediatric subspecialists, such as neurodevelopmental pediatricians, developmental-behavioral pediatricians, or child neurologists, can perform the developmental diagnostic evaluation, as can other early childhood professionals (early childhood educators, child psychologists, speech-language pathologists, audiologists, physical therapists, occupational therapists) in conjunction with the child's primary pediatric health care professional.

Given the shortage of subspecialists and the long waiting times at tertiary care developmental evaluation centers, a child's medical evaluation may be performed by the primary pediatric health care professional within the medical home. This medical evaluation is intended to determine underlying etiology and initiate related medical treatments. It should be a comprehensive medical evaluation, including review of the developmental and behavioral histories; review of newborn metabolic screening and growth charts; risk identification from the environmental, medical, family, and social histories; and a comprehensive physical examination. Vision screening and audiologic evaluation should be performed when indicated. Laboratory investigation is also recommended for identification of specific developmental etiologies. Guidelines for such investigations are available from the AAP, American Academy of Neurology, Child Neurology Society, and the American College of Medical Genetics.[25,53–58] Typical laboratory tests considered include neuroimaging, DNA analysis for fragile X syndrome, and chromosomal microarray (see Chapter 7).

The child whose developmental screening is of concern should also be referred for early intervention programming, which may include early childhood special education, physical therapy, speech-language therapy, occupational therapy, and/or behavioral therapy services, as appropriate. Such a referral can be made contemporaneously with initiation of the medical evaluation, as these services are not predicated on a diagnosis but instead are offered typically based on presence of a significant developmental delay, deviancy, or regression. Services delivered through local EIPs may also include service coordination, social work services, assistance with transportation and related costs, family training, counseling, and home visits.[24,59] As reported previously, the AAP Developmental Surveillance and Screening Policy Implementation Project recently reported that a diverse sample of practices could successfully implement developmental screening as recommended by the AAP. Unfortunately, however, these practices reported referring only 61% of children who failed screens, many practices struggled to track their referrals, and those that did track found that many families did not follow through with recommended referrals. Thus this study indicates that primary care practices are able to initiate developmental care through screening as recommended by the AAP, but barriers remain in terms of care coordination and monitoring for children who fail developmental screening.[52]

As briefly reviewed early in this chapter, the child identified with a developmental disorder, such as CP, intellectual disability, or autism, should be designated as a child with special health care needs within a family-centered medical home.[9–11] Children with special health care needs who receive care in a medical home receive improved quality of care, including improvements in the receipt of evidence-based care, minimized delays in receipt of appropriate health care, and improved health status and family functioning.[12] Providing a program of chronic condition management allows for regular health and developmental monitoring and proactive care. Such a program typically includes specialized condition-related office visits, written care plans (developed with families and that recognize and address specific family needs), explicit comanagement with medical specialists, appropriate patient education, and an effective system for monitoring and tracking. Community-based support services, such as respite care and advocacy organizations, may be helpful to the child and family, and parent-to-parent organizations (eg, Family Voices) and condition-specific organizations (eg, Autism Speaks, Autism Society of America, The Arc) offer further support, assistance, and information. It is useful and advantageous for offices to have lists of these resources available to distribute to families readily available in practice. Financial benefits are available for eligible children through supplemental social security income, public insurance, waiver programs, and state programs for children and youth with special health care needs[60](see Chapter 23).

Management of the Child With Behavioral Concerns

A comprehensive system and algorithm for screening for behavior disorders similar to that of developmental screening has not yet been developed, but the developmental screening system generalizes well for the screening and identification of behavior concerns and related disorders. While the screening of development is focused most on early identification during the infancy and preschool years, a strategy for identification of school-age developmental disorders and behavior disorders requires extension into later childhood. Key ages for behavioral screening have not been identified, but should be considered at the 30-month preventive care visit, the 4- or 5-year visit (prior to school entry), the 8-year visit (early elementary age), and other key ages during preadolescence and adolescence.

Several screening tests have been developed for behavioral screening. The Pediatric Symptom Checklist (PSC) is a 35-question checklist for completion by the parent for children ages 4 to 16, or by an adolescent in its Youth Report form.[61] It takes 5 to 10 minutes to complete and is available in multiple languages. A summary score is derived. If it is above a defined score, further behavioral evaluation by a medical or mental health professional is recommended. While not diagnostic, the PSC has been identified in its screening role as useful for the identification of moderate to serious impairments in children's psychosocial functioning.

The Ages and Stages Questionnaires-Social Emotional (ASQ-SE) are a parent-completed system for social-emotional screening of children from ages 3 to 60 months.[62] The ASQ-SE specifically targets personal-social skills in self-regulation, compliance, communication, adaptive functioning, autonomy, affect, and interaction with people. It takes 10 to 15 minutes to complete and score and is available in English and Spanish. Children scoring above predetermined cutoff scores are recommended for further behavioral evaluation.

The Brief Infant-Toddler Social and Emotional Assessment (BITSEA) is a 42-item parent-completed screening test for children between 12 and 36 months of age.[63,64] It has a 31-item problem scale, assessing problems such as aggression, defiance, overactivity, negative emotionality, anxiety, and withdrawal. Its 11-item competence scale measures empathy, prosocial behavior, and compliance. Cutoff points based on scores below the 15th percentile are used to indicate higher risk for social or emotional problems.

The Strengths and Difficulties Questionnaire is a 25-item parent-report form assessing emotional problems, conduct, hyperactivity and inattention, peer relationships, and prosocial behaviors, covering children ages 3 through 17 years.[65] It is available in more than 50 languages. Parent, teacher, and adolescent self-report forms are available. Specificity of 95% and sensitivity of 63% for psychiatric diagnoses have been reported, with particular strength around identification of conduct, hyperactivity, depressive, and anxiety disorders.

The National Initiative for Children's Healthcare Quality (NICHQ) Vanderbilt Assessment Scale, a component of the ADHD toolkit developed by the AAP and NICHQ, is a set of parent and teacher report scales aimed at identifying psychiatric disorders in school-aged children ages 6 and older, including ADHD, oppositional defiant disorder, conduct disorder, and anxiety and depression symptoms.[66,67] Associated school academic and behavioral performance are also screened. The parent scale consists of 55 questions and the teacher scale 43 questions. They are available in English, French, and German. The scales require 10 to 15 minutes to complete and score.

As recommended with these screening tests, and in accordance with the algorithm for developmental screening, children showing behavioral concerns on screening should have further evaluation by a medical or mental health professional. Behavioral evaluation may be completed by primary pediatric health care professionals within the medical home (see Chapters 4, 12, 16–19) or through referral to specialty physicians trained in such evaluation, including pediatric specialists in developmental and behavioral pediatrics and neurodevelopmental disabilities, as well as child and adolescent psychiatrists. Other mental health professionals who may be available in the community include psychologists, psychiatric nurse practitioners, and social workers. Behavioral evaluations are aimed at determining a diagnosis and

developing an effective treatment program of psychotherapeutic and/or psychopharmacologic management. Comorbid medical or developmental disorders may be identified and may require additional medical or developmental evaluation. As with children with developmental disorders, medical home strategies directed at chronic condition management should be implemented in the primary care medical home for children with behavioral disorders.

Conclusion

With the high frequency of developmental and behavioral disorders in children, the primary pediatric health care professional is challenged to identify these problems in the course of providing health care to children in the context of a medical home. Through creation of a systematic and algorithmic approach based on the universal health care concepts of surveillance, screening, and evaluation, early identification is feasible during the course of preventive health care. Routine developmental and behavioral surveillance allows for ongoing monitoring of a child's development, whether it is typical or there are problems present. Use of standardized objective screening tests at discrete, defined ages provides further support toward this process, ensuring acceptable levels of sensitivity and specificity for distinguishing the child with developmental or behavioral disorders from those without. The child identified with a developmental or behavioral concern through surveillance and screening can begin the process of diagnostic evaluation, both developmental and medical, with initiation of developmental, behavioral, and medical treatments targeting the identified conditions. Successful implementation of surveillance and screening is now achievable in the primary care medical home through use of a clinician-guided, office system–based approach supportive of the child and the family. Continued advances in the practice of surveillance, screening test development, developmental and medical evaluation procedures, and developmental and medical treatments will further assist in the early identification and treatment of children with developmental or behavioral disorders.

Related American Academy of Pediatrics Policy Statements and Resources

American Academy of Pediatrics. Caring for Children With Autism Spectrum Disorders: A Resource Toolkit for Clinicians. Elk Grove Village, IL: American Academy of Pediatrics; 2008

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Chapter 7

Neurodevelopmental Assessment and Medical Evaluation

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The American Academy of Pediatrics (AAP) has established a policy for developmental surveillance and screening in the primary care setting that, when applied consistently, will identify children who are not meeting expected milestones[1] (see Chapter 6). This policy statement recommends that children who are identified by screening as being at risk receive a developmental evaluation to confirm a developmental diagnosis, a medical evaluation to investigate the etiology of the developmental condition, and a referral to early intervention services. The AAP policy statement states that pediatric subspecialists, such as neurodevelopmental pediatricians, developmental and behavioral pediatricians, child neurologists, pediatric physiatrists, or child psychiatrists, can perform the diagnostic developmental evaluation for those children who fail screening. However, given the lack of access to such pediatric subspecialists and the long waiting lists at tertiary care child development centers, it is important to note that the AAP policy statement also states that the diagnostic developmental evaluation can be performed by early childhood professionals in conjunction with the child's primary pediatric health care professional. Thus this chapter will describe a neurodevelopmental assessment and medical evaluation process that primary pediatric health care professionals may consider adopting to follow a failed developmental screen. Through adoption of this process, and in conjunction with standardized developmental evaluations performed by early childhood professionals, primary pediatric health care professionals can contribute both to making a developmental diagnosis and attempting to establish a cause of the disorder.

Principles of Neurodevelopmental Assessment

Neurodevelopment is a dynamic and complex process that begins before birth and typically proceeds in a predictable sequence and at a predictable rate. The study of development has concentrated on the first years of life, as the observation of developmental markers in infants, toddlers, and preschoolers are rich in number and show little variability. Five areas or "streams" of development (developmental "stream"

is a term coined by Dr Arnold Capute as a moniker that reflects the fluid or dynamic process of development) that have been consistently studied, and are evaluated during neurodevelopmental assessments, include gross motor, visual-motor problem-solving, speech and language, social, and adaptive skills.[2] Each of the streams of development has milestones ascribed to specific ages of expected achievement. Milestones do not represent the process of development, but reflect the product of a developmental process. Precisely defined milestones are the measure by which normal or abnormal development is assessed.

Modern developmental assessments are based largely in the pioneering work of Dr Arnold Gesell, who established the first norms of milestones in the 5 streams of development.[3] Gesell assiduously recorded the timing and sequence of normal development of infants and children and observed that in typically developing children, development is an orderly, timed, and sequential process that occurs with such regularity that it is predictable.[3] The miracle of development, however, is not so much in the regularity of the acquisition of milestones within any given stream, but the awesome synchrony of development across streams.

In atypical or abnormal development, the timing, order, or sequence of the acquisition of milestones within a given stream or across several streams is disturbed. The assessment of abnormal development employs the principles of delay, deviation (or deviance), and dissociation. Developmental delay describes a phenomenon in which milestones within a given stream are attained in the typical sequence, but at a delayed rate. Delay can occur within a single stream or across several developmental domains. Deviation or deviance represents an uncustomary sequence of milestone attainment within a single stream of development. A child who is able to crawl before he or she is able to sit demonstrates developmentally deviant gross motor skills. Dissociation is a descriptive term that indicates differing rates of development among developmental streams.

The developmental quotient (DQ) is a measure of the rate of development within a stream, and it is the tool by which delay is determined. Using the age at which specific milestones are generally present, we can ascribe a functional developmental age equivalent to a child's development. For example, a child whose best gross motor function is sitting unsupported has a gross motor developmental age equivalent of 6 months (Table 7.1). The DQ is calculated by dividing the child's developmental age (DA) within a given stream by his/her chronological age (CA). The DQ then represents the percentage of normal development present at the time of testing, and is arithmetically represented by DQ = DA/CA X 100.

The application of the DQ to determine delay within a single stream, or dissociation between streams, can imply diagnoses that should be supported by historical and examination findings. A child with a gross motor DQ of 50, a history of delay in motor milestone acquisition, and abnormal findings on physical and neurologic

Table 7.1. Gross Motor Milestones[a]

Average Age, months	Motor Milestone
1	Head off table in prone
2	Chest off table in prone
3	Up to elbows in prone
4	Up to wrists in prone Rolls prone to supine
5	Rolls supine to prone Sits with support
6	Sits alone
8	Comes to sit Crawls Pulls to stand
9	Cruises
11	Walks with hand held
12	Walks alone
15	Runs

[a] Adapted with permission from: Capute and Shapiro 1985.[4]

examination should be suspected of having a diagnosis of cerebral palsy (CP). Delays in language and nonverbal/visual-motor abilities, with DQs of 70 or less in both domains, places a child at risk for a diagnosis of intellectual disability/mental retardation. However, delayed language skills in the presence of normal nonverbal/visual-motor skills (dissociation) implies a diagnosis of a communication disorder. Unlike delay and dissociation, deviancy is not used diagnostically. Rather it is a finding that suggests abnormal development within a single stream and should heighten the clinician's suspicions regarding underlying developmental pathology. For example, significant deviancy, such as identifying letters and reciting the alphabet while not using any words to communicate a request, could suggest an autism spectrum disorder (ASD).

Developmental Milestones

Gross Motor Milestones

Motor development is the easiest stream to observe because of the numerous achievements that occur within the first year of life (see Table 7.1 and Chapter 9). Newborns demonstrate little voluntary motor ability and are restricted by primitive (involuntary) reflexes. At birth, a newborn does not have enough motor control to lift his head off of a bed when lying in a prone position. Over the first 4 months of life, neck and trunk tone are acquired in a cranial-caudal direction. By 1 month of age, an infant in the prone position should have enough neck tone to lift his head from the bed, by 2 months with head and trunk tone he lifts his head and chest off of the bed, by 3 months he props on his elbows, and by 4 months he props on his wrists. After trunk tone is acquired, the ability to dissociate the shoulder and hip movement allows for derotative rolling at 4 to 5 months. By 6 months, trunk tone is sufficient to allow the child to be put into a sitting position and to maintain that position. Shortly after sitting, the child integrates trunk tone with the derotational abilities required for pivoting to allow him to transition to a sitting position independently at 8 months. Independent sitting gives way to the ability to assume a quadruped position, ultimately leading to reciprocal crawling and pulling to a stand at

8 months, walking with hands held at 11 months, and walking independently at 12 months.[4]

In addition to the information gleaned by taking a developmental history of milestone acquisition and observation of motor skills, the motor evaluation is enhanced by a neurodevelopmental assessment that includes a standard neurologic examination and observation or elicitation of primitive reflexes and postural reactions. Abnormalities of tone, primitive reflexes, and postural reactions can mitigate the sequence and rate of motor development. The infant neurologic examination should include observation of the posture of the infant at rest and the repertoire of spontaneous movement in the child in a supine resting position. Abnormal posture of the extremities or a paucity of movement in one or more extremities should increase the suspicions of the examiner. Flexor tone predominates in the newborn but diminishes over the first 4 months of life. Normal infant flexor tone is symmetrical, and despite the flexion, there is a normal passive range of movement across all joints. The persistence of flexor tone past the newborn period, or the absence of flexor tone (generally signaling hypotonia) during the newborn period, can interfere with expected motor development. Asymmetry of tone and exaggerated tone (increased or decreased) are abnormal neurologic findings, which are further supported by evaluation of deep tendon reflexes.

Primitive reflexes are automated patterns of movement that are present in utero and persist until 3 to 6 months of life. Primitive reflexes are well-described patterns that have qualitative and temporal characteristics that add valuable information to the classic neurologic examination of infants. The quality of the reflex and the ability or inability to suppress primitive reflexes with repeated elicitations and with advancing age may precede or accompany motor delay and deviancy.[5] Primitive reflexes that persist past the first 6 months of age are abnormal, and their presence is a useful marker in the detection of motor abnormality. Classical primitive reflexes include the Moro, Galant, and Landau reflexes, as well as the asymmetrical tonic neck reflex (ATNR), the tonic labyrinthine reflex (TLR), the positive support reflex, and the symmetrical tonic neck reflex. Each reflex may be observed in the spontaneous movement of the newborn or can be elicited by an examiner. For purposes of delineating the clinical usefulness of the primitive reflexes, the ATNR and TLR will be described.

The ATNR is a primitive reflex in which rotation of the head to either side causes extension of the extremities on the chin side and flexion of the extremities on the occipital side. In the normal child, rotation of the head to one side will elicit an ATNR, but if the head is kept to one side the flexion and extension patterns caused by the ATNR relax, and the extremities resume a neutral position. Similarly, if the head is rotated to one side to elicit the ATNR, successive head rotations produce progressively diminished responses to the head turn. The ATNR is considered obligatory or exaggerated if the child maintains the flexion/extension pattern of the

ATNR the entire time the head is turned to one side, or if the child does not exhibit diminished response to the ATNR with subsequent repetitions. The TLR may be elicited with the child in the supine position or in prone suspension. This reflex is produced with flexion and extension of the neck. The neck brought to flexion will cause flexion of all extremities, and when the neck is brought into extension, the infant extends all extremities and retracts the shoulders. As with the ATNR, the TLR should not be exaggerated or obligatory, and should not persist past 6 months of age. Obligatory or exaggerated responses are considered abnormal and should be factored into the assessment of the motor examination.

Postural reactions are responses of equilibrium and protection that appear midway through the first year, as the primitive reflexes disappear. Postural reactions allow for the development of functional movement, while keeping the head and body upright and oriented. Postural reactions include the Landau reaction; head righting; derotative righting; anterior, lateral, and posterior propping (which are required for sitting); and upper and lower extremity parachutes. Unlike primitive reflexes, which arouse suspicion of motor abnormality by their exaggerated or prolonged presence, postural reactions raise suspicion by their failure to appear at appropriate ages, or by asymmetry of their appearance.

The motor assessment, then, consists of the assessment of the motor age equivalent, gleaned by eliciting a history of motor milestone acquisition and direct observation; the motor DQ (calculated by using the DA equivalent); the neurologic examination; and the assessment of primitive reflexes and postural reactions. Motor DQs of less than 50 signify considerable motor impairment, most commonly CP, while DQs of 50 to 70 represent more mild motor delays, which are often based in hypotonia or motor coordination difficulties and are frequently associated with delays or deviancies in other streams of development.

Visual-Motor Problem-solving Milestones

The visual-motor problem-solving stream of development represents a domain of nonverbal abilities that are dependent on cognitive function, visual capacity, and fine motor ability. Like gross motor skills, visual-motor and fine motor abilities are acquired in a timed and orderly pattern (Table 7.2). Visual-motor development dominates the first 4 months of life: in the first month of life, an infant will use eye movement and slight head turning for visual fixation and following to the midline; during the second month, visual pursuit uses eye and head movement that extends past the midline and in both vertical and horizontal planes; by the third month, the eyes and head can visually follow in a circle and a visual threat response can be elicited; and by the fourth month, the eyes and head turning are coupled with directed upper extremity use.[6] The ability to visually attend and track provides targets for the upper extremity to engage.

Upper extremity movement, like trunk tone, develops in a proximal to distal fashion, beginning with coarse movement originating at the shoulders at 5 months and advancing to precise pincer movement of the forefinger and thumb at 11 months. At 4 months of age, the hands come together in midline; by 5 months, a child can reach up from the shoulders, grasp an object, pull it toward herself, and transfer it from one hand to the other. At 6 months, a child in a sitting position can reach out in front of her and grasp an object presented within reach. Grasp develops in an ulnar to radial fashion, initially requiring support of the ulnar border of the hand and forearm on a surface to secure an object with a rake-like movement initiated by the index and middle finger and employing 3 fingers and a thumb. The radial rake gives way to the immature pincer grasp at 9 months, in which the child can pick up a pellet-sized object with an overhand movement using only the thumb, index, and middle fingers. The mature pincer grasp of an 11-month-old child employs only the index finger and thumb in an overhand movement that is precise.

Table 7.2. Visual-Motor Problem-Solving Milestones[a]

Age, months	Milestone
1	Visually fixes
2	Visually follows in horizontal and vertical plane
3	Visually follows in a circle Responds to visual threat
4	Hands unfisted Hands to midline
5	Reaches for object Transfers object from one hand to another
6	Picks up large object with a radial rake
8	Picks up small object with a radial rake
9	Picks up small object with immature pincer Looks over edge for fallen toy
10	Uncovers hidden toy
11	Picks up small object with a mature pincer
12	Intentional release of objects
16	Scribbles in imitation
18	Scribbles spontaneously
21	Makes tower of 3 cubes
24	Makes horizontal 4-cube train
30	Copies horizontal and vertical stroke in appropriate orientation Makes 4-cube train with a smokestack
36	Copies a circle Draws a person with a head and one other body part Recognizes one color

[a] Adapted from: Accardo and Capute 2005,[15] with permission Paul H. Brookes Publishing Co., Inc.

Nonverbal problem-solving skills are the product of visual, fine motor, and intellectual abilities. Once the visual and fine motor milestones of the first year of life are acquired, cognition becomes the testable variable of the nonverbal domain. In a neurodevelopmental assessment, parents typically can better provide a history of gross motor and speech and language milestone acquisition, but a history of visual-motor problem-solving milestone acquisition might be more difficult to elicit. Thus, during a neurodevelopmental assessment, visual-motor problem-solving milestones should be demonstrated by the child in the examination setting. Most standardized instruments for evaluating visual-motor problem-solving skills employ test batteries modified from the early work of Arnold Gesell,[3] which was considerably shortened by Cattell,[7] and further adapted by many authors over the next few decades. To be clinically useful, tests of nonverbal abilities for young children must provide age equivalents in order to quantify development using a developmental quotient.

Abnormal visual-motor development can result from visual impairment, significant motor impairment, and/or intellectual disability. Impairments sufficient to preclude assessment are identified in the first few months of life in the case of visual impairments, and in the latter half of the first year of life in the case of significant motor impairment. Occasionally children demonstrate qualitatively poor fine motor abilities that do not prevent testing, but which may slow or diminish the accuracy of movement. In the absence of significant motor or visual impairments, delays in problem-solving skills should be considered a result of cognitive or intellectual disability. Developmental deviance in problem-solving may be observed in children with qualitatively poor fine motor skills (children with poor writing skills may be unable to accomplish age-appropriate graphomotor tasks but may do well with other age-appropriate tasks) and in children with attention deficits sufficient to interfere with task completion, despite the motor and cognitive capacity to perform at age-appropriate levels.

Speech and Language Milestones

Language development is an intricate process by which an infant, naive to verbal stimuli, establishes a repository or lexicon of words that becomes a vocabulary available for the understanding of verbally presented information and for communication (see Chapter 11). Normal language development requires the presence of adequate hearing, the cognitive ability to acquire vocabulary and build a word repository, the ability to attend to verbal information, and the desire to establish rapport with a speaker.[8] A child who has all of the prerequisites of language development will gradually store vocabulary to which he or she is exposed. This stored lexicon also records the visual images, word patterns, and affective associations with which the vocabulary is learned. Over time, the stored lexicon is sufficient for the understanding of very complex communication. This understanding of linguistic cipher is referred to in development as receptive language and is measured by a few milestones in the first year of life, but more plentiful milestones thereafter (Table 7.3).

Expressive language development is dependent on a rich word repository or receptive language skills from which information is drawn, as well as the neurologic and oral-motor skills to produce speech. Expressive language skills gradually unfold over the first 12 months of life, beginning with guttural language from 0 to 2 months and followed by pre-linguistic skills from 3 to 11 months.[6] Guttural language represents vocalizations that are largely physiologic noises devoid of social significance, such as crying, burping, coughing, and yawning. The guttural phase of language is followed by the pre-linguistic phase, in which there is a social intent to vocalizations, albeit at a pre-word level. Pre-linguistic skills begin with the social smile at 2 months and coo at 3 months of age and are followed by razzing, laughing, "ah-gooing," and babbling. Babbling begins at 6 months as a string of hard constants and vowel sounds (eg, "da-da-da-da" and "ba-ba-ba-ba") and progress to "dada," which is used nonspecifically at 8 months. Use of "dada" and "mama" to specifically identify Dad and Mom begins at 10 months of age and heralds the beginning of the linguistic phase of development. The linguistic phase begins at about 11 months of age when the first identifiable word is uttered and grows exponentially over the next 2 years of life. Between 12 and 18 months the child's vocabulary expands only to 7 to 10 words but swells to 50 words and 2-word sentences at 24 months and 250 words and 3-word sentences by 3 years of age.

Abnormal speech and language development may represent a primary disorder of language or may be a secondary consequence of intellectual disability, hearing impairment, autism, or motor speech disorder. Language impairments include expressive, mixed receptive and expressive, and pragmatic disorders. Expressive language impairment is a failure to demonstrate age-appropriate verbal skills and is manifest as limited single-word vocabulary, delayed connected language use, or poor speech production (articulation, phonation, or fluency difficulties). Expressive language disorder assumes a delay in expressive language and a dissociation between the delayed rate of expressive language development and normal ability in receptive language and cognition. Speech production disorders affect the quality of verbalizations, rather than the size of vocabulary or use of connected language. In the early linguistic phase, some children use word approximations, in which they may drop the beginning or ending sound of a word ("bott" or "ottle" for bottle) or use sound substitutions ("bobble" for bottle) but are quite consistent and specific with word use. Word approximations, when employed specifically and consistently, should be counted when estimating the size of a child's vocabulary. Similarly, articulation deficits, phonological disorders, or speech fluency disorders (stuttering) affect the quality rather than the content of expressive language. An expressive language disorder should be suspected in the child who is not babbling or using gesture language by 12 months, not using single words by 16 months, or not using 2-word phrases by 24 months of age (Table 7.3).

Receptive language requires the ability to process and understand spoken language precisely and efficiently. Receptive language disorder represents impairment in

comprehension that is unexpected given a child's cognitive ability, evidenced by a delay in receptive language and a dissociation between the delayed rate of language milestone acquisition and an age-appropriate rate of nonverbal cognitive milestone acquisition. Receptive language skills are a prerequisite for learning expressive skills and therefore when receptive language is delayed, there is co-occurring expressive language disorder or a mixed receptive-expressive language disorder. Receptive and expressive language disorders generally present because of concern about expressive language delay, but parents may also complain that their child does not seem to listen or is not following simple commands. There is a paucity of observed receptive language milestones in the first year of life, but a number of receptive milestones are defined in the second and third years, some of which may be demonstrated on examination, some of which are credited by parental history (Table 7.3).

Pragmatic language disorders can be thought of as inadequate understanding of communication at the non-word level. Pragmatic language is the ability to derive meaning from the tone or prosody of voice, rather than the word meaning, and is required for understanding humor, sarcasm, idioms, and other verbal references that require drawing inference. Pragmatic skills also include the understanding of non-verbal communication: facial expression, eye contact, and body language. Children with pragmatic language disorders appreciate only the pedantic meaning of utterances, thereby missing valuable nonverbal meaning of communication. Pragmatic

Table 7.3. Receptive and Expressive Language Milestones[a]

Age, months	Receptive Milestone	Expressive Milestone
1	Alerts to sound	
2	Social smile	
3		Coos
4	Orients to voice	Laughs
6		Babbles
8		"Mama" or "dada" nonspecifically
10	Understands "no"	"Mama" and "dada" specifically
12	Follows 1-step commands with a gesture	2-word vocabulary
18	Points to one picture Identifies >2 body parts	7- to 10-word vocabulary
21	Points to 2 pictures	20-word vocabulary, 2-word phrases
24	Follows 2-step commands	50-word vocabulary, 2-word sentences (noun/verb)
30	Understands the concept of "1" Points to 7 pictures	Uses pronouns appropriately
36	Follows 2-step prepositional commands	250-word vocabulary Uses 3-word sentences

[a] Adapted from: Accardo and Capute 2005,[15] with permission Paul H. Brookes Publishing Co., Inc.

language skill deficits manifest largely as social delays and are most often appreciated in children who are on the autism spectrum. Pragmatic language skills are clinically appreciated in the developmental evaluation but are formally tested by experienced speech-language pathologists.

Social and Adaptive Milestones

The last 2 streams of development, social and adaptive skills, are different from the other streams of development in that (1) they are dependent on motor, language, and cognitive abilities; (2) they are more subject to environmental and cultural influences; and (3) delays in social or adaptive skills do not directly infer diagnoses but provide further evidence for suspected diagnoses (see Chapter 12). Social skills require a combination of language and problem-solving (nonverbal) skills, but they are more heavily dependent on appropriate language, so much so that if language skills are delayed, social skills are likely to be delayed. A child of 3 years is expected to have reciprocal play skills but may continue to engage in parallel play if language skills are delayed enough to interfere with reciprocal play. Learning to play reciprocally and share requires the problem-solving skills to know, for example, that if there is only 1 toy, 2 children cannot each have it at once, and the language skills to establish the rules of playing fairly. Adaptive skills, or self-help skills, require both language and nonverbal skills but are most heavily dependent on nonverbal or problem-solving skills. Adaptive skills such as feeding, toileting, and dressing rely on adequate motor skills as well as cognitive ability (Table 7.4). Adaptive skills are so dependent on cognition that delays

Table 7.4. Social and Adaptive Milestones

Feeding Skills	Age, months
Holds bottle	5
Finger feeds	10
Uses spoon	15
Holds cup	15
Spears with a fork	30
Spreads with a knife	48
Dressing Skills	
Helps with dressing	10
Partially dresses/undresses	24
Dresses completely	48
Buttons	48
Puts shoes on correct feet	48
Ties shoes	60
Toileting Skills[a]	
Urinates independently (girls)	31–33
Urinates independently (boys)	34–37
Dry by day (girls)	32
Dry by day (boys)	35
Uses bathroom independently for bowel movement (girls)	34
Uses bathroom independently for (boys)	39
Social/Play Skills	
Social smile	1½
Laugh	4
Peek-a-boo or pat-a-cake	10
Solitary play	12
Parallel play	24
Interactive play	36

[a] Schum TR, Kolb TM, McAuliffe TL, Simms MD, Underhill RL, Lewis M. Sequential acquisition of toilet- training skills: a descriptive study of gender and age differences in normal children. *Pediatrics*. 2002;109:e48. Available at: http://www.pediatrics.org/cgi/content/full/109/3/e48.

in adaptive skill attainment must be documented for the diagnosis of intellectual disability or mental retardation, thereby affirming the diagnosis suspected by significant delays in language and nonverbal performance. Delays in social skills may be seen in children with cognitive delay or in children with normal cognition with communication disorders. Delays and atypicalities in social skills, in association with dissociated delays and deviation in communication development, are most pronounced in children with ASDs (see Chapter 8 and Chapter 13).

Presentation of Neurodevelopmental Disorders

The age at which children present for evaluation of developmental disorders varies with the stream of development in question and the severity of the delay. In general, disorders of greater severity present earlier and to medical professionals. Disorders of lesser severity (but often of greater frequency) present later and may present to nonmedical professionals. Sensory deficits, including hearing and vision impairment, are often detected in the first few months of life, when a child does not respond to sound in the first months or turn to voice at 4 to 5 months, or when a child fails to fix or follow in the first 3 months. Delays in language should always prompt the primary pediatric health care professional to refer for a formal hearing evaluation, and delays in visual-motor problem-solving should always prompt referral for formal assessment of vision.

Motor deficits typically present in the second 6 months of life, when children fail to sit, crawl, or pull up to stand. Walking is expected at 11 to 12 months, with 15 to 16 months accepted as the upper limit of normal variation. Children with motor DQs of less than 70 warrant a full neurodevelopmental assessment with a developmental history, physical examination, neurologic examination, and evaluation of other streams of development, even though motor delay can occur independent of delays in other domains. A motor DQ of less than 70 would include sitting at ages older than 8.5 months, crawling later than 11.5 months, and walking at later than 17 months. Motor developmental quotients of 50 or less are often associated with CP, and children with this degree of motor delay present at an average age of 12 months.[9] Delays in communication or cognition typically present by about 27 to 32 months of age, most often with failure to demonstrate expressive language skills.[9] Social impairments and behavioral disorders present over a wide range of ages, again depending on the extent to which the disorder is impacting the child or his or her caregivers. Attention-deficit/hyperactivity disorder (ADHD) may present in preschool years because of concerns that hyperactivity or impulsivity preclude normal family function, jeopardize preschool or child care placement, or compromise the child's safety. Attention deficits, however, often present later, when distractibility or poor concentration hinders academic success. Adaptive skill delays are usually not a common presenting complaint but are noted during the developmental history in children who present with other concerns. Disorders of learning generally do not present until school age, when children are put to the task of learning and are unsuccessful.

Neurodevelopmental Assessment

The 2006 AAP policy statement on developmental surveillance, screening, and evaluation recommends a developmental evaluation to confirm results of failed screening.[1] While primary pediatric health care professionals should routinely refer patients who fail screening to their local early intervention programs (EIPs) for standardized developmental evaluations (and may also refer for developmental evaluations to pediatric subspecialists, such as those specializing in neurodevelopmental disabilities or developmental and behavioral pediatrics), information gleaned from a neurodevelopmental assessment performed within the medical home may also assist in the diagnostic process, particularly in terms of making an etiologic diagnosis. Similar to the medical evaluation of other presenting complaints, the neurodevelopmental assessment of a child who fails developmental screening begins with a comprehensive history (most importantly a developmental history of milestone acquisition across developmental streams) that may be confirmed by direct examination.

History

The first step in a neurodevelopmental assessment that follows a failed developmental screen is to obtain (for a new patient in a primary care practice) or review and update (for an established patient in the primary care practice) a thorough history, including the birth, medical, developmental, behavioral, educational, social, and family histories. The complete history offers clues to developmental diagnosis, provides evidence of the child's developmental trajectory, and uncovers historical risks for atypical development. The failed parent-completed developmental screen should lead the primary pediatric health care professional to define the stream of development that is the focus of parental concern, the age of the child when the parents first became concerned, and the impact that the complaint is having on the child's function. Presenting complaints derived from failed screening are typically focused: a child is not sitting or crawling on time or is not speaking enough or clearly. Occasionally, presenting complaints derived from failed screens are curiously vague: "my son isn't like my other children" or "the teachers told me to have my son evaluated." In either case, a thorough evaluation is required to assess the stream of development in question and also to look for lesser concerns in other streams of development, which may not have been appreciated by the family. It is imperative, however, that primary pediatric health care professionals recognize that the presenting complaint derived from a failed screen may represent only the "tip of the iceberg," highlighting only a single, most obvious component of a more extensive developmental disorder (see Chapter 8).

Birth History

Children with significant complications in their birth histories should have their development more closely monitored from birth by their primary pediatric health care professionals, as they are at higher risk for future developmental disorders.

The birth history ascertains the maternal age and parity at the time of the child's gestation, paternal age, medical complications of pregnancy (bleeding, hypertension, gestational diabetes, infections, intrauterine exposures), complications of labor and delivery, and neonatal problems. Intrauterine exposure to certain infections (varicella, herpes, toxoplasmosis, cytomegalovirus) and toxins (alcohol, tobacco, drugs of abuse, and specific groups of prescribed medications) may exert negative effects on fetal neurodevelopment and place infants at risk for neurodevelopmental sequelae. Difficulties with parturition, including prolonged labor with fetal distress and significant neonatal depression or premature delivery, offer varying risks to immediate and long-term outcome. Infants born at gestational ages younger than 37 weeks are considered premature and comprise nearly 13% of the deliveries in the United States, with 1.4% of those children born weighing less than 1,500 g.[10] The risk for developmental disorders, including CP, intellectual disability, vision and hearing impairment, learning disability, and ADHD, are increased in the entire preterm population, but the risk of neurodevelopmental sequelae increases with decreasing gestational age and birth weight. To compound the risks of early or difficult birth, neonatal complications, including hyperbilirubinemia, seizures in the first 24 hours, intraventricular hemorrhage, periventricular leukomalacia, chronic lung disease, retinopathy of prematurity, sepsis, and necrotizing enterocolitis, contribute to the established risk for poor developmental outcome.

Medical History

Medical and surgical history explore established and associated risk of serious infections; head trauma; seizure disorders; and chronic medical, metabolic, or endocrine disorders and surgical procedures. The history of an acute illness, such as meningitis or head injury, or a chronic illness, such as hypothyroidism, will establish a risk for developmental disorders that guides the primary pediatric health care professional in the evaluation and workup of a child with failed developmental screening. Furthermore, the history can provide information that clues the clinician in to associated risks: a history of an uncomplicated submucosal cleft palate repair or a cardiac anomaly repair may not directly influence developmental outcome but should remind the evaluator of possible genetic syndromes associated with midline defects and associated neurodevelopmental disorders.

Family History

Family history of ethnicity; recurrent pregnancy losses; diagnosed genetic, neurologic, or psychiatric disorders; intellectual disability; delayed speech, language, or motor development; hearing or vision impairment; learning disability; ADHD; and autism establish varying degrees of risk of similar disorders in the child being evaluated following a failed screen. A history of hearing impairment diagnosed in an octogenarian in the family contributes little of relevance to the child; however, a family history of ASD is of greater interest because of the evidence of a genetic basis to autism[11] (see Chapter 13).

Developmental History

A careful developmental history offers the most critical contribution to the neurodevelopmental assessment. Each stream of development should be reviewed to ascertain the age at which specific milestones were attained, so as to formulate an estimate of past DQs within each stream, thereby approximating a relative rate of development over time. As an example, in the child that presents with motor delay, a history of rolling over at 8 months, sitting at 12 months, and crawling at 16 months provides retrospective evidence of a consistent motor DQ of 50 and infers a static rate of development over time, which implies continued delays into the future. The estimation of developmental rates using the timing of prerequisite milestone achievement in comparison to DQs acquired during direct neurodevelopmental examination offers powerful information about developmental trajectories within a given stream of development, and thereby assists with prognostication. The patterns of milestone achievement are of profound importance to the evaluation. The persistence of delay over time, or a static pattern of developmental delay, suggests that continued delay is likely, but a developmental plateau or a history of developmental regression requires special attention. A developmental plateau, in which a child fails to acquire new milestones after a previous pattern of steady milestone attainment, is seen most frequently in language development. The timing of the language plateau is informative: children with severe hearing impairment acquire language normally until they reach the babbling stage at 6 to 8 months, then fail to progress past babbling.[12] Also, as many as one-third of children on the autism spectrum either plateau in language or lose previously achieved language and social skills between 18 and 30 months of age.[13] Regression, or loss of previously achieved milestones, though common in the language and social domains of children on the autism spectrum, may require extra consideration in the etiologic workup. When regression includes motor or cognitive streams of development, a thorough neurologic evaluation, including imaging and laboratory studies, is warranted to rule out a neurodegenerative disorder.

Behavioral and educational histories are important components of the developmental history and offer qualitative information about the child's function in his or her environment and reveal evidence of associated deficits. Behavioral disorders may be found in the areas of social behavior, problems with compliance, aggression, disorders of attention and activity (ADHD), stereotypic behaviors, and self-injurious behavior. Abnormalities such as immature play skills (eg, parallel play at an age when reciprocal play is expected or lack of symbolic or pretend play not explained by developmental level), lack of desire to interact with other children, or awkwardness in or avoidance of social situations may be uncovered during the behavioral history and direct the evaluator to further explore ASDs, especially when the delay in social behavioral development is accompanied by stereotypic behavior and language delay. Attention-deficit/hyperactivity disorder commonly coexists with developmental disorders that may be raised in the presenting complaint or the initial observation of a child but is further explored by inquiring about attention, distractibility, impulsivity,

and hyperactivity. Noncompliance and aggression, when present, should prompt the evaluator to determine if these behaviors are secondary to frustration originating from language or cognitive demands that exceed the child's capability or if they are a primary behavior that requires behavioral modification. Motor stereotypies (eg, hand flapping, spinning, compulsive jumping, or pacing) and self-injurious behaviors occur with greater frequency in children with intellectual disability, ASDs, and sensory deficits.

Social History

The social history should evaluate the psychosocial and socioeconomic risks and supports that the child who fails developmental screening possesses within the family and the community. The assessment of psychosocial stressors that may impact the family and child either directly or indirectly include parental educational history and employment status; family financial status; marital discord or separations; single-parent households; frequent moves; substance use or exposure; history of physical, sexual, or emotional abuse or neglect; members of the household/caretakers with mental health or developmental disabilities; and quality of child care. Review of early intervention services or school services that a child requires or is receiving must be appraised in light of the child's needs, the resources and stressors of the family, and the services available in the community.

Physical Examination

The physical examination of a child who fails developmental screening will include an assessment of growth parameters, a general physical examination, and a neurologic examination. Measurements of head circumference, height, and weight at the time of the examination, as well as documentation of historical growth parameters, can prove to be of diagnostic importance. Uniformly small growth (height, weight, and head circumference) may be explained by family history/growth patterns and known risks, such as prematurity, fetal alcohol exposure, or intrauterine infection, or may alert a clinician to genetic disorders. Short stature with normal head circumference and weight also raises questions about familial stature, endocrine dysfunction, and genetic disorders. In a child with microcephaly (head circumference measurement under the second percentile), evaluation of the trajectory of head measurements may provide etiologic clues: a child with microcephaly at birth may have been compromised by infection or a cerebral disruption in utero, while a child with a normal head circumference at birth but progressively diminishing percentile measurements in the first few months of life may have suffered a peripartum event. However, diminishing head circumference measurements in the second half of the first year of life of a girl brings suspicions of Rett syndrome, particularly if associated with loss of developmental skills and in some cases certain behavior patterns, such as hand wringing. Macrocephaly (head circumference greater than the 98th percentile) is associated with congenital hydrocephalus, acquired hydrocephalus, a number of

genetic overgrowth syndromes, and metabolic syndromes such as Canavan's disease, all of which carry risks of associated developmental disorders. Tall and short stature must be considered first in light of familial growth patterns. If a child's height is 2 standard deviations from the mean, and discrepant from familial growth, etiologic considerations include disorders of endocrine function, exposures to the fetus (such as alcohol), and syndromes associated with short (such as skeletal dysplasias) or tall (Klinefelter, fragile X, Marfan syndromes) stature and overgrowth syndromes (height, weight, and head circumference are all 2 standard deviations above the mean, such as in Simpson-Golabi-Behmel, Bannayan-Riley-Ruvalcaba, and Sotos syndromes).

The general physical examination should be expanded in children who fail developmental screening to include a search for markers or unusual features that may provide an indication of an underlying genetic disorder. Examination with a Wood's lamp should be considered to assist in identification of neurocutaneous markers of neurofibromatosis (café-au-lait spots, unusual freckles) and tuberous sclerosis (hypopigmented patches, shagreen patches). Midline defects noted on general examination, including cleft palate (or submucosal clefts), cardiac defects, and genitourinary findings may be informative. Dysmorphic features of the head, face, ears, or hairline may be subtle but contribute to an overall constellation of features that might suggest a specific syndrome associated with intellectual disability (see Chapter 10). Dysmorphic features of the extremities also need to be recognized. For example, noticeable 2- to 3-toe syndactyly should prompt an evaluation for Smith-Lemli-Opitz syndrome, a not uncommon metabolic cause of hypotonia, developmental delay, and autism, which would have significant treatment and family counseling implications. However, it is also important to be aware that most genetic syndromes are in fact a constellation of physical and neurobehavioral features: no single dysmorphic feature or neurobehavioral pattern is diagnostic, and no syndrome requires a complete constellation of features for diagnosis.

Neurologic Examination

The neurologic examination is critical to the neurodevelopmental assessment of a child who fails developmental screening, as it may provide explanation for motor findings, as in the case of the infant with motor delay or the older child with coordination difficulties. The neurologic examination of the infant and young child must include a standard evaluation of cranial nerve function, tone, strength, and reflexes, as well as pathological, primitive, and postural reflexes and a thorough assessment of motor function. In the ambulatory child, the quality of gait, functional movement (climbing stairs, rising from a sitting or squatting position), and fine motor movement quality imparts additional information to the neurologic assessment. Finally, in older children who present with concerns regarding cognition, language, learning, or behavior, an examination of neurologic soft signs can add to the standard neurologic evaluation. Soft signs, such as mild axial hypotonia, synkinesia, or overflow activity

(which is observed when a child is asked to perform specific repetitive hand movements or walk with a stressed gait), often accompany disorders of communication, learning disability, and ADHD.[14]

Neurodevelopmental Examination

Children who fail developmental screening require referral to local EIPs or pediatric subspecialists for standardized developmental evaluation. However, primary pediatric health care professionals may consider expanding the traditional neurologic examination of a child who fails developmental screening to include a direct neurodevelopmental examination to confirm the developmental history provided by parents and other caretakers, thereby adding to the validity of developmental conclusions. As an informative, yet non-standardized, component of the direct observation component of developmental surveillance (see Chapter 6), the neurodevelopmental examination may minimally consist of direct observation of a child performing the developmental milestones listed in Tables 7.1 to 7.4 to confirm the developmental history provided by the parents. Alternatively, standardized instruments are available to those primary pediatric health care professionals with interest in direct neurodevelopmental examination. For example, the Capute Scales, consisting of the Cognitive Adaptive Test and Clinical Linguistic and Auditory Milestone Scale (CLAMS), is an example of a standardized neurodevelopmental examination measure designed specifically for primary pediatric health care professionals to use in the office setting that assesses language and visual-motor problem-solving skills in children from birth to 3 years of age.[15]

Neurodevelopmental Examination Tools Commonly Used by Pediatric Subspecialists

Most measures used in standardized neurodevelopmental examination require specific training and are typically reserved for pediatric subspecialists, such as neurodevelopmental pediatricians or developmental-behavioral pediatricians, or other early childhood professionals and, thus, cannot be routinely incorporated into primary care practice. Chapter 14 provides a comprehensive review of standardized developmental evaluation. However, the following paragraphs will review some neurodevelopmental examination tools commonly used by pediatric subspecialists that may assist primary pediatric health care professionals in interpreting reports about their patients referred for subspecialty consultation as a result of failed screening. The neurodevelopmental examination tools to be discussed are summarized in Table 7.5.

A plethora of assessment tools are commercially available that range in scope from comprehensive measures that evaluate several domains of development to those that measure specific domain functions, such as articulation, receptive language, pragmatic skills, fine motor skills, writing skills, etc. Specific tests are often chosen based on the age of the child being tested, the streams of development being explored, the psychometric properties of the test (validity, reliability, specificity, and sensitivity),

the time required for test administration, and the preferences of the examiner. Ideal tests are valid, reliable measures that provide quantifiable results in a specific scope of assessment, are expeditious, and have good specificity and sensitivity. Often, comprehensive developmental testing requires the employment of a number of narrow scope tools to verify information or complement information gleaned on broad tests of development or from behavioral observation.

Estimation of cognitive ability is the first order of testing, as academic, language, social, adaptive, and behavioral findings are all assessed in relation to the child's cognitive potential. Measurement of intellectual capacity in the verbal (language) and nonverbal (visual-motor problem-solving) domains is imperative, as by employing the principles of delay and dissociation; it will distinguish the child with intellectual disability from the child with normal nonverbal skills and a communication disorder. As concerns about language delay and global developmental delay become evident to parents in the second and third years of life, many of the test measures for early identification of children with such difficulties target children from birth to 3 years of age. The Battelle Developmental Inventory[16] and the Mullen Scales of Early Learning[17] are examples of valid assessment tools of language and visual-motor problem-solving skills in the infant, toddler, and preschooler that require varying degrees of expertise and time to administer. In the older child, an estimate of cognition becomes more difficult, as fewer general measures are available. The Fluharty Preschool Speech and Language Screening Test,[18] the Clinical Evaluation of Language Fundamentals-Preschool,[19] the Peabody Picture Vocabulary Test (PPVT),[20] and the Expressive One-Word Vocabulary Test[21] are practical, psychometrically sound measures of language capacity in children that produce quantifiable results. Nonverbal test measures for the older preschooler and young school-aged child are more difficult to find, and pediatric subspecialists often use subtests from standardized tests, such as Gesell's figure drawing and block designs[22] and Goodenough's Draw a Person.[23] These and other selected subtests are used to estimate nonverbal ability. Other standardized measures to assess nonverbal abilities that may be used by pediatric subspecialists include the Leiter-R, the Raven Coloured Progressive Matrices, the Test of Nonverbal Intelligence, and the Matrices subtest of the Kaufman Brief Intelligence Test. Caution in the interpretation of nonverbal skills that rely largely on pencil and paper tasks is prudent, as children with even mild neuromotor disorders may have sufficient graphomotor impairment to cause the evaluator to underestimate a child's nonverbal abilities unless a battery of measures that do not require pencil and paper assessments is also engaged. Formal intelligence testing performed by a school psychologist will confirm estimates of cognitive ability based on the neurodevelopmental examination of school-aged children.

In school-aged children referred by their primary pediatric health care professionals to pediatric subspecialists with concerns about educational success, measures of academic achievement levels, such as the Wide Range Achievement Test–Fourth Edition (WRAT-4),[24] are useful in elucidating academic underachievement and

potential learning disabilities in reading, spelling, and arithmetic. Difficulties in written language and composition are common in children with language-based learning difficulties and may be evaluated using measures such as the Test of Written Language (TOWL).[25] In academic testing, it is essential to assess the time required for children to perform specific batteries of tests as well as the quality of the work produced. Timed tests may alter performance by either limiting the quantity of work accomplished or by limiting the quality of work, and thereby underestimate the capacity of the child to complete work at a higher level if given more time. The distinction between poor performance because of inadequate time to perform and poor performance because of inability to perform should not be overlooked, as the interventions for the 2 scenarios are very different: the slower capable worker

Table 7.5. Neurodevelopmental Examination Tools

Test	Language	Nonverbal	Academics
Infants and Toddlers			
Battelle Developmental Inventory	√	√	
Capute Scales	√	√	
Mullen Scales of Early Learning	√	√	
Preschoolers			
Battelle Developmental Inventory	√	√	
Gesell Figures		√	
Gesell Blocks		√	
Goodenough Draw a Person		√	
Fluharty-2	√		
Peabody Picture Vocabulary Test-4	√		
Expressive One-Word Vocabulary Test	√		
School Age			
Peabody Picture Vocabulary Test-4	√		
Expressive One-Word Vocabulary Test	√		
Test of Written Language			√
Wide Range Achievement Test 4			√
Leiter-R		√	
Raven Coloured Progressive Matrices		√	
Behavioral Measures			
Childhood Autism Rating Scale			
Gilliam Autism Rating Scale			
Gilliam Asperger Diagnostic Scale			
Vanderbilt ADHD Scale			
Conner ADHD Scales			
DuPaul ADHD Rating Scale-IV			

Abbreviation: ADHD, attention-deficit/hyperactivity disorder.

requires accommodations to succeed, while the child with specific learning disabilities needs remedial or supportive services in the academic setting.

Behavioral Assessment

The assessment of behavior is often a challenging component of the neurodevelopmental assessment process for a child who fails screening. In some circumstances, a child with ADHD will demonstrate difficulties with attention or will show overt hyperactivity and impulsivity during the office visit. Similarly, a child presenting with concerns about early language delay, developmental delay, or an ASD may display classic signs of autism, such as gaze avoidance, poor joint attention, lack of response to verbal prompts, and stereotypic behaviors. Commonly, however, children show very few of the behavioral patterns about which their parents are concerned during an office visit. In either circumstance, behavior rating scales are valuable accompaniments to historical information and observation, as they often give relative quantification of severity and objective information from parents, teachers, or child care providers regarding the child in different social or academic environments. Behavior rating scales should never be used alone as diagnostic tools; they are simply measures that may assist in gathering information on symptoms of a suspected diagnosis. Numerous ADHD rating scales for parent and teacher reporting are commercially available and provide observations of the child in different settings. These scales are also very valuable as tools to measure differences in ADHD symptoms before and after interventions are initiated (see Chapter 16).

Specific rating scales are also available for observations of behaviors on the autism spectrum. There are both parent-completed rating scales and observer-completed scales that offer descriptive information of behavior in each of the 3 domains of autistic symptoms: social interaction, communication, and restricted or repetitive behavior (Table 7.5). Despite the capacity to quantify degree of likelihood of an ASD, these rating scales are quite subjective and, ultimately, an autism spectrum diagnosis should not be solely based on caregiver-completed rating scales but on the clinical judgment of an experienced clinician based on presenting clinical information, observation/elicitation of developmental skills and behavior, and results from standardized diagnostic instruments (see Chapter 13).

Interpretation of Neurodevelopmental Assessment: Diagnoses

The principles of delay and dissociation are ultimately applied in the interpretation of the findings within the 5 streams of development (Table 7.6), along with qualitative consideration of deviancy. Gross motor delay with a developmental quotient of 50 or less is consistent with a diagnosis of CP regardless of the developmental quotients in other streams (see Chapter 9). Delay (DQ <70) in both streams of language and visual-motor (nonverbal) skills is consistent with a diagnosis of intellectual disability (mental retardation) and is generally accompanied by delays in adaptive skills and perhaps social skills as well (see Chapter 10). Delay in the

language stream in the presence of normal visual-motor skills exemplifies dissociation between the cognitive streams of development and indicates a communication disorder (see Chapter 11). Communication disorders may be accompanied by deficits in social skills as well and should signal the need for a historical review of autism symptoms or behavioral rating scales for autism symptoms (see Chapter 8 and Chapter 13).

A diagnostic summary clarifies the findings of the child being evaluated following failed screening and also leads to consideration of specific associated deficits and future risks. For example, a child with motor delay and physical findings consistent with a hemiplegic pattern of CP should be evaluated for common associated findings, such as homonymous hemianopsia and seizures, while the child whose motor delay yields a pattern of spastic diplegia is at greater risk of comorbid intellectual disability. Similarly, the child with language delay and associated low tone may have co-occurring attentional deficits but should also be considered to be at increased risk for language-based learning disabilities and monitored for academic underachievement as he or she advances in the school environment. Knowledge of patterns of associated comorbidities of developmental diagnoses and estimation of future risk of subsequent developmental hurdles provides parents with appropriate expectations and anticipatory guidance that is specific to their child with atypical development. The diagnostic conclusions of the neurodevelopmental assessment, made in conjunction with results of the standardized developmental evaluation performed by local early childhood professionals, also help to establish more clearly the nature of intervention services needed and enable the primary pediatric health care professional to guide parents to appropriate local resources.

Table 7.6. Diagnostic Interpretation of Delay and Dissociation

	Cerebral Palsy	Intellectual Disability	Communication Disorder	Autism
Gross motor	DQ <50	Normal or delayed	Normal or delayed	Normal or delayed
Language	Normal or delayed	DQ <70	Delayed	Delayed
Visual-motor or nonverbal	Normal or delayed	DQ <70	Normal	Normal or delayed
Adaptive	Normal or delayed	Delayed	Normal	Normal or delayed
Social	Normal or delayed	Normal or delayed	Normal or delayed	Delayed

Abbreviation: DQ, developmental quotient.

Medical Evaluation

The diagnostic summary of a child's neurodevelopmental assessment, made in conjunction with results from a standardized developmental evaluation performed by the local EIP, produces a response to the parents' concern as to *what* is wrong (CP, intellectual disability, autism, etc), while the etiologic assessment targets *why* a child has the diagnosed developmental disorder. The 2006 AAP policy statement on developmental surveillance and screening recommends a diagnostic medical evaluation for all children who fail developmental screening.[1] This policy statement indicates that this evaluation can be performed by a pediatric subspecialist, such as a neurodevelopmental pediatrician, child neurologist, or developmental-behavioral pediatrician, or through referral to a pediatric geneticist. However, given concerns about access to such subspecialty expertise, the diagnostic medical workup can be performed by primary pediatric health care professionals within the medical home, often in conjunction with a comanagement plan with the specialist.

Etiologic considerations begin in the birth, medical, and family histories, and continue with physical findings. In gross motor impairments, careful history of gestational, intrapartum, or neonatal complications offers clues to etiology in many instances. Congenital infections, specific perinatal medications or toxic exposures, peripartum complications, neonatal seizures, multiple gestations, and known family history of hematologic or metabolic disorders can provide information as to potential etiologies of motor delay, which may be confirmed with appropriately directed investigations. Premature delivery alone raises the risk of neurodevelopmental sequelae, which is compounded by some neonatal complications associated with preterm birth, such as intraventricular hemorrhage, periventricular leukomalacia, severe chronic lung disease, and other neonatal conditions. Historical information that suggests an etiology for motor impairment directs the etiologic investigations, which may include neuroimaging studies of the head or spinal cord; hematologic studies for clotting disorders; metabolic testing; and studies for infectious origins, such as TORCH infections. When the history is not suggestive of an etiology, the approach to the investigation of motor delay will consider the head circumference measurements and trajectory of head circumference growth, the observations of midline defects, dysmorphic features, lumbar or sacral findings, and the overall neurologic examination findings. The American Academy of Neurology (AAN)[26] recommends neuroimaging studies for children with CP of unknown cause, with magnetic resonance imaging in preference to computed tomography scanning. Selective metabolic and genetic studies are to be considered for children in whom no structural defect is found on imaging studies and in whom additional historical or physical examination findings warrant further investigation. Further consideration of coagulation disorder studies in children with hemiplegic CP is suggested by the AAN.[26]

The etiologies of global developmental delay and intellectual disability are diffuse and often unexplained by the most detailed of medical histories. The AAN,[27] the AAP,[28] and the American College of Medical Genetics[29] have all published similar guidelines for the medical evaluation of children with global developmental delay or intellectual disability. Each expert group recommends a thorough medical history, with a 3-generation family history, a dysmorphology evaluation, and a neurologic examination as a first step for assessment. If history or physical examination suggests a particular etiology, appropriate investigations should be performed to confirm these suspicions. If no diagnosis is suspected after careful history and examination, a stepwise investigation is prudent. The laboratory investigations recommended by these expert panels begin with a high-resolution karyotype and fragile X DNA studies. However, currently the more advanced technology of whole genome microarray comparative genomic hybridization (CGH) should be considered in addition to fragile X DNA studies. The whole genome microarray CGH technology identifies deletions and duplications of chromosome material in a manner more efficient than previously recommended karyotyping and targeted fluorescent in-situ hybridization (FISH) studies. Consideration may also be given to testing for metabolic disorders, if history suggests periodic or cyclic changes; loss of skills; or precipitation of symptoms by an intercurrent illness. Magnetic resonance imaging of the brain is suggested to look for structural defects, especially in the child with motor findings, microcephaly, macrocephaly, or changing head circumference growth velocity. Electroencephalogram (EEG) is recommended only if there is clinical suspicion of seizure activity.

Successful determination of etiology of delayed development is variable and influenced by the severity of the delay, the domains in which the delays are found, and the presence of dysmorphic features or focal findings on examination. In general, the greater the severity of the developmental delay or intellectual disability, the more likely one is to determine etiology. When all modalities of etiologic determination are considered (history, physical examination, imaging studies, chromosomal or genetic evaluations, metabolic assessments, etc) it is estimated that as many as 40% to 60% of children with developmental delay will have a cause identified, with most of those with cause identified functioning in the moderate to severe range of impairment or delay.[30,31] Etiologic yield is also variable by stream of development, with motor delays having the highest yield (59%) and language delays providing the lowest yield (4%).[32]

High-resolution karyotype is estimated to provide an explanation for about 5% of children with developmental delay, and DNA testing for fragile X yields a positive result in 2.6% of cases.[27] Metabolic testing and EEG testing each provide about 1% positive findings. The addition of whole genome microarray CGH to a workup that had previously provided no etiologic yield provides an additional 10% diagnostic

yield, with study variability of 4.2% to 20%.[30] Although currently not a part of any consensus guidelines, at this time, whole genome microarray CGH should replace high-resolution karyotype and subtelomeric FISH probes as the first line of investigation in intellectual disability/mental retardation and ASDs. The only finding that high-resolution karyotype will detect that whole genome microarray CGH will not detect is a balanced translocation, which would not be the etiology of the disability if truly balanced. Specific DNA testing for fragile X syndrome should accompany whole genome microarray CGH testing for all children with intellectual disability/mental retardation or ASDs.

The etiologic evaluation of autism in children overlaps tremendously with the workup of developmental delay, which is appropriate, as the rate of intellectual disability in children with autism is so high (see Chapter 13). Current evidence-based recommendations by the AAN include 2 levels of evaluation. Level 1 recommendations include developmental screening, with further developmental evaluation for those who fail the screen, auditory evaluation, lead screening, and monitoring of siblings. Level 2 recommendations include a high-resolution karyotype and DNA studies for fragile X in children with autism and intellectual disability. As stated previously, whole genome microarray CGH should now be considered the first line of investigation in children with ASDs. Selective metabolic testing is recommended for children with associated cyclic vomiting, lethargy, or seizures and should be considered in children with regression of language and social skills. Unlike the recommendations for global developmental delay, there is no recommendation for neuroimaging studies in autism, despite the higher incidence of macrocephaly in this population. However, evaluation for *PTEN* gene mutations (associated with Cowden syndrome) has increased diagnostic yield in children with autism and macrocephaly and has significant implications for tumor surveillance and family counseling. Electroencephalogram evaluation is only recommended with the suspicion of clinical or subclinical seizures, or with a history of regression, and when requested, EEGs in the autism population should be sleep-deprived studies. There is no scientific evidence to support hair or stool analyses, allergy testing, immunologic assessments, or vitamin or micronutrient level measurements in the etiologic workup of children with ASDs.[33] The genetic evaluation yield for the cause of autism ranges from 6% to 15%,[33,34] but it is also likely to increase with increased use of whole genome microarray CGH technology.

The technology presently available for the assessment of the etiology of developmental disorders can provide information that would be unthinkable only a decade ago. Neuroimaging techniques offer clear information on structural defects and may be amended with spectroscopic or angiographic information. Fluorescent in-situ hybridization probes and microarray techniques offer more specific genetic information over the whole genome. However, clinicians should remember several points as they discuss launching a workup with families. First, technology is not a substitute

for clinical assessment, and a comprehensive evaluation by a clinical geneticist may be of tremendous benefit in the etiologic assessment of children, particularly when a child with clear dysmorphic features and associated neurologic or growth concerns is not diagnosed using conventional techniques. Second, the technology of today, which might not have been imagined a decade ago, will be replaced in the next decade with more advanced technologies, so reassessment of the etiology of developmental delay should be revisited frequently. Third, as the cost of etiologic workups can be tremendous, tests should be chosen with great consideration and performed in a tiered manner when the family chooses to pursue information regarding the cause of the disability (Box 7.1). Finally, the search for the cause of developmental delays should be offered to, rather than imposed on, parents. It should be made clear to families that even if an etiology is found, it is unlikely to provide a treatment at this time. The pursuit of etiology may relieve family anxiety regarding the cause of the delay, and in the case of an identified disorder, may provide information regarding the natural history and associated medical risks of the disorder, as well as family planning information surrounding the risk of conceiving a child with similar findings.

Box 7.1. Suggested Tiered Etiologic Investigations[a]

Language Delay	Global Developmental Delay	Autism Spectrum	Cerebral Palsy
History and physical examination Hearing assessment Genetic evaluation if indicated by physical examination	History and physical examination Hearing assessment Whole genome microarray CGH or high-resolution karyotype (if whole genome microarray CGH not available) Fragile X FISH for specific syndromes if suspected and whole genome microarray CGH not available Metabolic testing when indicated MRI (brain)	History and physical examination Hearing assessment Whole genome microarray CGH or high-resolution karyotype (if whole genome microarray CGH not available) Fragile X Lead level if history of mouthing or pica FISH for specific syndromes if suspected and whole genome microarray CGH not available Metabolic testing with history of regression	History and physical examination with classification of type of CP Assess for associated deficits (vision, hearing) Neuroimaging (MRI preferred to CT scan) Consider coagulation studies in children with hemiplegic CP Consider metabolic and genetic testing in children with atypical features

Abbreviations: CGH, comparative genomic hybridization; CP, cerebral palsy; CT, computed tomography; FISH, fluorescent in-situ hybridization; MRI, magnetic resonance imaging.
[a] Adapted from: Shevell et al 2003[27] and Moeschler et al 2006.[28]

Conclusion

The neurodevelopmental assessment of children who fail developmental screening is a structured, comprehensive assessment of all streams of development in those children who have been identified by screening as potentially having developmental delay. The neurodevelopmental assessment has 4 basic components: (1) a thorough history to assess historical and established risk for developmental tribulations; (2) a complete general physical examination and neurologic examination to record general medical, neurologic, and dysmorphic findings; (3) a direct examination of the 5 streams of development to confirm historical information and establish age equivalents and DQs in each developmental domain; and (4) a medical evaluation to attempt to establish an etiologic diagnosis. The conclusion of the evaluation should generate a summary of diagnoses, a hypothesis as to the etiology of the delay, and a strategy for providing appropriate interventions to optimize outcome.

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Chapter 8

Developmental and Behavioral Diagnoses: The Spectrum and Continuum of Developmental-Behavioral Disorders

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Developmental Versus Etiologic Diagnosis

Developmental and behavioral disorders are the most prevalent chronic medical diagnoses encountered by primary pediatric health care professionals.[1] Given their frequent, longitudinal contact with children and their families, primary pediatric health care professionals are ideally positioned to identify children with developmental delays and behavioral problems, refer for appropriate educational and therapeutic services at the earliest possible age, and make comprehensive diagnoses within the medical home. Such diagnoses are 2-fold—each child requires a descriptive developmental-behavioral diagnosis (eg, cerebral palsy [CP], intellectual disability [formerly called mental retardation], attention-deficit/hyperactivity disorder [ADHD], autism, etc), but it is also imperative to pursue an appropriate laboratory workup (eg, chromosomal analysis, metabolic studies, head imaging, etc) in an attempt to establish an etiologic diagnosis to account for the child's developmental-behavioral difficulties. For example, a child who is exhibiting a static pattern of severe global developmental delays will receive a descriptive developmental diagnosis of intellectual disability; however, laboratory workup may reveal the child to have a chromosomal deletion syndrome. The specific chromosomal deletion is the etiology to account for the child's intellectual disability. Similarly, a child described as evidencing the developmental and behavioral patterns that meet criteria for a descriptive developmental diagnosis of autism may be found on laboratory testing to have fragile X syndrome—the fragile X syndrome is the etiology of the child's autism.

The more severe the developmental-behavioral disorder, the more likely a specific etiologic diagnosis will be identified. However, across the spectrum of developmental-behavioral disorders, mild disorders predominate over severe disorders and, thus,

while all children with developmental-behavioral disorders will receive a descriptive diagnosis, the specific etiologic diagnosis remains unknown for most children. For example, in children with intellectual disability, it has been reported that a biomedical cause can be found in 43% of those with moderate to severe intellectual disability (IQ <50), while such a cause can be determined in only 13% of those with mild intellectual disability.[2] With rapid advances in genetic testing and neuroimaging techniques over time, more and more children with developmental-behavioral diagnoses will receive etiologic diagnoses to account for their disabilities in the future. While each child's longitudinal pattern of specific developmental strengths and weaknesses guides predictions about prognosis and recommendations for educational and therapeutic interventions, a specific etiologic diagnosis allows for genetic counseling for families, may lead to detection of associated anomalies or medical problems and prevent medical complications, and often provides parents peace of mind in knowing specifically why their child has a disability.

Although it is critical to make developmental-behavioral diagnoses at as early an age as possible, and the American Academy of Pediatrics (AAP) has recommended the use of standardized developmental screening instruments at specified ages to identify delays early,[3] it is important to note that mild developmental-behavioral disorders are much more common than severe disorders. Further, the more subtle the developmental-behavioral disorder, the older a child must be before his or her development or behavior is appreciably different from similarly aged peers, such that the developmental or behavioral difficulty can be reliably identified by screening. Alternatively, the more severe the disability, the younger the age at which it can be reliably identified by screening. For example, severe intellectual disability should be suspected by 1 year of age—at 1 year of age, children with severe intellectual disability would be expected to show developmental skills at the level of a 6-month-old or below. However, of all children with intellectual disability, approximately 85% will have mild intellectual disability[4]—many of these children may not be reliably identified until they reach at least 3 years of age, when their developmental skills would be expected to be at an approximate 2-year-old level or below, clearly behind their similarly aged peers. Thus a significant number of children with mild intellectual disability may not be identified by developmental screens performed in the first 3 years of life, when such screens are recommended at specified ages. In fact, it is likely that many individuals with mild intellectual disability are actually never identified. While statistically, based on a normal distribution of intelligence, the prevalence of intellectual disability should be between 2% and 3%, prevalence rates determined by ascertainment are closer to 1% due to methodologic problems in identifying individuals with mild intellectual disability as adults; many of these individuals blend in with and become indistinguishable from other members of the community.[4] Further, the most subtle of cognitive disabilities, the specific learning disabilities (LDs), such as dyslexia, certainly cannot be diagnosed in the first 3 years of life and are typically not diagnosed until a child begins to have difficulty keeping up with

peers at school. Similarly, while intensive early intervention strategies have been shown to benefit children with autism spectrum disorders (ASDs),[5] it is typically only the more moderate to severe ASDs that can be most reliably identified by screening in the birth to 3 population.[6] The behavior of children with milder cases of pervasive developmental disorder not otherwise specified (PDD-NOS) or Asperger syndrome might not be recognized to be significantly different from peers until older ages, when there are increasingly complex demands for peer social interaction and communication. Thus, while early recognition and intervention should always be the primary goal, it remains difficult to identify the much more common milder disabilities in the birth to 3-year age range with the use of standardized developmental screening techniques.

Given these limitations of screening, improved clinical judgment in making developmental-behavioral diagnoses may be gained through an appreciation of the spectrum and continuum of developmental-behavioral disorders. Although each developmental-behavioral diagnosis has specific diagnostic criteria that will be reviewed in subsequent chapters of this manual, it is hoped that the model presented in this chapter will lead to improved understanding of the developmental processes underlying these diagnoses and the significant comorbid and overlapping relationships among these entities. Increased awareness of this model should enhance the clinical judgment of primary pediatric health care professionals and assist them in confidently making developmental-behavioral diagnoses within the medical home.

Primary Developmental Diagnosis and Associated Deficits (Comorbidities)

As opposed to adult neurology, where acquired focal neurologic deficits derivative of etiologies such as cerebrovascular accidents are common, the neurologic pathology in developmental-behavioral disorders of childhood tends to be more diffuse. While each child with a developmental-behavioral disorder will typically receive a primary developmental-behavioral diagnosis, such as CP, developmental coordination disorder (DCD), intellectual disability, LD, autism, or ADHD, associated deficits or comorbidities are the rule, rather than the exception, in children with these disorders. For example, approximately 25% of children with intellectual disability also have CP[7] and 30% have ADHD.[8] Approximately 50% of children with CP also have intellectual disability.[9] Further, about two-thirds of children with autism also have intellectual disability.[10] In addition, about 60% of children with ADHD have LDs[11] and 50% have motor coordination disorders.[12]

Such diffuse neurologic dysfunction should not be unexpected, given the etiologic entities that typically cause developmental-behavioral disorders. For example, a child with a chromosomal abnormality, such as a trisomy or chromosome deletion syndrome, would evidence the chromosomal abnormality in every neuron of the brain. Thus diffuse neurodevelopmental dysfunction would be expected to result from such

diffuse brain involvement. Similarly, other hypoxic-ischemic, metabolic, infectious, or toxic causes of brain dysfunction or the effects of environmental neglect or understimulation on a developing brain would also be expected to cause more diffuse rather than focal developmental-behavioral impairments. Thus, while in this chapter a spectrum of developmental-behavioral disorders from mild to severe will be described within each of 3 streams of development (motor, cognitive, and social-behavioral), one must remember that there is also a continuum of developmental-behavioral disorders across developmental streams—children with primary motor disorders are likely to have associated cognitive, learning, or behavioral difficulties; children with primary cognitive disorders are likely to have associated motor or behavioral issues; and children with primary social-behavioral disorders are likely to have associated learning or cognitive differences.

Delay, Dissociation, and Deviance

Developmental-behavioral disorders occur along a spectrum and continuum from high prevalence, low morbidity conditions (such as DCD, LD, and ADHD), to low prevalence, high morbidity conditions (such as CP, intellectual disability, and autism). Capute and Accardo[13,14] have described a model for enhancing developmental-behavioral diagnosis through understanding this spectrum and continuum via analysis of 3 fundamental neurodevelopmental processes: delay, dissociation, and deviance. *Developmental delay* refers to a significant lag in the attainment of developmental milestones. As reviewed above, given that the etiologic entities that result in developmental disorders tend to produce diffuse neurologic dysfunction, developmental delay is most commonly represented by a more global delay across streams of development. *Developmental dissociation* describes a significant difference between the developmental rates of 2 streams of development, with one stream significantly more delayed. Developmental dissociation thus describes significant scatter or unevenness across different domains of development. Given that developmental delays are typically more global in nature, dissociations between developmental domains are less commonly encountered and should be recognized as atypical, even in a setting where dissociation exists without corresponding significant developmental delay. For example, specific LDs represent an example of developmental dissociation. In most states, specific LDs have been defined by a significant discrepancy, or dissociation, between intelligence and academic achievement. Learning disabilities may also be defined by a significant dissociation between verbal and nonverbal cognitive abilities—in language-based LDs (LLDs), a significant dissociation exists between relatively stronger nonverbal, visual problem-solving skills and relatively weaker language skills, while in nonverbal-based LDs (NVLDs), there exists a significant dissociation between relatively weaker nonverbal versus relatively stronger verbal cognitive abilities. *Developmental deviance or deviation* represents nonsequential unevenness in the achievement of milestones (ie, achieving more difficult developmental milestones in a typical developmental sequence prior to

achieving more simple milestones)—such deviant acquisition of developmental milestones would be considered atypical at any age.[13] While developmental dissociation refers to significant scatter or unevenness of developmental abilities *across* different developmental streams, developmental deviance is typified by significant scatter or unevenness of abilities *within* a single developmental stream. For example, children with autism may exhibit a 50-word vocabulary (expected at 24 months) prior to using a specific "mama" and "dada" to refer to their parents (expected at 10 months),[14] and many persist in using echolalia and confusing pronouns (atypical after 30 months), despite having higher age-level single-word vocabularies.

While every individual exhibits a unique pattern of developmental strengths and weaknesses the terms dissociation and deviance are applied to describe more significant scatter or unevenness of developmental abilities, both across and within developmental streams, than is typically encountered. This model postulates that developmental delay, dissociation, and deviance reflect underlying central nervous system (CNS) dysfunction. Consequently, the amount of delay, dissociation, and/or deviance encountered increases as one moves from the mild to severe end of the spectrum and continuum of developmental-behavioral disorders. Increasing levels of developmental delay, dissociation, and deviance reflect increasingly atypical development, and increasingly atypical development is typically accompanied by increasingly atypical behavior.

Capute's Triangle: Spectrum and Continuum

Capute and Accardo[13,15,16] have described a model for making developmental diagnoses that distills the highly complex functions of the brain into 3 primary developmental streams that provide 3 sides of a triangle that represents the spectrum and continuum of developmental-behavioral disorders (Figure 8.1). In an adaptation of this model, this chapter will review the spectrum of developmental-behavioral disorders within 3 primary developmental streams—motor, cognitive, and social-behavioral—as well as the continuum of developmental-behavioral disorders across these streams.

Developmental-behavioral disorders exist across a spectrum of severity, from mild to severe, ***within*** each of these developmental streams, with mild disorders predominating over severe disorders. There is also a continuum of developmental-behavioral disorders ***across*** these 3 developmental streams, as primary developmental-behavioral diagnoses in one stream are generally accompanied by associated deficits or comorbidities in one or both of the other developmental streams. In this model, the presenting developmental or behavioral concern typically represents only the tip of the iceberg of more diffuse underlying developmental-behavioral difficulties or comorbidities.[13]

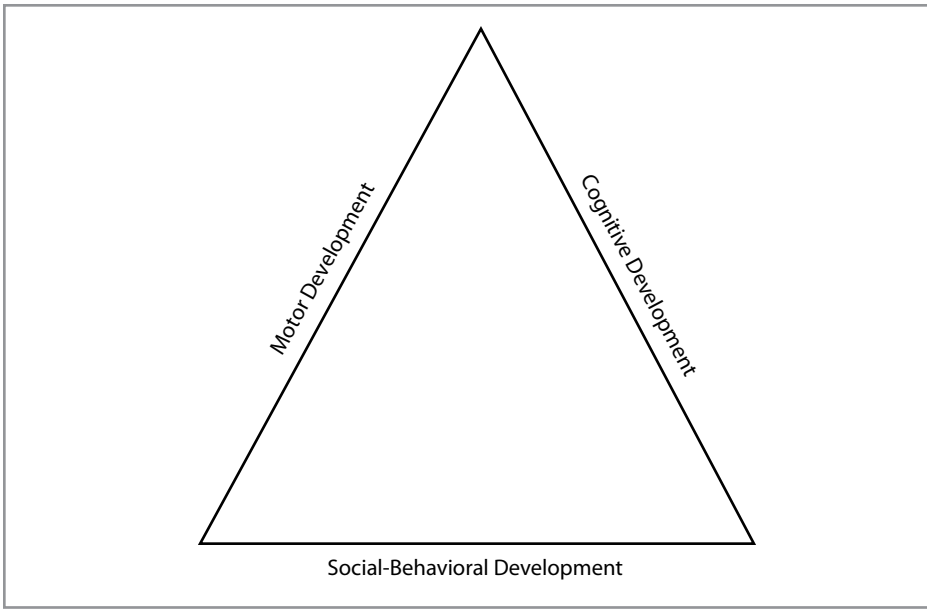

Figure 8.1. Capute's triangle. Adapted with permission from: Capute AJ. The expanded Strauss syndrome: MBD revisited. In: Accardo PJ, Blondis TA, Whitman BY, eds. *Attention Deficit Disorders and Hyperactivity.* New York, NY: Marcel Dekker, Inc.; 1991:28.

While the extreme ends of the spectrum and continuum of developmental-behavioral disorders are easily differentiated, the concepts of spectrum and continuum imply a lack of exact cutoff points and a blending in of diagnoses across the spectrum and continuum, with typically indistinct borders between specific developmental diagnoses. For example, in the spectrum of motor disability, it is easy to differentiate a mild developmental coordination disorder from severe CP, but it may be difficult to decide whether an individual with motor difficulties more in the middle of this spectrum is best described as evidencing a severe motor coordination disorder or mild CP. Similarly, in the continuum of developmental disabilities, it is easy to separate a child with language-based learning problems and associated ADHD from one with severe deficits in reciprocal social interaction and communication and repetitive and stereotypic behavior characteristic of autism, but it may be difficult to distinguish whether an individual in a more intermediate position in the spectrum/continuum has a severe language impairment with associated social-behavioral concerns versus a very mild PDD-NOS.[17,18]

An analysis of the degree of developmental delay, dissociation, and deviance that a child evidences based on the developmental history provided by his or her caretakers combined with the primary pediatric health care professional's direct neurodevelopmental examination of the child (see Chapter 7) directs the clinician in choosing the most appropriate developmental-behavioral diagnoses to apply to each child along

the spectrum and continuum described by this triangle. The remainder of this chapter will describe a framework for primary pediatric health care professionals to conceptualize the interrelationships between specific developmental-behavioral diagnoses. Of course, this model should not substitute for the use of currently accepted diagnostic criteria when a specific developmental-behavioral disorder is suspected. The chapters that follow (Motor Development, Cognitive Development, Social and Emotional Development, Autism Spectrum Disorders, Learning Disabilities, and ADHD) will specifically review current diagnostic criteria for each developmental-behavioral disorder. However, it is critical to acknowledge that viewing these entities in isolation may result in incomplete developmental-behavioral diagnoses and incomplete treatment. Thus it is hoped that exposure to the model to be described should improve primary pediatric health care professionals' clinical judgment in making comprehensive developmental-behavioral diagnoses.

The Spectrum of Motor Disorders

The motor stream of development includes the development of gross motor, fine motor, and oral motor (speech, chewing/swallowing) skills. There is a spectrum of motor disability within each of these motor areas, with mild motor disorders occurring more frequently than more severe disorders (Figure 8.2).

The mild end of the gross motor spectrum has in the past been described as clumsy child syndrome or developmental dyspraxia,[19] but it has been described in the *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV)* as DCD.[20] At this end of the spectrum, children evidence mild gross motor delays and difficulties with motor planning, and many evidence "soft" neurologic signs (such as synkinesis, choreiform movements, dysdiadochokinesis, and posturing of the upper extremities with stressed gaits).[21] As gross motor delay increases in severity within this spectrum, and is accompanied by "hard" neurologic signs (persistent primitive

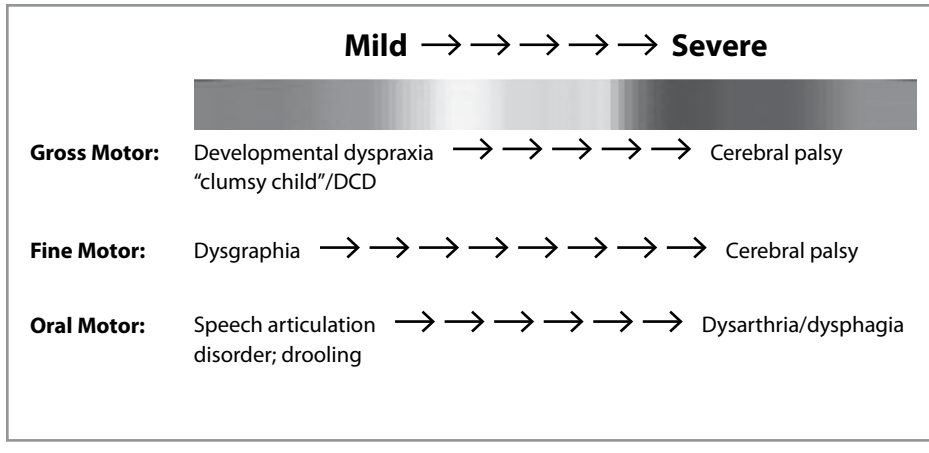

Figure 8.2. Spectrum of motor disorders. DCD indicates developmental coordination disorder.

reflexes, brisk deep tendon reflexes, spasticity, dyskinesia, ataxia, hypotonia), a diagnosis of CP becomes more appropriate. Minor fine motor deficits produce difficulties with handwriting (dysgraphia) and delays in accomplishing activities of daily living, such as buttoning and shoe-tying, while more severe fine motor delays are also seen in quadriplegic and hemiplegic forms of CP. From an oral-motor standpoint, the mild end of the spectrum includes milder feeding issues, drooling, and mild speech articulation disorders, while the severe end includes the dysarthria and dysphagia that frequently accompany more severe forms of CP. Consistent with disabilities within all streams of development, mild motor disabilities predominate over severe motor disabilities. There are many more children with gross motor clumsiness, handwriting difficulties, and speech articulation disorders (with an estimated prevalence of DCD of 10%)[19] than there are with CP (which has a prevalence of 3.6 per 1,000 or only 0.36%).[22]

The Spectrum of Cognitive Disorders

Global Cognitive Disorders

The spectrum of global cognitive disorders provides the most distinct diagnostic entities, as the diagnoses in this spectrum are defined by specific IQ and adaptive behavior scores in a normal distribution (Figure 8.3).

At the mild end of this spectrum, the Wechsler Intelligence Scale for Children-Revised (WISC-R) previously classified IQ scores between 80 and 89 as the "dull" range of intelligence.[23] Currently, while children who function in this range of intelligence are classified as "low average" by the WISC-IV,[24] they can more descriptively be referred to as "slower learners," as they typically experience significant difficulties keeping up in regular classroom settings.[25] As the global cognitive disorder becomes more severe across this spectrum, children with IQ scores between 70 and 79 are described as evidencing borderline intelligence, and those with IQ and adaptive behavior scores below 70 are defined as having intellectual disability. (See Chapter 10.) As with disorders across all developmental streams, mild global cognitive disorders predominate over severe disorders, given that 22% of the population evidences

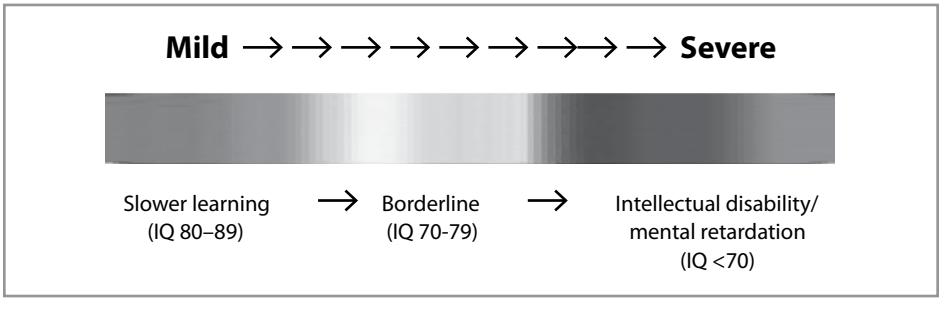

Figure 8.3. Spectrum of global cognitive disorders.

slower learning or borderline intelligence, while statistically only between 2% and 3% of the population would be expected to evidence intellectual disability, and only 1% of the population is actually identified as evidencing intellectual disability due to methodologic problems in identifying individuals with mild intellectual disability.[4] Within the spectrum of global cognitive disorders, there is no significant discrepancy or dissociation between cognitive domains. However, when there are discrepant and disproportionate delays in cognitive domains (language/communication versus nonverbal/visual problem-solving), there exist separate streams that describe a spectrum of dissociated language/communication and a spectrum of dissociated nonverbal/visual problem-solving disorders.

Dissociated Language/Communication Disorders

When children evidence discrepant and disproportionate delays in their language/communication development relative to their nonverbal/visual problem-solving development, their developmental diagnoses will lie within the spectrum of dissociated language/communication disorders (Figure 8.4). Of course, when a child presents with discrepant delays in their speech or language development, primary pediatric health care professionals need to first formally confirm the child's hearing status prior to making a diagnosis within this spectrum. As one moves from the mild to severe end of this spectrum, in addition to evidencing dissociated delays in language/communication relative to nonverbal problem-solving, increasing amounts of developmental deviance within the language/communication stream are observed.

At the mild end of this spectrum, the dissociation in language development involves a deficit in phonological processing, the primary neuropsychological deficit observed in dyslexia.[26] Children with phonological processing deficits have difficulty rhyming

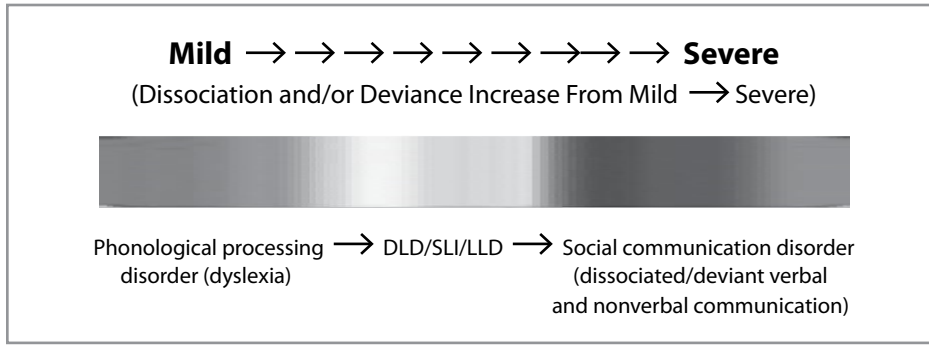

Figure 8.4. Spectrum of dissociated language/communication disorders. DLD indicates developmental language disorder; SLI, specific language impairment; LLD, language-based learning disability.

words, naming letters, and associating sounds with written symbols. They have difficulty with reading decoding, given their difficulty in sounding out words that they do not know, although they may be able to visually memorize lists of sight words. Most children with reading disabilities evidence underlying difficulties with phonological processing, although reading disorders are typically identified by schools based on a dissociation between IQ scores and discrepantly low achievement scores in reading. When comparing the mild end of the spectrum of global cognitive disorders (slow learning) with the mild end of the spectrum of dissociated language disorders (dyslexia), it is again confirmed that more diffuse or global cognitive dysfunction predominates over more focal or dissociated cognitive dysfunction, as more global slower learning affects 22% of the population, while discrepancy-based reading disabilities affect only 8%.[27]

As more diffuse receptive and expressive language processes become discrepantly delayed within this spectrum, an individual can be described as evidencing a developmental language disorder (DLD), which can also be described as developmental dysphasia or specific language impairment (SLI). When verbal reasoning is determined to be significantly below nonverbal reasoning on IQ testing, an individual can be described as evidencing an LLD. Rather than evidencing a relatively isolated difficulty in reading decoding as seen in dyslexia, individuals with SLIs or LLDs evidence more diffuse difficulties in listening comprehension, oral expression, reading comprehension, written expression, and math word problems.

As one moves toward the severe end of the spectrum of dissociated language/communication disorders, increasing developmental deviance, including deviant delays in both verbal and nonverbal aspects of communication and pragmatic language that negatively impact on social communication, is encountered. Thus individuals at the severe end of this spectrum can be described as evidencing language-based social communication disorders. In addition to evidencing discrepant and disproportionate delays in aspects of expressive and receptive language development relative to nonverbal visual problem-solving development, individuals with social communication disorders in this spectrum evidence significant deviance within the language/communication domain, including deviant delays in aspects of both nonverbal and pragmatic language (such as poor eye contact and lack of gestured language) as well as expressive and receptive language (such as persistent echolalia and pronoun confusion) despite upward scatter in other (usually more rote) aspects of their language functioning. When such dissociated language-based social communication difficulties occur in a setting of primary deficits in reciprocal social interaction, a diagnosis of autism needs to be suspected.[28] Confirming this spectrum concept, recent studies have shown a significant overlap and diagnostic confusion between individuals with SLIs versus those with the social communication disorders observed in autism.[17,18]

As with all other developmental streams, in the spectrum of dissociated language/communication disorders, mild disorders predominate over severe disorders. There are many more individuals with discrepant and disproportionate delays in language who do not evidence associated social communication deficits (dyslexia, SLI, LLDs—with discrepancy-based reading disabilities affecting 8% of children)[27] than there are individuals with the verbal and nonverbal social communication deficits seen in ASDs (which occur in only 0.91% of children).[29]

The Spectrum of Dissociated Nonverbal Disorders

When children evidence discrepant and disproportionate delays in their nonverbal/visual problem-solving development relative to their verbal/language development, their developmental diagnoses will lie within the spectrum of dissociated nonverbal disorders (Figure 8.5). Of course, when a child presents with discrepant delays in their visual problem-solving development, primary pediatric health care professionals need to first formally confirm the child's vision status prior to making a diagnosis within this spectrum. As one moves from the mild to severe end of this spectrum, in addition to evidencing dissociated delays in nonverbal/visual problem-solving abilities, increasing amounts of developmental deviance in the nonverbal domain are observed.

At the mild end of this spectrum, the dissociation in nonverbal/visual problem-solving development involves a relatively focal deficit in orthographic processing, which involves visual processing of letter identity and letter position within words.[30] Children with orthographic processing deficits have difficulty with visual-spatial feature analysis, which initially results in difficulty in remembering the shapes of letters and how to use the correct sequence of strokes to form letters, and goes on to result in letter reversals ("b" and "d") and reversals of whole words ("saw" for "was") in reading and writing.[31,32] Children with orthographic processing disorders have difficulty

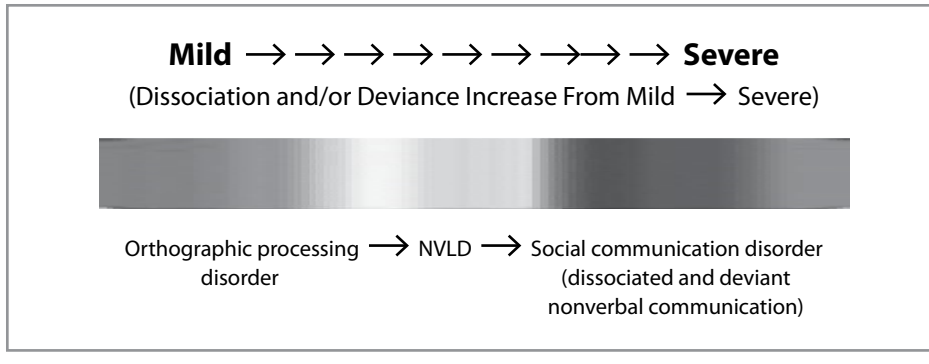

Figure 8.5. Spectrum of dissociated nonverbal disorders. NVLD indicates nonverbal learning disability.

with memorizing sight words, but they are able to sound out words that they do not know, and they can spell phonetically.

As more diffuse nonverbal/visual problem-solving processes become discrepantly delayed, an individual can be described as evidencing an NVLD, where typically verbal reasoning abilities significantly exceed nonverbal reasoning abilities on IQ testing. Rather than evidencing a relatively isolated difficulty in the orthographic processing of letters and words, individuals with NVLDs evidence more diffuse difficulties in visual-spatial, visual-perceptual, and visual-motor processing. Children with NVLDs have deficits in the discrimination and recognition of visual detail and in visual-spatial orientation, including evidencing difficulties in right-left orientation. These children also have difficulty understanding spatial relations, including appreciating appropriate boundaries of interpersonal space. From an academic standpoint, children with NVLDs have difficulties with the visual-spatial and conceptual aspects of mathematics (aligning columns for multidigit calculations, geometry), and their visual-motor deficits negatively impact on their writing.[32]

As one moves toward the severe end of the spectrum of dissociated nonverbal disability, increasing developmental deviance, including deviant delays in nonverbal aspects of communication, is encountered. Thus individuals at the severe end of this spectrum, despite their significant relative strengths in verbal ability and verbal reasoning, can also be described as evidencing social communication disorders as a result of their deviant delays in nonverbal aspects of pragmatic social communication. These nonverbal social communication deficits include poor eye contact, difficulty understanding and using gesture and facial expression, and difficulty visually "reading" social situations. Given their significant relative strengths in verbal reasoning and their lack of delays in structural verbal language, individuals with nonverbally based social communication disorders often have much to say, but they do not understand how to use their language to communicate effectively in social settings. These individuals experience difficulty with turn taking, maintaining topics of conversation, and understanding figurative language. They also tend to have one-sided conversing styles and to verbally perseverate on restricted interests. When such social communication difficulties occur in a setting of primary deficits in reciprocal social interaction, a diagnosis of Asperger syndrome needs to be suspected.

As with all other developmental streams, in the spectrum of dissociated nonverbal disability, mild disabilities predominate over severe disabilities. There are many more individuals with orthographic processing-based LDs who do not evidence associated social communication deficits than there are individuals with the nonverbal social communication deficits seen in more severe NVLDs or Asperger syndrome.

The Spectrum of Social-Behavioral Disorders

It is essential to appreciate the wide behavioral variation of typically developing children and the importance of always interpreting behavior within the context of a child's underlying developmental level and family/social/cultural environment. In the spectrum of social-behavioral development, the most appropriate diagnosis to apply to a child's behavioral difficulties begins with an analysis of the child's temperament. Temperament is considered to be an innate attribute that describes a child's behavioral style across 9 dimensions: quality of mood, attention span/persistence, distractibility, activity level, sensory threshold, intensity of reaction, adaptability, rhythmicity/regularity, and approach/withdrawal.[33] Normal variations in these temperamental domains often lead to behavioral problems secondary to a mismatch between the temperamental characteristics of a child and his or her caretakers. Further, given the wide range of behavioral variation in typically developing children, in the spectrum of social-behavioral disorders, it is often difficult to separate those with difficult temperaments from those with mild social-behavioral disorders.

Similar to the spectra of motor and cognitive disorders described previously, a spectrum of social-behavioral difficulties from mild to severe can also be conceptualized. In terms of social behavior (Figure 8.6), the shy child or child with a slow to warm up temperament might be considered to occupy the mild end of a spectrum of concerns about social development. At the mild end of this spectrum, the shy child with a slow to warm up temperament can be considered to be exhibiting a normal behavioral variation or possibly a behavioral problem that does not result in functional impairment. However, further along this spectrum, social-behavioral difficulties would be encountered that begin to result in some functional impairment, such as socially inappropriate behaviors that interfere with peer interaction (problems with turn taking, initiating and responding to interactions appropriately, aggression). Significant social anxiety could also be considered to occupy an intermediate position along this spectrum. At the severe end of this spectrum are the severely disabling deficits in reciprocal social interaction, joint attention, imaginative play, and empathy that are characteristic of ASDs (see Chapter 13).

Another example of the spectrum of social-behavioral disorders involves the related temperamental domains of attention span/persistence, distractibility, activity level, and sensory threshold. If a child presents to their primary pediatric health care professional with difficulties in these domains that are not resulting in functional impairment, the *DSM* for primary care suggests diagnoses such as "inattention variation" or "hyperactivity-impulsivity variation" and "inattention problem" or "hyperactivity-impulsivity problem."[34] However, further along this spectrum, when difficulties with inattention, poor impulse control, and excessive motor activity are present to a disabling and developmentally inappropriate degree that are causing significant functional impairment across multiple settings, then as per the *DSM-IV-R,* a diagnosis of ADHD should be considered (see Chapter 16). As one moves further

across this spectrum, in the place of the characteristic inattention and easy distractibility seen in ADHD, a child at the severe end of this spectrum evidences more significantly developmentally deviant and atypical attention—his or her short attention span may be present to such a degree that eye contact is difficult to maintain, while at the same time, the child becomes overfocused and either visually or verbally perseverative on restricted interests or repetitive behaviors.[16] This significantly atypical attention may also be manifested by overfocus or underfocus on sensory stimuli, such as evidencing a high pain threshold or intolerance to certain noises, tight-fitting clothes, or food textures. Rather than exclusively evidencing the excessive motor activity seen in ADHD, a child at the severe end of this spectrum may also evidence more atypical and developmentally deviant stereotypic motor activity, such as hand flapping, body rocking, or spinning (Figure 8.6).

As will be reviewed below in the section on the continuum of developmental-behavioral disorders, it is important to note that these more atypical behavioral manifestations of the spectrum of social-behavioral disorders are characteristically associated with the more atypical manifestations of the spectrum of cognitive disorders. Given that increasing developmental delay, dissociation, and deviance within the cognitive stream signal increasingly atypical information processing compared with those without developmental delay, dissociation, and deviance, it should be expected that the more atypically an individual processes information, the more likely that individual's behavior will be similarly atypical. Thus the repetitive and stereotyped behavior observed at the severe end of this spectrum is characteristically

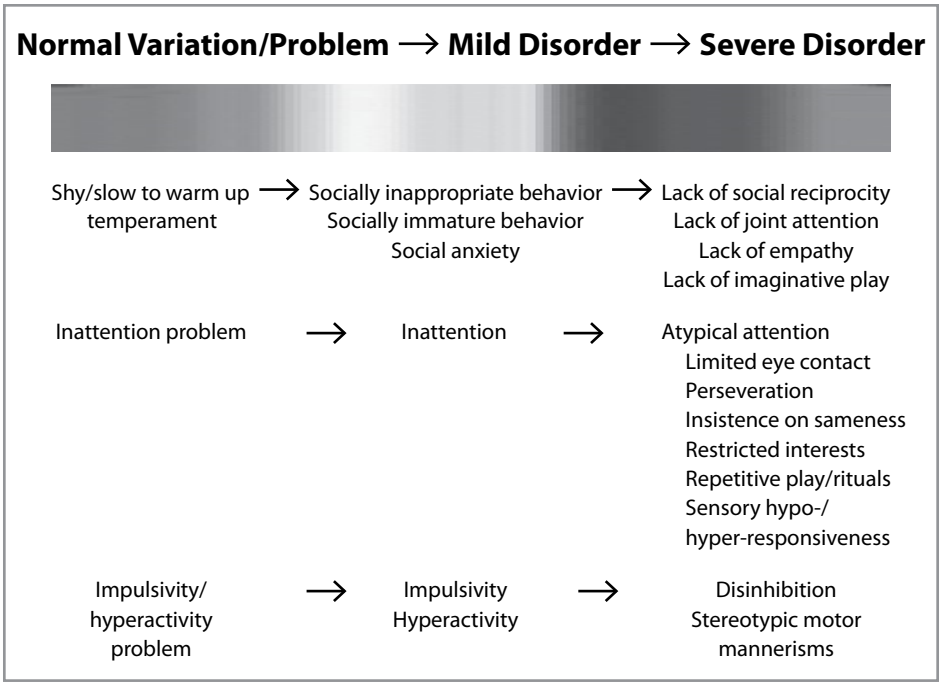

Figure 8.6. Spectrum of social-behavioral disorders.

observed in individuals with either severe to profound intellectual disability (significant delay) or social communication disorders (significant dissociation and deviance). Individuals with ASDs (autism, Asperger syndrome, PDD-NOS) evidence a primary deficit in reciprocal social interaction. However, these primary social deficits are typically accompanied by deficits in communication and the restricted interests and more atypical behaviors described at the severe end of this behavioral spectrum. This association of deficits across social, communicative, and behavioral streams of development observed in ASDs clearly illustrates the continuum of developmental-behavioral disorders.

The Continuum of Developmental-Behavioral Disorders

As reviewed previously, it is clear that diffuse developmental-behavioral dysfunction is more common than focal dysfunction among children with developmental-behavioral disorders—associated deficits or comorbidities are the rule rather than the exception. Thus, in addition to a spectrum of disorders from mild to severe within each stream of development, there also exists a continuum of disorders from mild to severe across developmental streams. The primary developmental-behavioral diagnosis within a single stream of development applied to a child's unique pattern of developmental delay, dissociation, and/or deviance is rarely unaccompanied by secondary diagnoses.

The continuum of developmental-behavioral disorders is most easily conceptualized by the continuum across motor, global cognitive, and behavioral streams of development at the severe end of each of these spectra (Figure 8.7). There is a significant continuum or overlap across these streams, as approximately 50% of individuals with

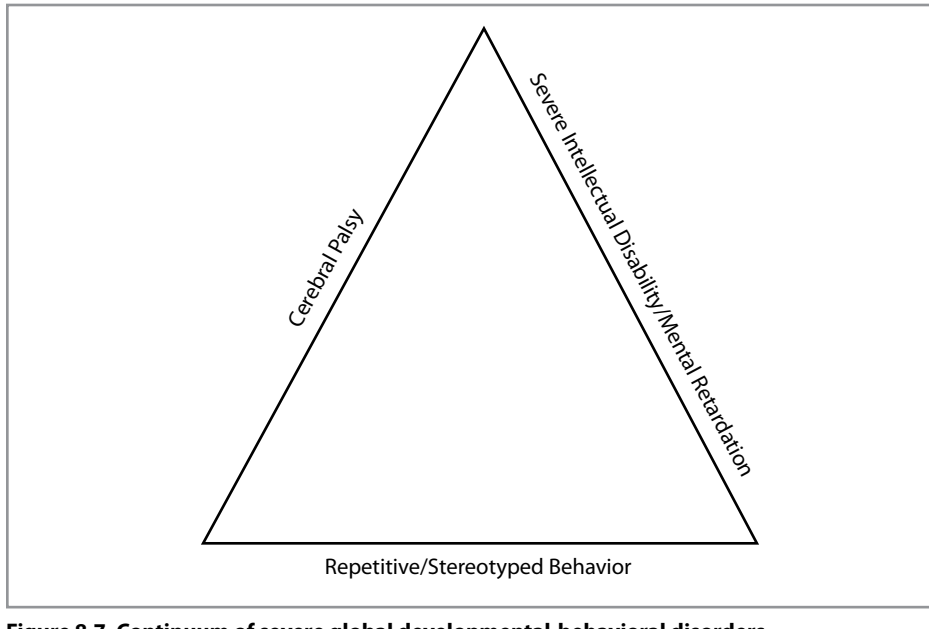

Figure 8.7. Continuum of severe global developmental-behavioral disorders.

CP also have intellectual disability, about 25% of individuals with intellectual disability also have CP (versus 3.6/1,000 in the general population), and most individuals with severe to profound intellectual disability evidence repetitive, perseverative, and stereotypic behaviors.[7,9,22] In addition, those individuals with CP who do not have intellectual disability are at higher risk than those without CP for other cognitive and behavioral difficulties, including slower learning, LD, and ADHD.[35]

There is also a continuum of disability across motor, dissociated cognitive, and social-behavioral streams of development at both the mild and severe ends of each of these spectra (Figure 8.8). Although an outdated term that has been generally discredited, the mild end of this continuum was previously described as representing *minimal brain dysfunction*.[36] This term implies mild but diffuse developmental dysfunction across developmental streams.[37] Epidemiologic research has supported this concept of mild but diffuse dysfunction as, for example, children with ADHD are much more likely than children without ADHD to have associated LDs, gross motor clumsiness, handwriting difficulties, and speech articulation deficits.[38] Certainly, not every individual with ADHD evidences all of these comorbidities, but it is rare to identify an individual with ADHD who does not evidence some comorbid developmental or behavioral diagnosis.

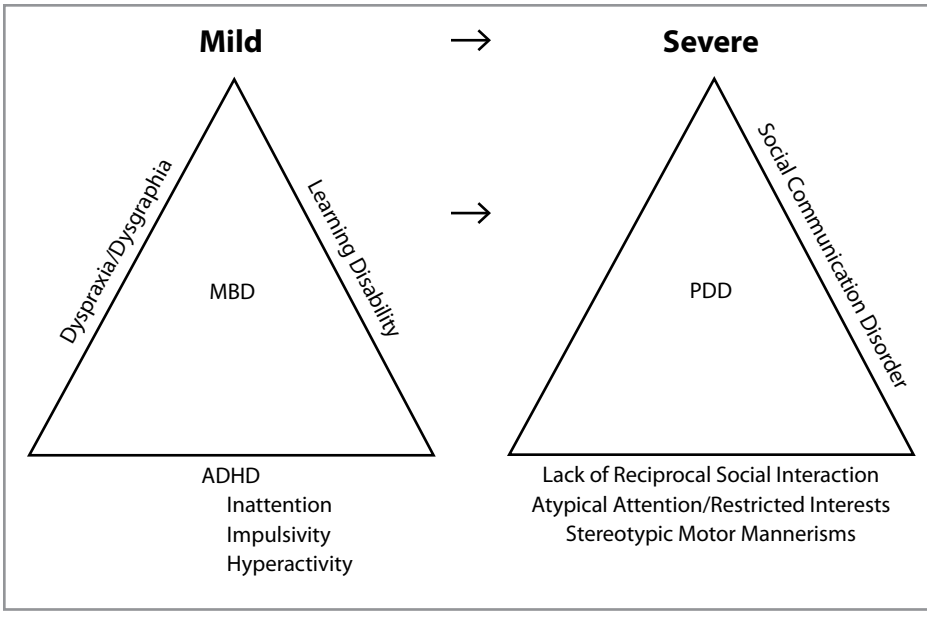

Figure 8.8. Continuum of dissociated developmental-behavioral disorders: mild to severe.
MBD indicates minimal brain dysfunction; PDD, pervasive developmental disorder.

At the severe end of the continuum of dissociated developmental-behavioral disorders lies what the *DSM* has termed the PDDs.[20] While these disorders share a pervasive primary deficit in social interaction, this term may be considered somewhat misleading, as the cognitive component of these disorders typically involves scattered, uneven, discrepant, dissociated, and deviant cognitive profiles, rather than pervasive cognitive delays across cognitive domains.[39,40] The pervasive developmental disorders, as defined in the *DSM-IV,* include the diagnoses of autism, Asperger syndrome, and PDD-NOS, which is a diagnosis to be used when an individual evidences subthreshold symptoms of one of the other disorders (see Chapter 13). All of the PDDs are characterized by deficits in reciprocal social interaction; however, within the continuum of developmental-behavioral disorders, in addition to their primary social deficits, individuals with PDDs exhibit associated deficits in communication at the severe end of the spectrum of dissociated and deviant cognitive disorders and tend to exhibit a repertoire of atypical behaviors described at the severe end of the spectrum of social-behavioral disorders (Figure 8.8).

Rather than relying exclusively on behavioral checklists and other screening instruments, primary pediatric health care professionals may develop improved clinical judgment in making diagnoses of autism and Asperger syndrome by adopting the model described throughout this chapter. Rather than existing without explanation in a "black box" of atypical behaviors, the behavior of children with autism and Asperger syndrome may be conceptualized through analysis of their characteristic developmental-behavioral profiles. In a continuum across cognitive and social-behavioral streams of development, the more delayed, dissociated, and/or deviant the cognitive development, the more atypical the information processing compared with children without delays, dissociation, or deviance. The more atypical the information processing, the more atypical the behavior should be expected to be.

In this model, in addition to their primary deficits in reciprocal social interaction, children with autism evidence the social communication disorder that lies at the severe end of the spectrum of dissociated language/communication disorders. Children with autism typically demonstrate discrepant and disproportionate delays and significant developmental deviance in their language/communication development relative to strengths in some aspects of their nonverbal/visual problem-solving development.[6,28,39–43] Thus their language/communication development is dissociated from their nonverbal development. They also evidence developmental deviance in their acquisition of language/communication skills, such as evidencing persistent echolalia and pronoun confusion when other aspects of their language development extend beyond a 30-month level (when most typically developing children are no longer using echolalia or confusing pronouns). Cognitive profiles with significant developmental dissociation and deviance (as markers of underlying CNS dysfunction) occur less frequently than more evenly developed cognitive profiles, and thus can be considered atypical compared with the norm. Thus, in addition to their primary deficit in reciprocal social interaction, children with autism can be considered

to evidence atypical patterns of cognitive development compared with those who evidence a more typically even and less scattered cognitive profile.[39] If their pattern of cognitive development is considered atypical, then they can be conceptualized as processing information in an atypical way, and if processing information in an atypical way, it should be expected that their behavior would be similarly atypical. The atypical behavioral pattern observed in children with autism may thus be conceptualized as being at least partially derivative of the discrepancy between how children with social communication disorders at the severe end of a spectrum of dissociated and deviant language/communication disorders process information visually versus verbally. As language is a primary medium for social interaction, it should be expected that children with dissociated delays and deviance in language understanding and pragmatic language usage would manifest a lack of interest in, and avoidance of, language confrontational social situations.[40] Alternatively, given their relative strengths in aspects of visual problem-solving, it follows that children with this type of dissociated cognitive profile would be attracted to more visually stimulating activities, such as lining, sorting, spinning, turning lights on and off, and opening and closing doors. Their strengths in aspects of visual problem-solving might also be expressed through abilities in art, direction sense (ie, noticing changes in usual car routes), and visual memory. Their stronger abilities in aspects of visual problem-solving might also impart a comfort with visual sameness that might underlie the ritualistic features, restricted interests, and need for routine that are often observed in children with autism. The developmentally dissociated and deviant cognitive pattern observed in children with autism may also contribute to conceptualizing the differential auditory and visual attention spans that are characteristically observed. Children with autism typically evidence inconsistent alerting to verbal and auditory stimuli and abbreviated eye contact in language confrontational social situations. At the same time, given strengths in aspects of their visual problem-solving, children with autism often evidence stereotypic perseveration or overfocus on a restricted number of visually stimulating activities, such as lining, sorting, or spinning.

It is important to note that most individuals with autism evidence a language-based social communication disorder in a setting of overall intellectual disability.[10] This increased prevalence of intellectual disability in association with autism is in harmony with this model, as developmental delay, dissociation, and deviance reflect underlying CNS dysfunction—it should thus be expected that dissociation and deviance (markers of increasing CNS dysfunction and characteristic of autism) should occur more commonly in a setting of increasing overall delay (intellectual disability). While their overall cognitive abilities may lie in the range of intellectual disability, these individuals still evidence a primary deficit in reciprocal social interaction and discrepant and disproportionate delays and developmental deviance in their language development relative to aspects of their visual problem-solving development. It has also been reported that greater than 75% of individuals with severe to profound intellectual disability may be categorized as autistic.[1,44] However, it is

important to note that repetitive and stereotyped behaviors are frequently observed in children with severe to profound intellectual disability, even if they do not have autism. A diagnosis of autism should be added only if, in a setting of severe to profound intellectual disability, the child evidences deficits in reciprocal social interaction and discrepant and disproportionate delays and developmental deviance in social communication.

Individuals with high-functioning autism evidence a primary deficit in reciprocal social interaction and a dissociated and developmentally deviant cognitive profile consistent with a language-based social communication disorder, but they do not evidence overall cognitive abilities in the range of intellectual disability. There has been controversy about whether high-functioning autism, defined as autism that occurs in a setting without associated intellectual disability, and Asperger syndrome represent the same or different entities. While both individuals with autism and Asperger syndrome evidence deficits in reciprocal social interaction and weaknesses in nonverbal aspects of social communication, those with autism do so in association with more global structural verbal/language difficulties and tend to exhibit relative strengths in aspects of their nonverbal/visual problem-solving development and engage in visually perseverative behaviors.[6,39–43] In contrast, individuals with Asperger syndrome tend to evidence strengths in their structural verbal/language development (and engage in verbally perseverative behaviors) and weaknesses in nonverbal aspects of social communication typically in association with more global nonverbal difficulties.[45,46] In this model, while both autism and Asperger syndrome share a primary deficit in reciprocal social interaction, the cognitive deficit in autism is a social communication disorder at the severe end of a spectrum of dissociated and deviant language disorders, while the cognitive deficit in Asperger syndrome is a social communication disorder at the severe end of a spectrum of dissociated and deviant nonverbal disorders. Thus, in addition to primary deficits in reciprocal social interaction, both children with autism and Asperger syndrome evidence atypical, scattered, and uneven dissociated and deviant cognitive profiles that should be expected to be associated with atypical behavioral manifestations. However, children with autism typically evidence relative strengths in aspects of their nonverbal development and relative weaknesses in their verbal/language development, while children with Asperger syndrome typically evidence the opposite discrepancy—they evidence strengths in their verbal development and weaknesses in their nonverbal communication, typically in association with other nonverbal learning deficits.

Studies have confirmed that most individuals with Asperger syndrome evidence a nonverbal learning disabled cognitive profile.[45,46] Thus, in addition to their primary deficit in reciprocal social interaction, as a result of their significantly dissociated and deviant cognitive profiles, which are less common than more evenly developed profiles, children with Asperger syndrome can be conceptualized as processing information in an atypical way compared with the general population. Given their atypical information processing, it makes sense that children with Asperger

syndrome should evidence atypical behavior. Given their relative verbal strengths, it is understandable that children with Asperger syndrome tend to perseverate verbally, engaging in pedantic, one-sided conversations that tend to list factual information about restricted interests, such as dinosaurs, train schedules, baseball statistics, or the weather. Despite their verbal strengths, but given their nonverbal weaknesses, in addition to the cognitive and academic difficulties seen in children with NVLDs, individuals with Asperger syndrome evidence deficits in nonverbal aspects of communication, including having difficulties with eye contact, understanding body language (gestures and facial expressions), and "reading" social situations.

Another way to appreciate the continuum of developmental-behavioral disorders and the difference between diagnoses of autism and Asperger syndrome has been suggested by Bishop.[47] In Bishop's model, 3 intersecting concentric circles represent language structure (form and content), social communication (pragmatic language use), and stereotyped/repetitive behavior (Figure 8.9). Autism represents the area that encompasses the overlap of all 3 of these circles. In this model, autism represents a more diffuse disturbance than Asperger syndrome, involving deficits in structural language, verbal and nonverbal social communication, and associated stereotyped/repetitive behaviors. Asperger syndrome represents the area of overlap

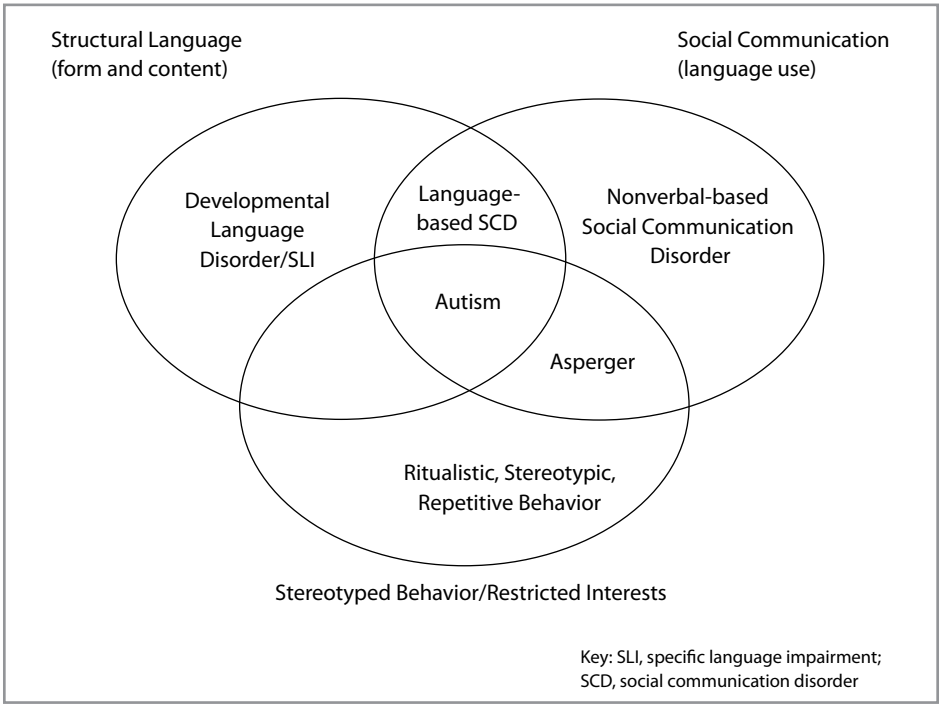

Figure 8.9. Autism spectrum disorders. SLI indicates specific language impairment; SCD, social communication disorder. Adapted from: Bishop DVM. What's so special about Asperger syndrome? The need for further exploration of the borderlands of autism. In: Klin A, Volkmar FR, Sparrow SS, eds. *Asperger Syndrome.* New York, NY: The Guildford Press; 2000: 273. With permission from The Guilford Press.

of the social communication and repetitive/stereotyped behavior circles. In this model, individuals with Asperger syndrome lack the deficits in language structure observed in children with autism. Thus, in this model, all individuals with autism evidence developmental dysphasia or DLDs, while all individuals with Asperger syndrome evidence deficits in nonverbal social communication but lack the dissociated structural language deficits observed in autism. A recent study provides support for both the Capute and Bishop models, as children with Asperger syndrome could be differentiated from children with high-functioning autism based on a lack of dissociated deficits in structural language development.[48]

In Bishop's model, the area of overlap in Figure 8.9 between the structural language and social communication circles, but lacking any overlap with the repetitive/stereotypic behavior circle, corresponds with the diagnosis of a social communication disorder that lies at the severe end of the spectrum of dissociated language/communication development in the Capute model. This region might alternatively be described as representing a semantic-pragmatic language disorder.[28,47] Individuals in this region evidence difficulties with both comprehension of language content and pragmatic language use, but no repetitive/stereotyped behavior. The area of the social communication circle that does not overlap with the other circles in the Bishop model corresponds to the social communication disorder that lies at the severe end of the spectrum of dissociated nonverbal development in the Capute model. Individuals in this region evidence difficulties with nonverbal aspects of pragmatic language, but they do not evidence any other deficits in language and in fact evidence a pattern of verbal strengths relative to nonverbal weaknesses. The area of the structural language circle that has no overlap represents the diagnosis of DLD/developmental dysphasia/SLI. Individuals in this region have difficulties with receptive and expressive language but no associated social deficits or repetitive/stereotypic behaviors.

A recent study provides support for the concept of a continuum of language/communication and social-behavioral disorders.[18] This study compared scores on diagnostic instruments for autism in children with autism versus those with SLIs. The key finding from this study was that the investigators found a continuum of pathology from isolated structural language abnormalities (as seen in individuals with SLIs), to individuals with SLIs with both structural and social abnormalities, to individuals with autism with both pragmatic impairment and language abnormalities.[18]

When reviewing the continuum of developmental-behavioral disorders, it is important to remember that diagnoses at the mild end of the continuum occur more commonly than those at the more severe end. For example, while LDs and ADHD occur in approximately 7% to 8% of the population, ASDs occur in only 0.91% of the population.[27,29,49] In addition, at the severe end of the continuum of developmental-behavioral disorders, epidemiologic studies have also confirmed that the more global cognitive dysfunction evidenced in intellectual disability is more common than the

dissociated and deviant cognitive dysfunction evidenced in the social communication disorders that accompany autism and Asperger syndrome. While statistically, based on the normal curve of IQ scores, the more global cognitive dysfunction seen in intellectual disability should occur in 2 to 3 per 100 individuals, the more dissociated and deviant cognitive dysfunction observed in ASDs occurs in only 1 per 110 individuals.[4,29]

Conclusion

Developmental-behavioral disorders are the most prevalent chronic conditions encountered by primary pediatric health care professionals. Unfortunately, there is a dearth of pediatric subspecialists in the overlapping fields of neurodevelopmental disabilities and developmental-behavioral pediatrics to which professionals can refer their patients, and long waiting lists at tertiary care developmental evaluation centers. Thus most children with developmental-behavioral disorders need to be identified and longitudinally managed by their primary pediatric health care professionals within the medical home. The AAP has recommended the use of standardized developmental screening tests at specified ages,[3] but it is important for primary pediatric health care professionals to know that while low-prevalence, high-morbidity developmental-behavioral problems should be identified by early screening, much more prevalent mild developmental-behavioral problems are still likely to be missed by screening at younger ages, even when using the best screening instruments available. In addition, while standardized screening and assessment instruments certainly facilitate making accurate developmental-behavioral diagnoses, expert panels continue to endorse that the clinical judgment of an experienced clinician is the gold standard for making developmental-behavioral diagnoses.[6] Given the prevalence of these conditions in primary care practice, primary pediatric health care professionals are ideally positioned to serve as such experienced clinicians. Certainly, in clinical practice, not every child with a developmental-behavioral concern always fits neatly into the diagnostic categories described in this chapter.

Box 8.1. Key Principles Underlying Developmental-Behavioral Diagnosis

- 3 primary streams of development
  - Motor
  - Cognitive (including language and nonverbal processing)
  - Social/behavioral
- Delay, dissociation, and deviance reflect underlying central nervous system dysfunction.
  - The more delayed, dissociated, and deviant the development, the more atypical the behavior should be expected to be.
- There is a spectrum of disorders within each developmental stream.
  - Mild disorders predominate over severe disorders within each stream.
- There is a continuum of developmental-behavioral disorders across streams.
  - More diffuse/global developmental-behavioral dysfunction predominates over more isolated/focal dysfunction (comorbidities are the rule rather than the exception).

However, it is the goal of this chapter that a thorough understanding of the key principles underlying the entire spectrum and continuum of developmental-behavioral diagnoses (Box 8.1) should enhance clinical judgment and improve the confidence and diagnostic acumen of clinicians in identifying children with developmental-behavioral disorders within the medical home at the earliest possible age.

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Chapter 9

Motor Development

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Concerns about the development of infants and young children fall into 3 broad areas: cognitive or intellectual problems (including speech and language), neuromotor and orthopedic concerns, and behavioral issues. This chapter focuses on the motor aspects of infant and child development.

The acquisition of motor milestones occurs in a predictable sequence with skills accomplished during an expected age range. Earlier motor milestones generally make up an infant's first developmental achievements and, as such, missed milestones may be the first indication of significant delays. The astute clinician will recognize that the presenting complaint of delayed motor skills could indicate a specific motor control problem or may be a symptom of a global disorder, such as central hypotonia, progressive neurologic disorder, or severe cognitive deficit.[1]

The ability to make a diagnosis of motor impairment depends in large measure on the type of disability. A diagnosis of spina bifida or club feet is apparent at birth. In the case of cerebral palsy (CP), diagnosis is delayed until a problem with motor development is recognized and investigated. The child with severe impairments is likely to be identified earlier (usually before 12 months of life) than the child with mild delays because the former will more obviously fail to achieve developmental milestones and will typically demonstrate pathologic neurologic signs. The key, then, to early identification lies with the primary pediatric health care professional's knowledge of normal developmental milestones and clinical comfort with the pediatric neurodevelopmental examination[1,2] (see Chapter 7). Because early indicators of motor delay may be ambiguous and early intervention is ideal, as described in detail in Chapter 6, the American Academy of Pediatrics (AAP) recommends that primary pediatric health care professionals provide longitudinal surveillance at each well-child encounter, specifically eliciting parent concerns and focusing on interval developmental history. In addition, formal developmental screening with a standardized tool should be administered at the 9-, 18-, and 24- or 30-month health maintenance visits to identify children at risk for developmental disorders, including disorders of motor development.

The natural variation of human development puts forth the confounding factor that even typically developing children will acquire motor milestones at different ages. This chapter will assist the primary pediatric health care professional in identifying and evaluating children who present with delayed motor development and will primarily address 3 topics

- Motor development and the clinical neuromotor assessment
- Static neuromotor encephalopathy (CP)
- Congenital neuromotor dysfunction due to spinal cord anomaly (spina bifida)

Motor Evaluation of the Infant

To make a meaningful statement about an infant's motor competence, the primary pediatric health care professional should organize data gathered from the history, physical examination, and neurodevelopmental examination according to the following schema: (1) motor developmental milestones, (2) the classic neurologic examination, and (3) markers of cerebral neuromotor maturation (primitive reflexes and postural reactions).[2]

Motor milestones are extracted from the developmental history as well as from observations during the neurodevelopmental examination. A reference table of sequential gross and fine motor milestones is needed (Tables 9.1 and 9.2). Milestone assessment is best summarized as a single (or narrow) motor age for the child. This approach eliminates the distracting practice of examining every milestone for its own unique "range of normal," and it also allows one to think of the child in terms of his or her level of motor function. The motor age can be converted to a motor quotient (MQ = [motor performance age/chronologic age] x 100), giving a simple expression of deviation from the norm. It is also convenient to adjust for other factors (eg, prematurity) at this point. An MQ above 80 is considered within normal limits, while an MQ below 70 is abnormal. Those falling in the 70 to 80 range are borderline but most likely within normal limits. The upper limit of normal is not of particular value except for the pleasure of identifying above-average performers. Whether truly gifted athletes can be recognized early by this method is thought-provoking but speculative at present.[3]

EXAMPLE

A 12-month-old boy is seen for well-child care. He is not walking alone, but he pulls up to stand (9 months), cruises around furniture (10 months), and will walk fairly well when held by both hands (10 months). This child has a gross motor age of 10 months at a chronologic age of 12 months. From a motor standpoint, he is behaving like an average 10-month-old. Is this 2-month discrepancy something to be concerned about, or is he "within normal limits?" (Mother may already be concerned, especially if her neighbor's baby is already walking!) To decide, one must proceed to calculate an MQ.

Motor age = 10 months
Chronologic age = 12 months
MQ = [10 months/12 months] x 100 = 83

Table 9.1. Gross Motor Development

Timetable	
Prone	
Head up	1 mo
Chest up	2 mo
Up on elbows	3 mo
Up on hands	4 mo
Up on hands and knees	7 mo
Crawl	8 mo
Rolling	
Front to back	3–5 mo
Back to front	4–5 mo
Sitting	
Sit with support ("tripod" sitting)	5 mo
Sit without support	7 mo
Get up to sit (unassisted)	8 mo
Walking	
Pull to stand	8–9 mo
Cruise	9–10 mo
Walk with 2 hands held	10 mo
Walk with 1 hand held	11 mo
Walk alone	12 mo
Run (stiff-legged)	15 mo
Walk up stairs (with rail)	21 mo
Jump in place	24 mo
Pedal tricycle	30 mo
Walk down stairs, alternating feet	3 y

Table 9.2. Fine Motor Development

Timetable	
Opens fists	3 mo
Brings objects to midline	4 mo
Transfers objects	5 mo
Reaches unilaterally and grasps	6 mo
Pincer grasp (mature)	10–11 mo
Releases voluntarily	12 mo
Displays handedness	24 mo

The motor age and the MQ are each good summary descriptors of the child and have more meaning than trying to plot each individual milestone. Since the lower limit is 70 to 80, this boy's motor development is within normal limits.

Motor milestones do not take into account the *quality* of a child's movement. The motor portion of the neurologic examination, including observations of station and gait, take qualitative features into better account. The neurologic assessments of tone, strength, deep tendon reflexes, and coordination are difficult in the infant because of their subjective nature compounded by the limited ability for cooperation. Clinical experience is essential to gaining accurate and useful information.[4] The Early Motor Pattern Profile is a useful tool, developed for use by primary pediatric health care professionals, that assigns scores for abnormalities in tone and primitive reflexes.[5] It serves to minimize the objectivity inherent in early neurodevelopmental evaluations and facilitate the early detection of CP.

The elicitation of reflexes requires patience and repeated, yet gentle, trial and error (one is most often swinging at a small moving target). Muscle tone (passive resistance) and strength (active resistance) are a challenge to distinguish in the contrary subject. The best clues often come from *observation,* not handling. Spontaneous or prompted motor activities (eg, weight-bearing in sitting or standing) require adequate strength. Weakness may be best appreciated from observing the quality of stationary posture and transition movements. The Gower's sign (arising from sitting on the floor to standing using the hands to "walk up" one's legs) is a classic example. The maneuver is indicative of pelvic girdle and quadriceps muscular weakness. Spontaneous postures (eg, frog-legs, scissoring—Figure 9.1) provide visual clues to hypotonia/weakness and spastic hypertonus, respectively. After 2 to 3 years of age, the neurologic examination becomes easier and more meaningful as cooperation improves.

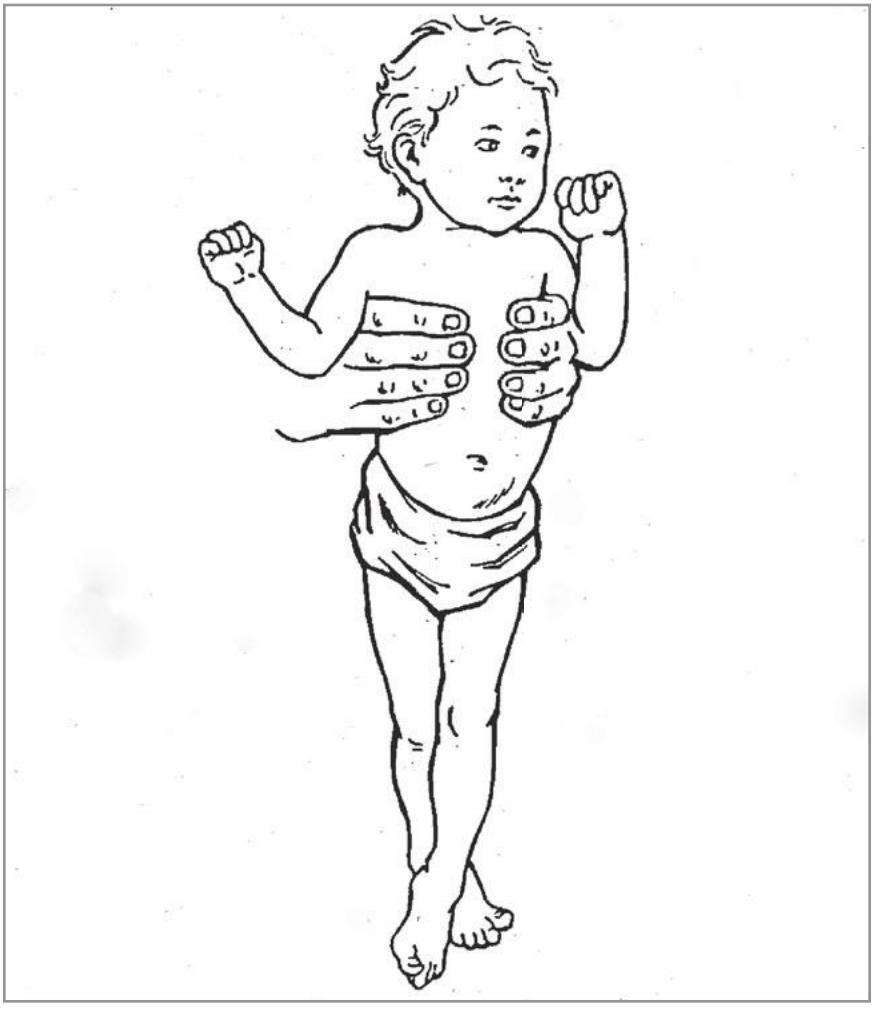

Figure 9.1. Scissoring. In vertical suspension the ankles cross one another. Note the fisting and tonic labyrinthine ("surrender") posture. From Blasco PA. Normal and abnormal motor development. *Pediatric Rounds.* 1992 Volume 1, number 2. Reprinted with permission.

Station refers to the posture assumed in sitting or standing and should be viewed from anterior, lateral, and posterior perspectives looking for body alignment. Gait, of course, refers to walking and is examined in progress. Initially, the toddler walks on a wide base, slightly crouched, with the arms abducted and elevated a bit. Forward progression is more staccato than smooth. Movements gradually become more fluid, the base narrows, and arm swing evolves, leading to an adult pattern of walking by 3 years of age.

The motor neuromaturational markers are the *primitive reflexes,* which develop during gestation and generally disappear between the third and sixth month after birth, and the *postural reactions,* which are not present at birth but sequentially develop between 3 and 10 months of age (Figure 9.2). The Moro, tonic labyrinthine, asymmetric tonic neck, and positive support reflexes are the most clinically useful (Figure 9.3). As with all true reflexes, each requires a specific sensory stimulus to generate the stereotyped motor response that follows. Normal babies and infants demonstrate these postures inconsistently and transiently, whereas those with neurologic injuries show stronger and more sustained primitive reflex posturing.[6]

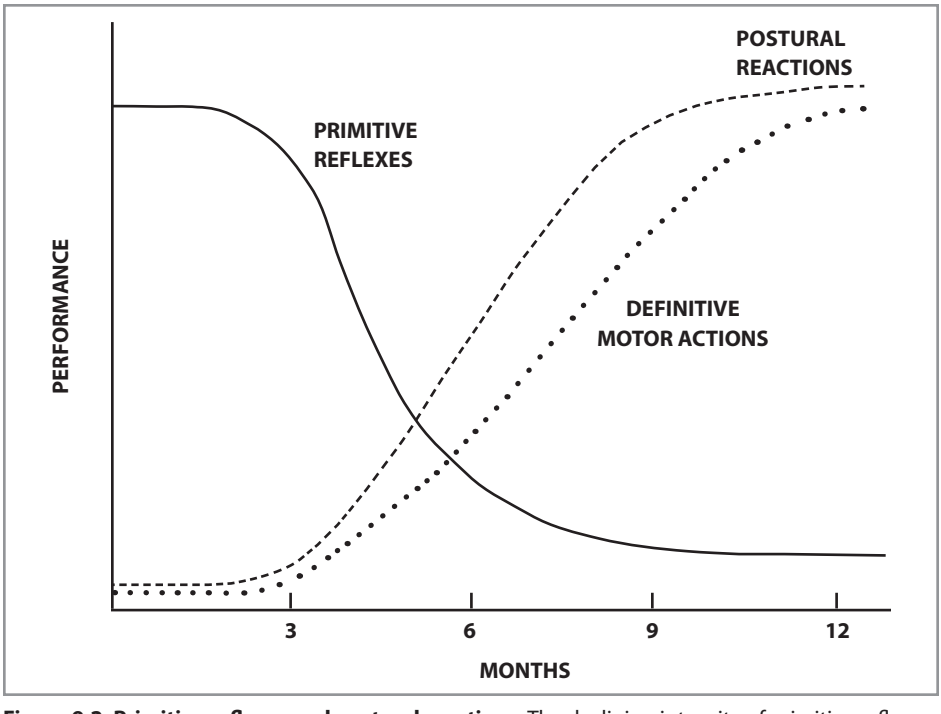

Figure 9.2. Primitive reflexes and postural reactions. The declining intensity of primitive reflexes and the increasing rate of postural reactions represent at least permissive, and possibly necessary, conditions for the development of definitive motor actions. From: Capute AJ, Accardo PJ, Vining EPG, Rubenstein JE, Harryman S. *Primitive Reflex Profile.* Baltimore, MD: University Park Press; 1978.

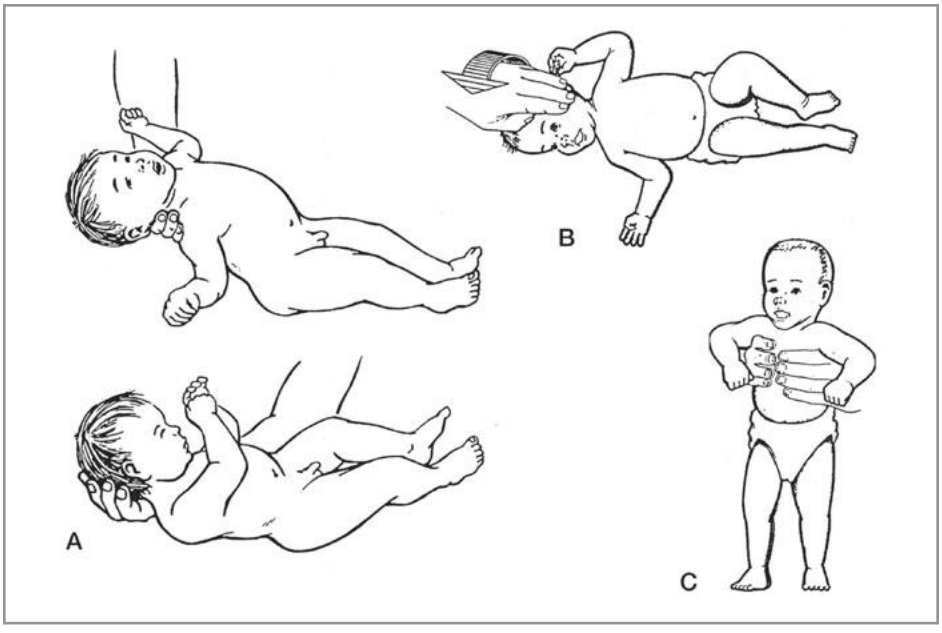

Figure 9.3. Clinically useful reflexes. A. Tonic labyrinthine reflex. In the supine position, the baby's head is extended gently to about 45 degrees below horizontal. This produces relative shoulder retraction and leg extension, resulting in the "surrender posture." With head flexion to about +45 degrees, the arms come forward (shoulder protraction) and the legs flex. **B.** Asymmetric tonic neck reflex (ATNR). The sensory limb of the ATNR involves proprioceptors in the cervical vertebrae. With active or passive head rotation, the baby extends the arm and leg on the face side and flexes the extremities on the occiput side (the "fencer posture"). There also is some mild paraspinous muscle contraction on the occiput side that produces subtle trunk curvature. **C.** Positive support reflex. With support around the trunk, the infant is suspended and then lowered to pat the feet gently on a flat surface. This stimulus produces reflex extension at the hips, knees, and ankles so the infant stands up, completely or partially bearing weight. Children may go up on their toes initially but should come down onto flat feet within 20 to 30 seconds before sagging back down toward a sitting position. From: Blasco PA. Normal and abnormal motor development *Pediatr Rounds.* 1992;1(2):1– 6. Reprinted with permission.

Primitive reflexes are somewhat tricky to gauge, even in expert hands. The appearance of postural reactions in sequence beginning after 2 to 3 months of age is easier to elicit clinically and can provide great insight into the motor potential of young infants. Postural reactions are sought in each of the 3 major categories: righting, protection, and equilibrium. These movements are much less stereotyped than the primitive reflexes, and they require a complex interplay of cerebral and cerebellar cortical adjustments to a barrage of many sensory inputs (proprioceptive, visual, vestibular—see Figures 9.4 and 9.5). They are easy to elicit in the normal infant but are markedly slowed in their appearance in the child with nervous system damage. When elicited, the primitive reflex and postural reaction movements or postures are typically quite

Figure 9.4. Postural mechanisms. The infant is seated comfortably, supported about the waist if necessary. The examiner gently tilts the child to one side, noting righting of the head back toward the midline, protective extension of the arm toward the side, and equilibrium countermovements of the arm and leg on the opposite side. From: Blasco PA. *Pediatr Rounds.* 1992;1(2): 1–6. Reprinted with permission.

Figure 9.5. Normal parachute reaction. The examiner has suspended the child horizontally by the waist and lowered him face down toward a flat surface. The arms extend in front, slightly abducted at the shoulders, and the fingers spread as if to break a fall. From: Blasco PA. *Pediatr Rounds.* 1992;1(2):1–6. Reprinted with permission.

symmetric comparing each side. Striking and consistent asymmetries should alert the examiner to a variety of possible abnormalities on the less responsive side. For example, a Moro reflex that is consistently full and typical on one side but blunted on the other could represent a fractured clavicle, a brachial plexus injury, or a hemiplegic brain injury. The reader is referred to Baird and Gordon[4] and Capute[6] for more detailed information on individual reflexes. Analysis of the information gathered in these 3 areas makes it relatively easy for the primary pediatric health care professional to reassure herself (and the parents) about a child's motor competence or to identify at an early age the motor-impaired infant.

Impairments of Motor Development

Individual therapists, teachers, nurses, physicians, and others trained in child development often disagree on what should be considered significant in terms of motor delay, neurologic examination findings, etc. There is no uniformly accepted method for determining this, especially for subjective parameters like muscle tone and coordination. The calculation of a motor quotient, as previously described, is most useful in approximating the significance of motor delay. This ratio is easiest to apply in children younger than 2 years and, of course, is not absolute. Nonetheless, it is a reliable and easy screen. When combined with other aspects of the child's neurologic examination, it becomes the basis for specific diagnosis of motor impairment.[3,7]

When motor abnormality is identified, several steps should be taken by the primary pediatric health care professional. Regardless of diagnosis, evaluation for associated delays is essential. This should be performed by the primary pediatric health care professional (see Chapter 7) and should be accompanied by a referral to the local early intervention program (see Chapter 5) to determine whether the child with a motor delay qualifies for early intervention services in the local community. If not, referral for private physical or occupational therapy services should be considered. In addition to identifying the motor and potential associated developmental deficits, the primary pediatric health care professional must both assess the nature of the motor disability and evaluate the child for associated medical complications related to the motor disability. Referral to a subspecialist in child neurology/neurodevelopmental pediatrics should also be strongly considered. The differential diagnosis of motor disability falls into 4 general categories: (1) static central nervous system (CNS) disorders, (2) spinal cord and peripheral nerve injuries, (3) progressive diseases, and (4) structural defects.

Static Central Nervous System Encephalopathies

Static CNS encephalopathies, by definition, indicate some type of nonprogressive brain disorder. Insults may occur during early brain development, resulting in a CNS anomaly. The anomaly could be the result of improper genetic information or a very early biochemical or mechanical insult permanently altering the anatomical development of the brain. Alternatively, a normally developing brain may be damaged before, during, or after birth by a variety of insults. Examples would include hemorrhage (in the premature infant), infection (meningitis, encephalitis), trauma, ischemia (as might occur in dehydration or a stroke), poisons (such as lead intoxication), metabolic diseases (as with phenylketonuria), and so forth.[8,9]

Brain injury resulting in neuron or axon destruction is permanent—little to no true neuronal repair or regrowth takes place. However, intact areas continue to develop, so that some functional improvements may continue to be seen throughout childhood. Maturation of motor areas in the brain reaches completion by 7 or 8 years of age, although some minor changes will continue to take place through adolescence.[10]

Cerebral Palsy

Cerebral palsy is the classic motor manifestation of a static encephalopathy. Depending on the location and degree of damage, CP may affect anywhere from one limb to the entire body. As would be suspected, children with greater body involvement who have suffered more significant areas of brain damage are more likely to have comorbid neurologic conditions, such as intellectual disability (formerly called mental retardation), seizures, and vision impairment, as well as significant morbidity arising secondary to their motor deficits.[8,9]

Cerebral palsy is a nonprogressive disorder of motor function caused by brain insult during the prenatal, perinatal, or postnatal period that significantly limits the functional activity of affected individuals. With an incidence of 1.5 to 3 cases per 1,000 children, CP is one of the most common serious motor disorders found in children.[9,11] Cerebral palsy is a blanket description of static motor impairment caused by CNS injury. The list of possible etiologies for the injury, however, is extensive.

Twenty years ago, the belief was that most CP was a consequence of birth-related injury to the brain. Obstetricians often took blame for causing CP. In 2003 the American College of Obstetricians and Gynecologists published a literature review studying the linkage of perinatal asphyxia and CP.[12] This study was endorsed by the AAP and other pediatric advocate groups. The report described that, in the absence of moderate to severe neonatal encephalopathy, birth asphyxia is not causally related to CP. In cases where encephalopathy is present in the newborn period, asphyxia should only be considered a possible etiology if there is severe metabolic acidosis at birth, and the subsequent CP syndrome is spastic quadriplegic or dyskinetic in nature. There has been a shift in emphasis to unknown prenatal events as the causative factors that both precipitate preterm birth and/or difficult deliveries and result in neurodevelopmental disability.[13]

What insights we do have about the underlying mechanisms of CP come from an understanding of the neuropathology and the epidemiologic correlation between certain antecedent events, such as preterm birth, and the ultimate development of CP. Premature birth and low birth weight (<1,500 g) are the most closely associated risk factors for the development of CP, with an incidence of 100 per 1,000 surviving infants born before 28 weeks' gestation.[14] Genetic abnormalities, maternal viral infection, and exposure to teratogens or drugs also contribute to antenatal injury causing CP. Postnatal factors account for approximately 10% of children with CP and include severe neonatal infection, cerebral infarction, metabolic disease, or trauma.[8,9,14]

Cerebral palsy is not usually evident at birth; therefore, children with significant risk factors warrant close monitoring throughout infancy. Children who are severely affected may be detected as early as 4 to 6 months on evaluation for delayed early motor milestones. Those with less severe CP may not present until later, when their milder motor impairments interfere with smooth acquisition of more skilled motor activities. Children with CP will display abnormalities in muscle tone, which are manifested functionally in abnormal posturing (poor head control, cortical thumbs, opisthotonus, scissoring of the lower extremities). Their neurologic examinations are abnormal as well, with retention or obligation of primitive reflexes, asymmetric reflexes, hyperreflexia, and/or sustained clonus.[8]

The diagnosis of CP is made by observation of the clinical signs listed above. Imaging is recommended in all children who are newly diagnosed with CP and, often,

infants with significant risk factors will have been imaged prior to diagnosis.[11] Confirmation of brain damage, such as periventricular echodensities seen on cranial ultrasound or periventricular leukomalacia noted on magnetic resonance imaging (MRI), can assist practitioners as they describe the diagnosis to families. Children with clear risk factors, especially infants born before 32 weeks' gestation, should be considered at risk for CP even in the absence of MRI abnormalities.[9] In addition, despite an exhaustive etiologic list, there continue to be children with CP who exhibit no clear risk factors. In such cases, imaging is essential. However, computed tomography and MRI studies report identification of a cause in only 77% to 89% of cases.[9,11] Additional workup, including genetic (when children display dysmorphic features) or metabolic (in the case of dyskinetic CP with no clear history of encephalopathy), should be performed on a case-by-case basis.[9,11]

The classification of CP is also a clinical task and implies which motor control system in the brain has been primarily damaged. The key categories of CP describe predominant motor characteristics, including (1) spastic, in which there are exaggerated stretch reflexes, muscle spasticity, and a strong tendency to develop contractures; (2) dyskinetic, with purposeless, uncontrollable movements or rigidity; and

Box 9.1. Cerebral Palsy: Associated Disorders

- Orthopedic Deformities
  - Muscle/tendon contracture
  - Bone deformities/malalignments
  - Joint dislocation/degeneration
  - Scoliosis
  - Osteoporosis and fracture
- Cognitive Deficits
  - Mental retardation/intellectual disability
  - Learning disability
- Poor Growth/Undernutrition
- Sensory Deficits
  - Sensation deficits
  - Visual impairment/oculomotor disturbance
  - Hearing loss (sensorineural or conductive [recurrent otitis])
- Oromotor Performance Impairments
  - Speech deficits (dysarthria)
  - Feeding dysfunction (dysphagia)
  - Drooling
  - Aspiration
- Gastroesophageal Reflux
- Bowel and Bladder Problems
- Seizures
- Cervical Neuropathy
- Behavioral/Emotional Disturbances
  - Organic
  - Acquired

(3) ataxic, in which the child has poor balance, poor coordination, and a staggering gait. In general terms (although somewhat oversimplified), damage in the corticospinal tract usually results in spasticity, and damage to the basal ganglia and cerebellum results in dyskinesia and ataxia, respectively. Approximately 20% to 25% of children with CP will have both pyramidal and extrapyramidal features. Additionally, CP can be described according to the pattern of limb involvement: (1) monoplegia, the one affected limb determined by the area of brain injury; (2) hemiplegia, affecting one arm and leg, often secondary to an infarct or unilateral intraventricular hemorrhage; (3) diplegia, in which the bilateral lower extremities are most significantly affected (although the arms are involved but to a much lesser degree); (4) triplegia, in which one limb is minimally affected compared with the other 3, which are substantially involved; and (5) quadriplegia, which involves motor limitations in the trunk and all 4 extremities. Establishing the type of CP has value in treatment planning and in prognosis.[8]

Medical Comorbidity in Cerebral Palsy

Cerebral palsy, by definition a disorder of motor function, is often associated with multiple and some severe comorbidities (Box 9.1). The additional morbidity is accounted for by additional areas of brain damage or by the underlying loss of central control over muscle tone and coordination. Physical problems associated with CP include deformities of the bones and contractures of the tendons and muscles that result from the influence of excessively high or abnormally low muscle tone. While diagnostic distinctions and comprehensive treatment plans often require coordinated efforts among the primary pediatric health care professional, orthopedic surgeons, physiatrists, neurologists, neurodevelopmental or developmental-behavioral pediatricians, nurse coordinators, and therapists experienced in the care of children with motor impairments, knowledge of these comorbidities, and the interventions for them are indispensable to the primary care provider.[9,14,15] As primary pediatric health care professionals have the most frequent and longitudinal contact with children with CP and their families, they are in a position to coordinate and decipher specialty clinic visits and provide assistance and guidance to families coping with disability. They are often first in line to identify possible comorbid conditions or common side effects of treatment.

Problems with growth and nutrition are commonly seen in children with CP and may be related to multiple underlying comorbid factors. The motor disability in children with CP frequently includes oral-pharyngeal motor dysfunction manifested by drooling, difficulty handling respiratory tract secretions, and disordered chewing and swallowing. All children with poor growth and coughing associated with feeds should be evaluated for swallowing dysfunction. In addition, even in the absence of coughing or choking, a high degree of suspicion should be kept for silent aspiration in children with CP, as bulbar dysfunction is common and unrecognized aspiration can lead to significant respiratory morbidity and mortality.

Many children with spasticity may have difficulties with weight gain simply secondary to increased energy expenditure. Families of children with CP benefit from dietary counseling on maximizing caloric and nutrient content without significantly increasing the volume of food required. Gastroesophageal reflux also commonly affects children with CP, and the associated pain may limit oral intake. Such patients may require either medical or surgical management to improve their weight gain.

As bowel motility is affected by CNS injury, children with CP often manifest complications secondary to impaired gastrointestinal motility. Noted above, gastroesophageal reflux is a significant complication of delayed gastric emptying. Although conservative interventions, such as upright positioning and smaller meals, are often helpful, medical intervention with acid-reducing medications like proton-pump inhibitors or h^2-blockers may be necessary. In more severe circumstances, surgical intervention to prevent aspiration and ensure proper nutrition should be considered. Constipation can be a result of gut dysmotility and/or physical inactivity and is almost universally seen in CP. It can result in symptoms of pain, increased spasticity, decreased oral intake, vomiting and, in very severe forms, bowel perforation. Symptoms of constipation can be elicited easily by history of infrequent, painful, hard, or very large caliber stools. If necessary, abdominal films can contribute to a diagnosis and assist in determining the intensity of intervention necessary. Depending on the degree of impaction at diagnosis, initial clean-out with enemas, suppositories, or larger doses of polyethylene glycol may be necessary. Long-term management with a daily or every-other-day bowel regimen may be needed. Therapeutic interventions should take into account the family and child's needs, circumstances, and relevant comorbid factors (ie, children at high risk for aspiration should not be administered high volumes of liquid stool softener).

Either secondary to their brain lesion/injury or as a result of the underlying condition that caused their motor impairment, children with CP are at risk for associated neurologic complications, including seizure disorders; intellectual disability; behavior problems; and sensory impairments, including problems with vision and hearing. Ensuring that children are appropriately evaluated and that education is targeted at a suitable level will help minimize school problems. As might be expected, children with more severe motor impairment are at increased risk for intellectual disability; however, it is important to recognize the very challenging task of determining cognitive ability in a setting of limited motor control and verbal output. In children with significant dysarthria, evaluation for potential augmentative communication device use may greatly improve both the ability to interact and quality of life. Also, identifying potential sensory impairments early through formal auditory and vision evaluations will help optimize learning.

Further common but significant medical comorbidities in children with CP include dental caries, skin breakdown, osteopenia, and urinary tract dysfunction. During regular health maintenance visits, primary pediatric health care professionals should

perform thorough systems reviews, focusing particularly on areas of known risk, and address identified concerns. Children with CP are among the most complicated patients in primary care practice, and comprehensive primary care, combined with subspecialty care when needed, is essential for children with CP to ensure that their broad spectrum of medical needs is addressed.

Management of Spasticity

Spasticity is managed with the purpose of improving function, maintaining range of motion, reducing pain, increasing the ease of caregiving, and preventing deformity. Although the degree of intervention is dependent on the degree of spasticity, less invasive approaches are generally applied first. First-line intervention involves specific stretching and activity exercises. These exercises are sometimes to be assisted by a physical therapist but almost always involve daily family participation. Depending on the nature of the patient's spasticity, exercises can be augmented by splinting and postural support to maintain a proper resting position and prevent contractures. Orthotic devices, such as hand splints fashioned from neoprene or plastic braces that lend support to the foot and ankle, when used in appropriate patients, may also augment functional ability.[16,17]

Medical management can be used for generalized spasticity. Common oral agents include baclofen, tizanidine, benzodiazepines, dantrolene, and gabapentin. Most of these agents work by increasing the effects of gamma-aminobutyric acid (GABA), an inhibitory neurotransmitter. Tizanidine works by decreasing presynaptic excitatory activity. Dantrolene works directly on the muscle by inhibiting calcium release and therefore decreasing the coupling reaction involved in muscle contraction. Unfortunately, side effects, including drowsiness and weakness, often limit optimum dosing of these oral agents.[17]

Children with significant lower extremity spasticity who are inadequately treated by oral agents may benefit from intrathecal baclofen. This modality is most useful in nonambulatory patients who have difficulty in hygiene or transfers secondary to extreme lower extremity spasticity. The technique involves surgical implantation of a pump device into the abdomen that continuously infuses baclofen through a catheter into the intrathecal space. Because of the direct application of medication, general side effects are minimized, although they still may occur. Baclofen withdrawal, most commonly associated with hardware malfunction of intrathecal baclofen pump systems, is the most serious and potentially fatal complication among the preceding treatments.[18,19] It is manifested by return of baseline spasticity, development of fever, altered mental status, seizures, and/or malignant hyperthermia. Rapid recognition of these symptoms and treatment in an emergency department or intensive care setting is important to prevent potentially life-threatening effects. Restoration of the intrathecal infusion is the ultimate treatment, but in emergent situations, oral or enteral baclofen or oral or intravenous benzodiazepines can be used.

In patients with focal spasticity, local treatment is preferable to systemic medication. Medical options in these patients include injections with either botulinum toxin (botox) or phenol. Botox works directly on the targeted muscle by blocking the release of acetylcholine at the neuromuscular junction. Although the blockade is permanent, patients who respond well to botox may require injections every 4 to 6 months due to reinnervation of the muscle groups. Studies thus far have only shown reliable improvement in spastic pes equinus.[20] However, the reduction of tone, combined with continued physical therapy, has shown improved passive function in patients, assisting with hygiene, dressing, etc.[17] Phenol injections offer another option for children with focal spasticity. Phenol is injected directly into the nerve, which causes neurolysis and decreased spasticity in all supplied muscle groups. Axonal regeneration again contributes to the need for repeated injections of phenol. Phenol and botox can be used simultaneously for additive effects.[17]

Despite aggressive spasticity management, fixed muscular contractures cannot always be avoided. In such cases, no amount of stretching or medication can reverse the deformity. For milder contractures, especially in younger children when postponement of surgery would be preferred, serial casting of contractured joints can be attempted. This procedure involves casting the joint in a position slightly closer to neutral for several consecutive weeks. Proper positioning and technique involves specially trained physical therapists, orthotists, and/or orthopedists. Although improved range of movement can usually be achieved through this procedure, surgery will often follow.[17]

Surgical intervention is not a last resort, but an adjunct to other modalities of treatment. Surgery should be judiciously applied when the need becomes apparent. The general goals of surgery are to maximize function, minimize deformity, and keep future interventions to a minimum. The first goal of surgery is to diminish muscle imbalance. This is accomplished by weakening muscles and decreasing spasticity (tendon lengthening and tendon release procedures) and by altering the direction of muscle force (specific muscle transfer procedures). Most muscle procedures are muscle shortening procedures, whereby muscle origins or tendons are released or the tendons are lengthened. This weakens the muscle by reducing the resting length and diminishes tension on the muscle spindle and tendon organs, thereby reducing spasticity.[21] The second goal of surgical intervention is to prevent bony and soft tissue deformity, such as hip dislocation. Where deformity has developed already, correcting the bony deformity and releasing soft tissue contracture become necessary. Most bone procedures involve realignment/angulation changes in order to achieve better position and better mechanical advantage. The resulting bone realignment and joint stabilization lead back to maximal function.[16]

Selective dorsal rhizotomy—cutting portions of the dorsal roots of the spinal cord—is another procedure performed occasionally on patients with CP and is aimed specifically at eliminating lower extremity spasticity.[22] By limiting the sensory input

through the afferent nerve fibers from the muscle spindles, spasticity can be greatly reduced. Ideally, this procedure is to be used in selected spastic diplegic patients to improve a gait pattern hindered primarily by lower extremity spasticity. Less often, this procedure could be used in severely spastic quadriplegic patients to assist in the ease of hygiene and caregiving. In most cases, spasticity is entirely or largely eliminated from the lower extremities with very little recurrence.[22] Contraindications to this procedure include patients with athetosis, ataxia, muscle weakness, and severe fixed contractures. Despite the significant reduction in spasticity, patients who have undergone rhizotomy may later require orthopedic surgery for preexistent or worsening lower extremity contractures.

The intervention choices described are considerable, and the patients involved are complicated. In addition, the evidence base for many of these interventions is sketchy.[23] The decision to recommend hands-on therapy, orthotics or serial casting, orthopedic surgery, muscle relaxant drugs, rhizotomy, a baclofen pump, a combination of these, or none of these is best carried out by an interdisciplinary spasticity management clinical team that includes both families and primary pediatric health care professionals in all clinical decisions. Though not directly involved in the administration of many of these therapies, it is essential for primary pediatric health care professionals to be aware of common adverse effects and complications associated with these therapies, so that appropriate interventions can be made when a problem is identified.

Spinal Cord and Peripheral Nerve Injuries

Spinal cord and peripheral nerve injuries are all static conditions, except for the rare instance of a growing spinal cord tumor. They differ from CP in that functional loss is much easier to predict, and there are different types of associated problems. In this category, the largest single group consists of children with myelomeningocele (spina bifida). This condition results from anomalous spinal cord development, whereby early neural tube closure fails to take place, causing an incomplete cord to develop below the level of failed closure. As this can occur anywhere along the length of the spinal cord, the resulting loss of motor and sensory function will vary greatly from person to person.[10,24] The morbidity and mortality of spina bifida is related to a large extent to the level of the spinal defect, with higher-level lesions causing more in the way of lower extremity deformities, scoliosis, neurogenic bowel and bladder problems, and risk for skin breakdown secondary to sensory loss.[25] In addition, developmental abnormalities typically occur at the rostral end of the neural tube, bringing another set of associated morbidities.

Spina Bifida

Open neural tube defects constitute one of the most common congenital human malformation syndromes, with an incidence of 0.5 to 1/1,000 pregnancies in the United States.[24] Neural tube defects evolve during the third and fourth weeks of

gestation, when the neural tube fails to fuse at some point along its length. As early as day 18 of gestation, the neural plate forms and quickly develops into neural folds. The bilateral neural folds, in turn, join to form the blueprint of the fetal nervous system called the neural tube. Fusion of the neural folds begins at the cervical region of the spinal cord and progresses cranially and caudally in a zipper-like fashion. This process is completed with closure of the anterior and posterior neuropores on days 24 and 26, respectively. Defects of anterior fusion result in encephalocele or, in extreme cases, anencephaly. Failure of proper fusion anywhere along the length of posterior closure results in failure of the spinal cord to properly form distal to the lesion and is referred to as spina bifida.[10,24]

The cause of spina bifida is unknown. Although most cases are spontaneous, 2% to 16% of cases occur in association with known chromosomal or genetic defects (such as trisomy 18). In cases where additional anomalies are present, the percentage associated with genetic disorders increases to 24%.[24] Spina bifida is considered multifactorial, and several strong risk factors have been identified. Maternal folate status exhibits the strongest identified link with spina bifida. Although the mechanism of association between neural tube defects and folate is not clearly understood, studies have shown that with preconception folic acid supplementation, women can decrease the chance of their fetus developing a neural tube defect by up to 70%.[23] In fact, since the fortification of cereal grain products with folic acid in 1998, the prevalence of neural tube defects in the United States has fallen 26%.

Despite the strong connection to maternal folate status, other important risk factors for spina bifida persist. Maternal medication exposure to anticonvulsants (such as valproic acid), warfarin, and vitamin A all carry strong associations with neural tube defects. Prepregnancy obesity and diabetes are lesser known, but no less significant, risk factors. Obesity alone (body mass index >29 kg/m^2) carries a 1.5 to 3.5 times greater risk to the fetus.[24] Combined with diabetes, the risk is compounded further. Having a previous child affected by a neural tube defect greatly increases the risk of having another child with spina bifida; however, this risk can also be decreased by supplementation with folate.

Spina bifida is usually identified prenatally and screened for via ultrasound and maternal serum alpha-fetoprotein levels at 15 to 20 weeks' gestation. A positive finding in one of these screens prompts confirmation by either amniocentesis, detailed ultrasound, or both. After diagnosis, ultrasound should be used to look for spontaneous lower extremity motion, Chiari II malformations, and hydrocephalus. Deformities such as limb contractures and clubbed feet may also be seen but are not emergent conditions. Prenatal identification of complicating factors can assist in the rapid management of potential life-threatening conditions, such as hydrocephalus.[24]

In utero repair of myelomeningocele was introduced in 1994 and has shown promise. Approximately 200 surgeries have been performed via hysterotomy with 8 infant deaths due to uncontrolled premature delivery. Infants treated prenatally show similar urodynamic and lower-extremity function to infants who undergo surgery following delivery. However, studies indicate that in utero repair offers protection from the acquisition of the Chiari II malformation, one of the leading causes of mortality for patients with spina bifida. Randomized, controlled studies are currently underway to examine such endpoints as the need for shunting, neurologic function, cognitive outcome, and maternal morbidity.[25]

The prenatal course is generally uncomplicated, with 80% of infants with spina bifida being carried to term. Although debated, the mode of delivery for infants with spina bifida has not been shown to affect outcome.[25] Children with very large deformities or who have undergone prenatal surgery should be delivered via cesarean section to prevent trauma and possible wound dehiscence.[25]

Corrective repair is typically performed within 48 hours following delivery to prevent infection of the open defect. If hydrocephalus is present at birth, a shunt is placed during the initial surgery. Care for the infant with spina bifida should include imaging of the entire CNS, with subsequent serial head measurements to monitor for progressive hydrocephalus, whether or not the infant has been shunted. In addition, ultrasound and urodynamic studies should be performed to assess urologic dysfunction secondary to neurogenic bladder.[24,25]

Nearly 100% of individuals with spina bifida have an associated Chiari II malformation. The defect allows herniation of the posterior fossa contents into the foramen magnum and places children with spina bifida at high risk for developing progressive hydrocephalus and brain stem compression symptoms. In fact, 85% to 90% of children with spina bifida require shunting. Early in infancy, rapidly increasing head circumference, apnea, feeding difficulties, and stridor are strong clinical indications of a symptomatic Chiari II malformation. Children who have not required shunting as an infant may present later in life with brain stem compression symptoms of headache, quadriparesis, and balance difficulties. Prior to 1950 and the identification of acceptable shunting techniques, complications of untreated hydrocephalus were the leading causes of death in patients with spina bifida. Since 1972, complications from the Chiari II malformation and shunt problems are again the leading cause of mortality.[24]

Further morbidity associated with spina bifida is closely related to the level of the spinal defect. All patients are at risk for some level of paralysis, sensory loss, and bowel and bladder dysfunction. Patients with lower lumbar or sacral-level lesions are more likely to have some preserved lower extremity function and sensation, decreasing the likelihood of significant orthopedic deformities and increasing the likelihood of functional ambulation. Patients with higher lumbar- or thoracic-level lesions lack useful lower extremity muscle function. They are at significant risk for

lower extremity deformity, as well as significant kyphosis or scoliosis secondary to decreased truncal muscle tone.[24,25]

Almost all patients, even those with sacral lesions, are at significant risk for neurogenic bowel and bladder dysfunction. Between 1950 and 1972, urinary tract disease was the primary cause of morbidity and mortality in this population. The clean intermittent catheterization technique was introduced in 1972 and has been incredibly effective in preserving renal function. Since infantile bladder dysfunction may not be readily recognized due to diaper voiding, early evaluation is essential. Patients at greatest risk for renal failure are those with increased leak point pressure and sphincter dyssynergia, which can cause vesicoureteral reflux (VUR) and upper tract damage. In these patients, catheterization should be started early. Patients without sphincter tone do not often develop renal disease, but they suffer from continence problems. Usually catheterization is an effective treatment for both low- and high-pressure neurogenic bladders. Anticholinergics are useful adjuncts to improve continence, decrease bladder spasms, and further control VUR. Surgical options are also available. To increase continence, surgeons may lengthen the urethra or inject collagen at the opening to increase the urethral pressure. Patients who suffer from significant VUR despite catheterization and anticholinergic medication may benefit from bladder augmentation surgery to decrease spasms and increase bladder volume.[25]

Disorders of bowel elimination, although not associated with mortality, greatly impact the quality of life of individuals with spina bifida. As infants, initiating a bowel regimen is not urgent, as the gastrocolic reflex is intact and assists in adequate elimination. As stools become more formed, initiation of a high-fiber diet, suppositories, or laxatives combined with some sort of manual stimulation, timed elimination, or enema regimens are usually effective to achieve continence. Some patients are good candidates for an antegrade continence enema procedure, which involves using the appendix to form an abdominal stoma through which an antegrade enema can be administered to achieve bowel continence.[25]

In addition to the multiple physical problems encountered, children with spina bifida have been noted to have deficits in intellectual functioning. Patients who have required shunting are more likely to exhibit intellectual disability; in shunted patients, the average IQ is 80.[24] The most common pattern of cognitive processing deficit reported in children with spina bifida and shunted hydrocephalus is a verbal IQ that is significantly higher than performance IQ. These children have been reported to have poor memory and retrieval abilities, decreased computational accuracy and speed, and difficulty with problem-solving. Thus, although the overall IQ may be relatively normal, this atypical intellectual pattern may significantly negatively impact on learning.[24,25]

Studies also indicate that children with spina bifida are more likely to exhibit symptoms consistent with attention-deficit/hyperactivity disorder (ADHD) and related

executive function deficits.[26] This propensity has been evidenced specifically in populations with shunted hydrocephalus. Typically, these children are more likely to have a profile suggestive of the inattentive subtype of ADHD rather than the hyperactive/impulsive or combined subtypes. Although some hyperactive ratings may be distorted due to lack of mobility, ratings indicating impulsive behaviors are also lower and more consistent with the general population.

Children with spina bifida require the collaboration of a multidisciplinary team of specialists to address their complex medical, physical, and psychosocial situations. Assisting the primary pediatric health care professional should be a team that includes neurosurgery, urology, orthopedics, neurodevelopmental or developmental-behavioral pediatrics, physiatry, physical therapy, a nurse specialist, and a social worker. Longitudinal neurologic, musculoskeletal, and urologic surveillance is essential throughout each patient's life. The team should work closely with patients and families on mobility issues, bowel/bladder management, and proper skin care. Appropriate neuropsychological evaluation should be performed and special educational interventions implemented as needed. With a well-coordinated, patient-centered approach, individuals with spina bifida will have the support and care available to become more successful and independent as they transition to adulthood.[24,25,27]

Although it is important for families and health care professionals to be aware of the potential medical and learning difficulties that may be encountered by children with spina bifida, it is equally important to inform families about the availability of social and community resources that can assist children and families to succeed in the face of these obstacles (see Chapter 23). Primary pediatric health care professionals are in a prime position to offer guidance to families as they find a balance between providing support and encouraging responsibility in their children. Recognizing the isolation often experienced by children with chronic health conditions, they can foster relationships with other community families and children who have had similar experiences as an important source of information, modeling, and support for families. In addition, encouraging children with spina bifida to take active roles in their own medical care, participate in social settings, and set future goals promotes self-esteem and persuades children and families to consider long-term planning earlier in life (see Chapter 24). A superb set of guidelines on evidence-based management practices for children (and adults) with spina bifida has been created and recently updated by professionals in concert with the Spina Bifida Association.[28]

Progressive Neurodegenerative Disorders

Progressive diseases of the brain, nerves, or muscles produce motor impairments that worsen with time. Suspicion for a progressive motor impairment can be readily identified through a careful developmental history when the child is on the deteriorating slope. It is a much greater challenge to discern at the earlier stage, when the child is slowing his or her developmental pace or beginning to plateau. Although the number of diseases in this category is large, each individual disease is extremely

rare. Therefore, the fraction of all motor-impaired children with progressive diseases is quite small. Knowing the particular diagnosis is important because it helps one predict the rate of progression and may have other prognostic implications. An appropriate diagnosis is also essential for accurate genetic counseling of families. Most of these conditions are incurable, and most will eventually lead to a fatal outcome. A few can be halted or reversed with medical treatment, and aggressive management of the rest is critical to maximizing function and minimizing psychological morbidity. Examples of the more common serious disorders are Duchenne muscular dystrophy[29] and infantile progressive spinal muscular atrophy type 1[30] (Werdnig-Hoffman disease).

Children with progressive conditions may initially experience a period of normal or near normal development, obscuring the underlying slow disease process. In Duchenne muscular dystrophy, for example, the diagnosis is usually not made until 4 to 6 years of age, when parents first note increasing clumsiness or difficulty keeping up with peers. The importance of early diagnosis in these cases cannot be overstated. Early diagnosis and intervention are crucial, not only to maximize function and improve quality of life, but to provide proper genetic counseling for families. Classically, males with Duchenne muscular dystrophy (an X-linked disorder) have died in their early 20s, and children with Werdnig-Hoffman disease (an autosomal recessive condition) rarely lived beyond 2 or 3 years of age. With advancements in medical management, such as immunizations and infection prophylaxis, and aggressive supportive care measures, like noninvasive ventilation with bi-level positive pressure, length and quality of life have been increased in both conditions.

Knowledge of a comprehensive center where diagnosis, genetic counseling, and ongoing interdisciplinary treatment for children with progressive neurodegenerative disorders can be obtained is of inestimable value to both primary pediatric health care professionals and families. Such centers often offer opportunities for patients to join clinical trial networks, enabling access to clinical trials and the benefits of ongoing research. Financial and psychosocial supports are typically built into these centers. Examples would include most disability-oriented clinics in major children's hospitals, the national network of Muscular Dystrophy Association clinics, and disease-specific parent/professional groups, such as the International Rett Syndrome Association.

Structural Defects

Structural defects refer to situations in which some anatomical structure is missing or deformed (eg, a limb) or in which some support tissues for the nerves and muscles are inadequate (eg, connective tissue defects, biochemically abnormal bone, etc). Structural defects are usually the most straightforward to understand of all motor-impairing conditions. Some conditions are progressive in nature, and many are extremely complex to manage. As examples, osteogenesis imperfecta and the many

varieties of childhood arthritis fit in this category. On the mildest end of the spectrum, there exists a wide variety of fairly common orthopedic deformities that may or may not impact on motor development (eg, club feet, developmental hip dysplasia). They present as visible deformities at birth or in early infancy, they cause aberrations in the *quality* of movement, and their early recognition hinges on clinical examination. Early treatment can make a big difference in outcome and in the gravity of interventions required to achieve correction.

The diagnosis of any of the above groups of motor disorders almost always warrants referral to a coordinated team of physicians and therapists experienced in the care of children with these static or progressive conditions. This team functions to firmly establish the primary diagnosis, identify associated problems, and assist the primary pediatric health care professional in longitudinally managing their patients within the medical home. In addition, he or she may assist the family by referring to appropriate community-based early intervention programs and ensuring their connection with a social worker who is experienced in advocating for and working with children with disabilities and their families.

Conclusion

Primary pediatric health care professionals are in the unique position to screen and evaluate patients longitudinally for abnormalities or delays in motor development. Through careful history, observation, and neurologic evaluation, the primary pediatric health care professional should be able to identify all children with motor delays and determine whether a child's motor function warrants further evaluation. Disorders of motor development have multiple etiologies and may be symptoms of global developmental delays, CP, or a progressive disorder. Children with significant motor impairments, as evidenced in CP or spina bifida, often have multiple medical needs and typically benefit from referral to a multidisciplinary team of specialists. Primary pediatric health care professionals are considered critical members of such multidisciplinary teams, as they are responsible for both routine health maintenance and the longitudinal management of medical comorbidities within the medical home. They are also essential resources to translate specialty information to families, counsel families in their decisions about medical and/or surgical interventions, and provide information about local community resources, all of which serve to optimize the quality of life for the children with disorders of motor development and their families.

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Chapter 10

Cognitive Development

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Cognitive Developmental Milestones

For the first 1 to 2 years of a child's life, when primary pediatric health care professionals are most often interfacing with young patients, cognitive development can be more difficult to appreciate than development in other domains, such as gross motor, fine motor, and language. This is because, in those early years, clues to a child's cognitive development are more indirectly expressed through the child's interactions with the environment. Developmental psychologist Jean Piaget described this phase of cognitive development as "sensorimotor intelligence." Attaining cognitive milestones during this stage is dependent on intact sensory systems (ie, vision and hearing), and children learn through sensory exploration. Children in this phase of development express mastery of cognitive milestones through physical manipulation of objects in their environment. Children with sensory impairments (blindness, deafness) or motor impairments, such as cerebral palsy, will therefore show lagging or notable variability in attainment of cognitive milestones during this early phase. This may not ultimately be a true cognitive deficit but instead a function of the sensorimotor focus of cognitive development during this young age. Concerns about lack of interaction with the environment or delay in attainment of early cognitive milestones should prompt the primary pediatric health care professional to further assess vision and hearing and give more thought to the impact that overall motor development may be having on a child's ability to physically approach and access objects in the environment (gross motor) or his or her ability to manipulate objects (fine motor). Correctly identifying such impairments as early as possible in order to provide aid to a child and/or modifications in the environment can foster the progression of cognitive development.

Cognitive Development in Infancy

In the office, primary pediatric health care professionals should observe a child's physical manipulation of objects in the environment and expect to see much visual inspection, banging, mouthing, and throwing of objects between 4 and 10 months of age. During this time, infants begin to appreciate the association of "cause and effect," wherein they recognize how their actions lead to a response in the environment. By 9 months of age, most infants will show mastery of this concept, realizing that they cause lights or sounds to occur when they activate a push-button toy, for example. Understanding cause and effect is a powerful first step for infants to realize the impact they can have in interacting with and causing change in their environment.

A major milestone during this sensorimotor phase of cognitive development is that of object permanence (Table 10.1). The understanding that an object exists even if one cannot see it is typically mastered by 9 months of age. Out of sight is NOT out of mind once object permanence is achieved, so that a child will look for a toy that has been completely covered from view. Once object permanence has developed, a child can retain a mental picture of his parents when they leave the room, so separation anxiety also emerges at about the same age. Mastery of object permanence can be observed during a child's play or quickly elicited with a "peek-a-boo" game during a clinic visit.

One should be able to appreciate, either by observation or parent report, the development of a child's ability to understand the functional use of objects. Knowing what the objects they encounter in the environment are used for is a cognitive milestone typically mastered by 12 to 15 months of age. If a child sees a hairbrush, she might put it on her head, she may hold a phone to her ear, or hold a key to a doorknob. A child whose thinking skills have progressed to a developmental level of 18 months or more will begin to demonstrate representational play. In this phase of development, a child may put a hair brush to a baby doll's head, pretending to brush its hair. Increased complexity comes at approximately the cognitive level of 2 to 3 years, wherein symbolic play emerges. Children are then capable of using objects to symbolize something else, so that they may place a stick to a baby doll's hair, pretending it is a hair brush. At this level of functioning, play can become more imaginary and pretend, with symbolic play allowing children to think beyond the objects in their immediate vicinity in order to develop their play themes. Although the ages of acquisition are not at all absolute, a child's play can be a window into overall cognitive level, with the complexity providing some clues to a child's overall thinking abilities.

Table 10.1. Cognitive Milestones

Milestone	Description	Approximate Age of Attainment
Early object permanence	Follows an object falling out of sight, searches for a partially hidden object	4–8 months
Object permanence	Searches for an object completely hidden from view	9–12 months
Cause and effect	Realizes his/her action causes another action or is linked to a response	9 months
Functional use of objects	Realizes what objects are used for	12–15 months
Representational play	Pretends to use objects functionally on others, on dolls	18 months
Symbolic play	Uses an object to symbolize something else during pretend play	2–3 years
Pre-academic skills	Knows letters, numbers, shapes, colors, and counts	3–5 years
Logical thinking	Understands conservation of matter, multistep problem-solving; realizes there can be differing perspectives	6–12 years
Abstract thinking	Able to hypothesize, think abstractly, draw conclusions	>13 years

Cognitive Development in Preschoolers

In preschool-aged children (ages 3–5 years), cognitive development can be appreciated through their mastery of "pre-academic" skills. Children in this age group begin to recognize colors, shapes, numbers, and letters. They begin to develop a concept of time and understand concepts such as big/little, up/down, before/after. They are typically counting objects, up to 10, by the age of 5 years, and respond correctly when asked their name, gender, and age. Preschool children remain egocentric in their thinking and understand the world primarily by how it relates to them. Where expression of cognitive development was largely dependent on sensory and motor systems prior to this age, the domain of language becomes a primary factor in the appreciation of cognitive development in preschoolers. Preschoolers ask many questions (particularly "Why?" questions) and express their thinking and problem-solving skills largely through verbal communication. Therefore, one might make the mistake of assuming that a language-impaired child is cognitively delayed at this age. For children who seem to be lagging in attainment of pre-academic milestones, primary pediatric health care professionals should pay specific attention to their acquisition of language milestones, and obtain more formal assessment when necessary.

One must also be careful presuming cognitive level based on mastery of pre-academic skills because children in environments lacking adequate developmental stimulation may have the cognitive potential to learn pre-academics, but lack exposure; therefore, assessing the environment is an important part of a clinician's conclusions regarding cognitive level. Primary pediatric health care professionals should encourage parents to provide exposure to and opportunity to exercise pre-academic skills, at home or in a preschool setting.

Cognitive Development in School-aged Children and Adolescents

In school-age years, thinking becomes less egocentric, and children can appreciate that other people have viewpoints different from their own. Typically developing school-aged children can think more logically, appreciate concepts such as conservation of matter, identify more subtle relationships, and consider multiple aspects of a problem they are attempting to solve. Children who are not progressing through this phase of cognitive development are likely to struggle in school, so that the cognitively impaired child may present to the primary pediatric health care professional because of school failure. Typically developing adolescents master more elaborate logical thinking but also develop the ability to think abstractly. Adolescents who are not progressing to that expected phase of abstract thought may struggle more with recognizing right and wrong in hypothetical scenarios and be less able to think and plan toward the future.

Intellectual Disability/Mental Retardation

The term *mental retardation* is currently being replaced by the term *intellectual disability* in an effort to demonstrate greater appreciation for the humanness and potential of the individual. Intellectual disability will be the term used herein. Current definitions of intellectual disability not only include a cognitive IQ that is 2 standard deviations (SDs) below the mean (IQ <70), but also adaptive function that is 2 SDs below the mean (Table 10.2). IQ and adaptive function are based on tests from a sample from the general population and should follow a "normal" or bell-shaped curve. Standard scores are based on a mean of 100 with an SD of 15. Most individuals (~95%) in a population will fall between a standard score of 70 and 130 (Figure 10.1). *Adaptive function* refers to how an individual functions in their environment and includes measures of socialization; communication; motor skills; and activities of daily living, such as dressing and toileting. The American Psychiatric Association (APA) defines intellectual disability by 3 criteria: (Criterion A) "significantly subaverage general intellectual functioning"; (Criterion B) "significant limitations in adaptive functioning in at least two of the following areas: communication, self-care, home living, social/interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health, and safety"; (Criterion C) "onset must occur before age 18 years."[1]

In 2002 the American Association on Intellectual and Developmental Disabilities (AAIDD), formerly the American Association on Mental Retardation, published a definition of intellectual disability similar to the APA: a disability characterized by significant limitations in both intellectual functioning and in adaptive behavior as expressed in conceptual, social, and practical adaptive skills.[2] Culture, language, communication, sensory, motor, and behavioral factors must be considered in an intellectual disability diagnosis. It is also important to recognize that limitations often occur with strengths, limitations are described in order to determine needed supports, and a person with intellectual disability can gain functional skills over time with appropriate support systems in place.[2,3]

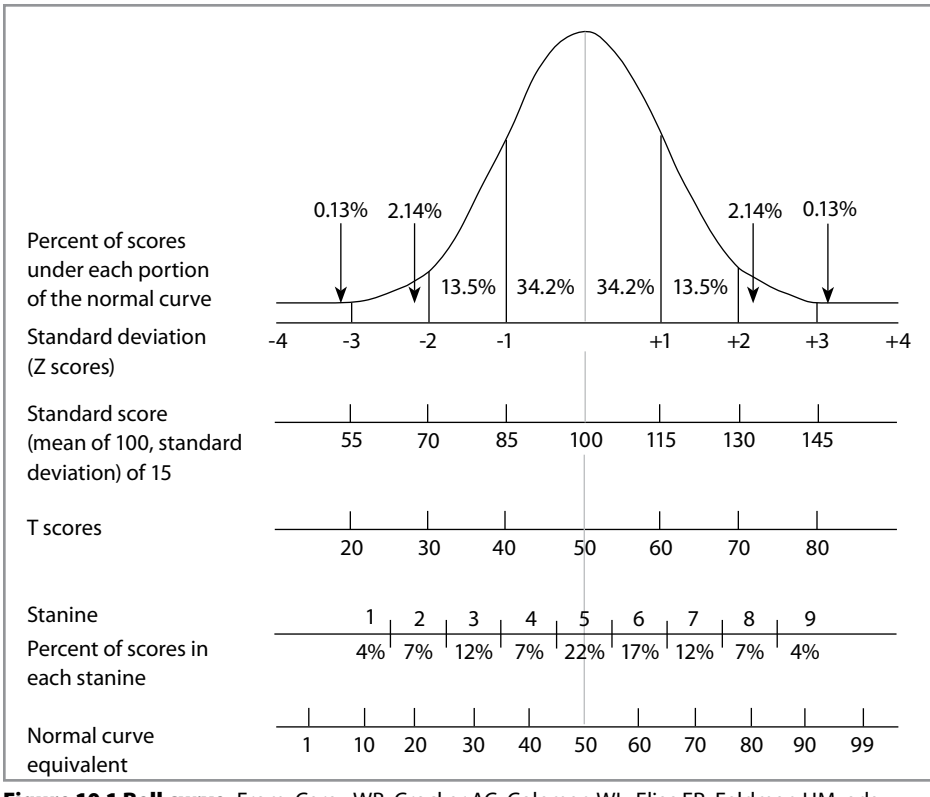

Figure 10.1 Bell curve. From: Carey WB, Crocker AC, Coleman WL, Elias ER, Feldman HM, eds. Developmental & Behavioral Pediatrics. Philadelphia, PA: Elsevier; 2009: 764. With permission from Elsevier.

Table 10.2. Categories of Intellectual Disability

Intellectual Disability Range	IQ	Level of Support Needed
Mild	50–55 to approximately 70	Intermittent
Moderate	35–40 to 50–55	Limited
Severe	20–25 to 35–40	Extensive
Profound	below 20–25	Pervasive

The APA defines levels of intellectual disability based on SDs from the mean on IQ testing. The AAIDD does not subclassify intellectual disability by IQ but focuses on whether the need for support is intermittent, limited, extensive, or pervasive.

Individuals with mild intellectual disability (IQ 2–3 SDs below the mean) may ultimately attain a sixth-grade level of academic functioning. As adults, they often possess social and vocational skills that allow them to function with intermittent supports. They may need supervision or guidance, especially during social or financial stress. They may live independently or in a supervised setting.[1]

Individuals with moderate intellectual disability (IQ 3–4 SDs below the mean) may ultimately attain second-grade academic functioning. They require limited support and may travel independently in familiar settings. They usually live and work in supervised settings and can be trained to do unskilled or semiskilled work.[1]

Individuals with severe intellectual disability (IQ 4–5 SDs below the mean) may learn to speak and to do some self-care skills. They may master some sight-reading, especially for functional words such as "stop" or "exit." They require extensive supports and typically live in a group home or with their family as adults.[1]

Individuals with profound intellectual disability (IQ >5 SDs below the mean) will require pervasive supports and supervision throughout life but may learn some self-help skills, such as self-feeding.[1]

The diagnosis of intellectual disability is usually reserved for children older than 3 years due to the lack of good predictive validity for developmental tests in children younger than 3 years. Many of the skills used to test development in young children assess sensorimotor skills. A child with poor motor skills or a hearing or vision impairment may not be able to show all of their abilities on these early measures of development.

A specific etiologic diagnosis can be made in up to 50% of individuals with intellectual disability.[4,5] In the past, the ability to identify the etiology increased when there were greater than 3 dysmorphic features or the level of functioning was lower.[6] With the advent of new technology, such as whole genome microarray comparative genomic hybridization (CGH), the etiological yield in the future is likely to increase. While the specific genetic workup to consider in individuals with intellectual disability is rapidly evolving, current expert consensus suggests that the initial diagnostic work up should include fragile X molecular genetic testing and whole genome microarray CGH. However, a medical genetics evaluation should be offered to all families of an individual with intellectual disability.[7] Making an etiologic diagnosis to account for an individual's intellectual disability allows for genetic counseling in regard to risk of recurrence in future pregnancies, alerts primary care clinicians to monitor for and prevent potential medical complications associated with a specific diagnosis, and often relieves parental anxiety about what they may have previously believed to have caused their child's cognitive disability.

Specific Etiologies of Intellectual Disability

In the following sections, the medical, developmental, and behavioral features of the most common genetic (Down syndrome), inherited (fragile X syndrome), and preventable (fetal alcohol syndrome) etiologies of intellectual disability will be described. Other genetic disorders with characteristic behavioral phenotypes will then be discussed. Many of these are detected by specific fluorescence in-situ hybridization probes or by whole genome microarray CGH, which is capable of detecting small deletions or duplications of genetic material.

Down Syndrome/Trisomy 21

Down syndrome/trisomy 21 is the most common genetic cause of intellectual disability, occurring in 1 in 600 births, with an increased incidence with increasing maternal age. It is caused by an extra copy of chromosome 21. The physical phenotype is well recognized and includes hypotonia, a small brachycephalic head, epicanthal folds, up-slanting palpebral fissures, Brushfield spots in the iris, flat nasal bridge, small mouth, small ears, excess skin at the nape of the neck, single transverse palmar creases, short incurving fifth fingers (clinodactyly), and a widened "sandal gap" between the first and second toes (Figure 10.2). Medical conditions, such as hypothyroidism, are commonly associated with Down syndrome, and patients should be monitored for these potential medical comorbidities[8] (Table 10.3).

The American Academy of Pediatrics (AAP) has published "Health Supervision in Children with Down Syndrome" (Table 10.4).[8] This policy statement is essential for

Table 10.3. Medical Conditions Commonly Associated With Down Syndrome[a]

Medical Condition	Down Syndrome Comorbidity (%)
Congenital heart defects	50
Gastrointestinal atresias	12
Hirschsprung disease	<1
Thyroid disease	15
Leukemia	<1
Hearing loss	75
Otitis media	50–70
Vision—severe refractive errors	50
Cataracts	15
Acquired hip dislocation	6
Obstructive sleep apnea	50–75

[a] From: American Academy of Pediatrics Committee on Genetics. Health supervision for children with Down syndrome. *Pediatrics*. 2001;107:442–449.

Table 10.4. Health Supervision for Children With Down Syndrome—Committee on Genetics*[a]

		Infancy, 1 Month to 1 Year					Early Childhood, 1 to 5 Years						Late Childhood, 5 to 13 Years, Annual	Adolescence, 13 to 21 Years, Annual
	Prenatal	Neonatal	2 Months	4 Months	6 Months	9 Months	12 Months	15 Months	18 Months	24 Months	3 Years	4 Years		
Diagnosis														
Karyotype review†	●	●												
Phenotype review	●	●												
Recurrence risks	●	●												
Anticipatory guidance														
Early intervention services	●	●	●	●	●	●	●	●	●	●	●			
Reproductive options	●‡	●‡	●‡				●				●			
Family support	●	●	●	●	●	●	●	●	●	●	●	●	●	●
Support groups	●	●	●			●								
Long-term planning	●					●							●§	●§
Sexuality													●	●
Medical evaluation														

* Assure compliance with the American Academy of Pediatrics "Recommendations for Preventive Pediatric Health Care." ● = to be performed; S = subjective, by history; and o = objective, by a standard testing method.
† Or at time of diagnosis.
‡ Discuss referral to specialist.
§ Give once in this age group.
¶ According to state law.
|| As needed.
** See discussion.
[a] From: American Academy of Pediatrics Committee on Genetics. Health Supervision for Children with Down Syndrome. *Pediatrics*. 2001;107:442–449.

Table 10.4. Health Supervision for Children With Down Syndrome—Committee on Genetics* (continued)

		Infancy, 1 Month to 1 Year					Early Childhood, 1 to 5 Years						Late Childhood, 5 to 13 Years, Annual	Adolescence, 13 to 21 Years, Annual
	Prenatal	Neonatal	2 Months	4 Months	6 Months	9 Months	12 Months	15 Months	18 Months	24 Months	3 Years	4 Years		
Growth		o	o	o	o	o	o	o	o	o	o	o	o	o
Thyroid screening		o¶			o		o			o	o	o	o	o
Hearing screening		o	S/o	S/o	S/o	S/o	S/o‡			S/o‡	S/o‡	S/o‡	S/o§	S/o
Vision screening		S/o	S/o‡	S/o	S/o	S/o‡	S/o			S/o‡	S/o	S/o	S/o‖	S/o
Cervical spine roentgenogram											o**			
Echocardiogram	●	o												
CBC		o												o
Psychosocial Development and behavioral	S/o	S/o	S/o	S/o	S/o	S/o	S/o	S/o	S/o	S/o	S/o	S/o	S/o	S/o
School performance										o	o	o	o	o
Socialization							S						S	S

* Assure compliance with the American Academy of Pediatrics "Recommendations for Preventive Pediatric Health Care." ● = to be performed; S = subjective, by history; and o = objective, by a standard testing method.
† Or at time of diagnosis.
‡ Discuss referral to specialist.
§ Give once in this age group.
¶ According to state law.
‖ As needed.
** See discussion.

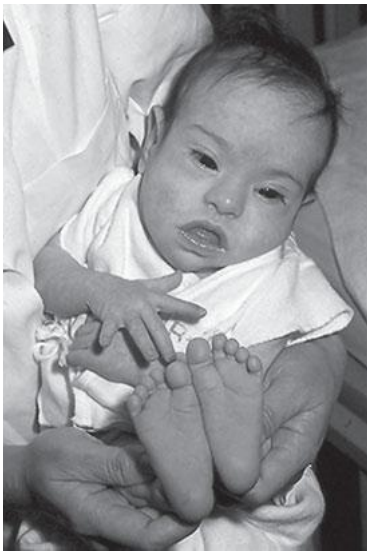

Figure 10.2. Down syndrome. A 6-month-old with Down syndrome with upslanting palpebral fissures, epicanthal folds, and protrusion of the tongue. From: Marion RW. Facial dysmorphism. In: McInerny, Adam, Campbell, Kamat, Kelleher, eds. AAP *Textbook of Pediatric Care.* Elk Grove Village, IL: American Academy of Pediatrics; 2009.

management of individuals with Down syndrome and includes growth charts specific for children with Down syndrome. There is a broad range of cognitive and behavioral outcomes for children with Down syndrome, although most will have intellectual disability in a mild-to-moderate range. Children with Down syndrome classically tend to have fewer severe behavioral problems compared with other children with similar levels of intellectual disability, although approximately 10% will meet criteria for an autism spectrum disorder. Individuals with Down syndrome have an increased risk for depression and Alzheimer's disease as they age.[9]

Fragile X Syndrome

The most common inherited cause of intellectual disability is fragile X syndrome. Fragile X syndrome is caused by a trinucleotide repeat expansion (CGG) within the fragile X mental retardation 1 *(FMR1)* gene.[10] The *FMR1* gene typically includes about 30 repeats. Individuals who carry the premutation have 54 to 200 repeats.[10] In the past, individuals with the premutation were thought to be unaffected; however, there is mounting evidence for a specific phenotype in these individuals. Women with the premutation have a higher incidence of premature ovarian failure.[11] Males with the permutation are at high risk for developing the fragile X–associated tremor-ataxia syndrome (FXTAS). Fragile X tremor-ataxia syndrome is a progressive neurodegenerative disorder that typically develops in male premutation carriers older than 50 years.[12] Individuals with the premutation have normal levels of *FMR1* protein but increased levels of messenger RNA.[13,14] Approximately 1 in 250 women and 1 in 800 to 1,000 men within the general population are premutation carriers.[15,16] The repeat number usually expands when passed from a woman with the premutation to her offspring. If the repeat size expands to 200 or more repeats, the gene will become methylated and silenced, which will result in a deficiency of *FMR1* and the classic fragile X syndrome phenotype.[17] Fragile X mental retardation 1 is an RNA-binding protein that is found in most cells in the body and is directly involved in brain development.[18]

The presentation of the full mutation (>200 repeats) varies between males and females because females have 2 copies of the X chromosome and experience random X inactivation. The portions of the X chromosome that are not present on the Y chromosome are randomly inactivated in each cell. The phenotype in females will

depend at least in part on the pattern of X inactivation.[19,20] If the pattern of inactivation is skewed in one direction or the other, the female may be minimally affected or more significantly affected. In general, females are more likely to be more mildly affected than males.[20] Most males will present with intellectual disability in the mild-to-severe range.

The physical phenotype in males is less apparent prior to puberty but includes macro-orchidism; protuberant ears; long, thin face; and a prominent jaw and forehead. Medical conditions frequently associated with fragile X syndrome include seizures, strabismus, otitis media, gastroesophageal reflux, mitral valve prolapse, and hip dislocation. The behavioral phenotype may include attention-deficit/hyperactivity disorder (ADHD), anxiety, sleep disturbance, perseverative language, hand flapping, gaze aversion, autism, and significant hypersensitivity to environmental stimuli.[21] Because the physical characteristics may not be present in young children, and because this is a relatively common disorder with a recurrence risk of 50%, specific DNA testing for fragile X syndrome is indicated in all children presenting with cognitive delays.

Genetic counseling is warranted in a family with a child with fragile X syndrome because of the 50% recurrence risk and implications for other family members. For example, the child's mother and maternal grandfather may be at risk for issues associated with the premutation, such as FXTAS or premature ovarian failure. Families should also be informed about support groups.

Fetal Alcohol Spectrum Disorders

The fetal alcohol spectrum disorders (FASDs) include a broad range of outcomes seen in individuals exposed to alcohol in utero. Fetal alcohol spectrum disorder occurs in about 1% of children. Several recommendations for diagnostic criteria have been developed.

Fetal Alcohol Syndrome

Fetal alcohol syndrome (FAS) refers to a full syndrome associated with prenatal alcohol exposure. The diagnosis can be made with or without confirmed maternal use of alcohol. The FAS diagnosis is made based on facial anomalies (at least 2 of the following: short palpebral fissures, thin upper lip, and smooth philtrum), poor prenatal or postnatal growth (height or weight ≤10th percentile), and abnormal brain growth (head circumference ≤10th percentile) or morphogenesis.[22–25]

Partial Fetal Alcohol Syndrome

A diagnosis of partial FAS can also be made with or without a confirmed history of maternal alcohol use. This diagnosis is made when at least 2 of the characteristic facial features and at least one of the following are present: poor prenatal or postnatal growth; poor brain growth or morphogenesis; or evidence of characteristic behavioral or cognitive disorders inconsistent with developmental level, genetic/family history, or environment.[24]

Alcohol-Related Neurodevelopmental Disorder

A diagnosis of alcohol-related neurodevelopmental disorder requires confirmation of prenatal alcohol exposure plus at least one of the following: poor brain growth or morphogenesis or evidence of characteristic behavioral or cognitive disorders inconsistent with developmental level, genetic/family history, or environment.[24]

Alcohol-Related Birth Defects

The diagnosis of alcohol-related birth defects requires confirmation of prenatal alcohol exposure, at least 2 of the characteristic facial features, and one or more major congenital malformations or dysplasias in cardiac, skeletal, renal, ocular, or auditory areas (ie, sensorineural hearing loss) or 2 or more minor anomalies, including hypoplastic nails, clinodactyly of or short fifth fingers, camptodactyly, "hockey stick" palmar creases (Figure 10.3), pectus carinatum/excavatum, "railroad track" ears (Figure 10.4), or refractive errors.[24]

Individuals with exposure to alcohol in utero typically have significant difficulty with complex cognitive tasks and executive function (planning, conceptual set shifting, affective set shifting, response inhibition, and fluency). They process information slowly. They may do well with simple tasks but have difficulty with more complex tasks. They have difficulty with attention and short-term memory. They are also at risk for social difficulties and mood disorders.[26] Functional classroom assessments can be a very helpful supplement to psychoeducational evaluations for children with FASD. Methods that have been found to be helpful for individuals with FASD are visual structure (color code each content area), environmental structure (keep

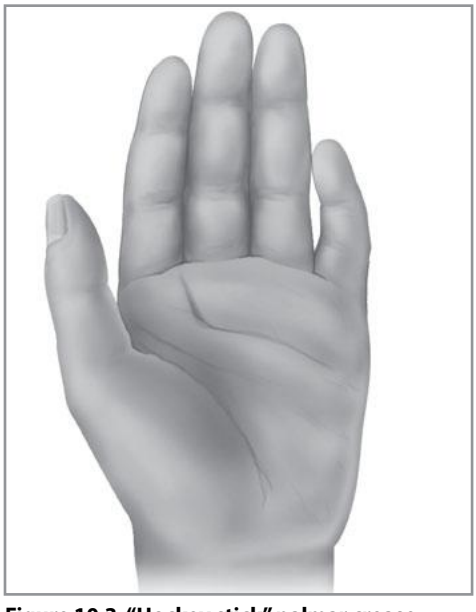

Figure 10.3. "Hockey stick" palmar crease and fifth finger brachyclinodactyly. Reprinted courtesy of Darryl Leja, NHGRI, NIH.

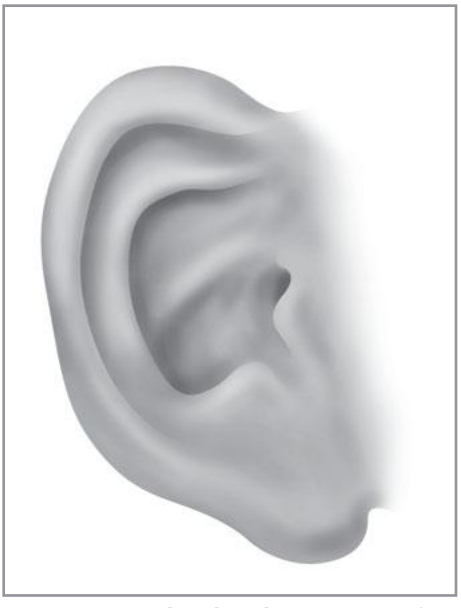

Figure 10.4. "Railroad track" ear. Reprinted courtesy of Darryl Leja, NHGRI, NIH.

work area uncluttered, avoid decorations), and task structure (clear beginning, middle, and end). Cognitive control therapy is an intervention that has shown promising results for children with FASD.[27] Primary pediatric health care professionals have a critical role in the prevention of FASD through education of families and adolescents.[28]

Prader-Willi Syndrome

Prader-Willi syndrome is caused by a microdeletion on chromosome 15q11.2-q13 in an area that is imprinted. Imprinting refers to a gene being turned on or off depending on the parent of origin. The Prader-Willi/Angelman critical region was the first region of the human genome described to be affected by imprinting. A deletion in the same area gives a completely different phenotype (either Prader-Willi or Angelman syndrome) depending on the parent of origin. In 75% of children with Prader-Willi syndrome, the symptoms are caused by a deletion on the paternal chromosome, in 20% there are 2 copies of the maternal chromosome and no copy of the paternal chromosome 15 (uniparental disomy), in 5% there is a translocation or other structural anomaly of chromosome 15, and in 1% there is a problem with the imprinting center.[29] As with Angelman syndrome, there is some variation in the phenotype with different mechanisms.

Infants with Prader-Willi syndrome initially have failure to thrive and hypotonia. By age 2 years, the children begin to develop obesity and evidence significant

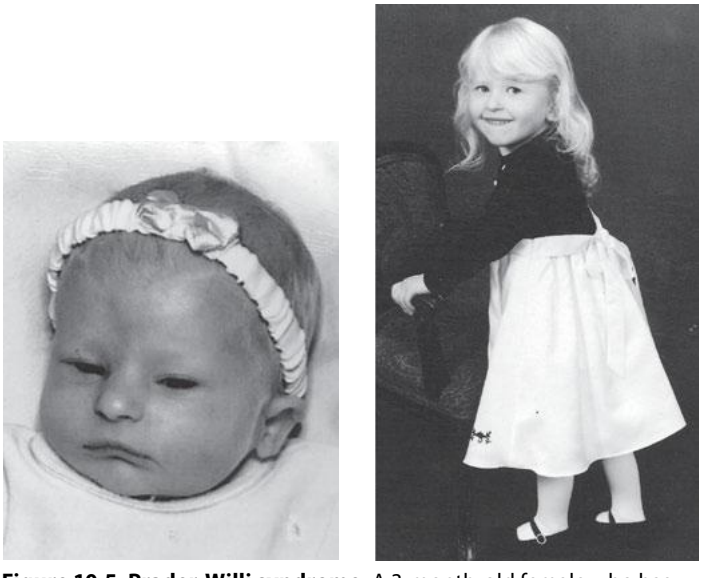

Figure 10.5. Prader-Willi syndrome. A 3-month-old female who has Prader-Willi syndrome (left). Note the almond-shaped eyes and down-turned mouth. The same patient at approximately 3 years of age (right). Used with permission from: Jonas JM, Demmer LA. Genetic syndromes determined by alterations in genomic imprinting pathways. *NeoReviews*. 2007;8;e120–e126.

hyperphagia. They have hypogonadism and short stature. Consultation with an endocrinologist is indicated to consider use of growth hormone, which increases growth, muscle, and bone mass, and decreases fat by about 10%. Sex hormone replacement is also sometimes recommended. Physical characteristics of children with Prader-Willi syndrome include almond-shaped eyes and a thin upper lip with hypopigmented skin, hair, and eyes (Figure 10.5).[29] Cognitive abilities in individuals with Prader-Willi syndrome may extend from a low average range to the range of moderate intellectual disability. Individuals with Prader-Willi syndrome often have obsessive-compulsive behaviors, including hyperphagia and skin picking. They can also have psychosis, a high pain tolerance, and disordered sleep.[29]

Angelman Syndrome

Angelman syndrome is also associated with the imprinted 15q11.2-q13 region on chromosome 15. There are 4 known mechanisms that lead to the Angelman syndrome phenotype: a deletion at 15q11.2-q13 on the maternal chromosome; paternal uniparental disomy, which means that the individual has 2 copies of the paternal chromosome 15 instead of one from each parent; imprinting defects; and mutations in the ubiquitin-protein ligase E3A gene *(UBE3A).*[30] The phenotype can vary depending on the genotype, and individuals with the deletion tend to have the most severe phenotype. Angelman syndrome is characterized by 4 routinely present features: severe cognitive impairment, expressive language more impaired than receptive language, movement and gait disorder, and a happy demeanor/frequent laughter. Features that are present in greater than 80% of individuals with Angelman syndrome are small head, seizures, and characteristic electroencephalogram (EEG) abnormalities[30] (Figure 10.6, Box 10.1).

Figure 10.6. Angelman syndrome. A female child with Angelman syndrome. Used with permission from: Jonas JM, Demmer LA. Genetic syndromes determined by alterations in genomic imprinting pathways. *NeoReviews*. 2007;8;e120–e126.

Box 10.1. Clinical Features of Angelman Syndrome[a]

A. Consistent (100%)

- Developmental delay, functionally severe.
- Movement or balance disorder, usually ataxia of gait, and/or tremulous movement of limbs. Movement disorder can be mild. May not appear as frank ataxia but can be forward lurching; unsteadiness; clumsiness; or quick, jerky motions.
- Behavioral uniqueness: any combination of frequent laughter/smiling; apparent happy demeanor; easily excitable personality, often with uplifted hand-flapping, or waving movements; hypermotoric behavior.
- Speech impairment, none or minimal use of words; receptive and nonverbal communication skills higher than verbal ones.

B. Frequent (more than 80%)

- Delayed, disproportionate growth in head circumference, usually resulting in microcephaly (2 SD of normal occipitofrontal circumference) by age 2 years.
- Microcephaly is more pronounced in those with 15q11.2-q13 deletions.
- Seizures, onset usually <3 years of age. Seizure severity usually decreases with age but the seizure disorder lasts throughout adulthood.
- Abnormal EEG, with a characteristic pattern, as mentioned in the text. The EEG abnormalities can occur in the first 2 years of life and can precede clinical features, and are often not correlated to clinical seizure events.

C. Associated (20%–80%)

- Flat occiput
- Occipital groove
- Protruding tongue
- Tongue thrusting; suck/swallowing disorders
- Feeding problems and/or truncal hypotonia during infancy
- Prognathia
- Wide mouth, wide-spaced teeth
- Frequent drooling
- Excessive chewing/mouthing behaviors
- Strabismus
- Hypopigmented skin, light hair, and eye color (compared to family), seen only in deletion cases
- Hyperactive lower extremity deep tendon reflexes
- Uplifted, flexed arm position especially during ambulation
- Wide-based gait with pronated or valgus-positioned ankles
- Increased sensitivity to heat
- Abnormal sleep-wake cycles and diminished need for sleep
- Attraction to/fascination with water; fascination with crinkly items such as certain papers and plastics
- Abnormal food related behaviors
- Obesity (in the older child)
- Scoliosis
- Constipation

Abbreviations: EEG, electroencephalogram; OFC, SD, standard deviation.

[a] Reprinted with permission from: Williams CA, Beaudet AL, Clayton-Smith J, et al. Angelman syndrome 2005: updated consensus for diagnostic criteria. *Am J Med Genet A*. 2006;140:413–418.

Williams Syndrome

Williams syndrome (also known as Williams-Beuren syndrome) is often recognized by characteristic facial features (periorbital fullness, short nose with bulbous nasal tip, long philtrum, wide mouth, full lips, and mild micrognathia) and a characteristic cognitive and behavioral profile. Williams syndrome is caused by a microdeletion on chromosome 7 at 7q11[23] and is found in about 1 in 7,500 children. Williams syndrome is associated with heart disease in about 80% (usually supravalvar aortic stenosis) and idiopathic hypercalcemia in 15%. The deletion on chromosome 7 includes the elastin gene, which is expressed in connective tissue and leads to the characteristic facial features, cardiovascular defects, bladder and bowel diverticula, hoarse voice, and orthopedic issues observed in individuals with this syndrome.[31] Table 10.5 lists the associated medical issues by age. Individuals with Williams syndrome have developmental delays, and 75% have intellectual disability. The most frequently reported cognitive pattern includes weak visuospatial skills with stronger language skills. While language is a strength, especially for concrete vocabulary and grammatical structure, pragmatic/social use of language may be weaker.[32] Children with Williams syndrome also tend to be overly friendly ("cocktail personality"), anxious, and empathetic.[33,34]

Smith-Magenis Syndrome

Smith-Magenis syndrome (SMS) is a syndrome caused by a deletion on the short arm of chromosome 17, involving band 17 p11.2. Smith-Magenis syndrome occurs in the general population at a rate of about 1 in 25,000. Smith-Magenis syndrome is typically associated with hyperactivity, aggressive and self-injurious behaviors, and significant sleep disturbances. Individuals with SMS have been known to pick or pull out finger and toenails and to self-hug.[35] A recent study of 58 subjects with SMS confirmed that most have moderate intellectual disability. Adaptive function was also found to be poor, with relative strengths in social skills and communication and relative weaknesses in daily living and motor skills. Cognitive and adaptive function were found to vary with the size of the deletion.[36] Physical features of SMS are brachycephaly, broad face, midface hypoplasia, prognathism, everted upper lip, short stature, short hands, scoliosis, strabismus, myopia, and cardiac and renal anomalies. Hearing impairment is found in about 70%, EEG abnormalities in 50%, and seizures in 20%.[36]

Table 10.5. Medical Problems in Williams Syndrome by Organ System and Age[a,b]

Organ System	Incidence, %	Age		
		Infancy	Childhood	Adult
Ocular and visual				
Esotropia	50	x		
Hyperopia	50		x	x
Auditory				
Chronic otitis media	50	x	x	
Hypersensitivity to sound	90	x	x	x
Dental				
Malocclusion	85		x	x
Microdontia	95		x	x
Cardiovascular				
Any abnormality (total)	80	x	x	x
Supravalvar aortic stenosis	75	x	x	x
Supravalvular pulmonic stenosis	25	x	x	x
Peripheral pulmonary artery stenosis	50	x		
Renal artery stenosis	45	x	x	x
Other arterial stenosis	20		x	x
Ventricular septal defect	10	x		
Hypertension	50		x	x
Genitourinary				
Structural anomaly	20	x	x	x
Enuresis	50		x	
Nephrocalcinosis	<5	x	x	x
Recurrent urinary tract infections	30			x
Gastrointestinal				
Feeding difficulties	70	x	x	
Constipation	40	x	x	x
Colon diverticula	30		x	x
Rectal prolapse	15	x	x	
Integument				
Soft lax skin	90	x	x	x
Inguinal hernia	40	x		
Umbilical hernia	50	x		
Prematurely gray hair	90			x

Table 10.5. Medical Problems in Williams Syndrome by Organ System and Age[a,b] *(continued)*

Organ System	Incidence, %	Age		
		Infancy	Childhood	Adult
Musculoskeletal				
Joint hypermobility	90	x	x	
Joint contractures	50	x	x	x
Radioulnar synostosis	20	x	x	x
Kyphosis	20			x
Lordosis	40		x	x
Awkward gait	60		x	x
Calcium				
Hypercalcemia	15	x		x
Hypercalciuria	30	x	x	x
Endocrine				
Hypothyroidism	2	x	x	x
Early puberty (but rarely true precocious puberty)	50		x	
Diabetes mellitus	15			x
Obesity	30			x
Neurologic				
Hyperactive deep tendon reflexes	75		x	x
Chiari I malformation	10	x	x	x
Hypotonia (central)	80	x	x	
Hypertonia (peripheral)	50		x	x
Cognitive				
Developmental delay	95	x	x	
Mental retardation	75		x	x
Borderline intellectual functioning	20		x	x
Normal intelligence	5		x	x
Impaired visuospatial constructive cognition	95		x	x
Behavioral				
Attention-deficit/hyperactivity disorder	70		x	
Generalized anxiety disorder	80		x	x

[a] From: American Academy of Pediatrics. Health care supervision for children with Williams syndrome. *Pediatrics.* 2001;107:1192–1204.

[b] Percentages based on the following: (1) review of rates of complications in several reports of series of patients with Williams syndrome and (2) database of 315 children and adults with Williams syndrome evaluated by Colleen A. Morris, MD.

Sex Chromosome Aneuploidies

Sex chromosome aneuploidies are the most common chromosomal abnormalities and occur in approximately 1 in 500 children. The most common conditions include XXY (Klinefelter syndrome) and XYY in males, and XXX (triple X) and 45,X (Turner syndrome) in females. Cognitive impairment in the intellectual disability range is rare in the trisomy conditions and in Turner syndrome; however, these groups have a high rate of learning disabilities. Interestingly, in XXY, XYY, and triple X syndromes, language-based learning disabilities are present in approximately 70% of patients with strengths in visual-perceptual skills, while in Turner syndrome, nonverbal learning disabilities occur in approximately 70% of patients with strengths in verbal skills.[37,38] Attention-deficit/hyperactivity disorder is also common across all of these conditions. Physical features of XXY, XYY, and triple X include tall stature, low muscle tone, mild hypertelorism, epicanthal folds, and clinodactyly. Males with XXY/Klinefelter syndrome also develop hypogonadism during puberty with findings of microorchidism, eunuchoid body habitus, gynecomastia (in 50%), and infertility.[38] Pubertal development and fertility are not typically affected in XYY and triple X. Girls with Turner syndrome present with short stature and can also have congenital heart abnormalities (40%), renal abnormalities (30%–40%), and ovarian dysgenesis/gonadal failure (90%).[37] The AAP has published health supervision guidelines for Turner syndrome.[39] Haploinsufficiency of the *SHOX* gene has been shown to be associated with the short stature in Turner syndrome, and thus overexpression of *SHOX* is postulated to be related to tall stature in the supernumerary conditions. Genes on the sex chromosomes related to the cognitive phenotypes have not yet been identified; however, in Turner syndrome, those with a small ring X-chromosome are at increased risk for intellectual disability.[37] Other rarer variations, such as XXYY, XXXY, XYYY, XXXXY, tetrasomy X, and pentasomy X, can also occur, and the degree of physical and cognitive problems increases as additional X and Y chromosomes are added.

Educational Programming for Children With Cognitive Impairments

All children are entitled to a free education, regardless of the presence of medical, emotional, or developmental disability. Federal law requires that education be provided in the learning environment that is most appropriate for that particular child. Schools are charged with accommodating or modifying for any disabling condition that might interfere with a child's learning in the school setting, but to also not excessively limit learning potential, so that education is provided in the *least restrictive environment* (LRE).[40] Per the Individuals with Disabilities Education Act (IDEA, formerly the Education for All Handicapped Children Act) children with identified disabilities are deemed eligible for special education and related services. Related services include, but are not limited to, speech/language, occupational, and physical therapies; counseling; and transportation services. Intellectual disability, learning

disabilities, autism spectrum disorders, physical/motor impairments, emotional/behavioral disorders, and hearing and vision impairments are examples of conditions that qualify under this legislation. In addition, a variety of medical conditions that have the potential to interfere with learning (ie, seizure disorder, ADHD) can be designated as "other health impairments" and therefore qualify for services under IDEA. Section 504 of the Rehabilitation Act is another legal avenue through which some children with disabilities may receive related services and/or modifications within the "regular" classroom setting, if they are not deemed to qualify for special education services under IDEA.

Specific to cognitive impairments, children who meet diagnostic criteria for intellectual disability qualify for special education and related services and, per IDEA, will have a written Individualized Education Program (IEP) developed for them by a multidisciplinary team. This team is comprised of teachers, special education personnel, therapists, and other professionals, and also includes the child's parents. Based on assessments of the child that document his or her profile of learning strengths and weaknesses and potential interferences for learning, the team develops an educational plan specific to that child. Primary pediatric health care professionals are encouraged to be a part of this team, and the AAP has described ways in which they can be involved in the IEP process.[41,42] Although schedules rarely allow for primary care providers to be able to physically attend IEP team meetings, they can still be active in the process through early developmental screening and identification, making referrals for additional assessments and IEP development, and reviewing IEPs to counsel families on plans that are developed (Box 10.2).

Primary pediatric health care professionals should, therefore, remain aware of federal legislation regarding special education and amendments as they occur. The most recent amendments to IDEA were enacted in December 2004 (PL 108-446, IDEA Improvement Act of 2004, HR 1350). Some changes to the law within the most recent amendment include clarification that the transition process for a student with a disability should begin by age 16 years, a requirement that an IEP in place in one school be implemented in a new school when a child transfers there until a new IEP can be developed, and an option for multiyear IEPs in some states.[42]

When developing an IEP, the educational conditions that provide the LRE remain a priority in decision-making, as a stipulation within IDEA. The intent of LRE is to provide children with disabilities the opportunity to be among their typically developing, nondisabled peers as much as possible during the school day. *Least restrictive environment* indicates that, whenever possible, children with disabilities will receive their education in the regular classroom environment, and "pull-out" services (such as part of the school day in a specialized educational classroom or all day in a self-contained classroom) is to be provided only when that is impossible. Children may receive accommodations (how a child accesses information or demonstrates mastery of a learning objective is adapted) or modifications (the learning material provided

Box 10.2. Recommendations for Primary Care Involvement in the Development of the Individualized Education Plan[a]

- Screen to identify children who might benefit from services under IDEA.
- Refer for additional evaluations and consideration for IEP team development.
- Diagnose conditions that determine eligibility for services through IDEA.
- Participate in the IEP team meetings and creation of the IEP, if time allows.
- Inform families in interpretation of the IEP and their rights under IDEA.
- Provide a medical home, including the coordination of medically related services described within the IEP.
- Provide liaison between family and school when necessary, remaining objective in the balance between realistic expectations and the family's hopes for outcome.
- Advocate regarding educational services for children with disabilities, including involvement in local and state advisory and interagency committees.
- Remain current regarding knowledge of medical and educational needs of children with disabilities and their legal rights.

Abbreviations: IDEA, Individuals with Disabilities Act; IEP, Individualized Education Program.

[a] Adapted from: American Academy of Pediatrics Committee on Children with Disabilities. The primary pediatric health care provider's role in development and implementation of an individual education plan (IEP) and/or an individual family service plan (IFSP). *Pediatrics*.1999;104:124–127 and American Academy of Pediatrics Council on Children with Disabilities. Provision of educationally related services for children and adolescents with chronic diseases and disabling conditions. *Pediatrics*. 2007;119:1218–1223.

is modified for the child) within the regular classroom setting. There are several models for combining disabled and nondisabled children in educational environments, and theoretical arguments and controversy[43–46] exist around classroom designs and terms such as mainstreaming, inclusion, and full inclusion; however, that debate is beyond the scope of this discussion.

A potential risk of including children with disabilities in regular educational environments is bullying of children with disabilities.[47] Bullying prevention programs within schools should include specific awareness regarding children with disabilities. Another concern about inclusion is the potential that the education of either group of children might be compromised by the presence of the other group because of different learning abilities.[48] This concern must be weighed against the potential benefits for both groups of children. Inclusion allows some normalization in the lives of children with disabilities. It has been shown to improve the academic performance of children with disabilities and to have a positive effect on their self-esteem and social skills.[43] Being among children with disabilities in the educational environment has also been linked to improved tolerance and awareness among the nondisabled students.[49,50]

Borderline Intelligence and Slow Learning

Intellectual disability and specific learning disability are diagnoses included within IDEA legislation. The child who will not qualify for special education under IDEA, but who has difficulty keeping up with average-performing peers in regular classroom environments, is the one with "borderline" or "low average" IQ scores (70–89).

These individuals do not have scores low enough (<70) to support the diagnosis of intellectual disability, and they also do not exhibit enough measured discrepancy between academic ability and cognitive level to qualify for the most commonly used definition of learning disability. Children with borderline to low average cognitive abilities (IQ scores between 70 and 89) represent 23% of the population and may be referred to as "slow learners."[51] Slow learners are a population at particular risk because their impairments are not deemed significant enough to qualify for an IEP or special education services, but they are not learning at the same pace as peers with average IQ scores. These children are often taught in the regular classroom setting, wherein the teacher is typically teaching at the pace of average learners, who have IQ scores in the average range of 90 to 110. An analogy primary pediatric health care professionals can use with families is the "miles per hour" metaphor, wherein the typical learner with an average IQ is learning at a pace of 100 mph, but the child with a borderline IQ is learning at a pace of 75 mph. The child with the IQ of 75 is learning, but not as "fast," so that it will take him or her longer to get to the destination, or to master the learning objective. In addition, there will be a limit to the complexity of academic material that children with slower learning can ultimately master. For example, an individual with an IQ of 85 would be expected to master academic material at only a ninth-grade level by the time of graduation from high school (Table 10.6).

Table 10.6. Projected Achievement Level for Slower Learners[a]

IQ	Age 14 (9th grade)	Age 17 (12th grade)
75	5th grade, 5 months	7th grade, 7 months
80	6th grade, 2 months	8th grade, 6 months
85	6th grade, 9 months	9th grade, 5 months

[a] Modified from: Dunlap HC. Minimum Competency Testing and the Slow Learner. *Educational Leadership.* 1979;327–329.

Children with IQ scores in the borderline to low average range require more time and practice to master academic material, and they need encouragement and emotional support to prevent them from losing motivation to succeed at school and to prevent school dropout. Primary pediatric health care professionals need to be aware that slow learners represent the largest group of children with learning problems in regular classroom settings and are at risk of being left behind in regular classrooms that do not provide accommodations and modifications to support their progress. Classroom accommodations that have had some success include scaffolding of information and reviewing the material in a more specific, systematic way for the slow learner (Table 10.7). Rather than grade retention, nongraded classrooms, learning in cooperative groups, basic and direct instruction of material, and direct instruction on study skills are more effective ways to support the learning of the child with borderline or low average intelligence.[52]

Table 10.7. Learning Difficulties Common for Slow Learners (IQ 70–89)[a]

Learning Difficulty	Descriptions of Learning Difficulties	Teaching Strategies That Might Be Effective
"Concrete" thinkers	Struggle to understand abstract concepts; understand only one dimension of a concept; cannot define, distinguish, analyze	Provide direct, systematic steps when teaching. Provide concrete, hands-on learning experiences.
Poorer auditory learning skills	Struggle to master information when provided initially (auditorally) in the typical classroom	Reinforce. Reteach. Use multi-modality teaching techniques (visual, tactile, kinesthetic).
Do not learn well "incidentally"	Do not generally "pick up" on learning unless explicitly taught	Carefully target purpose of learning activity. Teach studying skills directly.
Difficulty transferring information	Have trouble applying what is learned in one academic subject or within one learning concept to another subject or another situation	Point out connections within learning and within academic subjects. Assist in making associations in learning.
Difficulty "shifting mental gears"	Take longer to mentally "get going," to engage in the learning process, and to maintain attention to task	Allow frequent breaks. Review information. Develop subtle prompts in the classroom to assist with remaining on task.
Work more slowly	Have trouble organizing thoughts, tend to be slow readers (which is the way children's knowledge is most often assessed); mental effort takes longer	Modify tests. Shorten homework assignments. Modify grading process.
Difficulty following directions	Struggle to follow through on 2- and 3-step instruction	Prompt for active listening skills. Give 1-step instructions.
Difficulty recognizing the most pertinent information for study	Struggle to determine what to study; overwhelmed by textbook learning; waste time trying to filter through the material	Supplement the textbook learning. Provide study guides. Highlight important information.

[a] Adapted from: Johns K, Marshall C, Texas Education Agency. *The Slow Learner: An Advocate's View.* Austin, TX: Texas Education Agency; 1989.

Children with slower rates of learning are at increased risk for secondary social, emotional, and behavioral problems derivative of frustration caused by demands and expectations for their performance in regular classroom settings that exceed their abilities. Thus, when school-aged children present to their primary pediatric health care professionals due to behavioral concerns, it is critically important for the clinician to acquire information about a child's underlying cognitive abilities and to analyze whether the demands and expectations placed on the child at school are commensurate with their underlying abilities.

Gifted Children

Parents are often concerned when they interpret their child's developmental status as different from peers, and they will typically bring up those concerns for discussion with their primary pediatric health care professional. The concern can be that they see their child as lagging behind their peers, but it can also be that they believe their child has skills that surpass their peers or is "gifted" in one or more areas of development or learning. Of course, when counseling the parent whose child is gifted, primary pediatric health care professionals can and should be more optimistic and reassuring than when counseling the parent whose child is at the other end of the cognitive spectrum, with significant delay or impairment; however, gifted children have potential for their own set of challenges and complications that a primary pediatric health care professional should be aware of in order to provide the best support to the family of a gifted child.

Gifted is a broad term that is poorly defined. Historically, it has indicated intellectual superiority, as demonstrated by results in the superior range on intelligence testing (IQ >120).[53,54] It has been a term used to describe children with particularly high ability in domains of intelligence that are most related to learning in an academic environment (linguistic and logical-mathematic abilities), but may also include children with other talents, in areas outside of academics (music, arts, sports, etc).[55,56] Early on, gifted children are often advanced in language and verbal abilities and demonstrate intuition and creativity that is precocious for their age.[54,55,57] Their advanced conversational ability, interests, sense of humor, and intellect can make them appear older than their actual age but also can make their age-appropriate social and emotional development appear to be more immature, in contrast. The asynchronous nature of their development is one of several notable characteristics often seen in gifted children (Box 10.3) and, along with a tendency toward sensitivity and perfectionism, can lead gifted children to experience emotional problems, such as self-criticism and depression.[54,58] Gifted children may underachieve academically related to this, or intentionally do so in order to fit in better with peers. By making them aware of these characteristics, primary pediatric health care professionals can help parents to better understand their gifted child and be prepared to anticipate the

Box 10.3. Characteristics of Gifted Children[a]

- Asynchronous development across domains
- Advanced language and reasoning skills
- Conversation and interests like older children
- Insatiable curiosity and perceptive questions
- Rapid and intuitive understanding of concepts
- Impressive long-term memory
- Ability to hold problems in mind that are not yet figured out
- Ability to make connections between concepts
- Interest in patterns and relationships
- Advanced sense of humor for age
- Courage in trying new pathways of thinking
- Pleasure in solving and posing new problems
- Capacity for independent, self-directed activities
- Talent in a specific area (drawing, music, math, reading, etc)
- Sensitivity and perfectionism
- Intensity in feeling and emotion

[a] Adapted from: Robinson NM, Olszewski-Kubilius PM. Gifted and talented children: issues for primary pediatric health care providers. *Pediatr Rev.* 1996;17:427–434.

support and enrichment their gifted child will need. With understanding and support from family and peers, gifted children can be happy and well adjusted, with a positive self-image and good social relationships.[59]

Although the US Department of Education states that gifted children "require services or activities not ordinarily provided in the schools,"[60] there is no federal requirement for distinct education of gifted and talented students.[58] Most states provide educational programs for gifted and talented students, but what is provided varies significantly among states and within communities. Some gifted programs only include certain age ranges, some schools provide in-class enrichment services while others "pull out" gifted children for specialized education and activities. Parents will often consult primary pediatric health care professionals for advice on educational programming and on parenting specific to their gifted child. The fact that gifted children are such a heterogenous group makes such guidance a challenge to provide, but some general information that can be discussed with families is included in Box 10.4.

Box 10.4. Guidance That Primary Pediatric Health Care Professionals Can Provide Parents of Gifted Children

- **Provide enrichment specific to a child's talents, but do not push young children too hard to excel.** All children, regardless of any particular talent, should be provided choices and allowed to be creative and variable in their activities.[a]
- **Treat gifted children as "normal."** The same household rules of discipline apply, regardless of giftedness, and gifted children should not be excused from chores or other expectations. Gifted children can also have the same problems as non-gifted peers, such as learning disabilities and attention-deficit/hyperactivity disorder.[b,c,d]
- **Advocate to make the gifted child's education as individualized as possible.** The best approach will not be the same for every gifted child. Some may be emotionally and socially mature enough for acceleration, such as skipping a grade or attending a higher-grade classroom for certain academic subjects. Alternatively, enrichment is a way to add depth to learning for a subset within the same age group, and may include pull-out gifted programs, honors classes, or special interest clubs.[e,f]
- **Make connections with parents of other gifted children.** Such partnerships among families can bring some organization to advocating for gifted educational opportunities; allow for children with similar interests to find one another; and also allow shared learning regarding access to resources, effective parenting techniques, etc.
- **Be aware of the personality traits your gifted child may be prone to have, but do not worry excessively about them.** The odds are that a gifted child will grow up to be successful and well adjusted. Being aware that a gifted child tends to be a perfectionist, for example, just might make a parent more likely to praise a child when they try something new, even if the outcome is imperfect. Reminding them it is acceptable and actually interesting to be "different" will help build a positive self-image.
- **Maintain awareness of legal and educational developments regarding gifted programming via Web sites and community advocacy chapters.** Reliable Web sites include the National Research Center on the Gifted and Talented and the National Association for Gifted Children.

[a] Schechter NL, Reis SM, Colson ER. The gifted child. In: Levine MD, Carey WB, Crocker AC, eds. *Developmental-Behavioral Pediatrics*. 3rd ed. Philadelphia, PA: Elsevier Health Services; 1999:653–661.

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[f] Jaffe AC. The gifted child. *Pediatr Rev.* 2000;21:240–242.

Suggested Resources

The Arc is a nonprofit organization dedicated to supporting families and individuals with development disabilities: http://www.TheArc.org.

National Dissemination Center for Children with Disabilities provides information about special education, No Child Left Behind, disabilities, and a range of other sources: www.nichcy.org.

Family Voices is a support network for those involved with children who have special health needs: www.familyvoices.org.

Educational materials and parent support information may be obtained at www.FragileX.org or www.fraxa.org.

Reliable Web sites about gifted and talented children include the National Research Center on the Gifted and Talented (www.gifted.uconn.edu/nrcgt.html) and the National Association for Gifted Children (www.nagc.org).

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Chapter 11

Speech and Language Development and Disorders

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Language acquisition is one of the most important components of a child's development. Language represents objects or actions in symbolic form and communicates ideas, intentions, and emotions. Effective communication is necessary for learning, social-emotional development and interactions, and effective functioning in society. Speech and language disorders are the most common developmentally disabling disorders of childhood,[1] and 30% of parents voice concerns about language development during primary care visits.[2] Functions of the pediatric primary care medical home include promoting language development, alleviating concerns about language development, and/or detecting language development problems. Early recognition and intervention are necessary to provide children with speech and/or language disorders with the best possible outcome.

Communication skill development begins at birth. Infants communicate nonverbally through facial expressions and gestures and verbally through sounds and primitive words, and they soon learn that speech is a more efficient means of communication. Language development occurs in an orderly and predictable manner for most children; however, variations can occur. Virtually any disruption in brain function can affect language acquisition; therefore, a variety of conditions affecting the brain are associated with language problems. Delays in comprehension and/or expression not associated with other developmental or neurologic problems are found in 7.5% to 10% of preschool children with a significantly higher proportion of boys being affected.[3]

The pediatric practitioner should be careful not to attribute cultural or gender differences as reasons for delayed language development. Children who learn 2 languages simultaneously follow the same pattern of speech and language development as monolingual language learners. The child may have a period when he or she mixes the 2 languages, but this should gradually disappear as language skills develop.[4]

Studies have shown that girls are more talkative (have more total words) than boys at all ages, with significant gender differences found between 1 to 2.5 years of age.[5] Although some boys may develop expressive language more slowly than girls, it is generally only by a few months and still within the accepted time frame. Language development is almost never delayed because the child "doesn't need to speak" (eg, "her big sister always talks for her"). There is a tremendous motivation to improve communication, as the use of verbal labels allows the child to meet needs more efficiently than pointing.

The term *language delay* implies the delay will resolve and the child will catch up at some point. However, more than 40% of children whose early language delays show improvement have later reading or cognitive difficulties.[6] Preschoolers with language disorders are at higher risk for language-based learning disorders, and social and behavioral problems.[7,8] Speech and/or language concerns should not be dismissed with reassurance that the child will "catch up," given the possibility of future difficulties and better outcomes with earlier detection of these problems.

Dimensions of Speech and Language

Speech produces complex acoustic signals that communicate meaning and is the result of interactions between the respiratory, laryngeal, and oral structures. This acoustic signal varies with regard to vocal pitch, intonation, and voice quality. The symbols need to conform to the language code so they can be decoded as meaningful communication.

Language involves both expressive and receptive components. Expressive language involves the communication of ideas, intentions, and emotions. Receptive language involves understanding what is said by someone else. Receptive language includes auditory comprehension (listening), literate decoding (reading), and mastery of visual signing.

Language has several components, as outlined in Table 11.1. The simplest "units" of language are *phonemes,* or individual sounds. Phonemes are combined to produce *morphemes,* which are the meaningful units of sound combined to produce a word. The *lexicon* (vocabulary) is the collection of all of the meaningful words in a language. *Syntax* (grammar) is the order of words in phrases and sentences. *Semantics* are the individual word and sentence meanings. The literal interpretation of words can be modified by *prosody* or vocal intonation. The social use of language is known as *pragmatics.*

Table 11.1. Components of Speech and Language

Term	Definition
Speech	
Intelligibility	Ability of speech to be understood by others
Fluency	Flow of speech
Voice and resonance	Sound of speech, incorporating passage of air through larynx, mouth, and nose
Language	
Receptive language	Ability to understand language
Expressive language	Ability to produce language
Phoneme	Smallest units of sound that change the meaning of a word, eg, "map" and "mop"
Morpheme	Smallest unit of meaning in language, eg, adding –s to the end of a word to make it plural
Syntax	Set of rules for combining morphemes and words into sentences (grammar)
Semantics	The meaning of words and sentences
Pragmatics	The social use of language, including conversational skills, discourse, volume of speech, and body language

Typical Speech and Language Development

The concept of "critical periods" is generally accepted for speech development in infancy and early childhood.[9] Table 11.2 shows the typical ages for attaining language milestones, although variability exists. Some skills may be demonstrated by the child during the office visit, while others may rely on parent report.

Pre-Speech Period

Children's communication development begins shortly after birth by social interactions with adults, necessary for bonding and having the infant's needs met. Infants are able to distinguish their mother's voice and show preference for familiar adults from early on in the first few months of life.[10] By a few months of age, the infant realizes that some sounds are important and specifically reacts to them. By 6 months of age an infant recognizes the basic sounds of her native language and has clear self-driven imitation of other's speech, with a rich interplay between the infant and the older individuals in her life.

Vocal development begins with phonation in the first few months (guttural or throaty sounds), then progresses to primitive articulation or cooing between 2 to 4 months of age. This expands to full vowel sounds by 4 to 5 months of age, single consonant sounds by about 5 months (eg, "ah-guh"), and well-formed babbling (repeated consonant-vowel pattern, "bababa") at around 6 months of age. Receptive language skills and social routines also develop in the first year of life. Six-month-old

Table 11.2. Speech and Language Milestones

Age	Receptive Language	Expressive Language
0–3 months	Alerts to voice	Cries, social smile Coos
4–6 months	Responds to voice, name	Laughs out loud Blows raspberries, clicks tongue Begins babbling
7–9 months	Turns head toward sound	Says "mama" and "dada" indiscriminately
10–12 months	Enjoys "peek a boo" Understands "no" Follows 1-step command with gesture	Says "mama" and "dada" appropriately Waves "bye-bye" Begins to gesture Shakes head "no" 1st word other than mama/dada
13–15 months	Follows 1-step command without gesture Points to 1 body part	Immature jargoning Up to 5 words
16–18 months	Points to 1 picture Points to 3 body parts and to self	Mature jargoning with true words Up to 25 words Giant words: "all gone, thank-you"
19–24 months	Begins to understand pronouns Follows 2-step commands Points to 5–10 pictures	Up to 50 words 2-word sentences Early telegraphic speech
25–30 months	Understands "just one" Points to parts of pictures	Uses pronouns appropriately Uses plural Speech is 50% intelligible
3 years	Knows opposites Follows 2 prepositions	250+ words 3-word sentences Answers "what" and "where" questions Speech is 75% intelligible
4 years	Follows 3-step commands Points to 4 colors	Answers "when" questions Knows full name, gender, age Tells stories Speech is 100% intelligible
5 years	Begins to understand left and right Understands adjectives	Answers "why" questions Defines simple words

children may pause momentarily when they hear their name called, and by 8 months pause at the word "no." At approximately 10 months of age children begin to gesture, holding their arms up to be picked up, waving bye-bye, and engaging in social games such as peek-a-boo.

Naming Period

Around their first birthday, children respond appropriately to requests for identification of familiar people or objects. Pointing is also used in a variety of contexts and is

an important expression of nonverbal communication. A child uses *protoimperative pointing* to a desired object in order to get an adult to obtain the object for him; the child "imperiously" implies "I need that!" by pointing to the object. *Protodeclarative pointing* is used when a child attempts to get an adult's attention to look at something of interest to the child and is a key component of *joint attention.* A child may also point to an object and vocalize in a questioning tone in an attempt to have an adult name that object for him.

Formal vocabulary development usually begins with the first word by an infant's first birthday and may include immature words such as "ba" for bottle or "cu" for cup. Vocabulary steadily expands to approximately 200 words by 2.5 years of age to more than 10,000 words by the time a child enters first grade. A child's receptive vocabulary is much larger than the number of words he or she uses expressively.

At 13 to 15 months of age, a child begins to *jargon,* or mimic mature conversation by varying intonation and pitch but initially does not use true words with this immature speech. Parents may report their child sounds like he is "talking in a foreign language." As new words are learned, they are incorporated into the child's speech patterns *(mature* jargoning). Between 12 and 18 months of age, a child uses single words to communicate desires (eg, "more"), emotions (eg, "no"), and specific objects (eg, "baby").[11]

Word Combination Period

Around 18 months of age, when a child has a 10- to 15-word vocabulary, she begins to combine words into phrases. Initially, word combinations tend to be "giant words" (ie, words the child often hears used together, such as "thank you" and "let's go"). Next, the child combines words into novel phrases (eg, "big truck"), and then into 2-word sentences (noun + verb, verb + object; eg, "want ___").[11]

Sentences

A rough rule of thumb is that 90% of children use 2-word sentences at 2 years of age, 3-word sentences by 3 years of age, and 4-word sentences by 4 years of age.[12] Sentences become increasingly complex as the child's understanding of language develops. By 3 to 4 years of age, children are able to understand and use prepositions (eg, "under" and "on"), adjectives, and adverbs. They begin to ask and answer questions. Semantics (word and sentence meaning) and syntax (grammar) improve. The child's pragmatic language skills also develop as the child learns the rules of the social use of communication.

Promotion of Language Development

As with all areas of development and behavior, promotion of language development is a function of the primary care medical home and is a component of the strength-based approach to pediatric care. Optimal language development occurs when

children experience stimulating environments with predictable and developmentally appropriate responses from adults. Some families may need guidance in strategies to encourage language development. Parents should be encouraged to "make their house a language house"—in essence, talking throughout daily activities with their children, no matter how mundane the activity. Reading aloud to young children has known positive effects on language and later reading decoding skills, as evidenced by research on Reach Out and Read, a primary care–based literacy promotion program. The American Speech-Language-Hearing Association has handouts on language stimulation activities for young children that can be downloaded, reviewed, and provided to families (http://www.asha.org/public/speech/development/Parent-Stim-Activities.htm). Parents should be reminded that television is not a substitute for language and should be avoided in the first years of life. Babies and young children do not get the same language stimulation from television as they do from personal, verbal interactions.

Variations in Development

Children's speech and language development is generally an orderly process, but variations do occur. When a parent raises a concern about language, the child will ultimately fall somewhere on the continuum of language developmental *variation,* language *problem,* or language *disorder.* Administering a well-standardized general developmental screen or language-specific screen will help the clinician determine where the child lies on this continuum. If the child passes the screen or has a borderline score, then watchful waiting is an appropriate next step. This includes discussion of language stimulation activities and referral to early intervention or early childhood programs (Head Start, preschool), with close follow-up and repeat screening in the medical home.

A speech-language or communication disorder is defined as impairment in the ability to receive, send, process, and/or comprehend verbal, nonverbal, or graphic symbol systems.[13] The most common variation in language development is language *delay.* The word *delay* inherently implies that catch-up will occur. Of children with early language delays, approximately 60% will catch up by 4 years of age with no persistent problems.[14] Another variation is language *dissociation.* This can occur either within the domain, as seen when developmental rates differ between expressive and receptive language, or between different domains (eg, language and motor skills). *Deviancy* of language development occurs when language development deviates from the norm, for example when children learn more advanced language-based concepts before they have mastered early language milestones. An example of this is a child who is able to recite the alphabet or TV jingles but is not yet able to communicate needs using words and phrases. Deviant language development can often be a sign of autism spectrum disorders (ASDs).[15]

Speech Disorders

Speech disorders reflect problems with creating the appropriate sounds representing the language symbols (the words). These problems include phonological *(articulation)* disorders, dysarthria, apraxia of speech, voice disorders, and speech fluency disorders. Speech disorders may or may not also include impairments in expressive language.[13]

Phonological or Articulation Disorder

A phonological or articulation disorder is characterized by the substitution, omission, addition, or distortion of phonemes and represents most speech therapy referrals. Children master sounds at different ages depending on the difficulty in producing the sound. In the first 2 years, children master simple sounds, including all vowels and the consonants /b/, /c/, /d/, /p/, and /m/. More difficult sounds, such as the consonants /j/, /r/, /l/, and /v/ and blends (ie, sh, ch, th, st) may not be mastered until 5 or 6 years of age.

Dysarthria

Dysarthrias are motor speech disorders that involve problems of articulation, respiration, phonation, or prosody as a result of paralysis, muscle weakness, or poor coordination.[16] Dysarthric speech is characterized by weakness in specific speech sound production and is frequently associated with cerebral palsy. Dysarthric speech may also encompass problems in coordinated breath control and head posture.

Apraxia of Speech/Dyspraxia

Apraxia of speech or *dyspraxia* is a speech disorder involving problems in articulation, phonation, respiration, and resonance arising from difficulties in complex motor planning and movement. This results in a child having difficulty correctly saying what he or she wants to say. The child has problems putting syllables together to form words, and has more difficulty with longer words rather than shorter, simpler words. It is not due to weakness of the oromotor musculature as seen with dysarthria. Therefore, apraxia/dyspraxia can be differentiated from dysarthria by the lack of association with other oral-motor skills, such as chewing, swallowing, or spitting. Other neurologic "soft signs," such as generalized hypotonia, may be present on examination.[17] Acquired apraxia/dyspraxia commonly results from head injury, tumor, stroke, or other problem affecting the parts of the brain involved with speaking and involves loss of previously acquired speech. It may co-occur with dysarthria or aphasia, a communication disorder impacting understanding or use of words caused by damage to the language centers of the brain. Developmental apraxia of speech is present from birth. Individuals with apraxia or aphasia might both have difficulty with verbal expression; however, apraxia on its own does not present a problem with language comprehension. Apraxia of speech is differentiated from an expressive language delay, in that children with expressive language delay typically follow a normal language trajectory but at a slower pace. Because individuals with

apraxia of speech demonstrate similar language concerns as individuals with expressive language disorders, it is necessary for examiners to administer an oral-motor examination to help differentiate the 2 conditions.

Voice Disorders

Variations in pitch, volume, resonance, and voice quality can be seen in isolation or in combination with a language delay. Impaired modulation of pitch and volume can be seen in children with ASDs, nonverbal learning disorders, and some genetic syndromes. Hyper- or hyponasal voice quality suggests anatomic differences or sometimes neurologic dysfunction, with hypernasal speech occurring secondary to velopharyngeal palatal incompetence and hyponasal speech arising from air impeded by large adenoids.[18] Velopharyngeal palatine incompetence (insufficiency) can be a marker of velocardiofacial syndrome.

Fluency Disorders

A fluency disorder involves the interruption in the flow of speaking. Examples of dysfluent speech include pauses, hesitations, interjections, prolongations, and interruptions. This is common in early childhood (age 2.5–4 years) and at that time is categorized as *normal dysfluency of childhood.* Persistent or progressive dysfluency is more likely *stuttering,* which arises in the preschool years for most affected children. Red flags indicative of pathological dysfluency requiring speech therapy include repetitions associated with sound prolongations (eg, "ca-caaaaa-caaaaat"), multiple part-word repetitions (eg, "ca-ca-ca-cat"), hurried and jerky repetitions with associated self-awareness and frustration, associated articulation problems, or a home environment with a low tolerance for stuttering or high pressure for verbal communication.[19]

Normal dysfluency usually improves over time, and parents should be instructed to avoid bringing attention to the dysfluent speech by correcting the child or reminding him to slow down. Parents also should speak more slowly and spend time with the child individually so that he can express himself in a noncompetitive environment. Encouraging families to take turns and not interrupt conversations during family activities is also beneficial. However, referral to a speech-language therapist is indicated if parents continue to be concerned.

Language Disorders

A language disorder, or specific language impairment (SLI), is an impairment in the ability to understand and/or use words in context, both verbally and nonverbally. The disorder may involve the *form* of language (phonology, morphology, and syntax), the *content* of language (semantics), and/or the *function* of language (pragmatics).[13] Language disorders are also classified as *receptive disorders* (trouble understanding others), *expressive* disorders (trouble sharing thoughts, ideas, and feelings), or *mixed receptive and expressive disorders.*

Receptive Language Disorder

Deficits in receptive language almost always occur in conjunction with expressive delays. There are situations where a child may appear to have an isolated receptive delay, but on careful evaluation, deficits in both areas are present. For example, a child with an ASD may appear to have normal or advanced expressive language skills due to extensive use of echolalia, but their functional communication delays are similar to their impaired receptive skills. Children with hydrocephalus (congenital or acquired) may have superficially appropriate or advanced expressive language skills but exhibit poor content of expression; known as "cocktail party syndrome."[20] In this case, receptive language lags behind expressive language and is felt to be secondary to hydrocephalus and related effects on the language centers of the brain.

Auditory processing disorder (APD) is a set of purported functional deficits in information processing despite normal auditory thresholds and is characterized as a disorder in receptive language. Auditory processing is what happens when your brain recognizes and interprets verbal information and sounds. The "disorder" part of APD means that something is adversely affecting the brain's processing or interpretation of the information heard. Children with APD often do not recognize subtle differences between sounds in words, even though the sounds themselves are clear, and may have difficulty comprehending verbal messages, especially in noisy environments, when others are talking, or when listening to complex information. Auditory processing entails many different processes at all levels of the nervous system, and poor-quality acoustic environments; peripheral ear functioning; behavioral factors involved in listening; and problems with the cochlea, nerve, brain stem, and cortex can all be involved. Empirical research is scarce validating modality-specific auditory-perceptual dysfunction. The diagnosis is made using behavioral tests supplemented by electroacoustic measures. Audiologic assessment is recommended, and the audiologist will give a testing battery to determine how well the child recognizes sounds in words. A comprehensive assessment with a speech-language pathologist, audiologist, psychologist, and physician is often necessary to diagnose this disorder, as it is often associated with and must also be differentiated from other language, learning, and attention problems. Attention-deficit/hyperactivity disorder (ADHD) and APD especially present overlapping symptomatology, specifically inattention and distractibility. Children with ADHD have difficulty completing tasks that challenge the central auditory nervous system. Clearly these entities have an overlapping clinical profile, and controversy still exists as to whether they are clinically distinct entities.

Interventions for APD are directed toward improving the listening environment, the acoustic signal, and auditory skills, and using bypass strategies. More research is needed to clearly identify APD problems and the best intervention for each child.

Expressive Language Disorder

These disorders represent a broad spectrum of delays, including developmentally inappropriate short length of utterances, word-finding weakness, semantic substitutions, and difficulty mastering grammatical morphemes that contribute to plurals or tense. Signs of weakness in expressive language include circumlocutions (using many words to explain a word instead of using the specific term), excessive use of place holders ("um," "uh"), nonspecific words ("stuff" or "like"), using gestures excessively, or difficulty generating an ordered narrative. Isolated expressive language delays are generally less indicative of organic pathology if not also associated with anatomic abnormalities.

Mixed Receptive-Expressive Language Disorder

Unless formal language testing using standardized instruments supports the presence solely of an isolated articulation disorder or specific receptive or expressive weakness, a child with a history of "language delays" should be presumed to have had some combination of language understanding and expression weaknesses. A variety of receptive-expressive subgroups have been described, including *verbal auditory agnosia* (impairment in interpreting the phonology of aural information and resultant limited comprehension of spoken language), *phonological-syntactic deficit* (extreme difficulty producing language with variable levels of comprehension), *semantic-pragmatic deficit* (expressively fluent with sophisticated use of words, but poor comprehension and superficial use of conversational speech), and *lexical-syntactic deficit* (word-finding weakness and higher order expressive skills weakness).[21] Table 11.3 outlines the various linguistic features seen with these subtypes of mixed receptive-expressive language disorders.

Impairment in both the receptive and expressive language domains raises the possibility of a more serious pathologic process, including intellectual disability (mental retardation), autism and other communication disorders, and deafness. Milder impairment often correlates with less severe forms of these disorders. An audiology evaluation can uncover hearing impairment or auditory processing disorder. Evaluation of a child's nonverbal problem-solving and adaptive skills can determine whether the child may have an underlying cognitive impairment. If there are concerns related to a child's social relatedness and social interactions, an ASD should be suspected. There is considerable overlap among these underlying causes, especially considering the wide spectrum of severity in each area (Table 11.4).

Disorders of Pragmatic Language

Deficits in pragmatic skills involve the inability to use language appropriately for social communication. A child may be unable to regulate social interactions or reciprocal body language or appropriately modulate their voice. They may stand too close or too far away from people or have improper voice pitch or volume. They

Table 11.3. Subtypes of Developmental Language Disorders

	Receptive			Expressive				
	Phonology	**Syntax**	**Semantics**	**Phonology**	**Syntax**	**Semantics**	**Pragmatics**	**Fluency**
Verbal auditory agnosia	↓↓	↓↓	↓↓	↓↓	↓↓	↓↓	↓↓	↓↓
Phonological-syntactic deficit	↓	↓	↓	↓	↓	↓	↓	↓
Semantic-pragmatic deficit	NL	NL	↓↓	NL	NL	↓↓	↓↓	NL or ↓
Lexical-syntactic deficit	NL	↓	↓↓	NL	NL or ↓	↓↓	↓	NL or ↓
Verbal dyspraxia	NL	NL	NL	↓↓	↓↓	↓	NL	↓↓
Phonological production deficit (articulation)	NL	NL	NL	↓	↓	NL or ↓	NL	NL or ↓

Abbreviations: NL, normal; ↓, below average; ↓↓, significantly below average.

Table 11.4. Developmental Delays in Intellectual Disability, Autism, Deafness, and APD

	Intellectual Disability	Autism	Deafness	APD
Expressive language	↓	↓↓→↑↑	↓↓	↓↓
Receptive language	↓	↓↓	↓↓	↓↓
Nonverbal IQ	↓	↓→↑	NL	NL
Adaptive (self-help) skills	↓	↓→↑	NL	NL
Social relatedness	NL for DQ	↓↓	NL	NL
Audiology assessment	NL	NL or ??	ABNL	NL

Abbreviations: APD, auditory processing disorder; NL, normal; ABNL, abnormal; DQ, developmental quotient; ↓, below average; ↓↓, significantly below average; ↑, above average; ↑↑, significantly above average.

commonly have difficulty initiating, maintaining, or terminating a conversation; modifying a topic for an audience; or including others in conversation. Pragmatic language disorders are often found in children with ASDs and nonverbal learning disorders.[22]

Relationship Between Early Language Delays and Dyslexia

Developmental continuities exist between oral (including speech) and written (reading and written expression) language disorders. Specific language impairments affect fewer children than does dyslexia. By age 5 years, roughly 7.5% of children have oral language skills below age expectations,[3] and 25% of these children meet criteria for dyslexia in second, fourth, and eighth grades. Learning to read involves the association between the sounds (phonemes) of spoken language and the symbols (letters, or *graphemes)* of printed words. Oral language skills, including phonology, semantics, grammar, and pragmatics, are the foundation for reading. Dyslexia is therefore characterized by reading decoding problems with the core problem based in phonological (speech) processing problems. However, broader language skills including vocabulary and comprehension processes are involved and can modify the impact of phonological difficulties. Children with wider language problems typically are at higher risk for reading comprehension deficits. Children with persistent SLI at 8.5 years of age have been shown to have pervasive problems with spelling, word-level reading, and reading comprehension at 15 years of age.[23]

Evaluation of Children Suspected of Having a Speech or Language Delay

Screening all children for delays in any of the developmental domains should be conducted at periodic intervals and whenever parents voice concerns about their child's development. The American Academy of Pediatrics has established guidelines for developmental screening, and recommends screening for all children at ages 9, 18, and 24 to 30 months[24] or at any time concerns are raised by a caregiver or pediatrician, with additional screening for ASDs at the 18- and 24-month visit.[15]

It is important to take parental concerns about speech or language development seriously, as these concerns are valid up to 75% of the time.[2] The evaluation of a child suspected of having a speech or language delay should involve a thorough history and physical examimation to determine the nature and extent of the problem but also uncover the etiology whenever possible.

History

It is important to determine whether the delay involves expression alone or both expressive and receptive language abilities. Isolated delays in receptive language are extremely rare. Sometimes a child may appear to have normal or advanced expressive skills due to complex echolalia of entire sentences, but their spontaneous

(or functional) language is delayed at least to the same degree as their receptive language (ie, with autism). Parental concerns are often focused on a child's inability to express himself and they may not be aware of associated delays in comprehension. Asking parents about any articulation or intelligibility difficulties is important. Inquiry about prenatal and delivery history, hearing loss, multiple ear infections, excessive drooling or difficulty feeding, and delays in other developmental domains will further elucidate an underlying cause. A detailed social history may uncover environmental causes of mild speech delay, including regression after a stressful event (eg, divorce, birth of a sibling), lack of stimulation, or the over-anticipation of needs by older siblings and parents. As biological factors contribute to language development, a detailed family history inquiring about speech and language or learning difficulties is important. Twin and family aggregation studies have demonstrated high heritability of language disorders.[25]

Physical Examination

A simple conversation with the child may be all that is needed to determine the extent of their comprehension, expression, and deficits in speech delivery. This includes all attempts to communicate, whether it is verbal (eg, babbling, jargoning, words) or nonverbal (eg, facial expressions, gesturing or pointing, presence of joint attention, eye contact, and body posture). A neurologic examination focused on oromotor skills should be completed, especially if there are also feeding difficulties or suspected speech apraxia. The oromotor examination should include imitation of tongue movements in all directions, observance of palatal elevation on phonation, and evaluation of structural integrity of the oral cavity.

Surveillance and Screening

Along with other developmental domains, surveillance of speech and language milestones should be performed at every well-child visit. This includes eliciting and attending to parental concerns, updating attainment of speech and language developmental milestones, determining risk and protective factors, and making accurate observations of the child. A 25% delay in milestone attainment is cause for concern and indicates the need for more detailed screening and/or assessment of speech and language skills. Red flags for delayed language skills are outlined in Table 11.5.

Several screening measures are available for quick evaluation in the pediatric office setting (Table 11.6). Parent-completed questionnaires, such as the Parents' Evaluation of Developmental Status (PEDS),[26] the Parents' Evaluation of Developmental Status: Developmental Milestones (PEDS:DM),[27] and the Ages and Stages Questionnaire (ASQ),[28] are good "broad-band" screens designed to assess multiple developmental domains. All 3 screens specifically screen language, and the PEDS:DM offers an assessment-level version that produces age-equivalent scores in expressive and receptive language. Parent questionnaires specifically designed to evaluate language that can be administered in a busy primary care practice include the Receptive-

Expressive Emergent Language Test (REEL),[29] the MacArthur-Bates Communicative Development Inventories (CDI),[30] the Language Development Survey (LDS),[31] and the Communication and Symbolic Behavior Scales: Developmental Profile Infant-Toddler Checklist (CSBS:DP-ITC).[32] If there are concerns about language and a possible ASD, the CSBS:DP-ITC[32] screens for social, speech, and symbolic communication in children between 6 to 24 months of age and helps to delineate between communication concerns in those realms. Direct interactive screening assessments include the Clinical Linguistic and Auditory Milestone Scale (CLAMS)[33] and the Early Language Milestones Scale (ELMS)[34] for children birth to 36 months.

Evaluation of articulation disorders begins with good surveillance. A formula for the expected conversational intelligibility levels of preschoolers talking to unfamiliar listeners is: AGE IN YEARS / 4 x 100 = % understood by strangers:

Child aged 1.0 = 1/4 or 25% intelligible to strangers
Child aged 2.0 = 2/4 or 50% intelligible to strangers
Child aged 3.0 = 3/4 or 75% intelligible to strangers
Child aged 4.0 = 4/4 or 100% intelligible to strangers[12]

Any child older than 4 years with a speech intelligibility score of less than 66% (ie, less than two-thirds of utterances understood by unfamiliar listeners) should be considered a candidate for intervention.[35]

It may not be practical to formally assess specific language-based learning problems in a school-aged child in the pediatric office setting. However, a few surveillance questions may help identify the presence of difficulties in language-based learning (see Box 11.1).

Table 11.5. Red Flags for Delayed Language Development

Age	Milestone
6 months	No cooing responsively
10 months	No babbling
12 months	No basic gesturing (waving bye-bye, holding arms out to be picked up)
18 months	No words other than mama, dada No understanding of simple commands No pointing to what he wants
24 months	<50 words No 2-word phrases <50% intelligibility
36 months	No 3-word sentences <75% intelligibility
4–5 years	Not able to tell a simple story

Table 11.6. Screening Measures for Speech and Language Delays

Parent Questionnaires	Developmental Age Range	Web Address
General Screens		
Ages and Stages Questionnaire	0–5 years	www.agesandstages.com
Parents Evaluation of Developmental Status (PEDS)	0–8 years	www.pedstest.com
Parents Evaluation of Developmental Status: Developmental Milestones (PEDS: DM)	0–8 years	www.pedstest.com
Language-Specific Screens		
MacArthur-Bates Communicative Development Inventories (CDI)	0–3 years	www.brookespublishing.com
Language Development Survey (LDS)	18–35 months	www.aseba.org
Receptive-Expressive Emergent Language (REEL)	0–3 years	ags.pearsonassessments.com
Communication and Symbolic Behavior Scales-DP Infant Toddler Checklist (CSBS: DP-ITC)	6–24 months	www.brookespublishing.com
Directly Administered Screens		
Clinical Linguistic Auditory Milestone Scale (CLAMS)	Up to 36 months	www.brookespublishing.com
Early Language Milestones Scale (ELMS)	Up to 36 months	www.proedinc.com

Box 11.1. Surveillance Questions for Language-based Learning Problems

1. Does she/he have trouble expressing her/his thoughts?
2. Is it difficult for her/him to understand or follow directions?
3. Does she/he express herself/himself through gestures rather than verbally?
4. Does she/he have trouble finding the correct word? (word retrieval)
5. Does she/he confuse words that sound alike (eg, tornado for volcano)?

   (auditory discrimination)
6. Does it seem to take a long time for her/him to understand direction or answer questions? (processing speed)
7. Does she/he seem to have to repeat things (out loud or to self) in order to understand them? (processing speed)
8. Can she/he tell you the letter that comes after "s" without going through the alphabet? (could also use days of the week, months, etc) (sequential processing)

Other Tests

If a speech or language delay is suspected, the child should be referred for a formal audiology examination, as a child may not have apparent hearing deficits by history. Even mild hearing loss can cause language delays and may not be picked up by newborn hearing screening. If there are concerns for an ASD, an autism-specific screening tool should be used in conjunction with general developmental or language-specific screening.

If a child is suspected of having a language disorder and fails a language screening test, in addition to hearing testing, referral to an early intervention program (EIP) for evaluation by a speech-language pathologist is recommended. Referral to a developmental-behavioral or neurodevelopmental pediatrician is also recommended if there is a history of language regression or if there are delays in other areas. Specific disorders in the differential for language regression include ASDs, Rett syndrome, or Landau-Kleffner syndrome (seizures accompanied by acquired aphasia).[36]

Detailed genetic and neurologic evaluations for isolated speech and language impairments are of low yield, and an underlying etiology will be determined in less than 5% of cases. If hypernasality is noted with suspected velopharyngeal insufficiency, then further investigation for velocardiofacial disorder is indicated, including referral to otorhinolaryngology and florescent in-situ hybridization study for 22q11.2. If a child has dysmorphic features or is found to have global developmental delays, then a full evaluation is recommended. This evaluation varies with the risk factors and findings and may include brain imaging, electroencephalogram, genetic testing, and/or metabolic testing.[37]

Treatment

Even when there is a question regarding the underlying diagnosis, all children suspected of having speech or language impairment should be referred promptly to their local EIP. Early intervention programs enrich a child's language experience through both parent training and provision of language-stimulating preschool environments. In addition, immediate speech and language therapy, which can be provided as a component of an EIP, has been shown to improve auditory comprehension and phonological disorders.[38,39] Additionally, speech and language therapy may prevent further delays and help reduce behavior difficulties associated with frustrated attempts to communicate.

Treatment of speech-language disorders includes 3 components: causal, habilitative, and supportive. Causal treatment is focused on repairing defects, correcting dysfunction, or eliminating factors that contribute to the language problem (eg, cleft palate repair, hearing aids). Habilitative treatment is designed to directly improve the child's language skills (ie, speech-language therapy, counseling of parents to actively engage in the child's language development). Supportive treatment aims

to boost language acquisition (eg, training programs for speech-related skills, increasing social contacts).

Goals for treatment depend on the nature of the child's speech or language impairment. Children aged 0 to 3 years can obtain services through the Individuals with Disabilities Education Act, Part C.[40] School-aged children may receive services through the public school system. If additional services are needed, or if the child lives in an area where there is a shortage of therapists in the school system, therapy is also available on an outpatient basis. The overall goal is to communicate with others, whether by spoken language or nonverbally through the use of sign language or communication systems, such as the Picture Exchange Communication System or an augmentative communication device. Some parents may voice concerns that early use of sign language or another communication system will impair a child's ability to speak, but there is evidence that using these systems may actually enhance a child's speech and language development.[41]

In addition to therapy, parent education that focuses on language stimulation activities is essential. Structured/stimulating child care centers, preschool programs, or "Mother's day out" programs are also beneficial, particularly in children with isolated non-pathologic speech-language delay reflecting developmental variation or lack of a stimulating home environment.

While therapy may improve the degree of impairment and prognosis, many children do not "outgrow" speech and language disorders, although these disorders manifest in different ways over time. A child who has early language delays may have difficulty in reading, written expression, learning a foreign language, or learning appropriate social interactions.[42,43] Developmental promotion of language and early identification and intervention for speech and language disorders are vital to provide the greatest long-term functional benefits.

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Chapter 12

Social and Emotional Development

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Healthy social and emotional development sets the foundation for promoting all other domains of children's development. After all, children learn to think, reason, communicate, run, jump, climb, care for themselves, and play in the course of social interactions with caregivers and peers. Children's development progresses in the context of the dynamic transactions between their biological and genetic tendencies and their life experiences. Life experiences are rooted in social and emotional relationships. Children who grow up in supportive, predictable, and nurturing environments are better prepared for a healthy, productive, adulthood and healthy, life-long relationships.

During the first 18 months of life, the social and emotional areas of the brain grow and develop more rapidly than the language and cognitive areas.[1] These right brain nonverbal systems build from social-, relational-, and attachment-based experiences and create the emotional regulation and stress regulatory systems of the body that may last a lifetime. Therefore, the quality of the child's early social and emotional experiences may have lasting implications. The young child who is raised in a nurturing, predictable, and safe environment is likely to be more resilient when faced with stressful experiences later in life. Conversely, the young child who is faced with stressful, chaotic, or traumatic early experiences may suffer lifelong consequences.

As children grow and develop through early childhood, middle childhood, and adolescence, the quality of their interactions with caregivers, extended family, peers, teachers and coaches, and other community members have an ongoing impact on their social and emotional development. The monitoring of social and emotional development and family relationship patterns from infancy through adolescence is a vital component of health supervision.

America's Promise Alliance has identified 5 key resources or "promises" that correlate with success in youth and adulthood.[2] Children who grow up in homes with caring adults, in safe places where they have opportunities for constructive use of time, with a healthy start and healthy development, with effective education for marketable skills and lifelong learning, and with opportunities to make a difference

through helping others have a much greater chance of experiencing success.[2] Teens who receive 4 of these 5 promises are nearly two-thirds more likely than those who have only zero or one promise to be generous, respectful, and empathetic, and to resolve conflicts calmly. Younger children with 4 or more promises are twice as likely as their peers to be socially competent than their peers with zero or one promise. Unfortunately, research performed by America's Promise Alliance indicates that more than two-thirds of children from 6 to 17 years of age are not receiving sufficient resources to place them on a path for success.

Similarly, Maughn and McCarthy[3] have proposed that adverse childhood experiences, especially inadequacies in early parental care, are associated with higher rates of both acute and chronic psychosocial disorders in adulthood. Most long-term sequelae seem to depend on a series of shorter-term links, some related to continued elevated risks of environmental adversity, others related to psychological vulnerabilities and resiliencies and problems in intimate social relationships. Primary pediatric health care professionals are well positioned to work with families to promote social competence at individual, practice, and community levels, and potentially reduce the risk for long-term concerns in social and emotional health.

Key Points

- Social and emotional development progresses through predictable stages.
- Consider the developmental and environmental context when assessing any social-emotional concerns.
- Clearly identify and address the family's concerns.
- Perform surveillance using open-ended trigger questions.
- Attend to your observations of parent-child interaction and the child's interaction with you.
- Use standardized screening tools that match the level of risk in the population you serve.
- Explore options for offering evidence-based interventions in the medical home.
- Use evidence-based strategies for behavior management as part of your anticipatory guidance.
- Promote community-based linkages and resources to support families to promote resilience and to address social-emotional concerns.

Social and Emotional Milestones

Social and emotional development progresses through predictable stages in healthy children. Monitoring the milestones of social and emotional development is an important component of health supervision from infancy to young adulthood. Multiple observations will provide opportunities to identify variation from the expected path of development early, facilitating early identification and treatment. Failure to achieve expected milestones at any age should trigger further investigation and consideration of referral.

During the infant and toddler period, the major tasks of social and emotional development are to experience and regulate emotions, develop secure relationships, and begin to explore and learn. Even in the first few months of life, the infant's temperamental characteristics will emerge. Chess and Thomas[4] have described 3 functional constellations of temperament. The "easy" group includes 40% of children. These children demonstrate regularity; positive approach responses to new situations; high adaptability to change; and a mild to moderate, predominantly positive mood. About 10% of children have a "difficult" temperament. These children have irregularity in biological functions; negative withdrawal responses to new situations; poor or slow adaptation to change; and intense, often negative moods. The "slow to warm up" temperament is seen in 15% of children. This group combines mild, negative responses to new situations with slow adaptability after repeated exposure. With additional opportunities with new experiences, these children will eventually show quiet and positive interest. Not all children fit into 1 of these 3 groups. Others will have a mix of temperamental characteristics. It is vital that the child's temperament be considered when assessing social and emotional development and parent-child interaction. The most important factor may be the "goodness of fit" between the child's temperament and the temperaments of his caregivers.

Information on the stages of social and emotional development outlined below is adapted from Bright Futures.[5]

The newborn is most responsive during short periods in a quiet, alert state. She recognizes the unique smell of her mother, can hear and prefers her parents' voices, responds favorably to gentle touch and withdraws from unpleasant touches, and can imitate simple facial expressions from a distance of 7 to 8 inches.

Throughout the first 2 months, the infant becomes increasingly capable of consoling and comforting himself while becoming increasingly alert, smiling responsively, and responding to calming actions when upset. By 4 months, he smiles spontaneously, initiates social interactions, and shows an even greater capacity to comfort himself. He has discovered that he can control the movements of his hands and may use them to console and comfort.

By 6 months, she recognizes familiar faces and is beginning to notice strangers. She has sustained interactions and jointly attends to actions and objects of interest to her parents and regular caregivers. At 9 months, she has clear apprehension with strangers, and actively seeks out her parents for play, comfort, and assistance. By 12 months, she has a strong attachment with her parents and significant caregivers and shows distress on separation from her parents. The 12-month-old is playing interactive games like peek-a-boo and pat-a-cake and is using gestures to wave bye-bye and to indicate interests and needs. She will hand a toy or book to her parent when she wants to play or hear a story.

By 15 months, he is very interested in imitating whatever he sees. He may start to help with simple household tasks and will listen actively to stories. His interactions with his parent or caretaker should be robust, complex, continuous, and goal-directed. By 18 months, his temperament will be more and more evident in his approach to participating in new or familiar group settings. He may be interactive or withdrawn, friendly or aggressive. He will show increasing willingness to separate and explore on his own but will want his parent in close proximity for periodic reassuring check-ins or encouragement. He is spontaneous with affection and laughs in response to others.

The 2-year-old is becoming increasingly independent. She refers to herself as "I" or "me." She may have a special attachment to a book, a toy, or a blanket to help her make the transition to independence more smoothly. She will show an eagerness to share, show, and engage with the parent, to the parent's delight. She plays alongside other children and is showing more pretend play. At 2.5 years, imaginative play is clearly evident, and she is showing evidence of symbolic play, making an object into something new or different. Her play now includes other children. As she struggles with her newly found independence, she may be fearful of unexpected changes in daily life.

By 3 years, he will be showing much more elaborate imaginative play with themes and story lines. He enjoys interactive play and is delighted to show his parents his capacity for independence with feeding, dressing, and toileting. The 4-year-old is working to establish a comfortable place in an expanding world. Depending on his temperament and the history of the relationship patterns, he may be cooperative, friendly, and responsive, or withdrawn, aggressive, or defiant. Extremes of behavior may be seen at times of stress. He is also able to see himself as an individual yet equally enjoy demonstrating his relational capacity as being a communicator and entertainer. He knows his gender and age and can describe his interests and what he does well. He has favorite toys and favorite stories. He spends more time in fantasy play.

The 5- and 6-year-old is successful at listening, attending, and following simple rules and directions. However, she is also likely to test those rules. She is becoming comfortable with spending more time with a peer group. As she approaches 7 to 8 years of age, she more fully understands rules, relationships, and mores. She will consistently show cooperation and attention and is capable of taking on family responsibilities and chores. As her moral development progresses, her coping skills emerge. Her beliefs may be tested by her peers, as she turns more to them and other adults for new ideas and activities. She may have a best friend and will usually identify most with children of the same gender that have similar interests and abilities. By 9 to 10 years, her peer group takes on greater importance. Her growing need for independence from her family will often be a valuable incentive for her to make contributions at home in order to earn privileges with her friends. She will demonstrate

increasingly responsible and independent decision-making. Her feelings of self-confidence can be bolstered by descriptive praise, affection, and quality time in a nurturing relationship with her family.

The Association of Maternal and Child Health Programs and the National Network of State Adolescent Health Coordinators have identified critical developmental tasks that indicate healthy progression through adolescence from 10 to 24 years of age.[6]

In the context of caring, supportive relationships with family, other adults, and peers, a child will have increasing opportunities to engage in positive activities in his community that will promote a sense of self-confidence, hopefulness, and well-being. As the 11- to 14-year-old approaches adolescence, there is a greater drive for independence and growing commitment to the peer group. As a result, he may engage in risky behavior to impress his peers. He will respond well to authoritative parenting that is firm, accepting, and democratic. Social networks will form, break down, and then form again. He will usually cope well with these stressful experiences and should be encouraged and supported while making more and more independent decisions. As the adolescent approaches young adulthood, school and work and its activities are the central focus of his life. It becomes increasingly important to monitor for emotional problems and risk-taking behaviors.

By understanding the expected path of social and emotional development, from infancy through adolescence, the health care professional will be well positioned to anticipate areas for timely guidance and to identify variations in development that warrant intervention (Table 12.1).

The third edition of *Bright Futures: Guidelines for Health Supervision of Infants, Children, and Adolescents* outlines the components of health supervision and places special emphasis on promoting child development, mental health, and community relationships and resources as key themes to be addressed in well-child care visits.[5(pp210–218)] Bright Futures recommends that each preventive health visit include establishing a context for the visit, setting priorities for the visit, a review of the interval history, observation of parent-child interaction, surveillance of development, a physical examination, screening, immunizations, other practice-based interventions, and offering anticipatory guidance. The American Academy of Pediatrics (AAP) has published recommendations for developmental surveillance and screening of young children.[7] The components of surveillance include eliciting and attending to parents' concerns, maintaining a developmental history, making accurate observations of the child, identifying risk and protective factors, and documenting the process and findings. These components can be reviewed in the context of the Bright Futures framework. All of these elements can contribute significantly to the understanding of a child's social and emotional development and assist in identifying opportunities to build on strengths within the child, family, and community, as well as identifying concerns that may require additional support and intervention.

Table 12.1. Social-Emotional Milestones[5,6]

Age	Milestones
Newborn	Most responsive in quiet, alert state Recognizes the unique smell of her mother Prefers her parents' voices Responds favorably to gentle touch and withdraws from unpleasant touches Imitates simple facial expressions from a distance of 7–8 inches
2 mo	Consoles and comforts self Increasingly alert Smiles responsively Responds to calming actions when upset
4 mo	Smiles spontaneously Initiates social interactions Shows greater capacity to comfort self Control the movements of his hands and may use them to console and comfort
6 mo	Recognizes familiar faces and is beginning to notice strangers Sustains interactions Jointly attends to actions and objects of interest to her caregivers
9 mo	Clear apprehension with strangers Seeks out her parents for play, comfort, and assistance Plays interactive games like peek-a-boo and pat-a-cake Waves bye-bye Looks preferentially when name is called
12 mo	Proto-imperative pointing; uses gestures to indicate needs Hands a toy or book to her parent when she wants to play or hear a story
15 mo	Proto-declarative pointing to indicate interests Imitates whatever he sees Helps with simple household tasks Listens actively to stories
18 mo	Temperament will be more and more evident in new or group settings Willing to separate and explore on his own but will want his parent close by Spontaneous with affection Laughs in response to others
2 y	More independent Refers to herself as "I" or "me" May have a special object to assist with transition to independence Plays alongside other children Shows more pretend play
2.5 y	Shows imaginative play Shows symbolic play, making an object into something new or different Play includes other children May be fearful of unexpected changes in daily life
3 y	Shows much more elaborate imaginative play with themes and story lines Enjoys interactive play Independent with feeding, dressing, and toileting

Table 12.1. Social-Emotional Milestones[5,6] *(continued)*

Age	Milestones
4 y	Extremes of behavior may be seen at times of stress Sees himself as an individual Knows gender and age Describes interests and strengths Has favorite toys and favorite stories Spends more time in fantasy play
5–6 y	Listens, attends, and follows simple rules and directions Tests rules Spends more time with a peer group
7–8 y	More fully understands rules, relationships, and mores Shows cooperation and attention Takes on family responsibilities and chores May have a best friend Identifies most with same gender children with similar interests and abilities
9–10 y	Peer group takes on greater importance Increasingly responsible and independent decision-making
11–14 y	Greater drive for independence and growing commitment to the peer group May engage in risky behavior to impress his peers Forms and breaks down social networks Copes well with stressful experiences Makes more and more independent decisions
15–24 y	School, extracurricular activities, and work become central focus Forms caring, supportive relationships with family, other adults, and peers Participates in the community Demonstrates resilience with everyday life stressors Increases independence in decision-making Shows self-confidence and hopefulness

Context

When assessing a child's social and emotional development, it is important to consider a number of contexts. At any encounter, it is vital to consider the child's age, developmental level, and circumstances in order to accurately interpret any concerns regarding social and emotional development.

Squires et al[8(pp6–7)] recommend considering the following variables: setting/time, development, health, and family/cultural considerations. First, where and when a behavior occurs may lead to different interpretations of the behavior. A child who is hitting or biting his younger sibling may have that behavior accidentally rewarded when it draws attention from his mother. However, the same behavior may quickly disappear in a child care setting where the child is redirected and given descriptive praise for gentle touching and has models for appropriate play with peers. Timing makes a difference as well. A child who experiences a major traumatic event early in life may experience lasting effects, while the same experience for an adolescent with strong coping skills may lead to only a temporary setback.

Second, it is vital that expectations be matched to the child's overall level of development, including cognition, language, academic, and motor skills. Children with isolated or global delays in development may develop secondary behavioral or emotional symptoms due to frustration, or a tendency to avoid tasks that may be too difficult for them. There may be wide variation in achieving various milestones. Opportunities for serial observations allow the health care professional to establish whether the child is progressing as expected over time. However, the health care professional should not dismiss problems based on the assumption that they are simply due to normal variation.

Third, health variables may also affect children's social and emotional functioning. The child with obstructive sleep apnea may be irritable and inattentive at school. A child with atopic dermatitis may be so uncomfortable with pruritis that she may be noncompliant. Children with special health care needs may develop a pattern of learned helplessness because of their chronic illness and its accompanying challenges.

Finally, the patterns of interactions and communication within family relationships in the context of family values and culture have a significant role in determining children's social and emotional responses. Some cultures allow their emotions to flow freely and openly, while others expect children to hold their emotions inside. Various cultures have different expectations regarding when children should sleep on their own, be weaned, master toileting, and dress themselves.[9] Children who are growing up in families affected by parental mental illness,[10] marital discord and domestic violence,[11] high levels of stress, and poverty[12] may show changes in their social and emotional functioning in an attempt to cope with these challenges.

Priorities

The needs and concerns of the family should be given first priority during each visit. Since as many as 25% of children have social or behavioral problems,[13] it is likely that concerns about social and emotional development will be raised during many preventive health visits. It may be necessary to give up one's professional priorities for the visit in order to address the family and child's concerns. Bright Futures recommends that maternal and family functioning be a priority for all health supervision visits.[5] A focus on social and emotional functioning and parent-child interactive patterns should be emphasized during early infancy, between 18 and 30 months, and at the transitions to kindergarten, middle school, and high school. These times coincide with anticipated challenges to social and emotional competence.

Social-Emotional Surveillance and Interval History

When social and emotional development is identified as a priority for the visit, several open-ended questions can set the stage for a productive, targeted discussion. Parents' concerns about their child's development, behavior, and learning are

substantiated at least 70% of the time.[14,15] Therefore, beginning the interview by asking whether there are any concerns about the child's development, behavior, or learning can be very productive.

Additional trigger questions to assess social and emotional functioning include[16]

- How are things going for you as a parent?
- What changes have you seen in your child's development or behavior since the last visit?
- What does your child do really well? With you? At school? In the community?
- What kind of child is she? Tell me about your child's personality or temperament.
- What are your child's favorite play or recreational activities?
- What do you enjoy doing together?
- Have there been any significant changes or stressful events in the family since the last visit?
- What do you find most difficult about caring for your child now?
- How do you and your child solve problems?

The Classification of Child and Adolescent Mental Diagnoses in Primary Care: Diagnostic and Statistical Manual for Primary Care (DSM-PC)[17] provides a framework for assessing protective factors and environmental challenges. When faced with a child or family who appears at risk, this framework can provide a structured approach to identify opportunities to further support social competence and for remediation.

Once children reach the age of 3 or 4 years and through adolescence, these questions can be adapted and asked directly to the child. The health care professional may be reassured by the response or may feel a need to probe further.

Children have a full set of emotions by 3 years of age.[18] By 5 years of age, most children are skilled at describing their feelings. It is appropriate to ask school-aged children to describe examples of what leads them to feel various emotions.

- What makes you happy? Sad? Mad?
- What makes you afraid?
- What do you worry about? Do you have any worries about your body or about your health?
- Are there things you are afraid might happen to you or to people you care about?
- Have you ever thought about hurting yourself or running away? If so, have you made a plan? If you ever do think of hurting yourself, is there an adult you can talk to about your feelings?
- Do you have a best friend? What do you like to do with your friends? Are you being bullied?

- What would you like to change about yourself? What would you like to change about school? What would you like to change about your family?
- If you had three wishes, what would they be?

All of these questions can assist the health care professional in determining whether further history, evaluation, or referral may be indicated.

As children approach middle childhood and adolescence, it is often important to interview the child alone. This provides an opportunity to discuss risk-taking behaviors, or to identify any perceived risk within the family system.

Observation of Parent-Child Interaction

Beginning in the newborn period, and on through adolescence, observation of parent-child interaction can offer insight into family relationships, parenting style, and the child's social and emotional well-being. Often much can be learned by observing what is happening when the health care professional opens the examination room door. Is the infant held in the mother's lap in a loving embrace with good eye contact and nice vocalizations? Or is the child tearing apart the office and climbing on the cupboards while the father is intent on reading a magazine? Neither of these observations should lead to a diagnostic conclusion, but one might offer the opportunity to provide descriptive praise, while the other might serve as an opportunity for incidental teaching. Yet, it is within the family relational and behavioral patterns that one sees, firsthand, the strengths and vulnerabilities of social-emotional developmental competency.

It is also important to attend to the quality of the interaction between the child and the health care professional. Are the child's responses developmentally appropriate? Does the child make good eye contact? Is the toddler showing joint attention skills and proto-imperative and proto-declarative pointing? Is the child's mood and affect appropriate or flat or withdrawn? Is the child anxious about what is going to happen next? Is the parent or the child flushed or visibly nervous? These observations may add to the assessment of the child's social and emotional functioning.

Healthy children are better prepared to explore and learn. Therefore, fulfilling the recommended immunization schedule will help set the foundation for healthy social and emotional development. In addition, observation of the child's behavior during the immunization process can provide additional insight into her coping skills and into the parent-child interaction. The administration of immunizations may provide an opportunity for the health care professional to model and teach coping and relaxation skills.

Physical Examination

The physical examination offers an additional opportunity to observe the child's adaptation to and compliance with the health care professional's requests, and to

observe the parent's response as well. Is the child cooperative or overly anxious or defiant? Is the parent encouraging and supportive or overly protective or coercive? The child's vital signs might be an indicator of stress if the child's blood pressure or heart rate is high for no other apparent reason. It is important to look for any subtle dysmorphic features that may indicate a genetic disorder that may present with developmental delays, including social or emotional concerns. The neurologic and mental status examinations may contribute to an understanding of any social and emotional concerns. It is also important to look for any signs of accidental, intentional, or self-inflicted injuries.

Screening

Perrin and Stancin[19] have published a comprehensive review of behavioral screening that includes an extensive list of available instruments. They emphasize that a practitioner can use their own interview or questionnaire, but there are advantages to using a published standardized questionnaire. These include the ability to compare responses with national norms, compare responses from several observers, and quantify and document change over time. A combination of an interval history, as outlined above, with a standardized questionnaire, if concerns are identified, may be most beneficial. At the same time, it may be beneficial to routinely administer a standardized tool to assess social and emotional functioning of children and their caregivers at key well-child visits (Table 12.2). Universal screening may provide an opportunity to introduce a difficult topic and reduce the tendency to rely on normal variation to explain concerns. For example, the AAP has recommended that all children have a standardized screen for autism, a primary disorder in social relatedness, at 18 and 24 months of age.[20]

Caregiver Functioning

There is growing attention to the importance of screening for maternal depression, especially in the first 6 months when postpartum depression is prevalent, but also at any time there is a concern regarding a child's behavior. Observation without screening tools fails to identify most mothers with depressive symptoms. Pediatricians detect only 25% of mothers with depressive symptoms.[21]

Several tools are available to screen for maternal depression. The Edinburgh Postnatal Depression Screening is a widely used 10-item tool that includes anxiety symptoms as well as symptoms of depression.[22] The Center for Epidemiologic Studies Depression Scale (CES-D) is a 20-item measure of psychological distress.[23] The Patient Health Questionnaire-2 (PHQ-2) has 2 questions that assess the presence and frequency of concerns about mood and anhedonia in the past 2 weeks.[24] It has been validated in primary care and obstetric settings. Olson et al[25]evaluated the PHQ-2 in a pediatric setting and found that it added no more than 3 additional minutes to the length of the visit. Prolonged discussion was uncommon, requiring 5 to 10 minutes in 3% of well visits and more than 10 minutes in just 2% of well visits.

Table 12.2. Screening and Assessment Tools

Screening	Instrument	Age Range	Time	Source
Caregiver functioning	Edinburgh Postnatal Depression Screening	Adults	5 min	www.dbpeds.org/media/edinburghscale.pdf
	Center for Epidemiologic Studies Depression Scale	Adults	5–10 min	www.chcr.brown.edu/pcoc/cesdscale.pdf
	Patient Health Questionnaire-2	Adults	1 min	www.cqaimh.org/pdf/tool_phq2.pdf
	Depression, Anxiety, and Stress Scale	17 y and up	10 min	www.psy.unsw.edu.au/dass/
	Parenting Stress Index–Short Form	Parents of 1 mo–12 y	10 min	www4.parinc.com
	Adverse Childhood Experiences Score	Adults	5 min	www.acestudy.org/
Temperament	Carey Temperament Scales	1 mo–12 y	15 min	www.b-di.com
Infancy to early childhood	Ages & Stages Questionnaire: Social-Emotional	6–60 mo	10–15 min	www.brookespublishing.com
	Communication and Symbolic Behavior Scales: Developmental Profile, Infant Toddler Checklist	6–24 mo	5–10 min	www.brookespublishing.com
	Brief Infant-Toddler Social Emotional Assessment	12–36 mo	7–10 min	http://pearsonassess.com
Early childhood to adolescence	Eyberg Child Behavior Inventory	2–16 y	5 min	www4.parinc.com
	Pediatric Symptom Checklist	4–18 y	5 min	www2.massgeneral.org/allpsych/psc/psc_forms.htm
	Pictorial Pediatric Symptom Checklist	4–18 y	5 min	www2.massgeneral.org/allpsych/psc/psc_forms.htm

Table 12.2. Screening and Assessment Tools *(continued)*

Screening	Instrument	Age Range	Time	Source
Assessment				
Multidimensional	Infant-Toddler Social Emotional Assessment	12–36 mo	25–30 min	http://pearsonassess.com
	NCAST Parent Child Interaction Feeding and Teaching Scales	12–36 mo	Feeding: length of meal Teaching: 1–6 min	www.ncast.org
	CHADIS	Birth–adolescence	10–20 min	www.chadis.com
	Achenbach System of Empirically Based Assessments	1.5 y–adult	20–30 min	www.aseba.org
	Behavioral Assessment Scale for Children, 2nd Edition	2–21 y	10–20 min	http://pearsonassess.com
	Connors Comprehensive Behavior Rating Scales	3–17 y	10–20 min	www.pearsonassessments.com
	Child Symptom Inventories-4	3–18 y	20–30 min	www.checkmateplus.com
	Vanderbilt Parent and Teacher Assessment Scales	6–12 y	5–10 min	www.nichq.org
Single dimension attention-deficit/ hyperactivity disorder	Conners 3	6–18 y	5–20 min	www.pearsonassessments.com
	Attention Deficit Disorders Evaluation Scales	4–18 y	12–15 min	www.hes-inc.com
	Brown Attention Deficit Disorder Scales	3 years–adult	10–20 min	www.drthomasebrown.com
Single dimension anxiety/depression	Beck Youth Inventories	7–18 y	5 min	http://pearsonassess.com

Additional tools are available to assess broader dimensions of parental and caregiver functioning. For example, the Depression, Anxiety, and Stress Scale (DASS)[26] is a 42-item instrument designed to measure depression, anxiety, and tension/stress. It can be downloaded at no charge. The Parenting Stress Index[27] is available in 36-item and 120-item forms. The Parenting Stress Index is designed for the early identification of parenting and family characteristics that raise the risk for difficulties in social and emotional functioning and parent-child interaction. It can be used with parents of children as young as 1 month up to 12 years of age. The Adverse Childhood Experiences (ACE) score is used to assess the total amount of stress during childhood and has demonstrated that as the number of ACE increase, the risk for physical and mental health problems increases in a strong and graded fashion.[28]

Temperament

Parents will benefit from understanding their child's temperament and its influence on parent-child interaction as well as adaptation to new experiences. Medoff-Cooper and associates[29] have developed a number of questionnaires that can assist parents and health care professionals in understanding their child's temperamental characteristics as early as 4 months of age. When combined with the interval history and observation of the parent-child interaction, a temperament questionnaire can be a useful tool for better understanding the caregiver and child's individuality and behavior. A series of questionnaires are available for various age groups.[30]

Infancy and Early Childhood

Provence[31] suggested that asking parents to describe a typical day from arousal to bedtime can be a useful tool to identify sources of harmony and potential areas of difficulty. A number of standardized tools are also now available to screen specifically for social and emotional development. The Ages & Stages Questionnaire: Social-Emotional (ASQ:SE) is a parent-completed child-monitoring system for social-emotional behaviors in children from 6 to 60 months of age.[8] The areas screened include self-regulation, compliance, communication, adaptive functioning, autonomy, affect, and interaction with people. Questionnaires can be photocopied and take 10 to 15 minutes for parents to complete, and 1 to 3 minutes to score. The ASQ:SE can be used routinely at key well-child visits, such as 18 and 24 months, or as needed if surveillance indicates the need for standardized information. The Communication and Symbolic Behavior Scales Developmental Profile[32] identifies predictors of later language and communication problems by assessing social skills, including the use of gestures, emotions and eye gaze, communication, and object use. A 24-question checklist can be completed by the caregiver of a 6- to 24-month-old child in 5 to 10 minutes. Early concerns can identify those at risk for social and emotional or communication disorders. A more comprehensive questionnaire and a structured observation of behavior are available for further assessment when the initial checklist raises concerns.

The Brief Infant-Toddler Social-Emotional Assessment Scale (BITSEA)[33] is a first-stage screening of social and emotional development that is appropriate for all children from 12 to 36 months of age. Parents can complete the 42 items in less than 10 minutes. If concerns are evident, they can be further characterized by administering the more comprehensive Infant-Toddler Social Emotional Assessment (ITSEA).[34]

Early Childhood to Adolescence

The Eyberg Child Behavior Inventory[35] is designed to assess parental report of behavioral problems in children and adolescents ages 2 to 16 years. It measures the number of difficult behavior problems and the frequency with which they occur. It takes 5 minutes to complete and 5 minutes to score.

The Pediatric Symptom Checklist (PSC)[36] is a first-stage screening appropriate for all children from 4 to 18 years of age. It focuses on broad concerns that reflect general psychological functioning. Parents can complete the 35 items in less than 5 minutes. Cut-off scores are established, and it is available free of charge. It is also available in a pictorial format. The PSC is not a diagnostic instrument but rather a tool that will indicate whether further assessment of social and emotional development is warranted. It may be worthwhile to administer a tool such as the Eyberg Child Behavior Inventory or the PSC at annual visits from preschool through adolescence.

Multidimensional Tools

The Center for Promotion of Child Development through Primary Care has developed CHADIS,[37] a Web-based diagnostic, management, and tracking tool designed to assist health care professionals in addressing parents' concerns about their child's behavior and development. Parents complete online questionnaires from home that are based on the *DSM-PC.* CHADIS analyzes the responses and provides the health care professional with decision support, handouts, and community resources. Additional information is available at www.childhealthcare.org/chadis/.

Numerous, more extensive multidimensional scales are available to provide additional information when initial surveillance or screening tests raise concern. None of these tools should be considered a substitute for a thorough clinical assessment. Therefore, it may be appropriate to consider referral to a mental health professional when screening assessments raise concerns. Examples of multidimensional scales include the Achenbach System of Empirically Based Assessments[38]; the Behavioral Assessment Scale for Children, 2nd edition[39]; the Conners Comprehensive Behavior Rating Scales[40]; the Child Symptom Inventories-4[41]; and the Vanderbilt Parent and Teacher Assessment Scales.[42] The Vanderbilt Scales are in the public domain.

Single Dimension Tools

A number of single-dimension, problem-specific scales are available to assist with further identification of the child with disorders such as attention-deficit/hyperactivity disorder,[43–45] depression, and anxiety.[46]

Practice-based Interventions

A variety of practice-based interventions have been developed to assist the health care professional in promoting healthy social and emotional development and in addressing concerns. Options range from written materials for parents, Internet resources, and seminars, to innovative models for offering health supervision and collaborative interdisciplinary practice models for addressing social and emotional concerns.

The AAP publishes a variety of brochures, books, and other materials that focus on promoting healthy social and emotional development. American Academy of Pediatrics resources are available at the AAP Online Bookstore at www.aap.org. Zero to Three[47] has published a series of 9 two-page tip sheets on early child development that include guidance for parents about common concerns that arise in the first 3 years of life. These have been endorsed by the AAP and cover topics such as crying, sleep, temperament, and making friends. The Centers for Disease Control and Prevention has developed a series of 2-page downloadable handouts entitled *Positive Parenting Tips for Healthy Child Development*[48] that offer suggestions for parents to promote social and emotional development from infancy through adolescence. Additional brief handouts for parents are available from www.triplep-america.com. Triple P, Positive Parenting Program offers more than 50 tip sheets on common behavioral concerns from infancy through adolescence. The Ages and Stages: Social-Emotional Kit includes a series of activity sheets for parents that focus on promoting social and emotional development from infancy to 5 years of age.[8(pp111–129)]

Parent Training Programs

Among a number of parent training programs that have been developed, the Incredible Years and Triple P are 2 that have a strong body of evidence to support their implementation. Parent training programs can be offered in the health care setting or the health care professional can work with community partners to ensure that such services are available to the families they serve.

The Incredible Years series targets children from 2 to 10 years of age, their parents, and their teachers. The program is designed to promote emotional and social competence and to prevent, reduce, and treat children's behavioral and emotional problems.[49] Video-taped scenes are used to encourage group discussion, problem-solving, and sharing of ideas. The core program includes 12 to 14 two-hour weekly sessions. The program teaches parents how to play with children, help children to learn, give effective praise and incentives, use limit setting, and handle misbehavior. Supplemental sessions emphasize parents' interpersonal skills, including communication, anger management, problem-solving between adults, and enhancing parent support. The Incredible Years series also includes a child-training component, which seeks to improve peer relationships and reduce aggression. The teacher training program emphasizes classroom management skills.

Triple P, Positive Parenting Program is a tiered system of behavioral family intervention that includes a public health component and may benefit from application at a population rather than practice level.[50,51] There are 5 levels of Triple P services. The first level involves raising public awareness of the challenges of parenting and supporting families who are concerned about their child's social and emotional functioning to seek help early. The second and third levels of Triple P may be offered in the primary care setting or throughout the community. Brief, problem-specific advice is offered either as anticipatory guidance or to manage a discrete, short-lived, mild problem in an otherwise well-functioning family. The fourth level of Triple P is a more in-depth, broad-based curriculum for parents of children who have more longstanding and challenging behavioral concerns. Parents learn and practice the principles of functional behavioral assessment. Parents learn to monitor their child's behavior, determine the cause of the problem behavior, use strategies to promote social competence and self-control, and manage misbehavior. The fifth level offers assistance to parents who may be struggling with lack of partner support, high levels of stress, lack of coping skills, or difficulty with anger management. Families can enter Triple P at any level, so that sufficient services are offered to match the nature of the behavioral or emotional concern.

Expanding pediatric practice to include parent training can set the foundation for the possibility of developing more broad-based interdisciplinary services within pediatric practice. For example, the Healthy Steps for Young Children Program[52] is a model for enhanced well-child care that includes child development and family health checkups, home visits at key developmental points, written materials that emphasize prevention, a Healthy Steps telephone information line, parent groups, and linkages to community resources. An early childhood specialist works with primary pediatric health care professionals to deliver this enhanced approach.

Group well-child visits are another innovation that may facilitate the development of social competence in families. Osborn and Woolley[53] examined the use of group discussions of 45 minutes followed by brief, individualized examinations with 4 to 6 families. Mothers were more assertive, asked more questions, and initiated discussions of more topics. There was greater coverage of recommended content and a decrease in advice-seeking between visits.

Colocation and/or collaborative practice models with mental health professionals is another approach to offer interdisciplinary services to families to address social and emotional concerns.[54] An integrated model of health care may increase the likelihood that families will pursue mental health services and provides opportunities for support and collaboration among pediatric and mental health professionals.

Mental health professionals can offer parent training programs, as well as individual, group, and family therapy services, that will augment routine pediatric care. Services could include opportunities to address early concerns about temperament mismatch, attachment disorders, or family violence.

Mental health professionals can offer cognitive behavioral therapy to parents and children for depression and anxiety. The goals of cognitive behavioral therapy are to teach the adult or child to identify their stressors and experiment with new ways of thinking and doing. Examples of promising cognitive behavioral approaches include The Optimistic Child[55] and the Coping Cat program.[56] Similar manualized approaches are available to teach relaxation and stress reduction.[57]

Anticipatory Guidance

The content of anticipatory guidance will be determined by the context, priorities, surveillance of development, physical examination, and screening completed during health supervision. Bright Futures suggests that anticipatory guidance should be timely, appropriate to the child and family, and relevant.[5(p206)] Should concerns about social and emotional development rise to the surface, it is likely that the information in this section will prove helpful.

Carey[58] has suggested that information on temperament be part of basic education for all parents on child-rearing. Temperament refers to a child's behavioral style, or the "how" of behavior. Individual differences in temperament are real and have a significant impact on the parent-child interaction, including the parents' feelings of competency in child-rearing. Temperament cannot be changed, but it can be accommodated. It may be especially helpful in the first months of life to provide guidance related to the role of temperament in influencing children's social and emotional development.

The principles of positive parenting outlined in Triple P,[59–61] a prevention-oriented early intervention program that aims to promote positive, caring relationships between parents and their children and to help parents develop effective management strategies for dealing with a wide variety of childhood behavior problems and common developmental issues, can serve as a framework for providing additional anticipatory guidance regarding social and emotional development.

Teaching parents skills for promoting their children's development, social competence, and self-control with evidence-based strategies will enhance their self-sufficiency in managing children's behavior. Additional benefits should include a reduction in parents' use of coercive and punitive disciplinary methods, improved communication between parents, and less parental stress related to raising their children. Parents can be encouraged to ensure a safe and developmentally stimulating environment, create a positive learning environment using assertive discipline, have realistic expectations, and take care of themselves as parents.

A number of strategies can encourage positive relationships between parent and child. These include spending quality time with children and adolescents. Frequent, brief amounts of time, even for a few minutes, engaged in a child-preferred activity can be highly beneficial. These brief interactions offer opportunities for children to

self-disclose and practice conversational skills and for adolescents to enjoy parent contact and maintain a positive relationship. Talking to children and adolescents during brief conversations about an activity or interest of the child promotes vocabulary and conversational and social skills, and gives adolescents the chance to voice opinions and to discuss issues important to them. Showing physical affection (eg, hugging, touching, cuddling, tickling, patting) and being certain to avoid public embarrassment for adolescents provides opportunities to become comfortable with intimacy and physical affection in appropriate ways.

Examples of strategies for encouraging desirable behavior for all ages are summarized here. Using descriptive praise provides encouragement and approval by describing behavior that is appreciated. This might includes behaviors such as speaking in a pleasant voice, playing cooperatively, sharing, drawing pictures, reading, and compliance with instructions. Providing positive nonverbal attention (eg, a smile, wink, pat on the back, watching) will also encourage appropriate behavior. Additional strategies include arranging the child's physical and social environment to provide interesting and engaging activities, materials, and age-appropriate toys; creating opportunities for adolescents to explore and try out new social and recreational activities; and encouraging independent play and interests while promoting appropriate behavior when in community settings.

Parents can also benefit from guidance in teaching new skills and behaviors. By setting a good example, parents can demonstrate desirable behavior. Through modeling, children learn how to behave appropriately, especially in relation to interpersonal interactions and moral issues. Incidental teaching is a technique in which a series of questions and prompts is used to respond to child-initiated interactions and promote learning. This approach promotes language, problem-solving, cognitive ability, and independent play in children older than 1 year. "Ask, Say, Do" is another strategy to gradually build the confidence and skills of children 3 years and older. In a stepwise approach, children are first given verbal, then gestural, and then manual prompts to teach new skills.

Behavior charts are effective tools to provide social attention and backup rewards contingent on the absence of a problem behavior or the presence of an appropriate behavior. They are useful for children 2 years and older.

Adolescents can benefit by being coached in problem-solving skills. This helps them to deal with a problem in a constructive and effective way and promotes independence. Adolescents can also benefit from behavior contracts. An agreement can be negotiated to deal with a dispute or a distressing issue. This promotes development of personal responsibility. Family meetings can also be a valuable tool with this age group. By organizing a set time for family members to work together to set goals for change, adolescents learn compromise, decision-making, and personal responsibility. Scheduling a review session to check on how the strategies worked and offering suggestions for the future can reinforce the strategies that emerge from family meetings.

Parents of adolescents should be encouraged to establish networks with their friends' parents to assist with monitoring their children's behavior, identify opportunities to reinforce desirable behavior, and address problem behaviors.

Parents frequently seek guidance regarding managing misbehavior. It is important to keep in mind that focusing attention on promoting positive relationships and encouraging desirable behavior will greatly reduce the need to manage misbehavior. At the same time, numerous simple techniques are effective in managing misbehavior. Distracting the young child by drawing her attention away from a problematic activity to an acceptable activity is often beneficial. Children older than 3 years will respond to fair, specific, and enforceable ground rules that have been negotiated in advance. This will clarify expectations and avoid casual conflict. The use of directed discussion for rule breaking permits children to identify and rehearse the correct behavior after rule breaking. This is especially helpful for minor rule breaking and initial violations following the application of a new rule. Planned ignoring is a useful strategy for minor behavior problems in 1- to 7-year-olds. The parent can be advised to withdraw attention while the problem behavior continues and ignore attention-seeking behavior.

One of the most important suggestions for parents is to become competent in giving clear, calm instructions. Children older than 2 years will respond well to receiving a specific instruction to start a new task or to stop a problem behavior and start a correct behavior. It is critical that the child be told what to do rather than what not to do. Instructions should be backed up with logical consequences. The consequence should involve the removal of an activity or privilege from the child or the child from an activity for a set time. This is effective for children older than 2 years when dealing with noncompliance and mild behavior problems that do not occur very often. Quiet time for misbehavior can be a valuable tool for children 18 months to 10 years of age. The child can be removed from an activity in which a problem has occurred and be instructed to sit on the edge of the activity for a set time. This can be helpful for noncompliance and for children who repeat a behavior after a logical consequence. By staying at the edge of the activity, children have a chance to see what they are missing, increasing their incentive to demonstrate desirable behavior when they return to the activity. Time-out should be reserved for serious misbehavior in 2- to 10-year-olds. The child should be removed to an area away from others for a set time. The family may need to identify a safe place where all attention can be withdrawn from the child. It is important to consider the function of the problem behavior before deciding to use time-out. Time-out will generally be effective for attention-seeking behavior rather than avoidant behavior. If a child is acting out in order to avoid a certain request or activity, time-out may actually be an accidental reward. Time-out is helpful for children who refuse to sit in quiet time and for those having temper outbursts and aggressive or destructive behavior.

Parents of adolescents may be challenged by their child's emotional behavior. Parents may need assistance in helping their adolescent manage unpleasant or intense emotional responses that interfere with effective problem-solving or lead to more conflict and distress. For example, parents should be encouraged to acknowledge their adolescent's distress, ask what he wants his parent to do, and then coach him to problem-solve. If this is not effective, he may need a cooling-off period while setting a later time to talk again. The parent should be encouraged to stay calm and use a cooling-off period for herself, if necessary. It is also important for parents to consider the possibility that emotional behavior is being used to avoid something the adolescent should do. If that is the case, the behavior should be ignored.

Particular challenges come with addressing risky behavior in adolescents. It is important for parents to anticipate events that may lead to risk-taking behavior and prevent unexpected demands from leading to conflict or decision-making under pressure. Parents can be sure their adolescent has accurate information about the potentially risky behavior. Health care professionals can offer this type of information during a visit or with appropriate resources. Parents can role-play with their adolescent to give them an opportunity to practice the words they will use when pressured into a possible risky behavior. Parents should be sure their adolescent knows they will be available to rescue them if they find themselves in an uncomfortable situation. Parents can work with their child in advance to negotiate a fair set of rules with preordained consequences. This approach will ensure that important decisions are not made on inaccurate assumptions and that the adolescent has the chance to participate in peer activities with a clearly established plan to support her in avoiding risky behavior.

Promoting Community Relationships and Resources

Primary pediatric health care professionals are in a unique position to foster the development of community relationships to address the social and emotional development of children. In the course of daily practice, it is likely that assets that are available in the community to support children and families will become evident. This information can be shared on the practice's Web site, in information kiosks in the office, on bulletin boards, and in the course of a visit. In addition, opportunities for the development of new programs and services will become apparent through repeated contacts with families and community leaders.

There are numerous evidence-based programs that have been developed as preventive and intervention models related to addressing the social and emotional development of children. When opportunities arise to promote development of community relationships and resources, it is important to consider whether existing, proven, or promising programs from across the country and across the world may be applicable in the health care professional's local community.

The health care professional has an important role in linking families with early intervention services.[62] Programs like Connecticut's ChildServ and Help Me Grow[63] have trained providers to do effective developmental surveillance, created resource inventories of community-based services, and developed a referral and monitoring system to link young children and their families with early childhood services and support.

Several programs have included home visits as core components. The Nurse-Family Partnership is a prenatal and early infancy home visiting program that has demonstrated better infant emotional and language development and improved maternal independence.[64] The Infant Health and Development Program (IHDP)[65–67] was a multicenter comprehensive early intervention program for low-birth-weight (≤2,500 g) and premature (≤37 weeks) infants that has proven to reduce the infants' health and developmental problems. The IHDP was designed as a randomized clinical trial that combined early child development and family support services with pediatric follow-up for the first 3 years of life. The intervention services, provided free to participating families, consisted of 3 components: home visits, child attendance at a child development center, and parent group meetings. Infants participated in pediatric follow-up, which comprised medical, developmental, and social assessments, with referral for pediatric care and other services as indicated.

Early Head Start[68] is a federally funded community-based program for low-income pregnant women and families with infants and toddlers up to age 3 years. Its mission is to promote healthy prenatal outcomes for pregnant women, enhance the development of children from birth to 3 years of age, and support healthy family functioning. Services include child development services delivered in home visits, child care, comprehensive health and mental health services, parenting education, nutrition education, health care and referrals, and family support. Each community has the opportunity to select options for providing services that best meet their community's needs.[69]

Promoting First Relationships[70] is a curriculum designed to guide caregivers in building nurturing and responsive relationships with children from birth to 3 years of age. The program focuses on promoting the development of trust and security in infancy and promoting healthy development of self during toddlerhood, while assisting caregivers in understanding and intervening with children's challenging behaviors.

Big Brothers Big Sisters[71] is a program that matches unrelated mentors who serve as role models for children to promote positive development and social responsibility. This program has been shown to reduce rates of alcohol, tobacco, or illegal drug use and appears promising in reducing violent behavior and serious conduct problems and in performing at grade level.

A number of school-based programs are promising as well. For example, the Cognitive Relaxation Coping Skills program[72] is designed to increase sixth to eighth graders' ability to control their emotions. Students are taught relaxation methods and strategies for attitude change. By learning how to control their anger, they avoid frustrating situations. The Reaching Educators, Children, and Parents program[73,74] is a semi-structured, school-based skills training designed for children experiencing internalizing and externalizing problems. The primary goal of the program is to reduce the level of children's psychological problems, as well as preventing the development of more serious problems among children who are not referred for formal mental health services. The Social Decision Making/Problem Solving program[75,76] can be provided to any student, rather than target-ing those with special characteristics. The program seeks to develop children's self-esteem, self-control, and social awareness skills, including identifying, monitoring, and regulating stress and emotions; increasing healthy lifestyle choices; avoiding social problems, such as substance abuse, violence, and school failure; improving group cooperation skills; and enhancing the ability to develop positive peer relationships.

Additional information on the programs outlined above and on similar promising or proven programs is available at www.promisingpractices.net.

It is also important to consider that the media, including television and the Internet, may have broad applications for reaching large populations of people to promote healthy social and emotional development. In 2005 in the United Kingdom, a 5-episode television series, *Driving Mum and Dad Mad,* aired on prime-time television. In a project called The Great Parenting Experiment,[77] 500 families who viewed the series and had access to Web support and self-help materials reported improved confidence in managing their children's behavior. For some parents, television may be the only way they will access parenting information.

There are many evidence-based prevention and intervention strategies available. As health care professionals identify needs in their community, it will be important to investigate and advocate for implementation of strategies that have been shown to be effective in other settings.

Conclusion

Healthy social and emotional development is rooted in the quality of relationships with parents, extended family, caregivers, peers, teachers, coaches, and other adults. It is vital for health care professionals to consider the dynamic interactions between the child's genetics, biology, and temperament with the environment in which he is expected to grow and develop. By following the Bright Futures recommendations for promoting child development and mental health, while also promoting community relationships and resources, the health care professional will be well positioned to support children, adolescents, and their families in fostering healthy social and

emotional development. Each preventive health visit provides an opportunity to establish a context for the visit and to set priorities for the visit. Should these identify social and emotional development as a priority, then an appropriate review of the interval history, observation of parent-child interaction, surveillance of development, a physical examination, and screening will provide a fund of information to guide anticipatory guidance, office-based interventions, and referral to community resources.

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Chapter 13

Autism Spectrum Disorders

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In 1943 Leo Kanner eloquently described in the literature 11 children with "autistic disturbances of affective contact" characterized by profound lack of reciprocal social engagement; disturbances in communication ranging from mutism to echolalia, pronoun reversal, and literalness; and unusual responses to the environment, including insistence on sameness.[1] More than 6 decades later, diagnostic criteria and assessment instruments remain remarkably influenced by Kanner's original description.

According to the American Psychiatric Association's *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, Text Revision (DSM-IV-TR),* the broad diagnostic category "pervasive developmental disorders" includes 5 specific diagnoses: autistic disorder, Asperger's disorder (Asperger syndrome), Rett's disorder (Rett syndrome), childhood disintegrative disorder (Heller syndrome), and pervasive developmental disorder not otherwise specified (PDD-NOS).[2] The term autism spectrum disorders (ASDs) has been used to encompass 3 of the 5 pervasive developmental disorders: autistic disorder, Asperger syndrome, and PDD-NOS.[3,4] The current diagnostic criteria for these disorders are included in Box 13.1.[2]

Epidemiology

The prevalence of ASDs, as currently defined and operationally diagnosed, is at least 65 per 10,000 (1 in 154),[5–7] and possibly as high as 110 per 10,000 (1 in 91).[8] Pervasive developmental disorder not otherwise specified is diagnosed most frequently, followed by autistic disorder and then Asperger syndrome, with a prevalence ratio of approximately 3:2:1.[5] Boys are more commonly affected than girls, with an average male-to-female ratio of approximately 4:1. However, this ratio is affected substantially by cognitive impairment; the ratio among individuals without intellectual disability (formerly called mental retardation) is at least 5:1, and the ratio among those with intellectual disability is closer to 2:1.[7]

Box 13.1. *DSM-IV-TR* Diagnostic Criteria for Autism Spectrum Disorders[a]

Autistic Disorder (299.00)

A. A total of six (or more) items from (1), (2), and (3), with at least two from (1), and one each from (2) and (3):

(1). Qualitative impairment in social interaction, as manifested by at least two of the following:
   a. marked impairment in the use of multiple nonverbal behaviors such as eye-to-eye gaze, facial expression, body postures, and gestures to regulate social interaction
   b. failure to develop peer relationships appropriate to developmental level
   c. a lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (eg, by a lack of showing, bringing, or pointing out objects of interest)
   d. lack of social or emotional reciprocity

(2). Qualitative impairments in communication as manifested by at least one of the following:
   a. delay in, or total lack of, the development of spoken language (not accompanied by an attempt to compensate through alternative modes of communication such as gesture or mime)
   b. in individuals with adequate speech, marked impairment in the ability to initiate or sustain a conversation with others
   c. stereotyped and repetitive use of language or idiosyncratic language
   d. lack of varied, spontaneous make-believe play or social imitative play appropriate to developmental level

(3). Restricted repetitive and stereotyped patterns of behavior, interests, and activities, as manifested by at least one of the following:
   a. encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus
   b. apparently inflexible adherence to specific, nonfunctional routines or rituals
   c. stereotyped and repetitive motor mannerisms (eg, hand or finger flapping or twisting, or complex whole-body movements)
   d. persistent preoccupation with parts of objects

B. Delays or abnormal functioning in at least one of the following areas, with onset before 3 years old: (1) social interaction, (2) language as used in social communication, or (3) symbolic or imaginative play.

C. The disturbance is not better accounted for by Rett's Disorder or childhood disintegrative disorder

Asperger's Disorder (299.80)

A. Qualitative impairment in social interaction, as manifested by at least two of the following:

(1) marked impairment in the use of multiple nonverbal behaviors, such as eye-to-eye gaze, facial expression, body postures, and gestures to regulate social interaction
(2) failure to develop peer relationships appropriate to developmental level
(3) a lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (eg, by a lack of showing, bringing, or pointing out objects of interest to other people)
(4) lack of social or emotional reciprocity

B. Restricted, repetitive, and stereotyped patterns of behavior, interests, and activities, as manifested by at least one of the following:

(1) encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus
(2) apparently inflexible adherence to specific, nonfunctional routines or rituals
(3) stereotyped and repetitive motor mannerisms (eg, hand or finger flapping or twisting, or complex whole-body movements)
(4) persistent preoccupation with parts of objects

Box 13.1. *DSM-IV-TR* Diagnostic Criteria for Autism Spectrum Disorders[a] *(continued)*

Asperger's Disorder (299.80) *(continued)*

C. The disturbance causes clinically significant impairment in social, occupational, or other important areas of functioning.

D. There is no clinically significant general delay in language (eg, single words used by age 2 years, communicative phrases used by age 3 years).

E. There is no clinically significant delay in cognitive development or in the development of age-appropriate self-help skills, adaptive behavior (other than in social interaction), and curiosity about the environment in childhood.

F. Criteria are not met for another specific pervasive developmental disorder or schizophrenia.

Pervasive Developmental Disorder Not Otherwise Specified (299.80)

This category should be used when there is a severe and pervasive impairment in the development of reciprocal social interaction associated with impairment in either verbal or nonverbal communication skills or with the presence of stereotyped behavior, interests, and activities, but the criteria are not met for a specific Pervasive Developmental Disorder, Schizophrenia, Schizotypal Personality Disorder, or Avoidant Personality Disorder. For example, this category includes 'atypical autism'—presentations that do not meet criteria for Autistic Disorder because of late age at onset, atypical symptomatology, or subthreshold symptomatology, or all of these.

[a] From: American Psychiatric Association. *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, Text Revision (DSM-IV-TR).* Washington, DC: American Psychiatric Publishing; 2000. Reprinted with permission.

Most studies published prior to the late 1980s suggested a much lower ASD prevalence of approximately 4.5 per 10,000, and there has been much speculation about the reason for the nearly 15-fold increase. Heightened public and professional awareness, changes in special education laws, availability of services, and the development of screening and diagnostic tools are likely to have contributed to increased ascertainment of cases. Changes in the diagnostic criteria and, more importantly, the way the criteria are applied in medical and educational practice have contributed to diagnostic substitution, whereby children who would have been given a different diagnosis (eg, mental retardation or language disorder) in the past are now diagnosed with an ASD.[9,10] To a large degree, these changes in diagnostic tendency reflect evolving recognition that ASDs can occur in individuals of all levels of intellectual ability. Increased ascertainment and changes in diagnostic tendency undoubtedly are responsible for a large portion of the increase in prevalence, but presently there are insufficient data to determine whether these factors account for the entire increase or whether there has also been a true secular increase in the incidence of ASDs. The answers to these questions will come from well-designed, population-based prospective studies that examine rates in the same population and use the same classification methods over time, such as the Autism and Developmental Disabilities Monitoring Network established by the Centers for Disease Control and Prevention.[11]

Etiology/Pathophysiology

Autism spectrum disorders are a heterogeneous group of neurodevelopmental disorders without an identified unifying pathological or neurobiological etiology. Although research into the biological basis of these disorders is in its early stages, much progress has been made in the areas of genetics, neuroimaging, neurophysiology, and neuropathology.

Genetics

Defined genetic syndromes, mutations, and de novo copy number variants (microdeletions and microduplications) account for 10% to 20% of ASD cases, although no individual genetic etiology accounts for more than 1% to 2%.[12] Cytogenetic imbalances involving every human chromosome have been reported to be associated with ASDs, and a subset of individuals with virtually every known intellectual disability syndrome also meet criteria for an ASD. Some of the most common identifiable genetic etiologies are reviewed in Table 13.1. Inherited copy number variants may also strongly modulate ASD risk and contribute to the broader autism phenotype in relatives of affected individuals and the normal distribution of ASD-like features in the population.[12] Advanced paternal age is associated with increased risk of ASDs.[13] This may be due to men with features of ASDs, who carry more genetic liability, marrying and having children later or to mutation accumulation in older spermatocytes, or both.

Currently, most affected individuals do not have an identifiable etiology; they are considered to have non-syndromic, or idiopathic, ASDs. Lack of an identifiable genetic abnormality in these individuals does not imply that there is not a genetic etiology. In fact, twin and family studies provide unequivocal evidence for a genetic component to the etiology of idiopathic ASDs. For a couple with one affected child, the rate of recurrence in a subsequent child is approximately 5% to 10%, and concordance rates for monozygotic twins (70%–90%) are substantially higher than those for dizygotic twins (0%–10%).[12,14] The heritability estimate, based on the concordance rate in monozygotic twins compared to dizygotic twins and the recurrence risk in families, is at least 0.90, suggesting that 90% or more of phenotypic variance is due to genetic variance.[14] There is evidence suggesting that social behavior; communication; and rigid, obsessive behavior are each highly heritable but independent of each other, implying that mutations in different combinations of genes may be responsible for the triad of core impairments in ASDs and the wide variation in severity of each component.[15]

Environment

There is evidence that in utero exposure to medications such as valproate, misoprostil, and thalidomide, and other aspects of the prenatal environment such as testosterone level, alcohol exposure, and infections (eg, rubella, cytomegalovirus) may be associated with increased risk of ASDs, but epidemiologic data are lacking

Table 13.1. Genetic Abnormalities Most Commonly Associated With Autism Spectrum Disorders

Syndrome	Implicated Gene(s)	Primary Clinical Laboratory Test(s)
2q37 deletion	Unknown, possibly *CENTG2*	Microarray comparative genomic hybridization or subtelomere FISH
15q11-q13 duplication (supernumerary marker 15 or maternally inherited interstitial duplication)	*UBE3A, GABRB3,* possibly others	Microarray comparative genomic hybridization or specific FISH
16p11.2 deletion (and possibly duplication)	Unknown	Microarray comparative genomic hybridization
22q11.2 deletion	Unknown	Microarray comparative genomic hybridization or specific FISH
22q13.3 deletion	*SHANK3*	Microarray comparative genomic hybridization or subtelomere FISH
Fragile X syndrome (locus Xq27.3; abnormal expansion of CGG-trinucleotide repeat)	FMR1	Targeted mutation analysis (methylation-sensitive PCR, Southern blot)
Macrocephaly/autism syndrome, *PTEN* hamartoma tumor syndrome (locus 10q23.31)	*PTEN*	Sequence analysis, deletion/duplication analysis
Rett syndrome (locus Xq28)	*MECP2*	Sequence analysis/mutation scanning
Tuberous sclerosis (loci 9q34 and 16p13.3)	*TSC1, TSC2*	Sequence analysis

Abbreviations: *CENTG2,* centaurin gamma 2; FISH, fluorescence in situ hybridization; *FMR1,* fragile X mental retardation 1; *GABRB3,* gamma-aminobutyric acid A receptor beta 3; *MECP2,* methyl-CpG-binding protein 2; PCR, polymerase chain reaction; *PTEN,* phosphatase and tensin homolog; *SHANK3,* SH3 and multiple ankyrin repeat domains 3; TSC1, tuberous sclerosis 1; TSC2, tuberous sclerosis 2; *UBE3A,* ubiquitin protein ligase E3A.

or preliminary.[3,16] Perinatal factors such as prematurity, low birth weight, intrapartum hypoxia, and neonatal encephalopathy may also be associated with increased risk of ASDs.[13,17] However, more research is required to determine whether these are independent risk factors, since genetic susceptibility may be associated with both obstetric suboptimality and neonatal encephalopathy.

Assertions that toxic effects of the measles-mumps-rubella vaccine or a combination of thimerosal-containing vaccines are responsible for the increase in prevalence of ASDs have been soundly refuted by epidemiologic studies, and the weight of all of the scientific evidence overwhelmingly favors rejection of the hypothesis that there is a causal association between vaccines and ASDs.[5,18–20] The idea that a "second-hit" phenomenon, whereby genetic factors confer increased susceptibility to environmental toxins, infections, or other prenatal or postnatal insults, may play a substantial role in the etiology of ASDs, is attractive but remains speculative at this time.

Epigenetics

The potential role of epigenetic factors in the etiology of ASDs is also an active area of investigation.[21] Epigenetic modifications, such as DNA cytosine methylation and post-translational modification of histones, produce heritable changes in gene expression that do not involve a change in the DNA sequence. Epigenetic mechanisms are important in the pathogenesis of Rett syndrome and fragile X syndrome and are responsible for genomic imprinting, the process whereby gene expression is restricted to only one of the parental alleles. Interestingly, one of the most common cytogenetic abnormalities in patients with ASDs is maternally derived duplication of the imprinted domain on chromosome 15q11-q13. It has been speculated, but not proven, that alteration of gene expression by environmental factors may have a role in the etiology of ASDs. Epigenetic modifications can potentially be influenced by environmental factors, such as maternal exposures and postnatal experience, so they provide a tangible link between gene and environment.

Neurobiology

Although the neuropathology of ASDs has not yet been clearly defined, neuroimaging and postmortem neuropathological findings implicate pre- and postnatal developmental abnormalities involving multiple regions of the brain, including the cerebral cortex, cortical white matter, amygdala, brain stem, and cerebellum (Box 13.2).[22,23] Recent findings suggest that abnormalities in large-scale neural systems circuitry, rather than discrete regional abnormalities, may underlie many of the deficits associated with ASDs. Structural magnetic resonance imaging (MRI) morphometry and diffusion tensor imaging studies demonstrating regional differences in white matter volume; histopathological studies showing abnormalities in cortical minicolumns; and functional MRI studies documenting diminished functional connectivity among cortical regions involved in language, executive functioning, and social cognition suggest a fundamental problem of intrahemispheric cortical underconnectivity in ASDs.[23] However, some studies suggest that global underconnectivity may not be ubiquitous in ASDs, and factors such as clinical severity, cognitive strengths and weaknesses, experiential factors, and task demands may moderate functional connectivity findings.[24]

Cross-sectional and longitudinal studies suggest that as a group, children later diagnosed with an ASD have average or below-average head circumferences at birth, with presumed acceleration in brain growth during the first year of life leading to significantly above-average head circumferences and MRI brain volumes as toddlers.[25,26] Although brain volumes are increased by 5% to 12% in studies of very young children with ASDs (ages 18 months–4 years), the difference is only 1% to 3% (if a real difference persists at all) in studies of adolescents and adults, suggesting that a period of slowing or arrest of brain growth follows the early brain overgrowth.[22,25] The early childhood increase in brain volume, which involves both gray matter and white matter, seems to be regional rather than uniform. Sites of overgrowth include

Box 13.2. Neuropathological Abnormalities in Autism Spectrum Disorders (ASDs)

Structural Neuroimaging and Head Circumference Studies
Early brain overgrowth followed by early deceleration or arrest of growth
- Gray and white matter involvement
- Regional overgrowth in frontal and temporal cortices, cerebellum, amygdala, and caudate nucleus
- Increased volume of outer radiate white matter (intrahemispheric pathways), decreased corpus callosum volume (interhemispheric connection)

Functional Neuroimaging Studies
Reduced neuronal activity in regions that normally govern specific functional skill domains that are important in ASDs

Abnormal functional connectivity between brain regions involved in mediating complex language, selective attention, visual-motor coordination, social cognition, and executive functioning tasks

Histopathology
Reduced numbers of Purkinje cells in the cerebellum

Abnormal maturation of frontal and temporal lobe cortex and forebrain limbic system
- Atypical cortical minicolumns: more numerous, smaller (narrower), less compact in their cellular configuration, reduced neuropil space in the periphery
- Decreased number of neurons in the fusiform gyrus
- Reduced neuronal size, increased cell-packing density, decreased complexity of the neuropil in the hippocampus
- Decreased number of neurons in the amygdala

frontal and temporal cortices, cerebellum, and amygdala, but investigators have not yet charted a complete map of early regional overgrowth in ASDs.[25,27]

Electrophysiologic techniques, such as auditory brain stem response, electroencephalogram (EEG), and event-related potentials, allow precise temporal discrimination (in the range of milliseconds), and magnetoencephalography allows fine resolution in both time and space. Although the evidence is not yet conclusive, studies using these techniques have identified electrophysiologic correlates of abnormalities in auditory processing, imitation skills, selective attention, response to visual stimuli (including face recognition and response to facial expressions of emotion), somatosensory response, and neural connectivity.[28,29] Neurochemical studies have implicated abnormalities in serotonergic, cholinergic, GABAergic, glutamatergic, and dopaminergic neurotransmitter systems as well as dysregulation of neuropeptides, such as oxytocin and vasopressin.[30] The potential importance of autoantibodies and effects of the maternal immune response on the developing central nervous system are active areas of investigation.[31]

The cellular and molecular mechanisms of early regional brain overgrowth, histopathological abnormalities, and functional underconnectivity associated with ASDs are unknown. Just as there are multiple genetic etiologies, it is likely that there are

multiple different neuropathological processes involved in this group of behaviorally defined disorders. There is mounting evidence that abnormalities involving genes coding for neuronal cell-adhesion molecules, such as neurexins, neuroligins, and cadherins, are responsible for rare ASD cases, and common genetic variants on 5p14.1 may confer susceptibility to ASDs via this mechanism.[32] Cell-adhesion abnormalities resulting in abnormal axonal guidance, synaptic formation and plasticity, and neuronal-glial interactions may be a mechanism responsible for structural and functional underconnectivity.[32,33]

Clinical Features

Autism spectrum disorders are characterized and defined clinically by the core impairments, which are present in all or nearly all affected children but are relatively infrequent in those with other conditions. However, no single specific behavior or deficit is pathognomonic for ASDs. Although not part of the diagnostic criteria, other features such as cognitive and motor deficits, behaviors often attributed to sensory integration difficulties, maladaptive behaviors, and associated medical problems have a dramatic impact on adaptive functioning and are also important aspects of the clinical presentation.

Core Impairments

Autism spectrum disorders are characterized by distinctive impairments in reciprocal social interaction and communication that do not simply reflect associated intellectual disability, and by the presence of a restricted, repetitive behavioral repertoire (Box 13.1).[2] The clinical features vary with age, developmental level/intellectual ability, and severity of the condition. Boxes 13.3 through 13.5 provide additional details about the clinical features described in the *DSM-IV-TR* criteria (presented in Box 13.1). There is certainly overlap between social behavior and communication, and the difficulties that children with ASDs have with nonverbal communication, imitation, and imaginative play are sometimes considered to be examples of social deficits and sometimes communication deficits.[34] Therefore, some clinical features could be included in either Box 13.3 or Box 13.4; the decision is somewhat arbitrary, but in this case the intention was to correlate as closely as possible with the *DSM-IV-TR* criteria.

Social Interaction

Qualitative impairment in social interaction (relative to developmental level or mental age) is a defining feature of ASDs (Box 13.3).[2,3,34] As infants and toddlers, these children may not smile responsively or adopt a posture conveying anticipation and readiness or desire to be picked up. Young children with ASDs lack developmentally appropriate joint attention, which is defined as visually coordinating attention with a partner to an external focus, showing social engagement and an awareness of the partner's mutual interest for the purpose of "commenting" rather than "requesting" (Box 13.3).[35] Reciprocal social behaviors such as seeking to share enjoyment

with others, feeling genuine concern and offering comfort to other people, and forming caring friendships that go beyond classroom or parent-arranged interactions are absent or limited, even in older children.[34]

Wing and Gould[36,37] have described 3 types of social impairment exhibited by individuals with ASDs: aloof, passive, and active but odd. Individuals in the first group, whose social impairment is most easily recognized, are generally aloof and indifferent to others, especially peers, but may accept physical affection from familiar people and enjoy physical play. Those in the passive group do not socially interact spontaneously but passively accept and even appear to enjoy approaches from others. The active but odd group includes people who make active social approaches that are naïve, odd, inappropriate, and one-sided. Stability of the social interaction subgroup classification varies with level of cognitive ability. Children with higher levels of cognitive ability may move, for example, from the aloof group in early childhood to the passive or active but odd group in adolescence, whereas those with severe or profound intellectual disability are more likely to remain aloof and indifferent to others.[37]

Box 13.3. Clinical Features of Autism Spectrum Disorders: Social Interaction

Characteristics in Younger or More Severely Impaired Children	Characteristics in Older or Less Severely Impaired Children
Poor eye contact, gaze aversion	Decreased or atypical eye contact
Limited or absent responsive smiling	Difficulty forming developmentally appropriate friendships that involve a mutual sharing of interests, activities, and emotions
Decreased sharing of joyful affect	More socially immature and less independent than peers - May be viewed as odd, eccentric, or "weird" by peers - May initiate interactions in inappropriate, awkward, or stilted ways - Gullible, naïve, lacking "common sense"
Marked lack of interest in other people - Often aloof and indifferent, may prefer to be alone - More interested in objects than people - May bump into people as if they did not see them or climb on them as if they were furniture	Lack of modulation of behavior according to social context
Rather than being distant, some children display indiscriminate affection	Poor perspective-taking ability that may contribute to inappropriate, offensive behavior - Difficulty anticipating how another person will feel or what they might think - Lack of empathy - Difficulty knowing how to react to another person's behavior or emotions - Difficulty accepting that there might be multiple perspectives, not just a single correct perspective
Deficits in joint attention - Lack of response when name is called - Failure to follow the gaze and pointing gestures of others - Failure to spontaneously alternate gaze between an object and another person - Absent or limited attempts to draw the attention of others to objects or events for the purpose of sharing experiences (including lack of pointing to comment or show)	

Communication

The severity of language impairment ranges from profound (eg, verbal auditory agnosia) to relatively mild (eg, semantic-pragmatic deficit syndrome), and the clinical features vary with age and developmental level (Box 13.4).[3,38,39] Communicative speech is typically delayed or absent in children with autistic disorder, and comprehension is impaired. Nonverbal communication is also impaired, so there is little or no attempt to compensate by using gestures or pantomime.

Although children with Asperger syndrome and some with autistic disorder or PDD-NOS may not be delayed in attaining early vocabulary, grammar, and articulation milestones, other aspects of language, such as pragmatics (social, context-bound, functional language use), semantics (word and sentence meaning), and prosody (volume, rhythm, intonation), are impaired.[39,40] Word choice is often atypical, and these individuals have difficulty interpreting nonliteral communication such as figures of speech, humor, and irony.

Children with ASDs have difficulty with imitation of other people's actions, and there is a paucity of spontaneous (generative) symbolic play behavior relative to children with similar mental ages.[41] Symbolic play development is correlated with language and nonlanguage cognitive ability, but the deficit in play behavior typically exceeds what would be predicted based on these abilities.[41] Qualitatively, the play of children with ASDs tends to be repetitive, stereotyped, and excessively focused on sensorimotor manipulation of objects rather than varied, flexible, imaginative, and creative (Box 13.4).

Restricted, Repetitive Behavioral Repertoire

The behavioral repertoire in children with ASDs tends to be characterized by perseveration on narrow or atypical interests, behavioral rigidity (obsessive insistence on sameness, resistance to change, adherence to nonfunctional rituals or routines), and stereotyped movements (Box 13.5). These behaviors tend to be less frequent and less severe in older individuals than among younger individuals.[42] Stereotyped movements are more persistent in adolescents and adults with ASDs who also have intellectual disability. However, even in highly functioning individuals, unusual preoccupations and circumscribed interests often interfere with social interaction.

It is important to recognize that repetitive behaviors, including stereotyped movements and behaviors related to desire for sameness, occur in normal development and are common in individuals with other developmental disabilities or psychiatric conditions.[43–45] Richler and colleagues[45] found that repetitive sensorimotor behaviors were more common in 2-year-old children with ASDs than in typically developing controls and children with other developmental disorders, although these behaviors were relatively common in the latter group. Repetitive behaviors related to insistence on sameness were less common, and did not consistently differentiate among the 3 groups. Notably, some behaviors, such as self-injury, sensitivity to noise, and resistance to trivial changes in the environment, did not differ in prevalence or severity across the 3 groups of 2-year-olds.

Box 13.4. Clinical Features of Autism Spectrum Disorders: Communication and Play

Characteristics in Younger or More Severely Impaired Children

May not have any meaningful speech

Greater proportion of syllables with atypical phonation (eg, squeals, growls, and yells)

Delayed receptive and expressive language milestones

Increased idiosyncratic or inappropriate means of communication
- Self-injurious behavior, aggression, tantrums

Echolalia
- Immediate (eg, in response to questions)
- Delayed (scripted verses, reciting memorized dialogue)

Pronoun reversal (often "you" for "I" or "you" for "me")

May label objects or actions but not use those words to make requests or answer questions

Delayed, decreased, or absent use of gestures (eg, showing, waving, pointing, nodding head, depicting actions)

Mostly primitive motor gestures to communicate (eg, contact gesture of leading or pulling another's hand)

Early lack of interest in toys

Preoccupation with elementary sensory features of toys

Lining, spinning, arranging, hoarding, carrying

Later appreciation of symbolic meaning, appropriate functional use
- Sequences of appropriate actions
- Impoverished, never achieves typical creativity or variability
- Repetitious, mechanical
- Often precisely imitated from videos

Development of pretend play typically delayed and impaired

Parallel play, interactive physical play

Difficulty with turn-taking

Characteristics in Older or Less Severely Impaired Children

Unusual vocabulary for age/social group

Difficulty answering open-ended questions

Dysprosody (atypical intonation, inflection, rhythm of voice)
- Singsong or cartoonish
- Monotone

Lack of awareness of personal space

Difficulty with initiation and closure of conversation and topic changes
- Failure to provide enough background information
- Tangential remarks
- Abrupt topic changes
- Difficulty maintaining conversation by elaborating or requesting more information

One-sided conversations that revolve almost exclusively around the child's intense interest or include irrelevant detail
- Lack of reciprocal exchange (monologue-like)
- Inadequate clarification, vague references
- Topic preoccupation, perseveration

Scripted, stereotyped discourse

Overly literal; difficulty understanding nonliteral forms of communication, such as idioms, metaphors, humor/jokes, sarcasm, and irony

Difficulty recognizing and resolving communication breakdowns; unresponsive to partner's cues

Difficulty adapting style of communication to social situation
- Inability to infer expected degree of formality
- Excessively formal or pedantic style (especially in Asperger syndrome)

Good memory for details, difficulty understanding themes

Lack of appropriate use of facial expression, gestures and body postures to facilitate communication

Difficulty with flexible cooperative imaginative play, such as interactive role-playing with peers

Lack of creativity and imagination

Box 13.5. Clinical Features of Autism Spectrum Disorders: Repetitive Behavior and Restricted Range of Interests and Activities

Characteristics in Younger or More Severely Impaired Children	Characteristics in Older or Less Severely Impaired Children
Motor stereotypy - Hand flapping, finger flicking, rocking, spinning, jumping or prancing on toes, whole body twisting or posturing, etc	Special interests of abnormal intensity or focus - Obsession with facts, details, or collections
Fascination with objects that move or spin	Compulsions, rituals - Tapping, rubbing, sniffing, licking - Hoarding
Atypical use of peripheral vision or lateral eye gaze	Stereotyped motor mannerisms
Attachment to unusual objects - Often hard rather than soft - Category may be more important than the specific object (eg, attachment to batteries in general rather than one specific battery)	Desire to maintain sameness
Behavioral rigidity, difficulty accepting unexpected change, insistence on sameness	
Repetitive, stereotyped self-injurious behavior	
Extreme dietary selectivity	

Autistic Regression

Approximately 20% to 35% of children with ASDs begin to speak but then experience regression in language and broader interactive skills or, less commonly, in language alone.[46] Regression is commonly defined as loss of meaningful spoken language after acquisition of at least 3 to 5 words, often accompanied by loss of gesture communication, decreased use of eye gaze to regulate social interaction, social withdrawal, and loss of spontaneous imitation and symbolic play skills.[46,47] Autistic regression occurs most often between the ages of 15 and 24 months, can be gradual or sudden, and may occur following typical development or be superimposed on preexisting atypical development.[46,47]

Psychological Features

Historically, intellectual disability has been an associated diagnosis in 70% to 75% of children with ASDs, but more recent epidemiological studies suggest that the prevalence of intellectual disability is 40% to 55%.[7] Studies suggesting lower prevalence of coexisting intellectual disability include children with Asperger syndrome, in whom the diagnosis of intellectual disability is excluded, and those with PDD-NOS.

Children with autistic disorder tend to have higher performance IQ than verbal IQ and high intersubtest variability on standard intelligence tests. They often have difficulty with intellectual tasks that involve language, abstract reasoning, integration, and sequencing, whereas relative strengths include visual-spatial processing and rote memory abilities.[48] Individuals with Asperger syndrome often have a

different profile, with significantly higher verbal IQ than performance IQ, at least when strict *DSM-IV* or *International Classification of Diseases, Tenth Revision (ICD-10)* criteria (including absence of significant early language delay) are used for diagnosis.[49] Some studies suggest significant overlap between the neuropsychological profile associated with Asperger syndrome and a cluster of neuropsychological deficits and assets captured by the term *nonverbal learning disability.*[49]

Performance profiles also vary on standardized measures of academic achievement. Skills requiring primarily rote, mechanical, or procedural abilities are typically intact, but skills requiring more abstract reasoning, conceptualization, or interpretive analysis are deficient.[48] For example, a child with an ASD may have very good word and pseudoword decoding abilities but poor reading comprehension. Because the types of skills emphasized in the early elementary grades play to their strengths in memorization and mechanical procedures, such as sound-symbol correspondence and numerical operations, young children with ASDs may perform well academically. In later elementary and middle school grades, when comprehension, analytic interpretation, and abstract reasoning skills are emphasized, these children often struggle academically and require more supports. Even high-functioning children with ASDs typically have weaknesses in graphomotor skills, attention, and processing speed that interfere substantially with academic performance.[50]

Psychological features and theories that help to explain the behavior and psychological performance profile of individuals with ASDs are described in Table 13.2.[51–53] Recognition of the fundamental deficits in perspective-taking, cognitive flexibility, and ability to process complex information and form a coherent global picture is very helpful in understanding the behavior of children with ASDs and explaining it to parents and teachers.

Table 13.2. Psychological Features/Theories[a,b]

Feature/Theory	Description
Social motivation impairment	Early innate lack of responsiveness to social stimuli and motivation to socialize is theorized to lead to the other deficits that characterize ASDs. - Failure to orient to social stimuli, such as human sounds, and to prefer human to nonhuman speech in infancy. - Impaired face recognition, emotion recognition from faces and from voices, and matching facial and vocal expressions of emotions.
Theory of mind deficit (mindblindness, mentalizing deficit)	Impaired ability to take the perspective of others and understand that other people have intentions, knowledge, and beliefs that may differ from their own. - Necessary for developing the ability to identify other people's intentions (based on their gestures, expressions, and speech) and to understand deception.

Table 13.2. Psychological Features/Theories[a,b] *(continued)*

Feature/Theory	Description
Executive function impairment	Deficit in using executive functions to go beyond automatic activities and plan and carry out an integrated course of action. - Difficulty with creating strategies for behavior, making plans, shifting topics, maintaining a representation in working memory, solving tasks requiring ability to be flexible and innovative. - Deficits on tasks such as the Tower of London, Tower of Hanoi, and Wisconsin Card Sorting Test.
Weak central coherence	Limited drive for "central coherence" (meaningful wholes). - Tendency to focus on details and overlook broader contexts. - As a group, children with ASDs are good at finding figures embedded in larger forms, completing jigsaw puzzles, and reproducing patterns with blocks. - Approximately 20% have islets of special ability, or splinter skills.
Complex information processing impairment	Overall information processing is reduced relative to general cognitive ability level. Abilities and tasks most impacted are those that place the highest demands on information processing. - Difficulty with memory for complex information - Difficulty with higher-order interpretative aspects of language and concept formation - Intact basic attention, sensory perception, associative memory, and language encoding and decoding abilities The information processing disturbance is generalized, and includes nonsocial cognition. Many things that come naturally to typically developing children must be cognitively discovered by or explicitly taught to those with ASDs using compensatory strategies.
"Extreme male" systematizing/empathizing profile	Empathizing—capacity to predict and respond to the behavior of agents (usually people) by inferring their mental states (cognitive empathy) and responding with an appropriate emotion (affective empathy) Systematizing—capacity to predict and respond to the behavior of nonagentive deterministic systems by analyzing input-operation-output relations and inferring the rules that govern such systems In ASDs, there is a profile of relatively low empathizing (explaining social communication deficits), and high systematizing (explaining the narrow interests, repetitive behavior, and insistence on sameness)—a more extreme form of the typical male pattern

Abbreviation: ASD, autism spectrum disorders.

[a] Sources: Baron-Cohen et al 2005,[51] Minshew et al 2006,[53] Sigman et al 2006.[52]

[b] These theories and features are overlapping, not mutually exclusive.

Associated Clinical Features

In addition to the core impairments that define ASDs, there are other common neurologic findings, medical problems, and behavioral features that impact the clinical presentation and management of these conditions.

Neurologic Subtle Signs/Sensorimotor Symptoms

In addition to motor stereotypies, children with ASDs often exhibit less obvious motor abnormalities, including dyspraxia (developmental motor coordination disorder, clumsiness), idiopathic toe walking, mild hypotonia with associated increased joint mobility, decreased postural stability, and other subtle neurologic signs, such as slow speed and dysrhythmia on timed movements of the hands and feet and overflow movements during performance of stressed gait maneuvers and timed movements.[29,53,54] Tic disorders, including Tourette disorder, may also occur with increased frequency in association with ASDs.[55]

Sensory modulation symptoms have been defined as difficulties in regulating and organizing the type and intensity of behavioral responses to sensory input to match environmental demands and are categorized as follows: (a) over-responsivity, which is exaggerated, rapid onset, and/or prolonged reactions to sensory stimulation; (b) under-responsivity, which is unawareness or slow response to sensory input; and (c) seeking, which describes craving of and interest in sensory experiences that are prolonged or intense.[56] Some examples of behaviors that have been attributed by some researchers and clinicians to sensory modulation or other sensory processing dysfunction are included in Box 13.6.[29,56] Unusual sensory responses are described more commonly in children with ASDs than in typically developing children (effect

Box 13.6. Sensory Modulation Dysfunction

Over-responsivity
Distress, ear covering in response to loud noises or certain pitches
Gagging in response to smells or tastes or even the sight of foods with certain textures
Tactile defensiveness, aversion to social touch or certain textures
Gravitational insecurity (fear of backward movement, heights, being out of the upright position)

Under-responsivity
Lack of response to certain sounds, especially human voice
Unawareness of obstacles, walking over or into things
Apparent insensitivity to pain
Increased tolerance of spinning or being upside-down

Sensation-seeking
Smelling or licking people or objects
Lateral eye gaze, atypical visual inspection (especially while moving an object or fingers)
Craving deep pressure, squeezing into tight places
Prolonged rocking, jumping, or other rhythmic movements

size for under-responsivity > over-responsivity > sensation seeking).[56] However, the evidence that these symptoms differentiate children with ASDs from children with other developmental disabilities is quite limited.[56,57]

Epilepsy

Epilepsy occurs in approximately 20% to 35% of patients with ASDs, and most cases present in the first few years of life or in adolescence.[58] Although the prevalence is highest in individuals with severe intellectual disability and those with identified genetic etiologies, even the subgroup with idiopathic, high-functioning autism has a rate of epilepsy that is substantially higher than the rate in the general population.[58] Electroencephalographic abnormalities are common, but many children with abnormal EEGs do not have clinical seizures. The relationships among regression, epileptiform EEG, and epilepsy remain unclear.[58,59] Some studies suggest that epileptiform EEGs and/or epilepsy occur at greater frequency in children with a history of autistic regression, whereas others do not.[58] Whether subclinical seizures have adverse effects on language, cognition, and behavior is controversial, and no evidence-based recommendations can be made at this time regarding treatment of children with ASDs and epileptiform EEGs, with or without regression.[58,59]

Neurobehavioral Features/Coexisting Psychiatric Disorders

Maladaptive behaviors or psychiatric symptoms including irritability, tantrums, aggression, self-injurious behavior, property destruction, hyperactivity, impulsivity, inattention, repetitive behaviors (eg, perseveration, obsessions, compulsions), mood lability, anxiety, insomnia, and pica are commonly associated with ASDs.[60,61] In some cases, the diagnosis of a coexisting psychiatric disorder (eg, major depression, bipolar disorder, generalized anxiety disorder) can be reasonably made, although modifications of diagnostic criteria may be necessary. Although the *DSM-IV-TR* criteria prohibit the diagnosis of attention-deficit/hyperactivity disorder (ADHD) in the presence of an ASD, ADHD symptoms (inattention, distractibility, hyperactivity, and impulsivity) are commonly identified as significant problems by parents and teachers.[60,61]

Sleep problems are reported in 44% to 86% of children with ASDs, and they are common at all ages and levels of intellectual ability.[62] Sleep problems correlate with family distress and often have a significant impact on daytime behavior and quality of life.[4,62] Insomnia, which may include difficulty falling asleep, difficulty remaining asleep, or early morning awakening, is the most common problem. Behavioral factors, including inadequate sleep hygiene, maladaptive sleep associations (eg, holding the child until asleep), and problems with limit-setting, are common, but ASD-associated melatonin or GABA abnormalities, coexisting neurologic or psychiatric disorders (eg, epilepsy or anxiety), other medical problems (eg, gastroesophageal reflux), and adverse effects of medications may also contribute to disordered sleep.[62] Occasionally, other sleep disorders, such as obstructive sleep apnea, parasomnias, and periodic limb movements of sleep, are identified.

Gastrointestinal Problems

Some surveys suggest that gastrointestinal (GI) problems such as chronic constipation and diarrhea are more common in children with ASDs, but the relationship is not yet clear because most studies have not compared representative groups of children with ASDs to appropriate controls.[63] A retrospective study using complete medical records showed that there was no difference in the cumulative incidence of GI problems between a well-characterized population cohort of 121 individuals with autism and 242 matched controls.[64] Constipation and feeding issues/food selectivity where more prevalent in the group with autism, but there was no increase in diarrhea, abdominal bloating/discomfort, or gastroesophageal reflux/vomiting.[64]

In a large, well-characterized sample of children who participated in 2 Research Units on Pediatric Psychopharmacology (RUPP) Autism Network trials, the prevalence of GI problems was 22.7%, which is similar to the prevalence found in some population-based studies (17%–18.8%) and a convenience sample from an ASD clinic (24.1%), but substantially lower than the 68% lifetime prevalence of GI symptoms in another convenience sample ascertained through pediatric neurology and developmental pediatrics specialty clinics and even higher rates in gastroenterology clinic samples.[4,62–67] In the RUPP sample, children with GI problems had greater symptom severity on measures of irritability, anxiety, and social withdrawal relative to those without GI problems, and their irritability was less likely to respond to treatment with risperidone.[65] There is some evidence suggesting that reported GI symptoms, history of language regression, and family history of autoimmune disease are associated, but other studies suggest no link between GI symptoms and history of regression.[66,67]

Identification

Surveillance and Screening

Surveillance is the ongoing process of identifying children who may be at risk for developmental disorders, and screening is the use of standardized tools at specific intervals to support and refine risk. The American Academy of Pediatrics (AAP) has established guidelines for developmental surveillance and screening.[68] Surveillance is a continuous process that should occur at every preventive care visit during childhood and include the following components: eliciting and attending to the parents' concerns, maintaining a developmental history, making accurate and informed observations of the child, identifying the presence of risk factors and protective factors, and documenting the process and findings.[68]

The AAP recommends general developmental screening using a broad-band measure for all children at ages 9, 18, and 24 to 30 months of age or at any time concerns are raised by a caregiver or primary pediatric health care professional.[68] In addition, specific screening for ASDs is recommended at the 18- and 24-month visits and whenever risk factors are identified through general developmental surveillance and

screening (Figure 13.1).[3] The recommendation for universal screening for ASDs in toddlers was based on the recognition that symptoms can be observed in very young children and mounting evidence that early identification leads to early intervention and improved outcomes.[3,4] A variety of general developmental (broad-band), language-specific, and autism-specific screening tools have been shown to identify

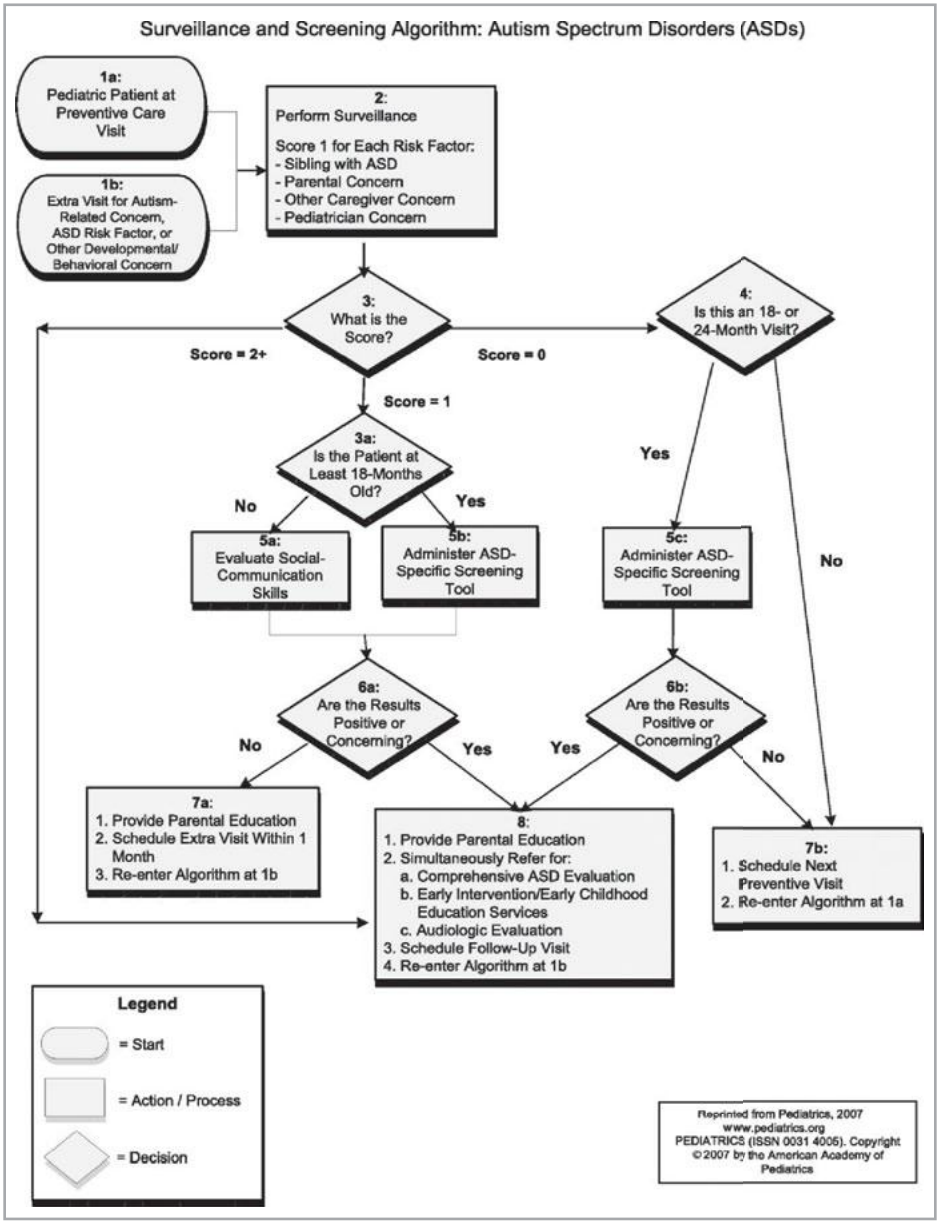

Figure 13.1. Surveillance and screening algorithm: autism spectrum disorders (ASDs).
From: Johnson CP, Myers SM, American Academy of Pediatrics Council on Children With Disabilities. Identification and evaluation of children with autism spectrum disorders. *Pediatrics.* 2007;120:1183–1215.

children of different ages who are at risk for ASDs.[3,69–71] Some examples of parent-completed screening measures that may be appropriate for use in primary care are described in Table 13.3. If a screening test is positive ("failed"), suggesting risk of an ASD, the child should be simultaneously referred to the early intervention program or school evaluation team for services and to a specialist or team for comprehensive diagnostic evaluation.[3] Audiologic evaluation is often warranted, particularly in a young child with delayed receptive and expressive language development.

Table 13.3. Examples of Parent-Completed Screening Instruments for Use in Primary Care

Screening Instrument	Age Range	Time to Complete, minutes
Infant-Toddler Checklist (ITC)[a]	9–24 months	5–10
Modified Checklist for Autism in Toddlers (M-CHAT)	16–48 months	5–10
Pervasive Developmental Disorders Screening Test-II Primary Care Screener (PDDST-II PCS)	18–48 months	10–15
Childhood Autism Spectrum Test (CAST)	4–11 years	10
Social Communication Questionnaire (SCQ)	4–18 years	10–15
Social Responsiveness Scale (SRS)	4–18 years[b]	15–20
Autism Spectrum Screening Questionnaire (ASSQ)	6–17	10

[a] The ITC is part of the Communication and Symbolic Behavior Scales Developmental Profile (CSBS DP).
[b] An adult version is also available.

Diagnostic Evaluation

The diagnostic evaluation will vary depending on the availability of local resources and clinician preferences. An interdisciplinary team specializing in ASDs and other developmental disabilities may be ideal, but often these are not available outside of major medical centers, and wait lists may be very long because of high demand. Many communities will have at least one expert who is capable of interpreting and integrating information from various disciplines and comfortable with making the diagnosis of an ASD even if there is not an interdisciplinary team that meets in person. This may be a developmental-behavioral or neurodevelopmental pediatrician, child and adolescent psychiatrist, pediatric neurologist, or pediatric psychologist.

The clinical standard of care for ASD diagnosis is expert clinician evaluation and application of the *DSM-IV-TR* or *ICD-10* criteria. In the hands of trained clinicians, diagnostic instruments (Table 13.4) help to operationalize the *DSM-IV-TR* criteria and inform clinical judgment.[3,72]

Table 13.4. Diagnostic Evaluation Tools

Instrument	Format
Autism Diagnostic Interview—Revised (ADI-R)	Structured caregiver interview
Diagnostic Interview for Social and Communication Disorders (DISCO)	Structured caregiver interview
Autism Diagnostic Observation Schedule (ADOS)	Semistructured direct observation/elicitation
Childhood Autism Rating Scale (CARS)	Combination interview and unstructured observation/elicitation

The diagnostic evaluation should ideally incorporate the following elements[3]:

1. *Caregiver interview,* with thorough medical history and review of systems, developmental and behavioral history, social history, and family history (at least 3 generations). Structured interview tools (eg, Autism Diagnostic Interview-Revised, Diagnostic Interview for Social and Communication Disorders) are informative but are often impractical in clinical settings because of time constraints.
2. *Direct clinical assessment,* including physical examination targeting neurologic examination and dysmorphic features, and developmental and/or psychological testing (appropriate for age and level of ability). Receptive and expressive language, nonverbal intellectual ability, and functional adaptive behavior should be measured. In addition, the use of standardized ASD-specific direct observation/elicitation instruments (eg, Autism Diagnostic Observation Schedule, Childhood Autism Rating Scale) is desirable.
3. *Determination of* DSM-IV-TR *(or* ICD-10) *categorical diagnoses.* Clinical judgment is informed by the caregiver interview and direct clinical assessment, often including information from ASD-specific diagnostic instruments, and by input from other sources (eg, narrative reports or standardized rating scales completed by a teacher, paraprofessional aide, or therapist).

There may be considerable overlap in symptoms of developmental and behavioral diagnoses, especially in very young children and those with intellectual disabilities or language impairments, and it is important for the diagnostician to consider the alternate explanations for observed symptoms.

Management

The role of the physician does not end with diagnosis. Comprehensive management of patients after the diagnosis of an ASD includes conducting an etiologic investigation; providing genetic counseling; promoting general health and well-being through effective longitudinal health care within a medical home; guiding families to effective educational and medical interventions; and alleviating family distress by providing

support, education, and access to resources. Primary pediatric health care professionals may take part in any or all of these duties in addition to making referrals and coordinating subspecialist care and other services.

Etiologic Evaluation

At this time there is no single approach to etiologic evaluation for children newly diagnosed with ASDs. Laboratory testing for children with ASDs is controversial, and in recent years the availability of new methods of genetic testing has progressed so rapidly that the publication of state-of-the-art guidelines by professional organizations lags behind. Therefore, the recommendations in this chapter reflect the authors' assessment of the current literature rather than strict adherence to the guidelines published previously by various professional organizations. The yield of an etiologic investigation may be more highly correlated with the presence or absence of coexisting intellectual disability or global developmental delay than with meeting criteria for an ASD.

Genetic Testing

All patients with ASDs who also have intellectual disability or global developmental delay should be offered genetic testing, including whole genome microarray comparative genomic hybridization (CGH) and fragile X DNA analysis, which will detect most of the common genetic etiologies outlined in Table 13.1 as well as others. Whole genome microarray CGH is more sensitive than the combination of high-resolution karyotype, subtelomere fluorescence in-situ hybridization (FISH) screening, and specific FISH probes for the 2q, 15q, 16p, and 22q regions that would otherwise be necessary to detect the most common cytogenetic abnormalities currently reported in individuals with ASD. Other specific genetic tests may also be appropriate, depending on the personal and family medical history and physical examination findings (Table 13.5). For example, phosphatase and tensin homolog (PTEN) sequence analysis is recommended in the case of marked macrocephaly (head circumference >2.5 standard deviations above the mean) even in the absence of typical skin findings of Bannayan-Riley-Ruvalcaba syndrome or Cowden syndrome.[73] Skin findings suggestive of tuberous sclerosis or neurofibromatosis may warrant other investigations, such as MRI of the brain and ophthalmologic examination, and occasionally confirmation of the diagnosis by genetic testing is necessary. It is likely that in the near future, cost-effective panels will be available to screen simultaneously for the known single gene causes as well as etiologic microdeletions and microduplications.

In patients with Asperger syndrome or high-functioning autism (HFA; defined as either autistic disorder or PDD-NOS without associated intellectual disability), especially those with average or above average cognitive ability, the appropriate etiologic investigation is less clear. The prevalence rates of chromosomal anomalies and fragile X syndrome in this population are unknown, but reports in the literature are relatively uncommon.[74] Fragile X syndrome and chromosomal deletions and

Table 13.5. Selected Clinical Findings in Genetic Conditions Associated With Autism Spectrum Disorders

Physical Findings	Condition	Confirmatory Testing	Importance
Characteristic facial features, 2–3 toe syndactyly, postaxial polydactyly, ptosis, microcephaly, cleft palate, hypospadias in males, growth retardation	Smith-Lemli-Opitz syndrome	7-dehydrocholesterol level (elevated); DHCR7 sequence analysis available	Genetic counseling (autosomal recessive inheritance) Potential role for treatment with cholesterol
Marked macrocephaly, skin hamartomas, pigmented macules of the glans penis	PTEN hamartoma tumor syndrome (includes Cowden syndrome, Bannayan-Riley-Ruvalcaba syndrome, Proteus syndrome, and Lhermitte-Duclos disease)	PTEN sequence analysis, deletion/duplication analysis	Genetic counseling (autosomal dominant inheritance with highly variable expression) Significant risk of malignancy
Hypopigmented macules, angiofibromas, shagreen patches (connective tissue nevi), ungula fibromas, retinal hamartomas	Tuberous sclerosis	Clinical criteria; TSC1 and TSC2 sequencing available	Genetic counseling (autosomal dominant inheritance) Associated problems requiring investigation or monitoring (CNS tumors, seizures, renal angiolyolipomas or cysts, cardiac rhabdomyomas and arrhythmias)
Multiple café-au-lait macules, axillary and inguinal freckling, iris Lisch nodules, dermal neurofibromas	NF1	Clinical criteria; optimized protein truncation testing and sequencing available but rarely required	Genetic counseling (autosomal dominant inheritance) Associated problems requiring investigation or monitoring (optic gliomas, other CNS tumors, peripheral nerve sheath tumors, vasculopathy, hypertension, orthopedic issues, osteopenia)

Table 13.5. Selected Clinical Findings in Genetic Conditions Associated With Autism Spectrum Disorders *(continued)*

Physical Findings	Condition	Confirmatory Testing	Importance
Deceleration of head growth velocity, acquired microcephaly, loss of purposeful hand use, prominent hand stereotypies (especially hand wringing or clasping), apraxia, hyperventilation or breath-holding, seizures	Rett syndrome	MECP2 sequence analysis	Genetic counseling (>99% de novo, <1% germline mosaicism) Associated problems requiring investigation or monitoring and anticipatory guidance (failure to thrive, gastroesphageal reflux, respiratory problems, osteopenia, sudden death)

Abbreviations: CNS, central nervous system; DHCR7, 7-dehydrocholesterol reductase; MECP2, methyl-CpG-binding protein 2; NF1, neurofibromatosis 1; PTEN, phosphatase and tensin homolog; TSC1, tuberous sclerosis 1; TSC2, tuberous sclerosis 2.

duplications are most often associated with intellectual disability in addition to the various particular physical phenotypes, so it is likely that the yield would be low in patients with Asperger syndrome or HFA, who by definition do not have intellectual disability. Unless future population-based studies reveal that a significant percentage of individuals with appropriately diagnosed Asperger syndrome or HFA have identifiable genetic abnormalities, it is the opinion of the authors that testing of individuals with an ASD without significantly subaverage general intellectual functioning should be limited to those individuals with specific indications, such as dysmorphic features or a family history suggestive of X-linked inheritance.[3,74]

Metabolic Testing

In rare cases, mitochondrial disorders or dysfunction and a variety of other neurometabolic disorders have been identified in association with ASDs.[75] Several studies have suggested that the yield of routine metabolic testing in patients with ASDs is very low. However, large population-based studies are lacking, so accurate prevalence estimates are not available and the evidence on which to base recommendations for testing is quite limited. Patients with primary mitochondrial disorders are likely to have multisystem dysfunction, including neurologic abnormalities such as cognitive impairment, significant neuromotor dysfunction, seizures, and aberrant behavior. The neurobehavioral abnormalities may include autistic features, but published evidence that mitochondrial disorders are a common cause of primary, uncomplicated ASDs is lacking.[75]

At this time, based on the available literature, no metabolic testing should be routinely recommended for all children with ASDs.[3] However, cognitive and motor regression, unusual prostration with illness, episodic encephalopathy, severe

hypotonia, ataxia, nystagmus, epilepsy, and severe or profound intellectual disability are factors that should raise the level of suspicion of an inborn error of metabolism. When indicated, a test battery that includes measurement of plasma amino acids, ammonia, lactate, pyruvate, and acylcarnitines, as well as urine organic acids and mucopolysaccharides, is likely to detect most of the pertinent neurometabolic disorders, as well as biochemical evidence of mitochondrial dysfunction. Clinical features may warrant measurement of 7-dehydrocholesterol (see Table 13.5), biotinidase, urine guanidinoacetate, cerebrospinal fluid 5-methyltetrahydrofolate, or other specific substances in isolated cases.[75]

Imaging and Neurophysiologic Testing

Although neuroimaging research has identified volumetric and other abnormalities in groups of patients with ASDs compared with controls, reliable markers for diagnosis or subtyping have not been identified, and routine clinical neuroimaging for individuals with ASDs is not recommended by the AAP.[3] However, in a subset of children, features such as focal seizures or neurocutaneous findings may warrant an MRI of the brain. The American College of Medical Genetics practice guidelines recommend MRI as a third-tier investigation, to be performed if the etiology is not determined by physical examination, karyotype, fragile X testing, microarray CGH, methyl-CpG-binding protein 2 (MECP2) analysis (in females), metabolic screening, and other genetic and metabolic testing as warranted based on specific features.[76] The practice parameters for the evaluation of the child with intellectual disability or global developmental delay published by the AAP Committee on Genetics and the American Academy of Neurology/Child Neurology Society both also endorse MRI of the brain if routine genetic testing is negative and the history and examination suggest features such as intrapartum asphyxia, microcephaly, focal neurologic findings on examination, cerebral palsy, focal seizures, or developmental regression.[77]

Electroencephalography is not routinely indicated for all individuals with ASDs, but should be considered in the setting of clinical spells that might represent seizures and possibly when there is isolated language regression after 24 months of age or broader autistic regression (including language, social and behavioral regression) after 36 months of age.[4,59]

Genetic Counseling

For a couple with one child with an ASD of unknown etiology, the rate of recurrence in a subsequent child is approximately 4% to 8% (4%–5% if the affected child is a girl, 7%–8% if the affected child is a boy). If there are already 2 children with autism, the risk of the couple having a third affected child may be as high as 25% to 30%.[14,76] Abnormalities identified on chromosomal microarray often require evaluation of parent samples to determine if the alteration is de novo or inherited. For certain identified genetic etiologies, the recurrence risk can be as high as 50% (eg, in the

case of a female carrier of a 15q11-q13 interstitial duplication) or very low (eg, in the case of a de novo microdeletion). Consultation with a clinical geneticist is suggested if a specific genetic etiology for the ASD is identified.

Educational Interventions

Treatment goals include (1) remediating or minimizing the core deficits (social impairment; communication impairment; and restricted, repetitive behavioral repertoire) and associated deficits; (2) maximizing functional independence by facilitating learning and academic achievement, acquisition of self-care and daily living skills, and development of play and leisure skills; and (3) eliminating or minimizing aberrant, maladaptive behaviors that interfere with functioning.[4]

Intensive, structured educational interventions, including behavior analytic strategies and habilitative therapies, are the cornerstones of management of ASDs.[4] Although most pediatricians are not experts on educational interventions for ASDs, they are often in a position to make referrals, provide resources, and guide families to empirically supported interventions. Characteristics of effective intervention programs are outlined in Box 13.7.[78–81] Treatment programs should be assessment-based, individualized, and intensive. The focus throughout childhood and adolescence is on achieving social communication competence, emotional and behavioral regulation, and functional adaptive skills necessary for independence.

A wide variety of methods have been used in educational programs for children with ASDs. All treatments, including educational interventions, should be based on sound theoretical constructs, rigorous methodologies, and objective scientific evidence of efficacy. There is some published evidence supporting the efficacy of the common intervention categories outlined in Table 13.6 for addressing communication, social skills, daily living skills, play and leisure skills, learning and academic achievement, or maladaptive behaviors in children with ASDs, although the quality of the research varies greatly and many methods, despite their popularity, have not been adequately evaluated.[4,80–82] Some methods, such as facilitated communication, have been proven ineffective, and the weight of the available evidence (or lack of evidence) does not support the use of interventions such as auditory integration training, dolphin-assisted therapy, holding therapy, interactive metronome, therapeutic touch, or vision therapy in the treatment of ASDs.[4,82,83]

Systematic reviews of the literature have concluded that early intensive intervention programs based on applied behavior analysis (ABA), particularly those based on the Lovaas model, are currently the most well-established, evidence-based treatments for young children with ASDs, although there is wide variability in response to treatment.[84–86] Rogers and Vismara[84] noted that a lack of strong study designs and independent replications or, in some cases, a lack of any peer-reviewed published data at all, prevented other well-known autism interventions from meeting preestablished,

Box 13.7. Components of Effective Educational Intervention for Children With Autism Spectrum Disorders[a]

Assessment of existing skills and formulation of individualized goals and objectives

Intensive, active engagement in systematically planned, developmentally appropriate educational activities designed to address identified objectives

Use of assessment-based, empirically supported methods to address
- Functional, spontaneous communication
- Social skills, including joint attention, imitation, reciprocal interaction, initiation, and self-management
- Symbolic play skills and leisure skills
- Cognitive skills, such as perspective-taking
- Functional adaptive skills that prepare the child for increased responsibility and independence
- Traditional academic skills
- Vocational skills
- Reduction of disruptive or maladaptive behavior using empirically supported strategies, including functional assessment

Low student-to-teacher ratio to allow sufficient amounts of 1:1 time and small group instruction to meet specific individualized goals

Incorporation of a high degree of structure through elements such as predictable routine, visual activity schedules, and physical boundaries to minimize distractions

Implementation of strategies to generalize learned skills to new environments and situations and to maintain functional use of these skills

Ongoing measurement and documentation of the individual child's progress toward educational objectives, resulting in adjustments in teaching strategies as necessary to enable students to acquire target skills

Promotion of opportunities for interaction with typically developing peers to the extent that these opportunities are helpful in addressing specified goals

Explicit transition planning (eg, from home-based early intervention to preschool services, preschool to elementary school, elementary school to middle school, middle school to high school, high school to work or postsecondary education, home to community living)

Inclusion of a family component, including parent training as indicated

[a] Adapted from: Myers, Pediatric Annals, 2009.[78] Reprinted by permission of Slack, Inc. Additional sources: National Research Council 2001.[79] Volkmar et al 2005.[80] Myers et al 2007.[4]

published criteria for the designations "well established" or "probably efficacious." This does not mean that they are ineffective, only that efficacy has not been established in adequately designed, replicated clinical studies.

Although the ABA-based interventions dominate the research literature, developmental models are widely used in early intervention programs and schools and, despite the difference in primary philosophical orientation, there is substantial integration of behavioral and developmental practices in community and research models.[4,84] There is good evidence that pivotal response training, a behavior analytic intervention that uses a developmental framework, is effective for teaching a variety

of skills necessary for further development of language and social interaction.[84] Evidence from several randomized, controlled trials suggest that other interventions focusing on communication (including the Picture Exchange Communication System) and joint attention may have significant impact on functioning of children with ASDs.[87,88] A randomized, controlled trial involving 48 toddlers with ASDs demonstrated the efficacy of the Early Start Denver Model, which integrates ABA with developmental and relationship-based approaches.[89]

In many communities, an "eclectic" treatment approach drawing on a combination of methods, such as ABA, structured teaching procedures, relationship-based strategies, speech-language therapy, sensory integration therapy, and general teaching practices, is used. The assumption is that assembling intervention techniques from various methods is advantageous due to tailoring of intervention to the learning and behavioral characteristics of each individual child. However, several studies comparing ABA programs to equally intensive eclectic approaches have demonstrated more favorable outcomes in the groups receiving behaviorally based interventions and raised questions about the general efficacy of eclectic approaches.[4,84] At a minimum, it is important for educators who choose to use portions of multiple intervention approaches simultaneously to carefully assess the underlying assumptions and goals of the various methods they plan to combine, build the program from empirically supported teaching practices, and monitor and document each child's progress.[84]

Table 13.6. Common Educational/Habilitative Methods[a]

General Category[b]	Examples	Comments
Applied behavior analysis (ABA)	Discrete trial training Naturalistic application of behavioral principles - Incidental teaching - Pivotal response training Applied verbal behavior Functional behavior analysis (functional assessment) Errorless teaching	ABA-based interventions have the strongest empirical support in the published, peer-reviewed research literature. In addition to their importance in teaching and maintaining new skills and desirable behaviors, behavior analytic methods are the primary treatment for problematic maladaptive behaviors such as self-injury and aggression.
Structured teaching	Treatment and Education of Autistic and Related Communication Handicapped Children (TEACCH)	The TEACCH approach includes an emphasis on improving skills of individuals and modifying the environment to accommodate their deficits. Physical organization, visual schedules, work systems, and task organization are emphasized.

Table 13.6. Common Educational/Habilitative Methods[a] *(continued)*

General Category[b]	Examples	Comments
Developmental models	Denver model Developmental, individual-difference, relationship-based model (DIR/Floortime) Relationship development intervention (RDI) Responsive teaching (RT) Social communication, emotional regulation, transactional support (SCERTS)	Developmental models focus on remediation of fundamental deficits in pivotal developmental skills that are thought to affect the development of social communication, emotional relationships, and cognitive abilities such as theory of mind.
Speech and language therapy	Didactic and naturalistic behavioral methods (applied behavior analysis) Developmental-pragmatic approaches Augmentative and alternative communication strategies - Sign language - Picture Exchange Communication System (PECS) - Voice output communication devices	A variety of methods are used to promote verbal and nonverbal communication. In addition to providing direct intervention, speech-language pathologists should train, support, and collaborate with teachers, educational support staff, parents/caregivers, and the child's peers to increase communication in all settings.
Social skills instruction	Social skills groups Joint action routines Social stories Visual cuing Social games Video modeling Social scripting Peer-mediated techniques Play and leisure curricula	Social skills curricula address skills such as initiating social interactions, responding to the social overtures of peers and adults, using a flexible and varied repertoire of responses, minimizing stereotyped or perseverative behavior, and self-managing new and established social skills.
Occupational therapy	Fine motor skills - Self-care - Academics Sensory integration (SI) therapy	Occupational therapy addresses fine motor deficits that have the potential to interfere with attaining academic, self-care, or prevocational goals. SI therapy is purported to remediate deficits in neurologic processing and integration of sensory information to allow the child to interact with the environment in a more adaptive fashion, although this has not been demonstrated objectively.

[a] Adapted from: Myers, *Pediatric Annals,* 2009.[78] Reprinted by permission of Slack, Inc. Additional sources: Volkmar et al 2005.[80] Myers et al 2007.[4] Simpson and Myles 2008.[81]

[b] Categories are not mutually exclusive; in practice, there is significant overlap.

Medical Management

Children with ASDs have the same basic health care needs as other children and benefit from the same health promotion and disease prevention activities, including immunizations. Some have additional health care needs related to associated medical problems, such as epilepsy, or to underlying etiologic conditions, such as those described in Tables 13.1 and 13.5. Efforts to optimize medical care are likely to have a positive impact on educational progress and quality of life, and it is important for primary pediatric health care professionals to provide longitudinal general health care, anticipatory guidance, and chronic disease management within a medical home.[4,78]

More time is often required for office visits, which may be challenging due to the patients' deficits in social interaction and communication, difficulty accepting novelty, and sensory aversions. Strategies such as familiarizing the patient with the office environment, staff, and routine during a "practice" visit; slowing down the pace of the visit; allowing ample time for talking before touching the patient; allowing the child to manipulate instruments and materials; keeping instructions simple; using visual cues and supports; exaggerating social cues; and having family and/or familiar staff available may be helpful in reducing resistance to physical examination and increasing family and patient comfort with the provider.[4,90]

Currently medical therapies are directed to specific symptoms or coexisting conditions rather than the ASD itself. For example, children with ASDs who have seizures are treated with anticonvulsant medications based on the same criteria that are used for other children with epilepsy, including accurate diagnosis of the particular seizure type.[58,91] Similarly, those who present with chronic or recurrent diarrhea or constipation should be evaluated and treated just as any other child with these symptoms would be evaluated and treated.[4,63]

Some children with ASDs have unusual food preferences based on texture, color, taste, smell, or temperature, and many develop very restricted diets. Overt malnutrition is uncommon; however, children on dairy restricted diets may have deficits in vitamin D and/or calcium.[63] Referral to a qualified nutritionist should be considered for children whose diets are significantly restricted due to extreme food selectivity or parental choice of dietary restriction as an intervention. Children with pica or persistent mouthing of objects or fingers should be evaluated for elevated blood lead concentrations, particularly if the history suggests potential environmental exposure. Evaluation for iron or zinc deficiency might also be indicated in children with pica.

Children and adolescents with ASDs often present with problematic emotional reactions and behaviors, such as irritability, tantrums, aggression, and self-injury.[60,61,92] In some cases, medical factors that result in discomfort may cause or exacerbate maladaptive behaviors (Table 13.7).[78,93] Recognition of these situations allows primary treatment of the medical condition, potentially eliminating the need for psychotropic medications or intensive behavioral interventions. For example, a child

Table 13.7. Treatable Medical Conditions That May Cause or Exacerbate Maladaptive Behaviors[a]

Medical Condition	Potential Treatment
Allergies - Atopic dermatitis - Environmental (rhinitis, conjunctivitis) - Food	Antihistamine therapy, anti-leukotriene therapy, topical or systemic steroid therapy, environmental interventions, dietary elimination
Dental discomfort - Abscess - Caries - Eruption - Impaction - Trauma	Antimicrobial therapy, analgesia, extraction or dental repair
Endocrine disorders - Hyperthyroidism, hypothyroidism - Premenstrual discomfort or dysphoria	Antithyroid medication, thyroid hormone replacement, analgesia, oral or injectable contraceptive therapy
Gastrointestinal disorders - Constipation - Diarrhea, cramping - Esophagitis, gastroesophageal reflux - Gastritis	Laxative therapy, fiber, acid-inhibiting therapy (proton pump inhibitors, histamine antagonists)
Infectious diseases - Otitis media - Otitis externa - Sinusitis - Pharyngitis	Antimicrobial therapy, analgesia
Medication side effects - Dietary supplements - Prescription medications - Over-the counter medications	Discontinuation of the offending agent
Musculoskeletal problems - Arthralgia - Strain or sprain - Occult fracture	Analgesia, anti-inflammatory agent, casting, splinting, rest, surgical intervention
Neurologic disorders - Headache (including migraine) - Seizures	Analgesia, abortive therapies, prophylactic therapies, anticonvulsant medication
Nutritional deficiencies - Iron deficiency - Protein-calorie malnutrition - Zinc deficiency	Nutritional supplementation
Ophthalmologic problems - Corneal abrasion	Analgesia, foreign body removal, temporary patching
Sleep disorders - Obstructive sleep apnea - Other sleep disordered breathing	Weight reduction, tonsillectomy and adenoidectomy, continuous positive airway pressure

[a] Adapted from: Myers 2008[93] and Myers 2009.[78] Reprinted by permission of Slack, Inc.

may present with sleep problems due to obstructive sleep apnea or gastroesophageal reflux or an acute onset or exacerbation of self-injurious behavior due to an occult source of discomfort, such as otitis media, constipation, or a dental abscess. The health care provider often must have a relatively high index of suspicion and complete a careful history and physical examination to detect these and other medical conditions in a patient who has limited ability to communicate and exhibits aberrant behaviors at baseline. Some behaviors may have their basis in a painful condition that has resolved. The challenging behavior can take on an operant, or learned, function that persists after resolution of the initial medical condition. In these cases, behavioral intervention may also be required.

It is also important to consider environmental factors that may precipitate challenging behaviors.[4,92,93] Parents, teachers, or other caregivers may inadvertently reinforce maladaptive behaviors, such as tantrums, aggression, self-injury, or elopement, by allowing these behaviors to become an effective means for the child to either escape from or avoid demands or situations that the child finds unpleasant or to gain access to attention or a tangible item, such as food or a preferred object.[94,95] In such cases, behavioral interventions are the most appropriate and effective treatment. Sometimes a mismatch between the educational or behavioral expectations and the cognitive ability of the child contributes substantially to the emergence of disruptive behavior (eg, when the degree of intellectual disability has not been recognized and the tasks are too difficult), and adjustment of expectations and demands is the most appropriate intervention. These situations can often be recognized by taking a careful history, but in more difficult cases formal assessment is necessary. A behavior specialist can complete a functional assessment and, if necessary, experimental functional analysis to identify the function or outcome of the behavior and develop a treatment program to reduce the rate of the maladaptive behavior and teach behaviors or skills to help the child achieve the same outcome through more appropriate means.[94,95]

While there may be biological differences in sleep architecture and melatonin release in children with ASDs, the caregiver response to inappropriate bedtime behavior, repeated night awakenings, daytime sleeping, and other sleep problems also frequently contributes to sleep disturbances. In addition to simple sleep hygiene measures and restriction of daytime sleep, specific behavioral interventions, such as bedtime scheduling, positive bedtime routines, bedtime fading, extinction, and graduated extinction, can be effective for sleep problems in children with ASDs.[96]

Psychopharmacology

Psychotropic medications have not been proven to correct the core deficits of ASDs. However, associated maladaptive behaviors or coexisting psychiatric conditions may interfere with educational progress, socialization, health and safety, and quality of life and, in some cases, these behaviors or symptoms may be amenable to treatment with psychotropic medication. Effective pharmacologic treatment may allow the child to

derive optimal benefit from educational and behavioral interventions.[4,78] After treatable medical causes and modifiable behavioral and environmental factors have been ruled out, a therapeutic trial of medication directed at specific target symptoms or a comorbid psychiatric diagnosis may be considered. Box 13.8 outlines principles to guide the clinical approach to psychopharmacologic management, from careful evaluation of target symptoms to initiation of treatment and adjustment of therapy based on monitoring of treatment response.[4,92,93]

Although a wide range of psychotropic medications have been used in children with ASDs, there is insufficient research literature to establish a consensus, evidence-based approach to psychopharmacologic management. However, in recent years, more information from randomized, double-blind, placebo-controlled clinical trials involving children and adolescents with ASDs has become available to guide clinical practice.[92,93] For example, there is substantial empiric evidence that risperidone and aripiprazole are effective for irritability and associated tantrums, aggression, and self-injurious behavior in children and adolescents with ASDs and that methylphenidate is effective for ADHD symptoms in this population. Preliminary controlled trials of atomoxetine for ADHD symptoms, alpha-2 adrenergic agonists for ADHD symptoms and irritability/outbursts, and valproate for repetitive behaviors are promising but require replication in larger studies.[92,93] Small double-blind, placebo-controlled randomized clinical trials of fluvoxamine in adults (n=30) and

Box 13.8. An Approach to Clinical Psychopharmacologic Management[a]

Evaluation of Target Symptoms

- Identify and assess target behaviors
  - Parent/caregiver interview, input from school staff and other caregivers
    - Frequency
    - Intensity
    - Duration
    - Exacerbating factors/triggers (time, setting/location, demand situations, denials, transitions, etc)
    - Ameliorating factors and response to behavioral interventions
    - Time trends (increasing, decreasing, stable)
    - Degree of interference with functioning
  - Baseline behavior rating scales and/or baseline direct observational data (eg, collect data on number of episodes of aggression or self-injury in a given time period)
  - Consider formal functional analysis of behavior
- Search for medical factors that may be causing or exacerbating target behavior(s)
  - Consider sources of pain or discomfort (see Table 13.7)
  - Consider other medical causes or contributors (obstructive sleep apnea, seizures, menstrual cycle, etc)
- Assess existing and available supports
  - Behavioral supports and services
  - Educational program, habilitative therapies
  - Family psychosocial supports, respite care
- Complete any medical tests that may have a bearing on treatment choice (eg, electroencephalography if possible seizures)

Box 13.8. An Approach to Clinical Psychopharmacologic Management[a] *(continued)*

Initiation and Monitoring of Psychopharmacologic Therapy

- Consider psychotropic medication based on the presence of
  - Evidence that the target symptoms are interfering substantially with learning/academic progress, socialization, health/safety, or quality of life
  - Suboptimal response to appropriate available behavioral interventions and environmental modifications
  - Research evidence that the target behavioral symptoms or coexisting psychiatric diagnoses are potentially amenable to pharmacologic intervention
- Choose a medication on the basis of
  - Likely efficacy for the specific target symptoms
  - Potential adverse effects
  - Practical considerations, such as formulations available, dosing schedule, cost, and requirement for laboratory or electrocardiographic monitoring
  - Patient's other medical conditions (eg, obesity, asthma, etc) that might be exacerbated by certain medications
  - Informed consent (verbal or written) from parent/guardian and, when possible, assent from the patient
- Establish a plan for monitoring of effects
  - Identify outcome measures for the target behaviors/symptoms
  - Discuss time course of expected effects and appropriate timing of follow-up telephone contact, completion of rating scales, reassessment of behavioral data, and office visits
  - Outline a plan regarding what might be tried next if there is a negative or suboptimal response or to address additional target symptoms
    - Change to a different medication
    - Add another medication to augment a partial or suboptimal therapeutic response to the initial medication (same target symptoms)
    - Add a different medication to address additional target symptoms that remain problematic
  - Obtain baseline laboratory data if necessary for the drug being prescribed and plan appropriate future lab monitoring
- Explore the reasonable dose range for a single medication for an adequate length of time before changing to or adding a different medication; titrate to optimal effect without intolerable side effects
- Monitor for adverse effects systematically
- Consider careful withdrawal of the medication after 6–12 months of therapy to determine whether it is still needed

[a] Adapted from: Myers et al 2007,[4] Myers 2008,[93] and Myers 2009.[92]

fluoxetine in children (n=39) suggest that these selective serotonin reuptake inhibitors (SSRIs) are superior to placebo for reducing repetitive behaviors associated with autism, but a larger trial (n=149) showed no difference between citalopram and placebo.[92–93,97] Currently, the evidence does not provide strong support for the use of SSRIs for the treatment of repetitive behaviors in children with ASDs, and further investigation is necessary to determine whether they are effective for other target symptoms, such as irritability and anxiety, in this population.

Clinical psychopharmacologic practice is also informed by well-designed studies conducted in other populations with overlapping target symptoms, such as obsessive-compulsive disorder, conduct disorder (with or without intellectual disability),

ADHD, anxiety disorders, depression, bipolar disorder, and schizophrenia. Some medication options for common target symptoms are listed in Table 13.8. Health

Table 13.8. Psychotropic Medication Options for Common Target Symptoms[a]

Target Symptoms	Medication Class	Disorders in Which Efficacy of Medication Class Has Been Established for Treatment of Similar Symptoms (Examples)	Agents With Published Evidence of Efficacy in ASD Population[b]
ADHD symptoms (hyperactivity, impulsivity, inattention, distractibility)	Psychostimulants	ADHD	Methylphenidate
	Selective norepinephrine reuptake inhibitors	ADHD	Atomoxetine
	Alpha-2 agonists	ADHD	Clonidine Guanfacine
Irritability, tantrums, aggression, self-injury	Atypical antipsychotics	Schizophrenia, bipolar mania, intellectual disability with conduct disorder	Aripiprazole[c] Olanzapine Risperidone[c]
	Alpha-2 agonists		Clonidine Guanfacine
	Selective serotonin reuptake inhibitors	Depression	Fluoxetine Fluvoxamine
	Anticonvulsants	Bipolar disorder	Valproic acid/divalproex sodium
Repetitive behavior, behavioral rigidity, obsessive-compulsive symptoms	Atypical antipsychotics	Schizophrenia, Tourette disorder	Aripiprazole[c] Olanzapine Risperidone[c]
	Anticonvulsants		Valproic acid/divalproex sodium
	Selective serotonin reuptake inhibitors	Obsessive-compulsive disorder	Fluoxetine Fluvoxamine
Anxiety	Selective serotonin reuptake inhibitors	Anxiety disorders (generalized anxiety disorder, separation anxiety disorder, social phobia)	Fluoxetine
Sleep dysfunction	Endogenous chronobiotic hormone with hypnotic properties		Melatonin

Abbreviations: ADHD, attention-deficit/hyperactivity disorder; ASD, autism spectrum disorder.

[a] Sources: Myers 2007[92] and Myers 2008.[93]

[b] At least one double-blind, placebo-controlled trial supporting efficacy in patients with ASDs.

[c] Risperidone and aripiprazole are currently the only medications with US Food and Drug Administration–approved labeling specific to autism (for the symptomatic treatment of irritability, including aggressive behavior, deliberate self-injury, and temper tantrums in children and adolescents with autism).

care providers should only prescribe medications with which they have sufficient expertise, including knowledge of indications and contraindications, dosing, potential adverse effects, drug-drug interactions, and monitoring requirements.[4,78,93]

Complementary and Alternative Medicine

Complementary and alternative medicine (CAM) interventions, defined by the National Center for Complementary and Alternative Medicine as "a group of diverse medical and health care systems, practices, and products that are not presently considered to be part of conventional medicine," are commonly used to treat children with ASDs.[98,99] For any intervention to be recommended, well-designed, appropriately controlled studies are required to prove that the observed effect is attributable to the intervention being studied. The most thoroughly evaluated CAM treatment for ASDs, intravenous secretin, has been unequivocally demonstrated to be ineffective.[100] Most CAM treatments have been inadequately evaluated and cannot be recommended for treatment of ASDs based on the available evidence (Box 13.9).[4,83,99] It is possible for a therapy initially deemed complementary or alternative to develop a sufficient evidence base to support its use in the treatment of a particular disorder. There is preliminary evidence from unreplicated controlled trials supporting some CAM interventions such as vitamin C and hyperbaric oxygen. However, at this time, there are no CAM therapies that have been adequately evaluated and determined to be effective for the treatment of ASDs.

Potential risks of CAM treatments include direct toxic effects of biological agents; adverse physical effects of manipulative techniques, presence of contaminants, interactions with prescribed medications, interference with appropriate nutrition, and interruption or postponement of empirically validated therapies, as well as unwarranted expenditure of time, effort, and financial resources.[83,99] The AAP has stated

Box 13.9. Examples of Treatments That Have Been Inadequately Evaluated and Cannot Be Recommended for the Treatment of Autism Spectrum Disorders Based on Currently Available Evidence[a]

Amino acids	Interactive metronome
Antibiotics	Intravenous immunoglobulin
Antifungals	Mineral supplements
Antivirals	Music therapy
Antioxidants	Omega-3 fatty acids
Auditory integration training	Oral immunoglobulin
Chelating agents	Therapeutic touch
Chiropractic manipulation	Transcranial magnetic stimulation
Craniosacral therapy	Trimethylglycine
Digestive enzymes	Vitamin A
Dimethylglycine	Vitamin B_6 and magnesium
Folic acid, folinic acid	Vitamin B_{12} (intramuscular or oral)
Gluten-free/casein-free diet	Vitamin C
Hyperbaric oxygen	Yoga

[a] From: Challman 2008,[83] Levy and Hyman 2008,[99] Myers et al 2007.[4]

that pediatricians should (1) critically evaluate the scientific evidence of efficacy and risk of harm of various treatments and convey this information to families, (2) help families understand how to evaluate scientific evidence and recognize unsubstantiated treatments and pseudoscience, and (3) insist that studies that examine CAM treatments be held to the same scientific standards as all clinical research.[4] The interested reader is referred to Chapter 21 in this volume for a more detailed discussion of CAM treatments in developmental and behavioral pediatrics.[101]

Clinical Resource

The AAP autism resource toolkit, *Autism: Caring for Children With Autism Spectrum Disorders: A Resource Toolkit for Clinicians,* was developed to support practicing health care professionals in the identification and ongoing management of children with ASDs within the medical home.[102] The toolkit includes resources such as AAP policy information, screening and surveillance tools, referral forms, sample letters, coding information, physician fact sheets reviewing important management issues, and family handouts on a variety of pertinent topics that can be provided to supplement discussion.

Outcomes

Although specific features change over time and outcomes vary, most children diagnosed with ASDs using *DSM* criteria remain within the spectrum as adolescents and adults. Regardless of their level of intellectual functioning, these individuals continue to experience difficulty with social relationships, independent living, employment, and mental/behavioral health.

Autism spectrum disorders are associated with increased morbidity, mortality, and health care utilization and costs. Mortality risk among individuals with ASDs is approximately twice that of the general population (standardized mortality ratio 1.9:2.6).[103,104] Female gender and epilepsy are associated with increased risk of death in this population.[103,104] Health care costs are increased, and it is estimated that the total economic cost to society to care for a person with an ASD is $3.2 million over his or her lifetime (including direct medical, direct nonmedical, and lost productivity costs).[105] Assuming a minimum prevalence of 60 per 10,000, the total cost of care for affected individuals in the United States is nearly $76 billion annually.[105]

Approximately 10% to 20% (range 3%–25%) of children with a well-documented ASD ultimately lose the diagnosis because they no longer fulfill diagnostic criteria and are reported to function within the broad range of normal cognitive, adaptive, and social skills.[106] It is not clear whether this is attributable to interventions, the nature of the original clinical presentation, intrinsic/genetic predisposition to a highly atypical developmental trajectory, or perhaps resolution of an interfering process. Preliminary evidence suggests that features associated with the potential

for this type of "recovery," or resolution of core ASD symptoms, include higher IQ and receptive language scores, better verbal and motor imitation, diagnosis of PDD-NOS rather than autistic disorder, earlier age at diagnosis, and earlier initiation of treatment.[106] Although it is difficult to draw firm conclusions from the available literature, there is evidence that individuals who no longer meet criteria for an ASD often continue to have other problems, including higher-order pragmatic language impairment, ADHD, anxiety disorders, depression, and tics.[106]

Most studies, including those focusing on high-functioning individuals, have found that a minority of individuals with ASDs achieve good adult outcomes, such as independent living in the community, competitive paid employment, and close social relationships, but there is evidence that outcomes are improving over time.[107,108] The intrinsic factors most consistently associated with prognosis are early language abilities and IQ. Useful speech by age 5 to 6 years, verbal IQ over 70, and nonverbal IQ over 70 are associated with better prognosis, but these factors alone are not enough to ensure a positive outcome.[107,108] Some evidence suggests that the degree of educational, vocational, behavioral, and social support provided may be as important as intellectual ability in determining the adequacy of function in adult life.[107]

Conclusion

Autism spectrum disorders are behaviorally defined conditions characterized by distinctive impairments in reciprocal social interaction and communication that do not simply reflect associated intellectual disability, and by the presence of a restricted, repetitive behavioral repertoire. Primary pediatric health care professionals play an important role in identification and management of children with these common neurodevelopmental disorders. Although no clinically useful biological markers have been identified, substantial advances have been made in characterizing the genetics, neurobiology, and neuropsychology of this heterogeneous group of disorders. There is a growing body of evidence demonstrating that certain educational and medical interventions and community supports ameliorate symptoms and improve functioning in children with ASDs. However, there is much research to be done to determine which interventions are most effective for children with ASDs, what variables moderate treatment effects and predict outcomes, and how much improvement can reasonably be expected. By providing their patients affected by ASDs with high-quality longitudinal medical care and guiding them to effective educational interventions, primary pediatric health care professionals can help to maximize important outcomes including functional independence and quality of life.

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Chapter 14

Psychoeducational Testing

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Interpreting Psychoeducational Testing

Approximately 20% of school-aged children have academic performance problems, the 2 major causes being learning disabilities (LDs) and intellectual or cognitive-adaptive disabilities (mental retardation). School failure results from a complex interaction of child, family, and school-related influences. The prevalence of LDs is 5% to 10%, while 2% to 3% have intellectual disabilities.[1] Other children underachieve academically for a variety of reasons. As with other developmental disorders, early identification of problems and implementation of interventions increases the likelihood of a successful outcome. However, learning difficulties can be subtle, may emerge in later grades, and/or may initially present as physical symptoms or emotional disturbance. Psychoeducational testing is a major component in identifying the cause of poor academic performance.

Primary pediatric health care professionals are often requested to review results of school testing and provide a plan of action for a child with academic difficulties. Unfortunately, they often lack the expertise to accurately interpret such findings. Moreover, they are at a loss as to what they can do to help a child with an LD. To better inform the clinician, this chapter will include a brief overview of issues regarding students with academic problems, a discussion of metrics encountered in psychoeducational testing, short descriptions of specific tests, and additional information to assist in evaluating reports or testing data provided by parents.

Federal Laws

Individuals with Disabilities Education Act

Federal mandates regarding special education services were launched under the Education for All Handicapped Children Act (PL 94-142; 1975), and then subsequently the Individuals with Disabilities Education Act (IDEA), which was reauthorized in 1997 and 2004 (http://idea.ed.gov). Under IDEA, approximately 2.9 million children receive special education services under a specific learning disability (SLD)

designation; this represents 5.5% of the school-aged population and one-half of all children receiving special education services.[1] Almost two-thirds of school-aged students with LDs are males, although this may reflect a referral bias.

For more than 3 decades, the routine procedure for categorizing a child as learning disabled involved documenting a discrepancy between a child's intelligence (aptitude) and his or her level of achievement. With this "legalistic" approach *(learning disability* is a term used to establish eligibility for services, versus an actual diagnostic entity), identification of LDs typically occurred from third to fifth grade, with children essentially having to "wait to fail" before they could receive services.[2] Moreover, testing children with the most severe LDs or those with other problems (eg, attention-deficit/hyperactivity disorder [ADHD]) often did not yield such discrepancies. Other problems with the aptitude/achievement discrepancy approach include distinct separation of special education from general education; eligibility determination procedures having a weak relationship to instructional interventions and causing delays in intervention; little emphasis on prevention and early identification of less complex, more easily resolved academic problems; Individualized Education Programs (IEPs) not implementing scientifically based instructions/interventions; and overrepresentation of minority students in special education (www.NASDSE.org).[3] Moreover, low-achieving students without aptitude-achievement discrepancies and those with a discrepancy suggesting an SLD have more similar characteristics than differences with regard to performance in areas such as reading difficulties; meta-analytic studies have indicated that IQ-discrepant and IQ-consistent groups do not necessarily differ with regard to effect sizes in academic achievement or cognitive abilities, response to intervention, neuroimaging, or prognosis.[4] There are also inconsistencies across states with respect to SLDs, with prevalence rates ranging from 2.8% in Kentucky to 7.4% in Rhode Island.[1] Differences exist across districts within states as well.

Under IDEA, a child with academic difficulty may be entitled to receive services under an SLD eligibility and should be reevaluated every 3 years. Problem areas qualifying for such interventions include oral expression, listening comprehension, written expression, basic reading skills, reading fluency skills, reading comprehension, mathematics calculation, or mathematics problem-solving. Attention-deficit/hyperactivity disorder was placed under the other health impaired (OHI) eligibility criteria in 1991, and since that time there has been a 280% increase in the number of students receiving services under this designation. However, the 2004 IDEA reauthorization (PL 108-446) no longer requires the IQ-achievement severe discrepancy as the sole criterion for SLD eligibility, and instead underscores response to intervention (RTI) for identification of LDs.[2] In fact, districts *must* adopt RTI, but *may* adopt discrepancy criteria.

Response to Intervention

Response to intervention is a process, not a single model. It allows for earlier identification, a stronger focus on prevention, and evaluation that is translatable into academic programming.[5] The stated purpose is to enhance education outcomes for all children by closer integration of general and special education. The major premise is that a student is identified as being learning disabled when response to a validated intervention is found to be inferior to that of peers. The inference is that a child who responds poorly to an effective intervention would have a disability that requires specialized treatment to produce successful learning outcomes. A major advantage is that the RTI procedure can help educators differentiate low achievement that is due to poor instruction versus that attributable to a true disability.[2] A potential disadvantage is that despite a "treat then test" approach, there may be a significant time lag before LD intervention services are offered, and the "treatment" may not fit the problem.

Most RTI models incorporate a 3- or 4-tier instruction/prevention system. Tier 1 is general education or primary prevention (routine educational programs). Generally, measures of academic skills that are sensitive to academic growth are administered to all students: (1) curriculum-based measures (CBMs), (2) curriculum-based evaluation (CBE), or (3) systematic behavioral observation. A CBM such as the Dynamic Indicators of Basic Early Literacy Skills (DIBELS)[6] (assessing literacy skills at the primary level) provides brief samples of academic performance (probes) that can be administered frequently in such areas as nonsense word fluency ("sim," "lum," "vit") and oral reading fluency (read passage). Growth-sensitive behaviors that are measured more frequently allow alteration of instruction or a heightened level of goals.[3] Administration of such instruments several times per year (eg, September, January, May) allows evaluation of the adequacy of the basic curriculum and identification of students who need further intervention.

If a child is unsuccessful academically or is deemed "at risk," then he would enter the LD identification process that begins with Tier 2 (secondary prevention). An instructional support team or a child study team should refer a child to Tier 2 because the child is not doing as well as most peers and needs extra help. This tier typically involves small-group tutoring and entails school-based problem-solving teams for functional assessments and managing interventions. Lack of improvement with this level of intervention is considered "unexpected failure" and establishes eligibility for tertiary intervention (which begins with a more detailed multidisciplinary evaluation).

The Tier 3 intervention level involves more intensive, individual programming with progress monitoring. Special education is not considered a place, but rather a service for children with academic difficulties. With appropriate progress, the student can reenter secondary or even primary prevention tracks.

Response to intervention systems may vary by school system. Some may combine Tiers 1 and 2; others might have several tiers exist between general education and special education services. There are differences in how children are identified: (1) a one-time universal screening with a norm referenced cut-point (eg, <25th percentile on a selected test instrument) or (2) a lack of change in performance after a specific time in a Tier 1 educational program (progress monitoring).[2] Deficits can be at the *level difference* (performance level is discrepant from peers) or *rate of learning difference* (slope of improvement or change over time is deficient). The type of intervention, classification of the student's RTI, and components of a multidisciplinary evaluation are other factors that may vary. Response to intervention services should be received more quickly than those based on the more traditional aptitude/achievement discrepancy, and may actually reduce the number of students in LD programs, particularly in grades kindergarten through third grade.

A criticism of RTI is the assumption (by default) that a child who does not respond to Tiers 1 and 2 has an SLD. Lack of response could be due to the wrong intervention, teacher, criterion measurement, or goal.[7] In fact, the US Department of Education[1] subsequently tempered the position that lack of response to early tiers in RTI is indicative of an SLD, stating that no single procedure (ie, lack or response to RTI) should be the sole criterion for an SLD designation. A true SLD is a brain-based learning deficit and not a delay. Clinicians should also realize the rate of adoption and stage of development of RTI varies by state (see www.RTInetwork.org).

Section 504

The Rehabilitation Act of 1973 (Section 504; http://www.ed.gov/about/offices/list/pcr/504faq/html) is a civil rights act (not education) prohibiting schools that receive federal funds from discriminating against children with disabilities. Here, a handicap is a physical or mental impairment that substantially limits one or more major life activities (MLAs), such as walking, seeing, hearing, or speaking. This act provides broader coverage, typically in the form of special accommodations that are tailored to the needs of the child. Basically the child must experience limitations to an MLA; learning also is an MLA. Under Section 504, fewer regulations are placed on testing, and there is no specified frequency of reassessment or defined role of outside evaluations. A written educational plan (Individual Accommodation Plan) is produced, and the child receives services in a regular classroom. Students receiving Section 504 accommodations typically have less severe problems than those found eligible under IDEA; accommodations for children with ADHD are often established under Section 504. Examples of accommodations include extra time to take tests, allowing verbal versus written responses, adjusting reading level, recording lectures, submitting homework done on computer, peer tutoring, multiple-choice versus short essay questions, and shorter homework assignments. The Americans with Disabilities Act (ADA)[8] is also a civil rights law that prohibits discrimination in employment, services, or programs due to a disability. This act provides similar protections to

students enrolled in private schools and is often applied in colleges. The 2008 amendments broadened the definition of what substantially limits MLA to include reading, thinking, and concentrating. An overview of IDEA, Section 504, and ADA can be found in ERIC Digest E606 (www.ERIC.ed.gov—search for ERIC #ED452627).

Psychoeducational Testing Terminology

The practitioner will encounter certain terms when reviewing data from group tests, individual psychoeducational testing, or less formal assessments. A primer of these terms follows (also see Aylward and Stancin[9]).

Standardized, Norm-Referenced Assessments

Standardized, norm-referenced assessments (SNRAs) are the tests most typically administered to children and adolescents who are experiencing academic difficulties. The SNRAs compare an individual child's performance to the performance of children in a reference group. This comparison is typically made on some standard metric or scale (eg, scaled score; standard score—defined subsequently). To allow for comparison of scores, tasks are presented in the same manner across different testings. However, if this format is modified, additional variability is added, precluding accurate comparison of the child's data and those of the normative group. The SNRAs answer the question: How does this student compare to his or her referent group? They do not answer the question regarding what the limits of the child's abilities are, regardless of comparison to a reference group. An alternative is *criterion-referenced* or *curriculum-based assessment,* which involves mastery of skills in specific areas or objectives to be achieved.[9] These assessments may be more applicable to RTI.

Descriptive Statistics

The *mean* (M) is a measure of central tendency and is the average score in a distribution of test scores. Because it can be affected by variations due to extreme scores, the mean can be misleading in a highly variable sample. The *mode* is also a measure of central tendency, being the most frequent or common score in a distribution. The *median* is defined as the middle score that divides a distribution in half when all the scores have been arranged in order of increasing magnitude. It is the point above and below which 50% of the scores fall. This measure is not affected by extreme scores and therefore is useful in a highly variable sample. In the case of an even number of data points in a distribution, the median is considered to be halfway between 2 middle scores.

Range is a measure of dispersion that reflects the difference between the lowest and highest scores in a distribution (highest score minus the lowest score + 1). However, the range does not provide information about data found between 2 extreme values in the test distribution, and it can be misleading when dealing with skewed data.

In this situation, the *interquartile range* may be more useful. Here the distribution of scores is divided into 4 equal parts, and the difference between the score that marks the 75th percentile (third quartile) and the score that marks the 25th percentile (first quartile) is the interquartile range.

The *standard deviation* (SD) is a measure of variability that indicates the extent to which scores deviate from the mean. The SD is the average of individual deviations from the mean in a specified distribution of test scores. The greater the SD, the more variability is found in test scores. In a normal distribution, 68% of the children taking a test will fall between +/- 1 SD. This is often defined as the normal range. This concept is critical in the development of test norms. In general, most psychoeducational tests that employ deviation quotients have a mean of 100 and an SD of 15. If a child falls 2 SDs or more below average on an achievement test (ie, standard score <70), the likelihood of an LD is increased.[9]

Skewness refers to test scores that are not normally distributed. If, for example an achievement test is administered to a population of children from a poor, inner-city school, the likelihood of more children scoring below average is increased. This is a *positively skewed* distribution (the tail of the distribution approaches high or positive scores). Probabilities based on a normal distribution will underestimate the scores at the lower end and overestimate the scores at the higher end. Conversely, if the test is administered to students from a more elite, college-preparation school, the distribution might be *negatively skewed,* meaning that most children did well (the tail of the distribution trails toward lower scores.) In this case, scores at the lower end will be overestimated, and those at the upper end underestimated. Skewness has significant ramifications in interpretation of test scores. This is why when reviewing group-administered testing, both national and local norm comparisons should be evaluated.

Transformations of Raw Scores

Linear Transformations

Linear transformations provide information regarding a child's standing compared with group means. Standard scores are raw scores that have been transformed to have a given mean and SD. Standard scores specify how many standard units above or below the normative mean a specific child's score falls (Box 14.1). The standard score provides a very precise method of pinpointing a child's performance.

The *z-score* is a standard score (standardization being the process of converting each raw score in a distribution into a z-score; raw score minus the mean of the distribution, divided by the SD of the distribution) that corresponds to an SD (ie, a z-score of +1 is 1 SD above average and a z-score of -1 is 1 SD below average). The mean equals a z-score of 0; therefore, scores between z-scores of -1 and +1 are in the average range. Stated differently, if a child receives a z-score of +1, she obtained a score higher than 84% of the population.

Box 14.1. Terminology

Standard Scores: Raw scores are transformed to standard scores with a specific mean (M) and standard deviation (SD). The M usually = 100, and SD = 15.

Z-Score: Standard score that corresponds to an SD. A Z-score of 0 = the mean; +1 is 1 SD above the mean, while -1 is 1 SD below the mean.

T-score: Standard score that is a Z-score x 10 +50. The mean T-score is 50, and the SD = 10. Average range is 40–60.

Percentile: Area transformation indicating the point at or below which the scores of a certain percentage of children fall. One SD below average is the 16th percentile; 1 SD above average is the 84th percentile. If a child falls at the 40th percentile, he/she is doing as well as or better than 40% of the population taking that test.

Quartile: Area transformation that divides the distribution into 4 equal parts, each being 25 percentile ranks in width and containing 25% of the normative group.

Stanine: Area transformation that divides the distribution into 9 parts. The M = 5 and the SD = 2. The average range is 3–7 (78% of scores fall in this range).

The *T-score* is another linear transformation, and can be considered a z-score times 10, plus 50. Therefore, the mean T-score is 50, and the SD equals 10. A z-score of 1 equals a T-score of 60. T-scores are often found in test instruments such as the Conners Rating Scales, or the Child Behavior Checklist (CBCL), where T-scores of 70 (ie, 2 SDs above average) or greater are considered to be clinically relevant (approximately the 98th percentile); these cutoffs are depicted in many scoring forms. Many clinicians consider a T-score between 60 and 70 (1–2 SDs above average) to be clinically relevant as well.

Area transformations

A *percentile* tells the practitioner where an individual child is in relation to a specified norm group. With a percentile score of 50, half of the children will score above this score, while half will score below. A score that is 1 SD below average is at approximately the 16th percentile; a score 1 SD above average is at the 84th percentile. Small differences in scores in the center of the distribution will produce substantial differences in percentile ranks, while greater raw score differences in outliers will not have as much of an impact on percentile scores. Frequently, the third percentile is considered to be a clinical cutoff. *Quartiles* are percentile bands that are 25 percentile ranks in width; each quartile contains 25% of the normative group. Percentiles require the fewest assumptions for accurate interpretation, and can be applied to virtually any shape distribution. This metric is most readily understood by parents and professionals and is recommended as the preferred way to describe how a child's score compares with a group of scores. For example, a Wechsler Individual Achievement Test-II (WIAT-II) reading comprehension standard score of 70 (2 SDs below average) indicates that fewer than 3% of children of a similar age score lower in this area; conversely, 97% or more of children taking the test will have a higher score.

The *stanine,* often encountered in group test output, is short for "standard nine"; this metric divides a distribution into 9 parts. The mean equals 5 and the SD is 2, with the third to seventh stanine being considered the average range. Approximately 20% of children will fall in the fifth stanine, 17% each in the fourth and sixth, and 12% each in the third and seventh stanines (78% in total). Stanines are frequently encountered with group-administered tests, such as the Iowa Tests of Basic Skills (ITBS), the Metropolitan Achievement Tests, or the Stanford Achievement Tests (SAT).

Age- and *grade-equivalent* scores are based on raw scores and portray the average age or grade placement of children who obtained a particular raw score. While these metrics are useful in explaining results to parents and make conceptual sense, age- and grade-equivalent scores are uneven units of measurement. These measures lack precision and, in some test manuals, the same standard scores can produce somewhat different age/grade equivalents. Both metrics assume that growth is consistent throughout the school year and tend to exaggerate small differences in performance. These measures also vary from test to test. Therefore, grade equivalents on one test should not be compared with those obtained from a different instrument.

Grade equivalents are often misinterpreted as a student's actual functional grade level. For example, if a second grader obtains a 4.7 grade equivalent on a word reading subtest, this does not mean her reading is similar to that of a student in the seventh month of the fourth grade and she could therefore successfully read at that level. What this score really means is that the student performs well in comparison to others in second grade who took that particular reading test. If an item gradient of a test is too steep (small changes in raw scores result in large changes in standard scores), grade equivalents become even more variable.

Furthermore, with achievement testing, one needs to know whether age or grade norms were used to obtain standard scores. For example, if age norms are used with a child who had been retained, he would be at a significant disadvantage because he would not have been exposed to the more advanced material. Conversely, if a child failed second grade and was being tested in early fall while repeating second grade, she may receive inflated scores if grade norms are used.

All tests contain measurement error. The *standard error of measurement* (SEM) estimates the error factor in a test that is the result of sampling or test characteristics, taking into account the mean, SD, and size of the sample. The larger the SEM, the larger the uncertainty associated with a given child's score. In 95% of cases, the interval of approximately 2 times (1.96) the SEM above or below a child's score would contain the "true" score—a 95% confidence interval (CI). Stated differently, a 95% CI indicates that if a test was given 100 times with different samples, scores will fall in this interval 95% of the time. The higher the degree of certainty or probability that an interval contains the child's true score, the wider the interval. Such estimates are

important in test-retest situations, or in the case of a child not receiving services due to missing a cutoff score by only a few points.

The *normal curve equivalent* (NCE) is often encountered in group-administered testing. The NCE is a type of standard score with a mean of 50 and an SD of 21.06 (range 1–99). Normal curve equivalents cover the same range as percentile ranks, with percentile ranks of 1, 50, and 99 corresponding to similar NCEs. A standard score of 100 equals an NCE of 50, a standard score of 70 equals an NCE of 8, and a standard score of 115 corresponds to an NCE of 71.

Intelligence Testing

Evaluation of cognitive abilities is often necessary in psychoeducational testing, although under the 2004 IDEA, this arm of testing is emphasized less than in previous iterations. IQ testing is necessary to establish an aptitude-achievement discrepancy, although this criterion is not mandated by the federal regulations. IQ testing is a component of determination of intellectual disabilities, which would meet different eligibility criteria for special education services. Some researchers[10] argue that cognitive evaluation should be brief and focused on establishing strengths and weaknesses as well as comorbidities. A listing of selected cognitive tests follows.

Kaufman Brief Intelligence Test, Second Edition (KBIT-2)[11]

The KBIT-2 is applicable for ages 4 years and older and provides a relatively quick estimate of IQ for screening purposes. The test produces a verbal, nonverbal, and composite IQ score (M=100, SD=15). The verbal scale consists of 2 subtests: Verbal Knowledge (measures receptive vocabulary and range of general information) and Riddles (measures verbal comprehension, reasoning, vocabulary knowledge, and deductive reasoning). *Matrices* is the nonverbal scale (uses meaningful [people, objects] and abstract [designs, symbols] stimuli). The KBIT-2 composite score is comparable to the Wechsler Intelligence Scale for Children Fourth Edition (WISC-IV) full-scale IQ.

Stanford Binet for Early Childhood-5 (Early SB5)/Stanford Binet Fifth Edition (SB5)[12,13]

The Stanford Binet for Early Childhood, fifth edition is applicable from age 2 to 7.25 years (the full SB5 extends to adulthood). Ten subtests comprise the full-scale IQ (FSIQ), and various combinations of these subtests comprise other scales. An Abbreviated Battery IQ (ABIQ) scale consists of 2 routing subtests: Object Series/Matrices and Vocabulary. The nonverbal IQ (NVIQ) consists of 5 subtests measuring nonverbal fluid reasoning, knowledge, quantitative reasoning, visual-spatial processing, and working memory. The verbal IQ (VIQ) contains 5 subtests that measure verbal ability domains in the same 5 factor areas. The SB5 Early Childhood also includes a Test Observation Checklist. An NVIQ, VIQ, and FSIQ are obtained (M=100, SD=15), as well as total factor index scores (sum of verbal and nonverbal scaled scores) for fluid

reasoning, knowledge, quantitative reasoning, visual-spatial processing, and working memory; scaled scores (M=10, SD=3) can be computed for each of the nonverbal and verbal domains.

Differential Ability Scales/Differential Ability Scales-II (DAS)[14,15]

The DAS is applicable from 2.5 to17 years of age but is frequently used in the early childhood/preschool range. The DAS is often considered to be an intelligence test, although it documents a range of abilities, not an IQ score per se. The composite score is based on reasoning and conceptual abilities and is termed the General Conceptual Ability (GCA) score (M=100, SD=15, range 45–165). There are 2 batteries: 2 years, 6 months to 6 years, 11 months, and older than 6 years, 11 months. The lower age range is subdivided into 2 groupings: 2 years, 6 months to 3 years, 5 months, and 3 years, 6 months to 6 years, 11 months. The DAS has 2 purposes: provision of the GCA as well as a diagnostic profile. Subtest ability scores have a mean of 50 and SD of 10 (T-scores). In addition, verbal ability and nonverbal ability cluster scores are produced for upper preschool-aged children (3.5 years onward). The DAS-II measures general intelligence with 11 core and 10 diagnostic subtests (the former contribute to the GCA). Diagnostic tests measure short-term memory, perceptual abilities, and processing speed. The special nonverbal composite is helpful in testing children who have communication problems.

Kaufman Assessment Battery for Children Second Edition (KABC-II)[16]

The KABC-II is applicable from ages 3 to 18 years of age and contains 18 core and supplementary subtests (the number of tests administered varies, depending on age). There is a simultaneous and sequential processing approach, along the lines of the Luria neuropsychological model. However, the test also uses the Cattell-Horn-Carroll (CHC) abilities model that includes fluid crystallized intelligence. As a result, interpretation and the number of scales produced are based on the model that is selected. The 5 areas assessed include (1) simultaneous processing, (2) sequential processing, (3) planning, (4) learning, and (5) knowledge.

At age 3 years, a Mental Processing Index (MPI) (Luria model) and a Fluid-Crystallized Index (CHC model) are derived. By age 7, the full array can be obtained, which includes the MPI, a Global Score, Fluid-Crystallized Index, and a Nonverbal Index. Subtest scale scores have a mean of 10 and SD of 3; the index score mean equals 100, SD equals 15.

Wechsler Preschool and Primary Scale of Intelligence Third Edition (WPPSI-III)[17]

The WWPSI-III contains 14 subtests with 2 age bands: 2 years, 6 months to 3 years, 11 months, and 4 years to 7 years, 3 months. In the first, an FSIQ, a VIQ, and performance IQ (PIQ) are obtained using 4 core subtests. Seven core subtests are applicable to the 4 years to 7 years, 3 months age range. Supplemental and optional subtests are

used to obtain a general language composite in younger children, and a processing speed quotient from age 4 years upward. The mean is 100 and the SD is 15 for IQs and composite scores; the mean is 10 and the SD is 3 for scaled scores.

Wechsler Intelligence Scale for Children Fourth Edition (WISC-IV)[18]

The WISC-IV is appropriate for children ages 6 years to 16 years, 11 months. It contains 15 subtests (10 core, 5 supplementary). The WISC-IV contains a Verbal Comprehension Index, a Perceptual Reasoning Index, a Working Memory Index, and a Processing Speed Index. Several subtests are optional. A measure of general intellectual function (FSIQ) is produced. The mean for index and FSIQ scores is 100, the SD is 15; the mean scaled score is 10, the SD is 3. A General Ability Index (containing 3 verbal comprehension and 3 perceptual reasoning subtests) can be computed; this is not influenced by working memory and processing speed and is more useful with children who have LDs or ADHD.

Wechsler Abbreviated Scale of Intelligence (WASI)[19]

The WASI is applicable from age 6 upward. A VIQ, a PIQ, and an FSIQ (FSIQ-4 [using 4 subtests] or an FSIQ-2 [using 2 subtests]) are obtained. Subtests are similar to those found in other Wechsler scales, but the actual items differ. Subtests include Vocabulary, Matrices, Block Design, and Similarities (Vocabulary and Matrices are used to compute the Full-2 IQ). T-scores are used for subtests (M=10, SD=5). The WASI is useful as a screening instrument. Scores are generally 3 points higher than the WISC-IV.

Achievement Tests

Different types of achievement tests are employed for various reasons. The first use is RTI, which employs CBMs, CBEs, or curriculum-based assessments.[2,3] These terms are often used interchangeably, although there are subtle differences among these approaches. These tests generally are short and are easy to administer. A second major area of use involves group-based, survey achievement batteries (allowing comparison of a student's achievement to local and national norms). The third category involves individually administered, more detailed evaluation of academic achievement, this often being used to determine the presence of low achievement and possibly LDs. Several tests from each grouping will be described below.

Curriculum-based Measures

The DIBELS[6] is a kindergarten through sixth grade screening of 3 major components of early literacy: (1) phonological awareness (initial sound fluency [identify/produce initial sounds of words] and segmentation fluency [produce individual sounds within a given word]), (2) alphabetic principle (nonsense word fluency [eg, bim, fik, etc.]), and (3) fluency with connected text (oral reading fluency of sentences). The DIBELS provides benchmark assessment scores that are indicators of the child's

skill development and tracks progress over the academic year (testing in September, January, and May). For example, with phonemic sequencing fluency, a child should decipher 35 or more phonemes per minute by the end of kindergarten, read 40 or more words per minute correctly by spring of first grade (oral reading fluency), and 90 or more words per minute by spring of second grade.[6] The DIBELS is Web-based. Probes typically take only several minutes for administration.

AIMSweb[20] is another achievement protocol that allows for benchmarking and progress monitoring. AIMSweb is applicable for grades kindergarten through eighth and enables measurement of basic skills in reading, early literacy, early numeracy, mathematics, spelling, and written expression in a paper and pencil administration. Reading involves 30 passages at each grade (from first grade onward), and the number of words a child reads in 1 minute is calculated. Reading comprehension (Maze) assesses reading proficiency by having a child select a missing word from 3 options at every seventh word in the passage that is read silently. The Test of Early Literacy (K–1) measures phonemic awareness (letters, names, sounds, and nonsense words) and phonemic segmentation (nonsense word fluency). Early numeracy (K–1) contains oral counting, missing numbers in a number line, number identification, and quantity discrimination. Mathematics assesses the child's proficiency in computational problems within a short time span, while spelling requires the child to write down dictated words over a 2-minute time span.

Survey Achievement Batteries

Survey achievement batteries are group-administered tests given to screen or obtain a general estimate of academic achievement across a wide range of skill levels. These tests are norm-referenced, and therefore pinpoint a given student's ranking or standing within a group. Some of the most frequently used tests are the Iowa Tests of Basic Skills (ITBS; K–8th grade),[21] Metropolitan Achievement Tests (MAT; K–12.9 grade),[22] SRA Achievement Series (K–12.9 grade),[23] Stanford Achievement Tests (SAT; 1.5–9.9 grades),[24] and California Achievement Tests/5 (CAT/5; K–12th grade).[25]

Although slightly variable with respect to content, most assess reading (vocabulary, word analysis, reading comprehension, spelling), mathematics (concepts, problems, computation), and language (capitalization, punctuation, usage, and expression). Composite basic (core) and complete battery scores are provided (eg, SAT, ITBS), while verbal and nonverbal totals are found in some tests (eg, SRA). Different content areas may be tested at different grade levels. Additional subtests that often are included in the batteries are reference materials, social studies, science, research skills, applied skills, and environment. National as well as local norms are often provided, and both should be reviewed.

Individually Administered Achievement Tests

These tests are often administered by psychologists as part of an LD evaluation. However, the tests often vary in terms of the presentation of information and responses that are elicited. As a result, scores for the same child may vary, depending on the test used. A general rule of thumb is that closed-end questions (fill in the blank) or ones in which the student can select from several alternatives will yield higher scores than tests using more of an open format (eg, read a passage and then answer questions). Similarly, some require pencil and paper responses, while others do not; thus, this requirement again influences scores. A comparison of several achievement tests can be found in a recent paper by Fletcher et al.[10]

Kaufman Test of Educational Achievement-II (KTEA-II)[26]

The KTEA-II is available in 2 formats: the Comprehensive (with parallel forms A and B) and Brief Form (M score = 100, SD =15). Noteworthy is the fact that the KTEA-II was co-normed with the KABC-II (a measure of aptitude). The Comprehensive Form, applicable from ages 4 years, 6 months to 25 years, assesses reading (letter/word recognition, comprehension), math (computation, concepts, and application), written language (spelling, written expression), and oral language (listening comprehension and oral expression). Several reading-related skill areas are also assessed (eg, phonological awareness). The Brief Form (for ages 4 years, 6 months to 90 years) measures reading word recognition and comprehension, math computation and application problems, and written expression (written language and spelling); a battery composite score is produced as well. Age and grade equivalents are provided. The test differs significantly from the original KTEA or the KTEA-NU (normative data update).

Peabody Individual Achievement Test-R/NU (PIAT-R)[27]

The PIAT-R/NU (normative update) is applicable for grades kindergarten through 12 (ages 5–19). This test differs from others in that spelling and math are presented in a multiple-choice format; in reading comprehension, the student selects a picture that best illustrates the sentence that was read. The test includes scores for general information, reading recognition, reading comprehension, total reading, math, spelling, written expression, written language, and the total test. The NU version is the same earlier version of the PIAT, with updated norms. Some argue that the multiple choice format may yield higher test scores because of the recognition versus recall format. In fact, paper and pencil are used only during the written expression section.

Wechsler Individual Achievement Test-II (WIAT-II).[28]

The WIAT-II is applicable for grades prekindergarten through 16. This is an updated form of the earlier WIAT. There are 4 composite scores: (1) reading (word reading, pseudo-word decoding, reading comprehension), (2) mathematics (numerical operations, math reasoning), (3) written language (spelling, written expression),

and (4) oral language (listening comprehension, oral expression). Standard scores (M=100, SD=15), age or grade equivalents, and quartile scores are reported. Reading rate can also be assessed, and the test form includes qualitative observations for various subtests. The test is linked to Wechsler IQ tests, and aptitude/achievement discrepancy tables are included. A newer version, the WIAT-III, has just been developed.

Wide Range Achievement Test-4 (WRAT-4)[29]

This is the eighth edition of the test and is applicable from ages 5 upward. There are 2 equivalent forms (Blue, Green) and each contains reading (read letters, pronounce words), spelling (write letters, words from dictation), and arithmetic (15 oral math and 40 computation problems). In addition, this newest version contains sentence comprehension (student fills in missing word in a sentence). The reading composite consists of the combined word reading and sentence completion standard scores. The WRAT-4 is normed by age and grade (fall, spring). The WRAT-4 is best considered as providing a quick screen of fundamental academic skills and standard scores, confidence intervals, stanines, and grade equivalents. An older WRAT-Expanded Version is also available[30] that contains a Group form (G) with reading/reading comprehension, math, and nonverbal reasoning, (some tests are multiple-choice) and an Individual form (I) that assesses reading, mathematics, listening comprehension, oral expression, and written language. The Expanded Version Group form is applicable to grades 2 through 12, the individual form, ages 5–24 years.

Woodcock-Johnson III Tests of Achievement (WJ III)[31]

The WJ III, applicable from grades kindergarten through 18, has 2 parallel forms (A and B) that are divided into a Standard Battery (12 subtests) and an Extended Battery (10 tests), yielding a total of 22 subtests. The latter provides the opportunity for more in-depth diagnostic evaluation of specific academic functions (eg, word attack, oral comprehension).

The WJ III contains (1) a reading cluster (letter/word identification, reading fluency [3 minutes, answer yes/no to a sentence, eg, the sky is green], passage comprehension [supply key missing word in passage]), (2) an oral language cluster (story recall, understanding directions), (3) a math cluster (calculation, math fluency [3 minutes], applied problems), (4) a written language cluster (spelling, writing fluency, writing samples), and (5) an academic knowledge cluster (content areas of science, social studies, humanities). Clusters are designed to correspond with IDEA areas. The Standard Battery provides 10 cluster scores, while the Extended Battery provides an additional 9 cluster scores. Broad reading, broad math, and broad written language are often used to provide an overview of the child's achievement. The WJ III is often used by school systems. Of note is the fact that the WJ III Tests of Achievement were co-normed with the WJ III Tests of Cognitive Abilities (another measure of aptitude) and are designed to be used in combination. Standard scores (M=100,

SD=15), percentile scores, and age and grade equivalents are the most helpful metrics. Computer scoring is necessary. There is also the Relative Proficiency Index (RPI) with a denominator of 90 and a numerator of 0 to 100. For example, an RPI of 45/90 indicates that the examinee is half as proficient on a task as are average age or grade peers. When others are predicted to show a 90% success, the examinee is predicted to show only 45% success.

Other Considerations

Purposes for testing a child who is having problems in school performance can vary. School systems must clarify whether a child meets the specific criteria for determination of an LD—this is defined by educational criteria. Clinicians often are more concerned with *why* the child is having a particular problem. This causes some professionals to differentiate a *learning disorder* (persistent deficit in terms of brain function) from a *learning disability* (determined based on meeting specific aptitude/achievement discrepancies). A child who has been tested and found to not qualify for special resource services still may have a learning disorder. Hence, ineligibility does not equal absence of a problem in school performance. In fact, school testing would not have been undertaken to begin with had the child *not* been experiencing school problems.

Primary pediatric health care professionals should also be sensitive to the scenario in which the child had previously received support services, but then the services were terminated because the child's performance was up to par. Unfortunately, without continued supplementary services, many children will show a decline in levels of achievement.

Qualitative Information

It is important to determine the child's area(s) of difficulty. These can include *academic subjects, functions* (memory, organization, visual motor integration), or *performance* (inconsistency, gives up quickly, does not complete work).[32] Unfortunately, many of these issues are overlooked when the aptitude/achievement discrepancy approach is employed or psychoeducational scores are reported. Qualitative observations or information gathered from parent, teacher, and child report are helpful additions to psychoeducational testing data.

Reading

Reading is the most frequent learning disorder; however, there are several possible reasons for a reading problem.[32] In fact, 80% of children with LDs have an LD in reading. In general, phonics is an early problem, while reading comprehension is apparent later. Dyslexia, a word level reading disorder, is considered to be a deficit in the phonological component of language. Basic reading problems can be *phonological/dysphonetic* (problems in decoding or word analysis; basic phonemic awareness), *orthographic/dyseidetic* (problem with the visual gestalt/memory for

whole words), or *combined phonological/orthographic.* However, the latter, sometimes called a surface reading problem, is controversial, with many experts indicating that this is a developmental lag, and is typically found only in younger children. Problems in reading decoding could be caused by the deficits listed above, as well as processing speed or poor instruction. Deficits in *reading comprehension* or in *reading rate (fluency)* are additional concerns. Reading comprehension could be related to poor verbal working memory, inattentiveness, poor vocabulary/expressive language, processing speed, deficient reasoning ability, or deficits in basic reading skills. Reading rate (fluency) is particularly receiving much emphasis and could be negatively affected by decoding or comprehension problems; it sometimes is a later, residual finding in children and adolescents with an earlier diagnosis of dyslexia.[33]

Children with a dysphonetic problem cannot sound out words, produce *semantic substitutions* ("laugh" is read as "funny"; "city" as "town") and/or *gestalt substitutions* ("house is read as "horse"), and display syllable reversals ("mawn lower" for "lawn mower"). Those with an orthographic (surface) reading problem have a deficit in visual memory, read slowly (as if reading even familiar words for the first time), have limited sight vocabularies, and can often read phonetic words up to their grade level (thereby not being detected on some achievement tests).[32] Dysphonetic problems are more prevalent than orthographic ones, and there continues to be debate as to whether the orthographic subtype is simply a delay in word recognition skills that is restricted to younger readers.[33]

When considering a possible reading disorder, the practitioner is advised to also evaluate the child's spelling. With a phonological reading disorder, misspellings are typically phonetically inaccurate (eg, "apple" spelled as "aplpe," "house" as "horse"); with an orthographic problem, misspellings are phonetically accurate (eg, "crkle" for circle), but children may not recognize simple words or may be able to read them in one sentence and not in the next. They may reverse letters (static reversals) or letter sequences in words (visuospatial reversals; eg, "keep" as "peek," "step" as "pets"). Some children with an orthographic problem can spell a phonetic word correctly, although they are unable to read it. Noteworthy is the fact that dyslexia is not diagnosed because of letter reversals or mirror writing problems. Similarly, eye exercises, behavioral vision therapy, or tinted filters or lenses are not recommended as evidence-based treatments.[34]

Mathematics

Mathematic difficulties can arise from several different causes: (1) *spatial organization* problems cause difficulty working with columns (eg, 29 minus 17 = 82 [incorrectly subtracting 1 from 9]), (2) problems with *visual detail* cause the child to mistake operands such as times for a plus sign, while (3) *procedural errors* might cause the child not to understand decimals, division, or fractions. Some children are weak in mathematics because of problems with automatic retrieval of number facts (simple

addition facts, times tables). Deficits in working memory or attention can cause difficulty with computation when numbers have to be borrowed or carried. Language weaknesses can make word problems difficult.

Generally, achievement scores 1 or more SD below average still are suggestive of potential academic problems, even though such scores may not qualify for the designation of an LD. Hence, scores at the 16th percentile or lower (or <3rd stanine) raise suspicion, although children with scores less than the 20th or 25th percentiles (standard scores of ~88 and 90, respectively) often are identified as having problems in academic performance. In such situations, in addition to comparing the child's score to national norms, local or school norms should also be used as a benchmark, particularly if the student is in a competitive academic environment.

With regard to psychoeducational testing, if a child is doing well in all academic areas except for one subject such as mathematics, the likelihood of a specific learning disorder is increased (provided the level of instruction is adequate). Conversely, if there is a flat level of achievement noted across the board, the list of possibilities is different (eg, intellectual disability, ADHD, emotional problems). Thirty percent of children with LDs have at least one other comorbid disorder.[4] Finally, school performance difficulties can be grouped into 4 possible etiologies: intellectual limitations (requiring administration of IQ tests), learning disabilities, ADHD (particularly ADHD-inattentive subtype), emotional/behavioral disorders, or some combination of these.[35] Unfortunately, the comorbidity rate is high,[36] making it difficult to separate out these causative factors without detailed testing.

Conclusion

It is recommended that the first step in pinpointing possible causes for academic problems is to review current and past report cards to identify historical patterns. If group-administered tests (surveys) were administered, review of these data is recommended. Similarly, CBMs and similar instruments could also be reviewed, with particular emphasis on change over time. Any individual achievement testing administered by either special education teachers or psychologists may also be perused. Scores may be compared and examined for patterns. IQ testing, if administered, could also be reviewed. This process could be considered a Level I approach.

Level II would involve consideration of more qualitative information, obtained using a scheme such as that provided by Kelly and Aylward.[32] This information may help to better identify underlying reasons for the low scores on testing. Level III would involve exploration of special education criteria and possible interventions, such as IDEA, Section 504, RTI, and ADA. The degree of involvement would depend on the nature of the primary pediatric health care professional's practice and interests.

While some practitioners may on occasion administer psychoeducational tests such as the WRAT-4 (eg, developmental-behavioral or neurodevelopmental pediatricians), all will have occasion to respond to inquiries from parents regarding their child's test performance or eligibility status derived from testing. The primary pediatric health care professional's role may include explaining test results to parents, acknowledging parental concerns and advocating for the child, providing direction and resources (eg, www.ldanetl.org, www.ncld.org, www.ld.org), or referring to other professionals.

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Chapter 15

Learning Disabilities

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Definition of "Learning Disability" and Scope of the Problem

The terms *learning disability* (LD) and *learning disorder* are often used interchangeably. The *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, Text Revision (DSM-IV-TR)* states that learning disorders are diagnosed "when the individual's achievement on individually administered, standardized tests in reading, mathematics, or written expression is substantially below that expected for age, schooling, and level of intelligence."[1] In other words, the child must demonstrate a significant discrepancy between achievement scores in reading, writing, or math in comparison to measures of cognitive ability (IQ scores). The child must demonstrate an impact of these discrepant measures on academic achievement in the classroom. Furthermore, according to *DSM-IV-TR,* learning disorders should not be diagnosed when delays in academic achievement are primarily attributable to impaired hearing or vision.[1] A child may meet the diagnostic criteria for one or more learning disorders (reading, mathematics, and/or written language).

Discrepancy Versus Low Achievement Definitions of Learning Disability

The *DSM-IV-TR* and previous federal special education laws emphasize the concept of a "significant discrepancy" between measurements of academic achievement and cognitive ability, typically defined as a 1 to 2 standard deviation or more difference in academic achievement and full-scale IQ scores.[1] However, recently there has been a trend toward LD definitions that emphasize low academic achievement among children with at least low average cognitive skills.[2] This is a particularly important concept for children who may, for example, have IQ scores in the low average range (80–90). In order to meet a discrepancy-based definition of LD, such a child would have to have academic achievement standard scores of 50 to 60, representing delays of several grade levels or more, in comparison to peers. In contrast, a child with an IQ of 100 or more would be required to have an achievement score of 70 in order to meet a discrepancy-based LD definition. In both instances, the child would be underachieving to a degree that would significantly impact his or her ability to

succeed in school, yet the child with low average intelligence would be required to have far lower achievement scores in order to be eligible to receive special education assistance, if only discrepancy definitions of LD are employed.

Legal Definition of Learning Disability as per the Individuals with Disabilities Education Act

The recent reauthorization of the Individuals with Disabilities Education Act (IDEA), the federal legislation that governs special education services in our public schools, reflects the trend away from discrepancy-based definitions of LD. In fact, the legislation states that LD criteria "must not require the use of a severe discrepancy for determining whether a child has a specific learning disability…."[3] Further, the law states that the presence of LD may also be defined by a child's failure to improve academic achievement "in response to scientific, research-based intervention" while also continuing to allow other "research-based procedures" to determine LD status.[3] Thus the law continues to allow children to qualify for LD services if they manifest a discrepancy between academic achievement and intellectual ability, but now also allows children who meet "low achievement" definitions of LD to receive special educational assistance. This should help to ensure that more children who are struggling with academic achievement will receive appropriate services. It is also likely that these new regulations will diminish the number of children who fulfill medical diagnostic criteria for LD yet who do not qualify for LD services in public schools. Most importantly, a child is deemed to have a specific LD if he or she "does not achieve adequately" for age or meet grade-level standards, assuming that appropriate instruction has been provided. Specific LDs can be reflected in problems with the following, specific academic tasks: oral expression, listening comprehension, written expression, basic reading skills, reading fluency skills, reading comprehension, mathematics calculation, and mathematics problem-solving.[3] It is important for primary pediatric health care professionals to be familiar with the new terminology in the reauthorization of IDEA, particularly response to intervention (RTI). Special education services must be supported by "scientifically based research…accepted by a peer-reviewed journal or approved by a panel of independent experts through a comparably rigorous, objective, and scientific review."[3] (See Chapter 14 for further information on RTI.) This new stipulation should help to ensure that unproven, non–evidence-based practices are gradually eliminated from special educational programs. The federal law requires that each state enact special education laws that are consistent with the federal law. This leaves the states considerable latitude in special education laws, which is likely to continue the historic tendency for significant state-by-state variations in criteria required to receive special educational services, as well as the nature of the services themselves. Unfortunately, despite the fact that this legislation was passed in 2004, many states have yet to operationalize IDEA by changing relevant state laws.

Epidemiology of Learning Disability

Learning disorders are among the most common neurodevelopmental disorders in childhood, and it is therefore essential for primary pediatric health care professionals to understand the important role that they play in identification, diagnosis, intervention, and advocacy for their patients with LD. The *DSM-IV-TR* includes prevalence estimates of 4% for reading disorder and1% for mathematics disorder, while noting that "disorder of written expression is rare when not associated with other learning disorders."[1] However, recent research has clearly demonstrated that all forms of LD are far more common than the *DSM-IV-TR* suggests. Estimates for the incidence (likelihood of developing LD during childhood) of reading disorder range from 5.3% to 11.8% and for mathematics disorder from 5.9% to 13.8%.[4,5] For each type of LD, the highest incidence rates are obtained with "low achievement" definitions, while discrepancy-based definitions lead to considerably lower estimates. It is also important to recognize that epidemiologic studies demonstrate that boys are 2 to 3 times more likely than girls to manifest any type of LD.[4–6]

Identifying Children With Learning Disability

Children at Risk for Learning Disability

As with other neurodevelopmental disorders, primary pediatric health care professionals play a crucial role in the early identification of children with LDs. This begins with recognition of medical, genetic, and psychosocial conditions that place children at greater risk for development of an LD. They also have a unique opportunity to contribute to early identification of children at risk for LD based on their knowledge of their patient's family and medical and psychosocial history. Learning disabilities are clearly familial, with genetics contributing substantially to a child's risk for LD.[7] The family history should include information about learning and other developmental disorders. Two categories of medical risk factors for LD deserve special attention: prematurity and cyanotic congenital heart disease. Premature infants are at significantly higher risk for not only global developmental delays, but also for LD.[8] In particular, children born at less than 32 weeks' gestation or who experience perinatal and postnatal complications such as prolonged ventilation, intracranial hemorrhage, sepsis, seizures, prolonged acidosis, or hypoglycemia are at higher risk for neurodevelopmental sequelae. Similarly, children now surviving previously fatal congenital cardiac anomalies are another group at high risk for LDs.[9] Children living in poverty are certainly at risk for academic underachievement, while their risk for LD may be less clear. Nevertheless, such children certainly warrant increased vigilance for not only developmental delays but also LD. Several genetic disorders have been linked to risk for various forms of LD. In particular, children with Klinefelter syndrome, Turner syndrome, velocardiofacial syndrome, and spina bifida with shunted hydrocephalus have all been shown to be at significant risk for LD.[10,11] Some studies have suggested that children with Turner syndrome and children with spina

bifida and shunted hydrocephalus may be specifically at risk for problems with visuospatial cognitive skills and math achievement.[10,12] However, given the limitations in the available literature, it is more appropriate to view these children as being at risk for LD in general, rather than for a specific type of learning problem.

Male gender is also a risk factor for LD. While some authors have suggested that boys and girls are at equal risk for certain types of LD, epidemiologic studies have consistently demonstrated that boys are at greater risk for all LDs.[4–6] Girls certainly deserve to be monitored in the context of well-child care, but boys are at significantly higher risk for LD.

When one of these risk factors is identified, the child should be monitored more carefully, with a low threshold for referral for comprehensive assessment, either privately or through public early intervention and school-based programs.

Early Development and Risk for Learning Disability

The importance of careful developmental surveillance and screening cannot be overestimated, not only in order to identify developmental delays that should be addressed in the toddler or preschool-aged child, but also to identify children at risk for later problems with language-based learning (see Chapter 6). In addition to formal developmental surveillance and screening, certain "red flags" suggest that a child may be at increased risk for later reading problems. These include delays in receptive and expressive language and speech articulation in toddlers and young preschoolers. Later, children may have difficulty learning simple songs or rhymes.[13] Unfortunately, less is known about early indicators of risk for math LD.

The Prekindergarten Checkup

Aside from immunizations and hearing and vision testing, primary pediatric health care professionals may question the utility of the prekindergarten preventive care visit. However, this visit provides an ideal opportunity to identify children at risk for LD. Milder delays in language development and speech articulation should be apparent at this age. At the prekindergarten visit, identification of at least 4 random letters that are not in alphabetical order is strongly associated with appropriate phonological processing skills needed for reading decoding. Early indicators of risk for math LD, such as difficulty learning to count or understanding the concept of one-to-one correspondence, may be detected. Children who have difficulty drawing simple shapes (circle at 3 years, square at 4 years, or triangle at 5 years) at the prekindergarten checkup may be at risk for difficulties with writing. In addition, several good developmental screens that include early academic learning skills are available, including the parent-completed Ages and Stages Questionnaire (ASQ) and the directly administered Brigance Screens-II and Parents Evaluation of Developmental Status—Developmental Milestones (PEDS:DM). Children who appear to be at risk for LD may be scheduled for reassessment during or toward the end of the kindergarten year. At that time, if problems are noted with acquisition of

basic number and letter identification or counting, or if teacher concerns are reported, it may be appropriate to refer the child for further assessment (see below).

School Age: The Report Card Visit

While schools are mandated to evaluate children whose academic performance suggests the presence of an LD, children often "fall through the cracks" and are not assessed until their academic achievement lags far behind their same-aged peers. During school-aged well-child visits, primary pediatric health care professionals have an opportunity to assess academic progress and identify children who warrant referral for more comprehensive assessment. This requires that a few, brief questions are asked about progress in reading, math, and writing: Has the first-grade child learned all of the letters and letter sounds, numbers, and beginning addition and subtraction facts? Does the child have poor memory for spelling words or numbers? Has the teacher expressed any concerns about academic progress?

Federal educational policy requires frequent, standardized assessment of student progress. Often the results of these standardized tests are used to gauge the overall performance of a school or school district. However, standardized tests can also help to identify children at risk for LD who require further assessment, but only if parents understand how to interpret the tests. Parents can be instructed to bring their child's standardized test reports to every well-child visit. The professional can then quickly identify children who score low on math, reading, and written language tests and who warrant further assessment. Similarly, parents can be instructed to bring their child's most recent report card to every well-child visit. Again, a quick review of the report card can assist in identifying worrisome academic performance and teacher comments that suggest a possible LD. All school-aged well-child visits should include this brief review of standardized test scores and report cards as a routine practice.

Referring Children for Evaluation by Local School or Private Agency

While it is the responsibility of the school to determine whether or not a child qualifies for special education services, primary pediatric health care professionals can guide parents to formally request an evaluation for their child. Once a formal, written request for evaluation is made, the applicable state special education rules take effect, and the evaluation process begins. Often a brief note from the professional, outlining the concerns that prompted the referral, will be helpful in getting the evaluation process started. If the quality or result of the school-based assessment is not satisfactory, the professional can assist the family by requesting additional assessment at school or making a referral to a qualified psychologist to complete additional testing. This may be especially important for children who have more complex learning problems, or who have not demonstrated adequate progress with school-based services. It should be noted that schools are not required to accept the

findings from private assessments, although some school districts will accept such assessment reports. However private assessment may still help to ensure that a child's needs are appropriately addressed—even if the school chooses to complete their own testing to verify the findings from private assessments.

Comprehensive Assessment for Learning Disability

Primary pediatric health care professionals have an important role to play not only in early identification and referral, but also in the evaluation of children with suspected LD. This begins with a complete medical history, physical examination (including formal hearing and vision testing, lead and iron deficiency screening), and a thorough neurologic examination. Primary pediatric health care professionals will also be familiar with the child's family and psychosocial history. The latter is particularly relevant as stressful or frankly neglectful home environments can lead to academic underachievement and school failure. A medical assessment can also identify behaviors suggestive of emotional and behavioral problems, such as depression or oppositional behavior, that can interfere with school performance. Finally, problems with attention and concentration associated with attention-deficit/hyperactivity disorder (ADHD) should be assessed as potential primary contributors to academic underachievement or as a comorbid condition with LD.[14] Of course, assessment for LD requires psychometric testing, including administration of individual standardized measures of cognitive ability (intelligence tests) and academic achievement. These assessments may be completed by school psychologists as part of an assessment of eligibility for special education services or by private psychologists.

What Causes Learning Disability?

A lengthy review of the genetic, neurologic, and neuropsychologic factors that underlie LD is beyond the scope of this chapter. However, it is important for primary pediatric health care professionals to have some basic information on these topics.

First, and most important, is an understanding that LD is in large measure genetically determined.[7] This does not diminish the important contribution of environment to a child's developmental progress, including the development of pre-academic and academic skills. Nevertheless, it does highlight the importance of the family history in identifying young children at risk for LD. A thorough family history, obtained at the time of an infant or young child's first well-child visit, is therefore an essential component of early identification of LD in primary care pediatric practice.

Recently, functional magnetic resonance imaging (fMRI) technology has helped researchers begin the process of understanding the neurologic underpinnings of LD. This has been particularly true for reading LD, with studies showing clear differences in fMRI activation patterns between individuals with normal reading skills and those with reading LD.[15]

These differences in central nervous system function correlate with deficits in phonological processing, a skill that has been shown to be essential for efficient reading decoding and that is impaired in individuals with classic "dyslexia."[16] Phonological processing refers to "awareness that words can be broken down into smaller segments of sound."[15] While children may have deficits in reading that are caused by a variety of other issues (eg, poor reading comprehension due to problems with receptive language deficits), impaired phonological processing seems to be the most common underlying problem for children who experience difficulty with the basic process of decoding (ie, "reading") written words.

Studies of the neurophysiologic and neuropsychologic deficits associated with underachievement in math and written language are few in comparison to the reading LD literature. Math LD presents a particular challenge, since learning math is dependent on a number of factors, including visual information processing, language, and memory, among others. New information on the etiology of math and written language LDs will undoubtedly be forthcoming in the next 5 to 10 years.

Learning Disability Subtypes/Comorbidities

Multiple Learning Disabilities

While reading LD (or "dyslexia") tends to get the most attention in both the research literature and the classroom, it is important to recognize that many children have more than one type of LD. A recent study of math LD demonstrated that, depending on how LD is defined, 35% to 56.7% of children with math LD also had reading LD.[5] The key for professionals is to recognize that children who are found to have problems in one area of academic achievement often have problems in other areas; hence, such children should be carefully assessed in all areas of academic achievement to ensure that they receive appropriate intervention.

Learning Disability and Attention-Deficit/Hyperactivity Disorder

Epidemiologic studies have demonstrated that children with ADHD experience multiple school-related problems, including academic underachievement, as well as increased rates of absenteeism, grade retention, and school dropout.[14] Comorbid LDs account for at least some of the observed academic underachievement in children with ADHD. This association is so common that every child with LD should be considered to be at risk for ADHD and vice versa. Primary pediatric health care professionals should at least screen for ADHD among their patients with known LD by asking about symptoms of ADHD and considering obtaining ADHD-specific rating scales from the child's parents and teachers. Children with learning problems often exhibit secondary attention deficits, or attention problems secondary to the underlying learning disorder. That is, it can be very difficult for a student to maintain focus on tasks that are difficult for him or her to understand. It is sometimes difficult to distinguish "secondary" attention deficits from primary attention deficits.

Language-based Versus Nonverbal

Most children with LD have problems with language-based learning. This is evident in the "classic" profile of psychometric test results in children with LD, with verbal cognitive measures typically being lower than nonverbal measures. Children with language-based LDs can be expected to have the greatest difficulty in reading and written expression. Teachers and special educators understandably tend to be most experienced in meeting the needs of children with language-based LDs.

In contrast, a smaller though uncertain proportion of children manifest nonverbal LD.[10] Such children have nonverbal cognitive measures that are significantly lower than their verbal scores. Children with nonverbal LD experience problems with math computation, organization (particularly in middle and high school), and higher order math and science concepts. In addition, these children often manifest significant problems with social perception and social interaction that contributes to their negative experiences in educational settings. It is important to distinguish between language-based and nonverbal LDs, both because of the differing profile of academic challenges and differences in associated problems and intervention strategies. School personnel tend to be less familiar with approaches that are successful for children with nonverbal LD; hence, primary pediatric health care professionals can play an important role in advocating for these children.

Intervention and Advocacy

Understanding Special Education Laws

As described earlier, LD is defined in the federal special education law. Each state, in turn, must enact laws that are consistent with the federal statute. As a result, the precise definition of LD varies from state to state, making it important for professionals to familiarize themselves with their own state's special education laws. Your state department of education Web site will typically provide a summary of relevant laws to enable you to understand the system and thus to be knowledgeable advocates for your patients.

In addition to the terminology described earlier, several other important abbreviations are worth noting.[3] The law stipulates that a child's special education plan will be described in detail in an Individualized Education Program (IEP) plan. Primary pediatric health care professionals can play a critical role for families as an independent resource to review school-based psychometric testing reports and IEPs with families to ensure that the IEP provides services appropriate to meet each child's needs.[17] In preparing an IEP, schools must adhere to the principle of a free and appropriate public education (FAPE). This means that the child must be provided with an appropriate array of accommodations and services to meet their basic education needs. This raises an important distinction between services provided under the federal special education law and services that may be available on a private basis. According to the FAPE principle, the child's intervention plan must be appropriate,

but it is not required to be "optimal." States have a great deal of latitude in defining an appropriate level of service, and professionals can play an important role in assessing the extent to which school services are sufficient or should be supplemented by privately available educational services.

The law also requires that educational programs are provided in the *least restrictive environment* (LRE). For example, a child with an LD may be able to receive sufficient support in a regular classroom to allow him to succeed, while another child may require more intensive services in a separate special education classroom for a certain period of the day or certain academic subjects. The law requires that the services are always provided in the setting that most closely matches the typical setting for a child of that age and grade placement.

Interventions: School-based and Private Services

One of the greatest challenges in the LD field has been to ensure access to evidence-based interventions for children with LDs. Recently multiple studies have clearly demonstrated that reading curricula that include explicit teaching of phonics are more effective. This is not surprising, since phonological awareness has emerged as a critically important prerequisite for the development of good basic reading skills. Functional magnetic resonance imaging has evolved as a powerful new research tool for the study of brain activation patterns during reading. This technique has revealed differences between dyslexic and non-dyslexic readers.[15] Reading skills in young children are positively correlated with activation in the left occipitotemporal area, a region of the brain that seems to be responsible for the most rapid, efficient reading skills.[15] Furthermore, when dyslexic children were exposed to an empirically validated educational intervention, they demonstrated increased activation of left-sided, occipitotemporal systems, making them functionally more similar to non-dyslexic children.[18] These studies illustrate the potential application of functional imaging techniques to elucidate the underlying neurophysiology of learning disorders, and to assess the impact of intervention on brain function in a direct manner. Unfortunately, far less is known about the most effective interventions for children with math LD.

For children who qualify for special educational services, the IEP must list specific learning goals, as well as the nature and intensity of services to be provided. Some children may require direct, individual, or small group instruction with a special education teacher in a special education resource room. In other cases, support from the regular or special education teacher, or a paraprofessional assistant, may be provided in the regular classroom. According to federal law, interventions must be provided in the LRE (see above) while still meeting the child's educational needs. In addition to direct instructional services, IEPs can include accommodations, such as shortened assignments, increased time to complete tests (so as not to penalize children for slower reading of questions), or oral administration of tests for children

with reading problems. The key is to ensure that services and accommodations specifically match the demonstrated needs of the child based on the results of individual assessment of learning strengths and weaknesses.

Some children who lag behind their peers in math or reading may not fulfill state and federal criteria to receive formal special educational services through an IEP. For such children, another option may be Title 1 reading and math support. Title 1 is a federal program, originally enacted in 1965 and revised in the No Child Left Behind Act of 2001, designed to provide additional support in reading and math for economically disadvantaged children.[19] Title 1 services are available to all students in a school building only if a sufficient percentage of students who attend that building are economically disadvantaged, based on receipt of federally subsidized free lunch services.

Some children may have delays or deficits in academic achievement that are significant, but not severe enough to qualify for any additional services in the school. In these instances, the only options are private services provided by individual teachers, tutors, or tutoring agencies. The primary pediatric health care professional can play an important role in directing families to high-quality tutoring services in the community.

Unfortunately, unproven, ineffective "interventions" and treatments are available, often at significant cost in both time and money. The issue of nonstandard therapies for children with developmental and behavioral disorders is addressed in detail elsewhere in this book (see Chapter 21). However, one specific, supposed "intervention" for reading LD deserves mention. Although there is no empiric evidence to support it, "vision therapy" is available in many communities and is typically provided by optometrists. This intervention is based on the non–evidence-based belief that reading problems can be corrected by "eye exercises" aimed at somehow improving the child's ability to process the written word. This clearly contradicts all of the research evidence that demonstrates that reading skills depend on language-based cognitive processes, such as phonological awareness. In a joint policy statement, both the American Academy of Pediatrics and the American Academy of Ophthalmology concluded that scientific evidence does not support the efficacy of eye exercises, behavioral vision therapy, or special tinted filters or lenses for improving the long-term educational performance in children with reading LD and, thus, these vision therapies are not endorsed and should not be recommended for children with reading LD.[20]

Consequences of Failure to Intervene

Recent changes in the federal special education laws were intended, in part, to prevent situations in which children were "required" to fall so far behind their peers before qualifying to receive special educational services that intervention was available too late to make a meaningful difference in learning outcomes. Children who do not receive timely intervention are at risk not only of academic failure, but also

for school dropout and the psychosocial morbidities that accompany limited academic achievement, such as unemployment, substance abuse, and juvenile delinquency.[21–23]

For reading LD, research has clearly demonstrated that intervention must be provided early, at least before third grade, in order to provide an opportunity to remediate reading problems.[24] Thus early identification and timely access to evidence-based reading intervention is essential to ensure the best possible outcome. For math LD, we do not yet know the critical age by which problems must be identified, although it is reasonable to assume that there may be a similarly limited window of opportunity to ensure adequate academic outcomes in math.

Historically, children who demonstrated inadequate academic achievement were often retained a grade, based on the assumption that another year at the same grade level would allow the child to "catch up" to their peers. For many children, repeating a grade led to a delay in assessment that would have revealed an LD, and initiation of appropriate remediation. We now know that grade retention is almost universally unsuccessful and is in fact associated with poorer long-term school outcomes.[25] If there is a role for grade retention, it is only for a limited number of children in the very early school years (kindergarten, first grade) and should only be considered after a thorough evaluation for specific LDs or other conditions that may account for the child's academic underachievement.

Box 15-1. Key Points

- Know your state's special education laws and local school district polices and procedures.
- Advise parents to request an evaluation if concerns are present.
- Write a brief note to the school requesting an evaluation.
- Get to know private psychologists who can evaluate the child if necessary.
- Chronic illnesses can affect academic achievement (uncontrolled asthma is an excellent example).
- Rule out hearing and vision impairment.
- Rule out neurologic disorders.
- Rule out common comorbidities of LD (especially attention-deficit/hyperactivity disorder).
- Be aware of services available at school and in the community (for kids who do not qualify for special education services at school).
- Refer parents to local advocates to review special education decisions and plans (Individualized Education Program [IEP]). The Learning Disabilities Association of America (www.LDA.org) and the National Center for Learning Disabilities (www.ncld.org) are great sources of information and support.
- Be aware of the impact of LD on the child and family and look for areas of strength to help minimize the impact of LD on self-esteem.
- Counsel families to avoid unproven approaches for LD (diet, vitamins, visual training, EEG biofeedback).
- Advocate for good reading instruction in your community; specifically, programs that include direct teaching of phonological awareness skills.
- Promote literacy in your practice and your community.

Abbreviations: LD, learning disability; EEG, electroencephalogram.

Advocacy

Throughout this chapter, the role that the primary pediatric health care professional can play is to ensure that children with LD are identified in a timely fashion and referred for appropriate assessments and that evidence-based intervention has been highlighted (Box 15.1).

Starting the Assessment Process

If professionals incorporate early identification and ongoing monitoring for LD into their routine practice, they will be in a position to direct their patients to timely assessments, either through the local school or private psychologists. This is particularly important for families that may lack the resources to monitor and understand information about their child's academic progress.

Reviewing Evaluation Reports and Individualized Education Programs

Similarly, the professional can assist the family by serving as an objective reviewer of assessment reports and making referrals for additional, private evaluations when school assessments have not adequately addressed the child's needs. Once the evaluation is complete, the professional can meet with the family to review the IEP to ensure that goals match the demonstrated learning needs of the child.

Get to Know Local Resources

Primary pediatric health care professionals are respected members of the community and recognized advocates for children under their care. It helps to develop a direct working relationship with local special education directors, school principals, and superintendents when possible. These relationships will help to ensure that the professional's input is considered when concerns arise with regard to school services. Similarly, it is helpful for the professional to become familiar with local providers and agencies that offer high-quality assessment and intervention for children who require private services to supplement programs that are provided in the public school.

Advocating for Evidence-based Interventions and Curricula

Finally, the primary pediatric health care professional is in a position to help ensure that special educational and private interventions employ evidenced-based interventions, particularly for children with reading LD.

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Chapter 16

Attention-Deficit/Hyperactivity Disorder

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Introduction

Overactive children were described in the medical literature over a century ago. In 1902 George Still observed a pattern of behavior in children that consisted of restlessness, inattentiveness, and overarousal with an inability to internalize rules and limits. As a reflection of the Victorian era, he attributed the condition to a defect in moral character.[1] Children who recovered from influenza encephalitis following the endemic of 1917–1918 often displayed symptoms of restlessness, inattention, impulsivity, easy arousability, and hyperactivity. This was described as a post-encephalitic behavior disorder.[2] Research in neuropsychology, coupled with clinical observations, led to progressive changes in the name of the condition from hyperkinetic impulse disorder, to attention deficit disorder, and most recently to attention-deficit/hyperactivity disorder (ADHD).

The core symptoms of ADHD are inattention, hyperactivity, and impulsivity. Attention-deficit/hyperactivity disorder is one of the most common and most extensively studied behavioral disorders in school-aged children. It is a chronic condition and persists into adolescence and adulthood in 60% to 80% of individuals diagnosed with ADHD during childhood.[3,4]

Demographics/Epidemiology

The prevalence rate of ADHD varies depending on diagnostic criteria, the population studied, and the number of sources used to establish a diagnosis. The absence of biological markers to establish a diagnosis of ADHD and the need to depend on parent and teacher reports of behavior is a challenge to epidemiologic research. In a recent national study, 7% of children met *Diagnostic and Statistical Manual, Fourth Edition (DSM-IV)* criteria for ADHD.[5] In this study, fewer than half of children identified with ADHD received either a diagnosis or regular treatment for ADHD. Poor children were more likely to meet criteria for ADHD, whereas wealthier children

were more likely to receive regular medication treatment. In studies that use clinical samples, there is a male predominance of ADHD with a male:female ratio of 3:1 for the combined type and 2:1 for the predominantly inattentive type.[6] In community samples, predominantly inattentive ADHD is the most prevalent subtype (about 1.5 times more common than the combined type).[6] School-aged and adolescent girls are more likely to comprise the inattentive subtype.[6] Attention-deficit/hyperactivity disorder does occur in preschool children, although the diagnosis is more challenging.[7]

Etiology

Attention-deficit/hyperactivity disorder is a heterogeneous disorder with a multifactorial etiology. A diverse set of biobehavioral pathways can lead to the behavioral expression of the core symptoms of ADHD.[8] Genetic, epigenetic, and environmental factors interact to give rise to the ADHD phenotypes. A multifactorial model integrates genetic, neural, cognitive, and behavioral mechanisms. Behavioral disinhibition has been proposed as the major core deficit in ADHD.[9] In this model, children with ADHD are found to have difficulty mobilizing delayed gratification, the ability to interrupt ongoing responses (such as stopping playing a video game because it is time for homework), and interference control (eg, not reacting to a friend walking past the classroom door while you concentrate on a math problem).

Imbalances in dopaminergic and noradrenergic regulation mediate the core symptoms of ADHD.[10–13] These neurotransmitters may increase the inhibitory influences of frontal cortical activity on subcortical structures. Deficits in frontal lobe functioning and subcortical connections with the frontal lobes, particularly the caudate, putamen, and globus pallidus, have been found in neurobiologic and neuroimaging studies.[14,15] Stimulant medications and other medications found effective in ADHD treatment seem to increase the inhibitory influences of frontal lobe activity through these dopaminergic and noradrenergic influences.

Recent evidence suggests that cortical development in children with ADHD lags behind typically developing children by years but follows the normal sequence of brain development. This observation has led to the conclusion that ADHD represents a delay rather than a deviance in cortical brain maturation.[16] Cortical delay is most prominent in the lateral prefrontal cortex, an area that supports the ability to suppress inappropriate responses, executive control of attention, evaluation of reward contingencies, higher order motor control, and working memory. These are the domains of neuropsychological functioning that have been found to be impaired in children with ADHD.

Attention-deficit/hyperactivity disorder has strong familial associations. Parents and siblings of a child with ADHD carry a 2- to 8-fold increase in the risk for ADHD. Twin studies have found that 75% of the variance in ADHD phenotype can be attributed to genetic factors. If one identical twin has ADHD, the other twin has a greater than 50% chance of having ADHD.[17] Associations have also been made between

ADHD and the dopamine transporter gene. The gene most strongly implicated in ADHD is the human dopamine receptor D4 gene.[18,19] In support of this observation, stimulant medications are primarily dopamine reuptake inhibitors.

Biological and psychosocial factors also contribute to ADHD. Prenatal exposure to alcohol, cocaine, and nicotine are associated with ADHD phenotypes.[20] Psychosocial adversity is also an important risk factor. Chronic family conflict, decreased family cohesion, and parental psychopathology have been found to occur more commonly in families of children with ADHD than in controls.

Functional Impairment

The diagnosis of ADHD and subsequent treatment require *evidence of impairment in functioning.* Children with ADHD have been found to have significant functional impairment in the areas of academic achievement, family relationships, peer relationships, self-esteem and self-perception, accidental injuries, and overall adaptive function.[21–23] They are likely to underachieve in school regardless of whether they have comorbid learning disabilities. They are also more likely to qualify for special education, repeat a grade, receive more suspensions, and drop out of school. Families who have a child with ADHD are more likely to experience impairment in parental harmony, parenting distress, perceived incompetence in parenting, and parent-child interaction problems. Children with ADHD often receive high levels of negative peer rankings of social standing. The self-esteem of children with ADHD is often lower than their peers, although they may initially report inflated levels of self-regard.[21]

Core symptoms of ADHD challenge school-related activities and tasks, relationships, and other functions. Cognitive impairments include lack of impulse control and deficits in attention, memory, organization, time management, and judgment. Activity limitations include difficulties in learning and applying knowledge (reading, writing, mathematics), problems with carrying out single or multiple tasks, studying, and self-managing behavior. Attention-deficit/hyperactivity disorder also impacts interpersonal interactions; communication and self-care; adjusting to and succeeding in educational programs; leaving school to enter work; and establishing a community, social, and civic life. It is these broad functional disabilities that should become targets for intervention in individuals with ADHD in addition to the core symptoms themselves.[24]

Coexisting Conditions

Most children and adolescents with ADHD have coexisting conditions. The most prevalent conditions include other disruptive behavior disorders (oppositional defiant disorder [ODD] and conduct disorder), anxiety disorder, depressive disorders, and learning disabilities. Sleep disturbances are also common. Each of these conditions adds its own elements to the functional impairment of individuals with ADHD (Table 16.1).

Table 16.1. Prevalence of Conditions in Community Samples With and Without Attention-Deficit/Hyperactivity Disorder (ADHD)

Condition	Coexisting With ADHD[a]	In Non-ADHD Populations[b]
Oppositional defiant disorder	35%	2%–16% (males)
Conduct disorder	25%	6%–16% (males); 2%–9% (females)
Anxiety disorder	25%	5%–10%
Depressive disorder	18%	2% (child); 5% (adolescent)
Learning disorder[c]	15%	7%

[a] Green M, Wong M, Atkins D, et al. *Diagnosis of Attention Deficit/Hyperactivity disorder. Technical Review 3.* Rockville, MD: US Dept of Health and Human Services; 1999. Agency for Health Care Policy and Research; AHCPR publication 99-0050.
[b] Lewis MB, ed. *Child and Adolescent Psychiatry: A Comprehensive Textbook.* 3rd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2002.
[c] American Academy of Pediatrics. *The Classification of Child and Adolescent Mental Diagnoses in Primary Care: Diagnostic and Statistical Manual for Primary Care (DSM-PC) Child and Adolescent Version.* Elk Grove Village, IL: American Academy of Pediatrics; 1996.

The presence of a coexisting condition can substantially change predictors of outcomes and influence the targets for treatment. For example, children with ADHD and coexisting ODD are at risk for developing conduct disorder, which can be a gateway to adolescent substance abuse.[25] Children with ADHD and coexisting mood disorders may have a poorer outcome during adolescence than children with ADHD alone. Children with coexisting anxiety disorders may differ in their response to stimulant medication and, in some cases, may respond just as well to behavioral treatments as to medication management.[26] Children and adolescents with ADHD and coexisting academic problems may benefit from accommodative services under Section 504 of the Rehabilitation Act or for more intensive special education services under the Individuals with Disabilities Education Act (IDEA), depending on the extent of their academic problems.[27]

Prognosis

The long-term outcome for children with ADHD is related to the severity and type of symptoms, coexisting conditions (eg, mental health disorders and learning disabilities), intelligence, family situation, and treatment. Seventy percent to 85% of children with ADHD have symptoms persisting into adolescence and adulthood.[28] Hyperactivity tends to diminish over time, but impulsivity and inattention often persist. Adolescents with ADHD often display immature peer interactions.[28,29]

It is important to recognize that most follow-up studies of children with ADHD do not evaluate the effects of treatment strategies. Adolescents with ADHD have more driving tickets and more motor vehicle accidents, including accidents with fatalities. They also initiate intercourse sooner, with more sexual partners and evidence less use of birth control, more sexually transmitted infections, and more teenage pregnancy. Teenagers with ADHD smoke at a younger age and have a higher prevalence of smoking. Those with conduct disorders are at increased risk for substance abuse.

The risk of substance use disorders over the lifespan is up to twice as great in individuals with ADHD. Adolescent girls with ADHD have more depression, anxiety, poor teacher relationships, an external locus of control, and impaired academic performance compared with their peers. Compared with boys with ADHD, they are more impaired by self-reported anxiety, distress, and depression, and an external locus of control.

Attention-deficit/hyperactivity disorder is associated with academic underachievement in reading and impaired school functioning.[30] Children and adolescents with ADHD have greater rates of school absenteeism, grade retention, and school dropout. They are more frequent users of school-based services, and have increased rates of detention and expulsion.[21] Coexisting learning disabilities and psychiatric disorders add to the magnitude of poor school outcomes. Treatment with stimulant medication is associated with better long-term school outcomes, although medication treatment does not necessarily improve standardized test scores or ultimate educational attainment. Stimulant treatment has been associated with a significant decrease in the rate of substance abuse disorder,[31] although some recent studies have challenged this effect of treatment.[32]

Studies of adults with ADHD suggest that they have lower socioeconomic status, more work difficulties, and more frequent job changes, as well as fewer years of education and lower rates of professional employment. Adults with ADHD also report more psychological maladjustment, more speeding violations and suspension of driver's licenses, poorer work performance, and more frequent quitting or being fired from jobs.[33]

Diagnosis and Evaluation

Hyperactivity, impulsivity, and inattention are observed in many children and adolescents at times during typical development. Attention-deficit/hyperactivity disorder is considered only when the symptoms are persistent, pervasive (present in multiple environments), and impair critical functions of learning and social development consistent with a child's developmental age. Most ADHD studies include only school-aged and adolescent patients. The diagnostic challenge to define ADHD in preschool children is significant in that, to some degree, all behaviors associated with ADHD are part of normal development in the preschool age group.

Unlike most medical conditions, but similar to other behaviorally defined disorders, there are no medical tests or imaging studies that can be used to diagnose ADHD. Instead, the diagnostic criteria from the *DSM,* an empirically based classification system of behavior disorders, are recommended as the framework for primary pediatric health care professionals' clinical assessment of ADHD. The *DSM* facilitates communication among professionals and patients, provides information relevant to treatment and prevention, and encourages research in understanding behavioral problems that impact development.

Diagnostic criteria for ADHD in school-aged children and adolescents include documentation of the following criteria:

- At least 6 of the 9 behaviors described in the hyperactive/impulsive domain and/or
- At least 6 of the 9 behaviors described in the inattentive domain
- Symptoms occur often and to a degree inconsistent with the child's developmental age
- Presence of these behaviors in 2 or more major settings (eg, home and school) for at least 6 months
- Presence of some symptoms of ADHD (by history) prior to 7 years of age
- Significant impairment in learning and/or social interactions
- Symptoms are not attributable to another mental health condition

Eighteen specific behaviors must be ascertained as a part of the diagnostic process (Box 16.1). Three subtypes of ADHD (predominantly hyperactive-impulsive type, predominantly inattentive type, and combined type) are delineated.[3]

The diagnostic process must include ascertainment of how many of the 18 ADHD-associated behaviors occur frequently and in most situations. The *DSM* for primary care is a guide to distinguish normal developmental variation from the behaviors associated with ADHD.[34] Documenting that the behaviors occur frequently is the first step toward good practice. One significant shortfall of the *DSM* is that it is not developmentally sensitive. Not all of the *DSM* criteria (eg, often runs about or climbs excessively in situations in which it is inappropriate) are applicable to teenagers. Preschoolers display many more of these criteria as part of their normal development, and adolescents display less (eg, hyperactivity during preadolescence may disappear or be quite diminished during teen years.)

A clinician must establish whether the behaviors are limited to a particular environment or situations only, or whether the behaviors are present in a variety of situations. There must be evidence that core ADHD behaviors occur across a child's major environments, including home and school. Knowledge about behaviors in social activities outside of school and home (eg, during sports, camp, scouting, or religious activities) may also be useful. If a child is only exhibiting ADHD symptoms at school but not at home or in any other settings, the symptoms may not represent ADHD but might be secondary to a primary language, learning, or intellectual disability. Alternatively, if the child is only exhibiting the ADHD symptoms at home, but not at school or any other setting, a parental-child interaction problem, developmentally inappropriate parental expectations or limit setting, or parental psychopathology may be the primary cause of these symptoms. Inattentive behaviors may also be the result of problems with hearing or vision or secondary to anxiety or depression. Ascertaining that the duration of symptoms is longer than 6 months is crucial. Many of the 18 ADHD symptoms may occur in response to life event changes (eg, marital discord, divorce, economic stress, a family move, a new school,

Box 16.1. *DSM-IV-TR* Criteria for Attention-Deficit/Hyperactivity Disorder (ADHD)—Behaviors Associated With Core ADHD Symptoms[a]

Inattention

Six or more of the following symptoms have persisted for at least 6 months to a degree that is maladaptive and inconsistent with developmental level.

- Often fails to give close attention to details or makes careless mistakes in schoolwork, work, or other activities
- Often has difficulty sustaining attention in tasks or play activities
- Often does not seem to listen when spoken to directly
- Often does not follow through on instructions, fails to finish schoolwork, chores, or duties in the workplace (not due to oppositional behavior or failure to understand instructions)
- Often has difficulty organizing tasks and activities
- Often avoids, dislikes, or is reluctant to engage in tasks that require sustained mental effort (e.g., school work or homework)
- Often loses things necessary for tasks or activities (e.g., toys, school assignments, pencils, books, or tools)
- Is often easily distracted by extraneous stimuli
- Is often forgetful in daily activities

Hyperactivity-Impulsivity

Six or more of the following symptoms have persisted for at least 6 months to a degree that is maladaptive and inconsistent with developmental level.

Hyperactivity

- Often fidgets with hands or feet or squirms in seat
- Often leaves seat in classroom or in other situations in which remaining seated is expected
- Often runs about or climbs excessively in situations in which it is inappropriate (in adolescents or adults, may be limited to subjective feelings of restlessness)
- Often has difficulty playing or engaging in leisure activities quietly
- Is often "on the go" or often acts as if "driven by a motor"
- Often talks excessively

Impulsivity

- Often blurts out answers before questions have been completed
- Often has difficulty awaiting turn
- Often interrupts or intrudes on others (e.g., butts into conversation or games)

[a] Reprinted with permission from: *Diagnostic and Statistical Manual of Mental Disorders.* 4th ed. Text rev. Copyright 2000. American Psychiatric Association.

an illness in a family member, etc) or during the early stages of a disease process (eg, posttraumatic encephalopathy, petit mal seizures, an acquired hearing loss, adrenoleukodystrophy, etc). Chronic problems such as living in poverty or enduring ongoing physical abuse or significant emotional neglect may also produce symptoms indistinguishable from ADHD.

Symptoms of ADHD that are not associated with impairments in schoolwork or with successful social relationships do not meet the diagnostic criteria for ADHD. An inadequate assessment of functional impairment is a common cause of overdiagnosis. For example, the hyperactivity, impulsivity, or inattentiveness in some

school-aged children is either not severe enough or is situational in an educational or social environment but not at home. Overactivity and situational inattentiveness in a school-aged child who is doing well in the classroom, achieving academically, and socially engaging is not ADHD. Clinical judgment is important in assessing the effect of the core ADHD symptoms on academic achievement, classroom performance, family life, social skills, independent functioning, self-esteem, leisure activities, and self-care. Asking a parent or teacher, "Do you think that Billy's inattention and hyperactivity are impairing his school performance or peer interactions?" can help to establish the presence of functional impairment.

An evidenced-based practice guideline for the diagnosis of school-aged children with ADHD has been published by the American Academy of Pediatrics (AAP).[35] Following is a summary of the guidelines.

Recommendation 1: In a child 6 to 12 years old who presents with inattention, hyperactivity, impulsivity, academic underachievement, or behavior problems, primary care clinicians should initiate an evaluation for ADHD.

Early recognition of ADHD by primary pediatric health care professionals is ensured when screening for core symptoms and problems in school and social relationships occur during health supervision visits. The AAP practice guideline suggests the following screening questions:

1. How is your child doing in school?
2. Are there any problems with learning that you or the teacher has seen?
3. Is your child happy in school?
4. Are you concerned with any behavioral problems in school, at home, or when your child is playing with friends?
5. Is your child having problems completing classwork or homework?

Recommendation 2: The diagnosis of ADHD requires that a child meet *DSM-IV* criteria.

Recommendation 3: The assessment of ADHD requires evidence directly obtained from parents or caregivers regarding the core symptoms of ADHD in various settings, age of onset, duration of symptoms, and degree of functional impairment. Use of ADHD-specific scales is a clinical option when evaluating children for ADHD. A toolkit for clinicians, *ADHD—Caring for Children With ADHD: A Resource Toolkit for Clinicians,*[36] contains one of the available scales. It can be downloaded from the AAP Web site for use in the office or clinic. The traditional pediatric clinical interview with a child and a parent (together and separately) provides rich information, some of which may not be apparent or emphasized on a behavioral rating scale. The clinical interview also begins the therapeutic alliance between the clinician, parent, and child, a process that is crucial to later adherence to a treatment plan.

Recommendation 4: The assessment of ADHD requires evidence directly obtained from the classroom teacher (or other school professional) regarding the core symptoms of ADHD, duration of symptoms, degree of functional impairment, and coexisting conditions. This can be accomplished with an ADHD-specific rating scale complemented by a teacher narrative where a clinician requests a written response to the following: "Tell me about _____in your classroom. Tell me about his/her learning style and behaviors."

Recommendation 5: Evaluation of the child with ADHD should include assessment for coexisting conditions. The AAP toolkit[36] includes a scale specific for ADHD behaviors that also ascertains coexisting ODD, conduct disorder, symptoms of anxiety and depression, learning problems, and level of impairment. Primary care clinicians may have difficulty diagnosing some coexisting conditions, and may need to consult with a developmental-behavioral pediatrician, neurodevelopmental pediatrician, psychologist or psychiatrist. A primary care toolkit, *Addressing Mental Health Concerns in Primary Care: A Clinician's Toolkit,* is available from the AAP.

Recommendation 6: Other diagnostic tests are not routinely indicated to establish the diagnosis of ADHD. These include hematocrit, blood lead, thyroid hormone levels, brain imaging studies, electroencephalography, and computerized continuous performance tests.

A complete physical examination should be done to rule out findings that might produce symptoms mimicking ADHD or that suggest syndromes with a high prevalence of ADHD behaviors. The examination should include visual acuity; an audiogram; measurements for height, weight, head circumference, and blood pressure; and a neurologic examination. Dysmorphic features that suggest a syndrome with a high prevalence of ADHD (eg, fetal alcohol syndrome or fragile X syndrome) should be assessed.

Expanding the traditional neurologic examination to include some neurodevelopmental screening items can informally assess a variety of components of neurologic function that may be associated with ADHD or coexisting conditions. Examples of some informal neurodevelopmental screening tasks that can be easily incorporated into a primary care office visit can be found in Table 16.2.[37] While these neurodevelopmental screening tasks are not standardized, they can provide clues to a specific learning disability or language disorder that requires further standardized assessment. The mild stress induced by these tasks may be associated with hyperactivity (fidgetiness, getting up from seat, constant motion, etc) and inattentiveness (distractibility, not on-task, daydreaming), signs that emerge in the office only during this part of the examination.

Table 16.2. Informal Neurodevelopmental Screening Tasks

Task	Function
Ask child to write a sentence.	Written expression and dysgraphia
Ask child to tell you about a movie or video seen recently.	Oral expression, memory, sequencing
Ask child to read a paragraph.	Reading fluency and comprehension appropriate for age (Grey Oral Reading Test, 4th edition)[a]
Ask child to copy geometric figure or do a draw-a-person test.	Fine motor and visual-spatial skills
Ask child to repeat a series of random numbers in both a forward and reversed manner.	Attention, short-term memory, sequencing, working memory
Ask child a multiple-step task to complete in order given.	Attention, memory, auditory processing

[a] Wiederholt JL, Bryant BR. *Gray Oral Reading Test.* 4th ed. San Antonio, TX: Pearson Psychological Corporation; 2001.

Another method to engage a child during the evaluation is to ask them to draw a picture of their family doing something. These kinetic family drawings within the context of a primary pediatric health care professional's office visit can be valuable in several ways. They may reduce the stress of the visit by giving the child something enjoyable to do while you are talking to a parent. The drawing can be used to assess fine motor and visual-perceptual skills that are important in learning. They also provide an opportunity to open new areas of communication with both parents and children. Ask the child, "Tell me about the drawing? What's going on? Where is _____?" Then give the parent an opportunity to comment. One cautionary note: Such drawings should not be overinterpreted; they are best used in the primary care pediatric practice for a source for enhanced communication with a child and parent.[39]

Similar to other chronic conditions, the diagnosis of ADHD often requires more than a single visit to a primary pediatric health care professional. Data gathering, a clinician interview and examination, and summary of results for parents and child require a significant time commitment. An adaptation to primary care is to schedule several 30-minute visits in order to accurately assess each patient.

Treatment

Medication and behavioral therapy techniques based on behavior modification have been found to be effective treatments for children and adolescents with ADHD. Educating the family (including the child and caregivers) about ADHD is the first step to ensure a good outcome. A discussion should include an understanding of ADHD as "brain based," not due to poor parenting or intentional misbehavior, amenable to change through medication and behavioral therapy, and responsive to accommodations in the home and classroom. Most parents appreciate an empathetic clinician who makes a clear statement about how difficult it must be to raise a child with

ADHD and clarifies the primary pediatric health care professional's role as a partner in the coordination of care between the family, school, and medical office.

The AAP clinical guideline on the treatment of the school-aged child with ADHD supports this approach.[40] Salient clinical points follow.

Recommendation 1: Primary care clinicians should establish a management program that recognizes ADHD as a chronic condition. The chronic disease model of care includes parent/patient education, continuous availability for questions and counseling, coordination with other services, setting specific goals, and monitoring.

The care of ADHD is best achieved by establishing a medical home model—the provision of comprehensive primary care in a high-quality and cost-effective manner. In a medical home for a patient with ADHD, a primary pediatric health care professional works in partnership with the family and patient to ensure that all of the medical and nonmedical needs of the child are met. Through this partnership, the pediatrician can help the family access and coordinate specialty care, educational services, out-of-home care, family support, and other public and private community services that are important to the overall outcome.[41]

Recommendation 2: The treating clinician, parent, and child, in collaboration with school personnel, should specify appropriate target outcomes to guide management. For the child with ADHD, target outcomes should reflect key symptoms the child manifests and the specific impairments these symptoms cause. Impairments and problem behaviors differ greatly from child to child.

Target outcomes (eg, complying with parental commands at home, or finishing homework and handing it in) are reflections of the functional impairment that are the result of core behaviors. It is these functional impairments that should be the foci for specific interventions.

Recommendation 3: The clinician should recommend medication and/or behavior therapy as appropriate to improve target outcomes in children with ADHD.

In general, medication management alone has been found to be a stronger intervention than behavioral treatment alone, particularly for the core symptoms of ADHD. There are particular circumstances, such as ADHD with comorbid anxiety disorders, where the 2 interventions may be equally effective, and other circumstances, such as difficult parent-child interactions, where the combined treatment with both medication and behavioral therapy is more effective than either intervention alone.

Recommendation 4: When the selected management for a child with ADHD has not met target outcomes, clinicians should evaluate the original diagnosis, use of all appropriate treatments, adherence to the treatment plan, and the presence of coexisting conditions.

Recommendation 5: The clinician should periodically provide a systematic follow-up for the child with ADHD. Monitoring of the ongoing treatment plan should be directed to target outcomes and adverse effects of medication by obtaining specific information from parent, teacher, and child.

Medication

Methylphenidate, amphetamine, atomoxetine, and extended-release guanfacine are approved by the US Food and Drug Administration (FDA) for use in children and adolescents on the basis of evidenced-based safety and efficacy studies. Other medications with limited evidence from randomized, controlled studies but without FDA approval for use in ADHD include buproprion, clonidine, short-acting guanfacine, and tricyclic antidepressants.

Stimulants

The most widely prescribed medications for children and adolescents with ADHD are the psychostimulants, including preparations derived from both methylphenidate and amphetamine.[42] Well over 200 scientific studies support their value for children with ADHD. Stimulants act as dopamine and norepinepherine reuptake inhibitors by increasing the available dopamine and norepinepherine in the caudate nucleus and prefrontal cortex.[43]

Stimulant medications are currently available in short-acting (3–6 hours), intermediate-acting (6–8 hours), and long-acting (10–12 hours) preparations (Table 16.3). Behavioral effects are seen within 30 to 45 minutes. Some parents report an intense wear-off period (rebound) as the behavioral effects decline. Children and adolescents do not develop tolerance to stimulants. Stimulant dosing is not based on milligrams per kilogram, but rather on the dose that works best without significant side effects up to the FDA-recommended maximum doses (see Table 16.3 and Chapter 20).[44] Primary pediatric health care professionals often settle for stimulant doses that are helpful but suboptimal. Stimulants should be titrated in dose to achieve the best response rather than using the dose that produces the first noticeable changes. Pharmacodynamic effects differ with specific targets. Disruptive behaviors are affected rapidly following a dose of stimulants, while increased attention to subjects like math may take up to 1.5 hours after administration. Stimulants work at school to increase on-task behavior and decrease interrupting and fidgeting. At home, they improve on-task behavior, parent-child interactions, and compliance. They also improve peer perceptions of social standing and increase attention while playing sports. Stimulant medications are also effective in preschool children with ADHD; however, in this age group, side effects are more frequent and severe.[45]

The most frequent side effects of stimulants include stomachaches and headaches (these symptoms typically resolve spontaneously after the first week), decreased

Table 16.3. Stimulant Medications—First-Line Treatments[a]

Generic Class (Brand Name)	Daily Dosage Schedule	Duration of Behavioral Effects, hours	Prescribing Schedule
Methylphenidate			
Short-acting			
(Ritalin, Metadate, Methylin)	BID to TID	3–5	2.5–20 mg BID to TID
(Focalin)	BID to TID	3–5	2.5–10 mg BID to TID
Intermediate-acting			
(Ritalin SR, Metadate ER, Methylin ER, Ritalin LA)	QD to BID	3–8	10–40 mg QD or 40 mg in the morning and 20 mg early afternoon
Long-acting			
(Concerta, Metadate CD)	QD	10–12	18–72 mg
(Focalin XR)	QD	10–12	5–30 mg QD
Daytrana (transdermal patch)	QD	9	10–30 mg QD
Amphetamines			
Short-acting	BID to TID	4–6	5–15 mg BID or 5–10 mg TID
(Dexedrine, Dextrostat)			
Intermediate-acting	QD to BID	6–8	5–30 mg QD or 5–15 BID
(Adderall, Dexedrine Spansules)			
Long-acting			
(Adderall XR)	QD	10–12	10–40 mg QD
Lisdexamfetamine	QD	12	30–70 mg QD

Abbreviations: BID, twice a day; QD, every day; TID, 3 times a day.

[a] Modified from American Academy of Pediatrics Subcommittee on Attention-Deficit/Hyperactivity Disorder and Committee on Quality Improvement. Treatment of the school-aged child with attention-deficit/hyperactivity disorder. *Pediatrics.* 2001;108:1033–1044.

appetite, difficulty with sleep initiation, and jitteriness. Motor tics may also occur in some children. These side effects often occur early in treatment; are usually mild; and can often be ameliorated by alterations in dose, timing, or the use of alternate stimulant medications. There are no differences in potential side effects among stimulants. A child or adolescent with a specific side effect on one stimulant medication may not experience it on another. While there is no evidence that chronic use of stimulants causes significant impairment in adult height, the 3-year follow-up of the Multimodal Treatment Study of Children with ADHD (MTA)[46] reported that growth in newly treated children averaged 2.0 cm less in height and 2.7 kg less in weight than growth in unmedicated children. The reduced growth occurred mostly in the first year of treatment and was not seen in the third year.[47]

Treatment of adolescent patients with ADHD with stimulant medication has been associated with a reduction in risk for subsequent drug and alcohol use disorders.[48] Use of stimulants is contraindicated in cases of previous sensitivity, glaucoma, in cases of drug abuse, in patients who are taking a monoamine oxidase inhibitor, or in patients who are actively psychotic. Stimulants are not contraindicated in cases of tics, and stimulants do not seem, as previously thought, to lower the seizure

threshold. While stimulant medications increase heart rate and blood pressure, the controversy concerning an association between the use of stimulant medication and sudden cardiac death is under investigation. The AAP recommends a careful assessment of children starting stimulants. Screening should include a targeted cardiac history, including a patient history of previously detected cardiac disease, palpitations, syncope, or seizures; a family history of sudden death in children or young adults, hypertrophic cardiomyopathy, and long QT syndrome; and a cardiovascular physical examination.[49] In general, stimulants are considered quite safe and effective for treatment of ADHD. Height and weight should be monitored regularly. Blood pressure and heart rate should be checked before and during treatment with stimulant medications. If the family history or child's history includes sudden unexplained death, severe heart palpitations, exercise intolerance, fainting spells, or chest pain, a cardiology consultation is recommended before initiating stimulant medications. Routine electrocardiograms are not indicated prior to stimulant use.

The development of long-acting stimulants was a response to problems observed by parents, teachers, and clinicians. Gaps in the behavioral effects of multiple daily doses of 4-hour preparations (immediate-release stimulants) occur at some of the least structured times of the day (bus rides, lunch time, and recess). In addition, many children feel embarrassed to take medication in school under the scrutiny of the school nurse and their peers, and compliance then becomes an issue. In some cases, homework time remains uncovered and may require an afternoon dose of an immediate-release stimulant.

Non-Stimulants

Stimulants remain the first-line psychopharmacologic treatment for ADHD. It is estimated that at least 80% of children will respond to one of the stimulants if they are used in a systematic way. Non-stimulants remain an option for children and adolescents for whom stimulants are not effective or cause significant adverse side effects or exacerbation of other coexisting disorders, or where non-stimulants are a preferred option for treating ADHD and a coexisting disorder with a single medication.

Atomoxetine (Strattera) is a non-stimulant medication that is approved for use in school-aged children, adolescents, and adults with ADHD. It is a norepinepherine reuptake inhibitor and blocks the presynaptic norepinepherine transporter in the prefrontal cortex. It has been found to have a beneficial effect on children and adolescents with ADHD but to have lower efficacy than stimulants.[50–53] Atomoxetine may provide symptom relief during the evening and early morning hours. Motor and verbal tics associated with atomoxetine have not been reported. In addition, atomoxetine may have less effect on delayed sleep onset compared with stimulants. The side effects are otherwise similar to stimulants, but atomoxetine may be associated with more fatigue and nausea compared with stimulants, and there have also been reports

of liver toxicity in association with atomoxetine use. Atomoxetine may be effective in children with ADHD and coexisting anxiety. The initial studies of atomoxetine reported a 2-fold increase compared with control subjects in suicidal ideation, usually occurring in the first month of treatment; actual suicide attempts were not increased. Dosing of atomoxetine is on a per kilogram basis in contrast to stimulants.

Methylphenidate, amphetamine, and atomoxetine have somewhat different efficacies. Reported effect sizes of atomoxetine are moderate (about 0.7 in children and 0.4 in adults compared with stimulants, which have approximately 1.0 effect sizes in clinical trials).[54] Among those who do not respond sufficiently or who develop intolerable side effects to the first ADHD medication tried, about half will respond to one of the other medicines. There is variability in dose response between patients; it is recommended to begin with a small dose of stimulants and titrate upward every 2 to 4 weeks to reach the optimal effect while monitoring side effects. Most children are treated with medication daily including weekends.[55]

The tricyclic antidepressants (TCAs) include imipramine, desipramine, and nortriptyline. They work by inhibiting norepinepherine reuptake. Desipramine produces a response rate for ADHD symptoms comparable to stimulants, as well as fewer tics in patients with coexisting tics and Tourette disorder.[56] Around 20 randomized, controlled trials have supported the efficacy of TCAs for treating ADHD. The use of TCAs, however, is limited by their cardiac side effects, the need for cardiac monitoring, the monitoring of plasma levels, and the possible association with sudden death reported in a small number of 6- to 14-year-old patients taking desipramine.

Bupropion is an amino-ketone antidepressant with both noradrenergic and dopaminergic properties. Improvement in core ADHD behaviors on bupropion is generally not as complete as improvement with stimulants. Its use in ADHD is supported by only one multisite study that found it significantly better than placebo but not as effective as stimulants.[57] It has been found to improve ADHD with coexisting depression.

Clonidine and guanfacine are presynaptic, central-acting alpha-2 adrenergic agonists that work by affecting norepinephrine discharge rates in the locus ceruleus, which may indirectly affect dopamine. A new long-acting form of guanfacine (Intuniv) was approved by the FDA in 2009 for the treatment of ADHD. These medications are considered second-line treatments for ADHD and should be considered only in cases where stimulants and norepinepherine reuptake inhibitors are not effective or contraindicated. Clonidine has been used clinically to counteract the stimulant side effect of delayed sleep initiation, and for children and adolescents with ADHD who also have significant aggressive behavior. Guanfacine may also be effective in children with ADHD, tics, and aggression.

Behavioral Treatment

Effective psychosocial treatment for ADHD employs the principles of behavior therapy. These principles include the use of behavior modification techniques and social learning theory. They emphasize contingency management and the shaping of children's behaviors through observing and modeling appropriate behaviors, attitudes, and emotional reactions of others. Parents and teachers can be trained in these behavior management principles, and there is good evidence that behavioral interventions are effective for children with ADHD.

The goals of parent training are to help parents learn to achieve consistent and positive interactions with their children, gain a better understanding of what behaviors are developmentally normal, help them cut down on negative interactions with their children (such as arguing or constantly having to repeat commands), teach parents to provide appropriate consequences for their child's behaviors and become more empathetic to their child's viewpoint, and help children to improve their abilities to manage their own behaviors.[58] Many parents have fallen into the trap of almost solely providing negative attention in response to negative behaviors. Using behavior therapy principles, parents learn to conceptualize "discipline" as teaching self-control rather than as punishing negative behaviors. They are taught what behaviors can be reinforced by praise and extinguished by active ignoring, using appropriate punishments only for intolerable or dangerous behaviors[59] (see Chapter 4).

A sound parent training program teaches parents how to

- Deliver and follow through on clear commands.
- Shape behaviors in gradual increments.
- Use daily contingency charts (star or "happy face" charts).
- Institute procedures such as time-out, token economies (earning rewards and privileges contingent on performing desired behaviors), and response cost (losing tokens or privileges for noncompliance).

The same principles of clinical behavioral therapy have been used effectively in training teachers in classroom behavior management. Once parents and teachers have been trained, they can learn to implement systems that provide continuity from home to school, such as daily behavioral report cards that can be used to report on specific target behaviors that are being monitored daily and allow parents to provide either positive reinforcement or consequences at home.

Behavioral interventions alone are often insufficient for effective treatment of core ADHD behaviors. For parents who are hesitant to use prescribed medications to treat their child, initiating treatment with a behavior therapy program alone should be supported with frequent monitoring.

Combined Medication and Psychosocial Treatments: The MTA Study

The short-term benefits of medications and behavioral therapy have been well established. Few studies have also looked at medication treatment and behavioral therapies in head-to-head comparisons. The National Institute of Mental Health's MTA study followed 579 children, aged 7 to 9, in 6 sites over a 14-month period. The children were randomly assigned to 1 of 4 groups: (1) intensive medication management (MedMgt), (2) intensive behavioral treatments (Beh), (3) combined MedMgt and Beh (Comb), and (4) study diagnostic procedures and then routine care in the community (CC). Medication management was initiated with methylphenidate (MPH), using alternative stimulants or non-stimulants for MPH non-responders.[26] The intensive behavioral treatments included 8 sessions of parent training with concurrent teacher consultation and an intensive child-focused treatment consisting of an 8-week, daily summer school treatment program. In addition, a school-based treatment program was carried out involving 10 to 16 teacher behavior management consultation sessions, 12 weeks of a part-time paraprofessional aide working with the child, and daily home/school report cards.

For the core symptoms of ADHD (inattention, impulsivity, hyperactivity) the MTA intensive MedMgt was superior to routine CC, in spite of the fact that more than two-thirds of the children receiving CC were being treated with stimulant medication. It was determined that children in the MedMgt group were on significantly higher doses of stimulant medication, as after a month of blind trials of different doses, they were assigned to the dose that led to the optimal symptom control without limiting side effects.

The Comb treatment did not yield significantly greater benefits than the MedMgt alone for the core symptoms of ADHD, but outcomes in the Comb group could sometimes be achieved at lower medication doses than the group that received only MedMgt alone. Combined treatment was superior to MedMgt alone for many non-ADHD domains of functioning, including oppositional or aggressive symptoms, symptoms of depression and anxiety, parent-child relations, and reading achievement. Based on a composite of parent and teacher ratings, 68% of the Comb children "normalized" by the end of the 14-month study, compared with 56% of the MedMgt alone, 34% of the Beh management alone, and only 25% of the CC children. The benefit seen in the medication groups persisted at 24 months but not at 36 months, regardless of original treatment assignment.[46] Factors associated with worse outcomes at 36 months were initial symptom severity, male gender, oppositional/disruptive behaviors, public assistance, and ADHD in a parent.

Results of the MTA study are limited by the absence of a control group and the use of short-acting stimulant mediations (long-acting formulations were not available). For clinicians, an important implication of the study results is that systematic monitoring with a plan for follow-up office visits is a critical component of successful treatment for children with ADHD.

School Interventions

Comprehensive pediatric care for children and adolescents with ADHD is achieved when the primary pediatric health care professional communicates effectively with the school. With the parents' approval, it begins with informing the teacher about the diagnosis and treatment. A monitoring system for transferring information to the clinical office about the patient's school behavior and educational achievement should be in place. Communication works best when parents partner with the school and the medical home.

Federal laws support school interventions for children with ADHD. For mild cases, Section 504 of the Rehabilitation Act requires that a school provide classroom accommodations to improve learning. These accommodations can include preferential seating; reduced assignments and homework; assisting the teacher in formulating a classroom behavioral program; and providing an "ADHD coach" to assist students in organizing, planning, homework completion, homework flow, and getting assignments submitted in a timely manner. For more severe cases, the IDEA recognizes ADHD under the "Other Health Impairment" category and requires schools to provide comprehensive educational testing followed by an Individualized Education Program (IEP) plan with measurable behavioral and academic achievement goals. Periodic monitoring and reassessment is incorporated into the IEP.

Daily home-school report cards are an effective method to monitor classroom behaviors. Parents and teachers decide on 3 to 5 behaviors that impair success in school. Each behavior is monitored daily and a daily home-school report card is sent home with the child. The daily report card is attached to an award system (eg, privileges or prizes) to encourage compliance. This system allows for frequent, immediate feedback that can be motivating to the child, parents, and teacher. An example of a daily report card with directions can be downloaded (http://ccf.buffalo.edu/pdf/daily_report_card.pdf).

Complementary and Alternative Therapies

Many parents seek complementary or alternative therapies either alone or in combination with evidenced-based medication and behavioral management.[60] Most of these treatments have not undergone randomized, controlled trials and cannot be recommended (see Chapter 21). Some alternative therapies have side effects and may be harmful to children; others are safe. For the parent who is set on trying an alternative treatment or who has already found it helpful, many clinicians incorporate the alternative plan with the more evidence-based proven treatment. If the treatment is not dangerous and does not interfere with other aspects of the primary pediatric health care professional's and parents' collaborative management strategies within the medical home, it is best not to alienate a parent, as the goal should be one of partnership in doing what is best for the child.

Conclusion

Attention-deficit/hyperactivity disorder is a common, chronic neurobehavioral condition associated with a variety of functional impairments in educational achievement and social development. The diagnosis of ADHD in children and adolescents can be established with confidence by primary pediatric health care professionals with the use of the AAP evidenced-based guideline. Inclusion of screening procedures for coexisting psychological conditions, learning disabilities, and psychosocial stressors is an important component of the diagnostic process. Effective treatment strategies include parent/patient education about ADHD, FDA-approved medications, and behavior management techniques. Ongoing communication with teachers; a system of communication with the medical home that parents can access with ease; a care plan that systematically empowers parents, children, and adolescents; and a plan for follow-up office visits ensure effective monitoring of a treatment plan for a child or adolescent with ADHD. This approach promotes the best opportunity to ameliorate or prevent academic, social, and occupational sequelae associated with ADHD.

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Chapter 17

Disruptive Behavior Disorders

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Definition of Disruptive Behavior Disorders and Scope of the Problem

The term *disruptive behavior disorder* describes "socially disruptive behavior that is generally more disturbing to others than to the person initiating the behavior."[1] Disruptive and oppositional behaviors occur on a continuum, with normal toddler resistance and tantrums at one end and more severe, maladaptive behaviors warranting a medical diagnosis at the other end.[2] While it is very important for primary pediatric health care professionals to screen for and address any challenges related to disruptive behaviors, the focus of this chapter will primarily be on those behaviors significant enough to constitute a medical diagnosis. In the *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, Text Revision (DSM-IV-TR),* the diagnoses included under the category of disruptive behavior disorders are oppositional defiant disorder (ODD), disruptive behavior disorder not otherwise specified, and conduct disorder (CD).[3] Although attention-deficit/hyperactivity disorder (ADHD) is often also included in this category, it is discussed in detail in Chapter 16.

Oppositional Defiant Disorder and Disruptive Behavior Disorder Not Otherwise Specified

The *DSM-IV-TR* states that a child who meets criteria for a diagnosis of ODD must have "a pattern of negativistic, hostile, and defiant behavior lasting at least 6 months."[3] In addition, the child must display 4 or more defiant behaviors, which are listed in Box 17.1. The child's age and developmental level must be taken into consideration, and the disturbance in behavior must cause clinically significant impairment in academic, social, or occupational functioning. A child is precluded from being diagnosed with ODD if the behaviors occur exclusively during the course of a mood or psychotic disorder, or if the child also meets criteria for either CD or antisocial personality disorder.[3]

The diagnosis of disruptive behavior disorder not otherwise specified describes conduct or oppositional defiant behaviors that do not meet criteria for ODD or CD but do cause clinically significant impairment.[3] Most published research addresses the diagnoses of ODD or CD, rather than disruptive behavior disorder not otherwise specified. Nevertheless, much of the information applicable for ODD or CD can be useful in caring for a child who is diagnosed with disruptive behavior disorder not otherwise specified. Therefore, this chapter will primarily focus on ODD and CD.

Conduct Disorder

The *DSM-IV-TR* describes the diagnosis of CD as "a repetitive and persistent pattern of behavior in which the basic rights of others or major age-appropriate societal norms or rules are violated."[3] Children with CD must meet criteria in at least 3 or more of the following domains: aggression to people and animals, destruction of property, deceitfulness or theft, and serious violations of rules (Box 17.1). The behaviors must result in clinically significant impairment in academic, social, or occupational functioning, and criteria cannot be met for antisocial personality disorder.[3]

In the *DSM-IV-TR,* the diagnosis of CD is coded based on age at onset. An individual with "CD, childhood-onset type" is described as having the presence of at least one criterion characteristic of CD prior to 10 years of age. An individual with "CD, adolescent-onset type" is described as having the absence of any criterion characteristics of CD prior to 10 years of age. An individual with "CD, unspecified onset" has an unknown age of onset.[3]

Epidemiology of Disruptive Behavior Disorders

It may not be possible to determine the prevalence of disruptive behaviors since many do not cause sufficient impairment to warrant a medical diagnosis. However, noncompliant and oppositional behaviors comprise some of the most common concerns among parents and are the most frequently reported behavior problems seen by primary care pediatricians.[4] Overall, about 5% of children between the ages of 6 to 18 years meet *DSM-IV* criteria for either ODD or CD at any given time.[5] The lifetime prevalence rates of each are slightly higher, ranging from 9% to 13% for ODD and 3% to 16% for CD.[6,7] The diagnosis of ODD or CD is more common in boys, although oppositional behavior per se may be equally common between both genders.[8]

Assessment of Children With Disruptive Behaviors

Primary pediatric health care professionals care for children with varying levels of disruptive behaviors. Therefore, it is important for clinicians to evaluate the child's behaviors and parent responses to determine if the parent-child interaction may be inadvertently perpetuating the child's negative behaviors. Parents often place demands on their children, such as "Clean up your toys," and some of these

Box 17.1. *DSM-IV-TR* Criteria for the Diagnoses of ODD and CD[a]

ODD

Must have ≥4 of the following present:

1. Often loses temper
2. Often argues with adults
3. Often actively defies or refuses to comply with adults' requests or rules
4. Often deliberately annoys people
5. Often blames others for his/her mistakes or misbehavior
6. Often touchy or easily annoyed by others
7. Often angry and resentful
8. Often spiteful or vindictive

CD

Must have presence of ≥3 of the following criteria in the past 12 months, with at least one criterion present in the past 6 months:

Aggression to people and animals

1. Often bullies, threatens, or intimidates others
2. Often initiates physical fights
3. Has used a weapon that can cause serious physical harm to others
4. Has been physically cruel to people
5. Has been physically cruel to animals
6. Has stolen while confronting a victim
7. Has forced someone into sexual activity

Destruction of property

8. Has deliberately engaged in fire-setting with the intention of causing serious harm
9. Has deliberately destroyed others' property

Deceitfulness or theft

10. Has broken into someone else's house, building, or car
11. Often lies to obtain goods or favors or to avoid obligations
12. Has stolen items of nontrivial value without confronting a victim

Serious violations of rules

13. Often stays out at night despite parental prohibitions, beginning before age 13 years
14. Has run away from home overnight at least twice while living in parental or parental surrogate home
15. Often truant from school, beginning before age 13 years

Abbreviations: CD, conduct disorder; *DSM-IV-TR, Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, Text Revision;* ODD, oppositional defiant disorder.

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demands may seem "aversive" to the child. The child may respond to this "aversive" event by displaying a "coercive" response, such as yelling or having a tantrum. Parents may respond to the "coercive" response by removing the original "aversive" demand, and this negatively reinforces the child's noncompliant behavior. The child is likely to continue to yell or tantrum when future demands are placed because he has learned that he gets his way by doing so. Another response the parent could make to the yelling or tantrum is to try to comfort the child and explain the reason for the request. This parental attention positively reinforces the child's noncompliant behavior, and thus the child may continue to manifest these "coercive" responses because he has learned that he will get attention for them. Alternatively, the parent may respond to the child's "coercive" response by presenting another "aversive" event, such as yelling or loudly repeating the request. The child may ultimately respond to the parent's demand, but the child learns to only respond to the parent's more "aversive" demands, such as yelling or stating commands loudly. If any

of these parent-child interactions are continually used, a persistent and worsening pattern of noncompliance and defiance may develop.[9] When the primary pediatric health care professional recognizes that the parent-child interaction is heading in a negative direction, he or she can play a crucial role in providing guidance and offering effective techniques that the parent can use. A more effective parent response to a child's protests may include a simple and single repetition of the request, followed by verbal acknowledgment that complying with the request may be difficult from the child's perspective, and firm limit setting on worsening behavior (see Chapter 4).

In addition to intervening when problem behaviors are described, the primary pediatric health care professional should evaluate the disruptive behaviors to determine if a medical diagnosis such as ODD or CD should be made. While specific antisocial acts occur in up to 80% of youths,[10] children who meet criteria for a diagnosis of ODD or CD display a persistent history of multiple problem behaviors.

Important Points to Consider When Taking the History

Although children may present to the primary pediatric health care professional with any number of disruptive behaviors, the most common referral symptoms for disruptive behavior disorders are fighting, stealing, lying, cruelty, fire-setting, substance abuse, and sexual misconduct.[1] When taking the history, it is important to obtain information from several sources, including children, parents, and teachers. The primary pediatric health care professional should learn as much as possible about the parenting style, parent-child interactions, and the child's strengths, as this information can be valuable for treatment planning and implementation of interventions.[5] The child's age and gender must be taken into account. At different ages, children display different types of disruptive behaviors, with property and status offenses more prevalent at older ages.[8] Males are more likely to use physical attack and females are more likely to use indirect, verbal, and relational violence.[11]

Important Questions to Ask in the History

In assessing a child with disruptive behaviors, it is helpful to use the criteria described in the *DSM-IV-TR* for the diagnoses of ODD and CD to guide the history. For example, in a child who displays aggression, it is important to determine what type of aggression the child shows, such as verbal, physical, etc, and to whom the aggression is directed, such as parents, other children, animals, etc. The clinician should ask how long these behaviors have been manifest and if there was any significant change in the child's life prior to the onset of the disruptive behaviors. It is also important to ask parents, teachers, and other caregiving adults how disruptive these behaviors are. Individuals with CD are often unable to appreciate other's welfare and have little or no remorse about harming others.[12] Therefore, they may not be able to understand and report how their behaviors are negatively impacting others.

Primary pediatric health care professionals may find the following brief questions helpful in determining whether criteria for the diagnosis of ODD are likely to be

met. Research has shown that a positive response for all 3 is 91% specific for meeting *DSM-IV* criteria on full interview and a negative response is 94% sensitive for ruling out ODD.[13,14]

1. Has your child in the past 3 months been spiteful or vindictive, or blamed others for his or her own mistakes? (Any "yes" is a positive response.)
2. How often is your child touchy or easily annoyed, and how often has your child lost his temper, argued with adults, or defied or refused adults' requests? (≥2 times weekly is a positive response.)
3. How often has your child been angry and resentful or deliberately annoying to others? (≥4 times weekly is a positive response.)

It is important for primary pediatric health care professionals to be sure to perform a thorough assessment of the child's psychosocial functioning to assess for comorbidities or alternate diagnoses. Areas to assess include attention, level of activity and impulsiveness, social interactions, and communication skills. Other factors to consider when evaluating a child with disruptive behaviors include anxiety, mood disorders, cognitive and/or learning problems, substance abuse, and history of physical or sexual abuse.

Supplements to Clinical Interview

Standardized questionnaires can aid the primary pediatric health care professional in evaluating disruptive behavior. Two commonly used rating scales for ADHD: the Conners Rating Scales-Revised and the Vanderbilt AD/HD Diagnostic Parent and Teacher Rating Scales, include information about oppositional and disruptive behaviors, and thus can be useful.[15] In addition, more broad-based behavioral rating scales, such as the Eyberg Child Behavior Inventory,[16] a parent rating scale, and the Child Behavior Checklist,[17] a child rating scale, can serve as an adjunct to taking a clinical history. Finally, the Modified Overt Aggression Scale[18] is another useful supplement to the clinical interview in evaluating disruptive behavior.

The diagnoses of ODD and CD are made clinically. Outside of research purposes or a medical history and physical examination indicating abnormal neurologic status, neuroimaging is not recommended. No laboratory work is routinely performed. If active substance abuse is suspected, a urine drug screen should be considered. In addition, if sexual abuse or unprotected sexual activity is present, testing for sexually transmitted infections may be warranted.[5]

Risk Factors for the Development of Disruptive Behavior Disorders

The many known risk factors for the development of disruptive behavior disorders can be classified into the following categories: biological, individual, family, and social/school (Box 17.2).[5] Rather than one risk factor acting in isolation, it seems that the accumulation of risk factors may be critical to the development of disruptive

Box 17.2. Risk Factors for the Development of Disruptive Behavior Disorders

Biological ■ Genetic ■ Antenatal and perinatal complications ■ Brain injury, brain disease ■ Male sex ■ Environmental toxins, such as lead	**Individual** ■ Cognitive impairment ■ Difficult temperament ■ Aggressiveness ■ Hyperactivity, impulsivity ■ Attention problems ■ Language impairment ■ Reading problems
Family ■ Single parent or divorce ■ Domestic violence ■ Lack of permanent family ■ Parental substance abuse or antisocial behavior ■ Child maltreatment or neglect ■ Parent-child conflict ■ Excessive parental control ■ Lack of parental supervision ■ Maternal depression or anxiety	**Social/School** ■ Low socioeconomic status ■ Rejection by peers ■ Association with deviant peers ■ History of being bullied ■ Neighborhood violence ■ Disorganized or dysfunctional school ■ Intense exposure to violence via media

behavior disorders.[19] Disruptive behavior disorders most likely have a multifactorial etiology, including some degree of genetic vulnerability and environmental and/or social contributors.[20] Overall, some of the most commonly cited risk factors include low socioeconomic status; history of rejection or abuse; and parental challenges, including antisocial behaviors, substance abuse, and dysfunctional parenting.

One interesting study evaluated the effect of poverty on the prevalence of oppositional and conduct disorders. In this study, 9- to 13-year-old rural children, of whom one-quarter were Native American and the remaining were predominantly white, were given annual psychiatric examinations for 8 years. Halfway through the study, a casino opened up on the reservation, which gave every Native American family an income supplement, moving 14% of study families out of poverty. The non-Native American families were unaffected. Reducing poverty among Native American families resulted in a reduction of oppositional and conduct problems in their children. The mechanisms by which this occurred seemed to be related to fewer demands on parents' time, fewer single parents, and better parental supervision, rather than purely related to income increase.[21]

While many risk factors are well known, the anatomical basis for disruptive behavior disorders it less well established. However, neuroimaging studies have found the frontal lobe to be associated with violence and aggression.[19] In addition, atypical frontal lobe functioning, as detected via electroencephalogram, has been suggested

as the basis for the negative affective style displayed in children with ODD.[22] Neurotransmitters have also been investigated to ascertain what role they play in relation to disruptive behaviors. Serotonin has been linked to aggression, though research remains ongoing to further elucidate its precise relationship to ODD and CD.[19]

Comorbidities of Disruptive Behavior Disorders

Disruptive behavior disorders are often comorbid with other conditions. The most frequently reported comorbid conditions are ADHD, major depression, and substance disorders. Attention-deficit/hyperactivity disorder occurs 10 times more frequently in children with disruptive behavior disorders, and major depression occurs 7 times more frequently, compared with children who do not have a diagnosis of a disruptive behavior disorder.[13] A child between the ages of 11 to 14 years with a diagnosis of CD has a 4-times increased likelihood of substance disorders by age 18 years.[23] Many other psychiatric and developmental disorders are more prevalent in children with disruptive behavior disorders compared with the general population. These include mood disorders and anxiety disorders,[7] as well as learning and cognitive disorders.[24] Given the high number of comorbid conditions associated with disruptive behavior disorders, it is important for primary pediatric health care professionals to routinely assess attention, mood, substance use, and school functioning in children and adolescents presenting with disruptive behaviors.

Treatment

Children who meet criteria for the diagnoses of ODD or CD generally require the care of a psychiatrist or psychologist who can work in conjunction with the primary pediatric health care professional.[1] Disruptive behavior disorders tend to be chronic conditions, and this should be taken into account when planning treatment. Treatments are more effective if initiated early. A structured psychosocial intervention should be the first line of treatment for ODD and CD. Treatments should involve the parents and, in most cases, core goals of treatment include improving parenting skills and parent-child interactions.[5]

Psychosocial Treatments

Psychosocial treatments have the best evidence for treating disruptive behaviors. Evidence-based psychosocial treatments include parent management training, multimodal interventions (such as multisystemic therapy), and individual interventions (such as cognitive behavior therapy), each of which will be described in this chapter (also see Chapter 19).

Parent management training is an effective and well-studied intervention.[25] The theory behind this intervention is that the disruptive behavior, at least in part, is due to a maladaptive parent-child interaction. Thus a parent is taught a new set of skills to use with the child, including ways to reward prosocial behaviors and ways to

combat noncompliant behaviors.[24] A challenge with parent management training is that it requires a committed parent, and it may not be successful with more dysfunctional families.[5]

Multimodal interventions include a more comprehensive system for change. While parent management training does not necessarily enhance the child's ability to make friends or perform well in school, multimodal interventions may include enabling the child to improve his problem-solving skills and working with the child's teacher to improve the child's educational functioning.[5] Some types of multimodal programs are multisystemic therapy, which targets adolescents with severe CD, and Families and Schools Together, which targets children with CD who are starting school.[5] While programs such as wilderness camps and boot camps may sometimes be considered to provide multimodal interventions, there is no solid evidence for their effectiveness. The main concern with these programs is that the skills children develop in these isolated settings will not generalize, and these settings enable antisocial peers to congregate together.[5]

Individual interventions involve providing children with problem-solving skills training. Children are taught to find adaptive solutions to their problems through the use of teaching in areas such as anger coping and social skills. Techniques such as role playing, structured activities, modeling behavior, stories, and the use of games facilitate teaching. In some cases, modified cognitive-behavioral therapy and day treatment may be part of the intervention.[6] Individual interventions are most effective when used in conjunction with a broader treatment program that also addresses parenting, social, and school interventions.[19]

Pharmacotherapy

Pharmacotherapy should never be the primary treatment for disruptive behavior disorders, although medication might be considered in children with comorbid conditions, such as ADHD, anxiety, or depression.[5,19] When using stimulants in children with ADHD and disruptive behavior disorders, careful monitoring and supervision should be implemented, given the high prevalence of substance use and abuse in this population.[26]

Prognosis

Although ODD and CD seem to be closely related, their relationship is complex. Some children presenting with a diagnosis of ODD in childhood ultimately evolve into a diagnosis of CD, often after puberty. Other children may exhibit disruptive behaviors for a short time and/or will continue to carry a diagnosis of ODD, though it will not evolve into CD. Boys with ODD are more likely to later be diagnosed with CD compared with girls.[27] Some children with CD will go on to develop a profile consistent with the diagnosis of antisocial personality disorder. In fact, the *DSM-IV-TR* criteria for antisocial personality disorder requires evidence of CD prior to the

age of 15 years.[3] However, most children diagnosed with CD will not develop antisocial personality disorder in adulthood.[5]

Predictors of Outcome

In general, studies have shown that the higher the number of disruptive behaviors a child exhibits, the worse the long-term outcome.[5] Aggressive, antisocial symptoms; fire-setting; family dysfunction; and substance abuse are associated with a poor prognosis.[1] Among children who have ODD, those with temperamental traits of oppositionality have a poorer prognosis compared with children in whom the oppositional behavior occurred as an outcome of an acute event.[1] Adolescents with severe externalizing behaviors are more likely to leave school early and report global adversity throughout life, compared with those with few or no externalizing behaviors.[28] It is hard to determine if their experiences in adolescence (eg, getting into trouble, turbulent family relationships) lead to more difficulties in life, or if the adult difficulties are primarily due to the underlying disruptive behaviors.

Conclusion

Disruptive behaviors and their associated diagnoses of ODD and CD are a common reason that parents seek medical care from a primary pediatric health care professional. Therefore, it is helpful to understand the risk factors, assessment, and treatment involved in caring for children with disruptive behaviors. While disruptive behavior disorders are often chronic, early identification and intervention of problem behaviors can often improve the functioning for affected children and families.

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Chapter 18

Anxiety and Mood Disorders

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Busy primary pediatric health care professionals are often the first providers approached by families to assess mood, anxiety, and behavioral difficulties in their children and adolescents, yet it is challenging to implement time-efficient methods of evaluation and treatment in busy clinical settings. Barriers such as time constraints, lack of mental health care referral resources, lack of reimbursement for behavioral health care, and lack of training in the identification of psychosocial issues exist at the primary health care provider level, precluding appropriate mental health screening and treatment.[1] However, a basic understanding of methods for assessment and treatment of anxiety and mood disorders in children is essential for all primary pediatric health care professionals, as psychiatric symptoms are common and can occur at any time. Rates of psychosocial disorders are as high as 13% in preschoolers and 27% in children and adolescents from 4 to 16 years of age, but given the barriers to mental health screening, only about 12% to 15% of children with these disorders are identified.[2] A recent longitudinal 4-year community study of children from 9 to 13 years of age growing up in the 1990s found that 13.3% of these children met *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV)* criteria for a mental disorder at any time, and 36.7% of these children met criteria for at least one disorder within the 4-year study, with 4.6% of these meeting criteria for anxiety and mood disorders.[3] Comorbidity can also be common in pediatric populations, with 11% to 69% of anxious youth having an additional depressive disorder and 15% to 75% of depressed youth having an additional anxiety disorder.[4]

When a parent or caregiver presents to a pediatric health care setting with a behavioral or mood concern about their child, the role of the primary pediatric health care professional is to rule out anxiety or mood problems as contributing to the complaint. It is well established that these disorders can drastically impair a child's functioning, can lead to significant morbidity and mortality and, when left untreated, can contribute to an increased risk of future anxiety, depression, illicit drug use, somatic complaints, educational underachievement, adolescent and adult violence and crime, suicide attempts, and psychiatric hospitalization.[2,5] Approximately

50% to 75% of children who commit suicide have had a mood disorder (with as many as 95% having had some mental disorder), yet less than half of these youth received psychiatric care.[6] The associated morbidity of these disorders emphasizes the importance of early identification and treatment, and primary pediatric health care professionals are key in the timely recognition of these concerns.

There is now evidence that both genetic vulnerabilities and adverse life events can lead to anxiety and depression. There is also evidence that resilience to adverse life events may be determined by both genetic vulnerabilities and the presence or absence of a supportive adult figure.[7] The presenting histories can be difficult to understand, as information related to undiagnosed family psychiatric disorders and psychosocial issues (eg, violence in the home or community, a family history of suicide, parental discord or substance use, and bullying or teasing at school) are often not volunteered.[8] Although no single cause of anxiety or depression has been identified, interactions among genetic, environmental, and psychosocial factors play a role. There is evidence that extreme environmental stressors can contribute to neurobiological changes that pose an increased susceptibility for anxiety and depression.[9] It is very important for the primary pediatric health care professional to convey that these biological brain-based disorders are very treatable, there is trust and confidence in the mental health professional who is scheduled to see the child, and evidence-based studies are demonstrating what types of therapy are best for certain disorders. Families may be much more likely to follow up on treatment if the disorders are explained as biological disorders involving the brain, and that just like diabetes, hypertension, or an infection, treatment is needed for the most rapid improvement.

Tips for Early Identification

It is essential that mood or anxiety symptoms be examined within a context of overall impairment, as some symptoms are transient and/or in response to an acute stressor that might resolve without intervention. However, symptoms of anxiety and depression cannot be taken lightly, as for some children, these symptoms are not transient and can represent a significant disorder with associated impairment and risk for morbidity and mortality. Symptoms can be a result of adjustment to negative environmental, medical, or life situations. Parents are often reluctant to raise concerns about psychosocial issues, or they often do not recognize the significance of their child's distress. The primary pediatric health care professional must depend on the history as presented by the child and parent. Family dysfunction and parental psychopathology can influence the presentation (and reliability) of the parent's report of the symptoms. Unfortunately, not all families volunteer some of the most key historical or family history facts that have contributed to the distress of the child. Input from other pertinent people in the child's life, such as teachers, can be extremely useful if parents provide permission to contact such individuals.

There are known risk factors that may need to be considered when thinking about adjustment issues. For example, children with chronic disease are about twice as likely to have a psychosocial disorder (those suffering from epilepsy are at highest risk).[10,11] These issues are especially important for the primary pediatric health care professional, as poor psychosocial functioning has a negative impact on medical adherence and is associated with higher utilization of primary health care services.[12] In addition, children often present with symptoms that may initially seem to meet criteria for attention-deficit/hyperactivity disorder (ADHD), but these symptoms are actually secondary to another mental health disorder, given the overlap in diagnostic criteria between ADHD and anxiety, mood, and other psychiatric disorders.[13]

Tips for Surveillance and Screening

A family, child, or adolescent will typically not share their concerns if their primary pediatric health care professional does not convey an accepting and interested attitude. Caregivers can accurately report on behaviors that are observed, but the child or adolescent is best qualified to report internalizing concerns, such as mood or anxiety symptoms. After screening has targeted a child's concern, a private conversation with the pediatric patient alone may be needed. Standardized psychosocial or psychiatric screening is supported by the American Academy of Pediatrics (AAP) Task Force on Mental Health (TFOMH) and enables primary care clinicians to identify problems more quickly and efficiently as well as to provide treatment or a referral for treatment at an earlier, more effective time. Screening tests are not designed for the purpose of making a diagnosis or labeling a child, but they assist in determining whether there is a problem present that needs to be more formally addressed.[14] Although research has indicated high levels of psychosocial disorders in children and adolescents, there is ongoing concern about underdiagnosis of these problems in pediatric primary care offices. A large study demonstrated that it is rare for primary care clinicians to use checklists to help with the identification of psychosocial concerns, more than 50% never use tools for screening, and only 20% use screens after a primary behavioral or mood disorder is recognized.[15] However, when used correctly, standardized psychosocial screening tools can add to efficient screening and improve time management. A concerning or failed behavioral screen needs to be followed by a thorough diagnostic assessment, which can help to determine the diagnosis and lead to the implementation of appropriate treatment using well-established clinical standards and evidence-based treatment protocols.

Tips for Use of General Screening Instruments

The Pediatric Symptom Checklist (PSC),[16] a 35-question, parent-reported checklist of psychosocial symptoms, and the shorter PSC-17[17] have potential to be a means of reducing the number of undiagnosed children with psychosocial disorders. Both scales have been designed for use in the primary health care setting and consist of subscales to measure internalizing, externalizing, and attention behaviors. It is recommended that such scales be used as part of routine health supervision visits,

but Borowsky et al[2] found that patients screened during a nonroutine visit (eg, for illness or injury) were more likely to score higher on the PSC-17. This suggests that this screen should be considered for use during acute care visits as well. The AAP TFOMH recommends that mental health surveillance questions be asked at all health care visits (eg, "How is school going?" or "How does he/she sleep?"). However, the use of screening questionnaires is not practical in most short acute visits. If the responses to surveillance questions indicate a potential problem, or if the physical problem could have a mental health basis, then asking the parents to complete a screen is appropriate either at the same visit or a future follow-up visit.

Due to the high rates of psychosocial problems and the developmental consequences of missing relevant diagnoses, surveillance for mental health disorders should be completed at all pediatric primary care visits.[2,18] Use of the PSC instrument takes only a few minutes for a parent to complete. Specific questions can be used in brief interviews as well (ie, question 17 on the PSC 35—"Seems to be having less fun"—is a useful screen for anhedonia associated with depression). In addition, 2 questions added to the PSC that are not included in the score are useful in identifying parental concerns: "Does your child have any emotional or behavioral problems for which he/she needs help?" and "Is your child currently seeing a mental health counselor?" These questions were designed to measure parental/emotional concern and current involvement in counseling, respectively,[1] and have been found to be useful in identifying parent concerns.

Given that the impact on overall functioning is an important factor in determining the need for intervention, brief questions about a child's self-esteem can be useful. There are multiple useful screening tools contained in *Bright Futures: Mental Health* that was published in 2005, and the *Bright Futures Mental Health Tool Kit.*[19] By having the parent or primary caregiver answer simple questions from the *Bright Futures Mental Health Tool Kit* (eg, "What does your child think he does well?"), the physician or caregiver can gain insight into the child's perceived strengths. Questioning how the child responds to failure and new challenges allows a parent to assess his or her independence and resilience. It is important for children to know that they have the ability to make choices that will affect the outcome of events in their lives.[8]

General Behavior/Emotional Screens (see Table 18.1)

1. The Achenbach Child Behavior Checklist (CBCL) is an instrument used to gain a caregiver's perception about their child's competencies and behavior problems.[20] The CBCL measures the magnitude of social withdrawal, somatic complaints, anxiety/depression, social problems, thought problems, attention problems, delinquent behavior, and aggressive behavior. In addition to the individual score for each symptom, the CBCL allows for total scores of externalizing and internalizing problems. Scores from the CBCL are designed to be comparable with the Teacher Report Form (TRF) and Youth Self-Report Form (YSR ages 11–18).[20]

Table 18.1. Screening Tools for the Identification of General Behavior/ Emotional Functioning, Anxiety Disorders, and Mood Disorders

Area of Screening	Scale	Description
General behavior/ emotional functioning	Achenbach Child Behavior Checklist (CBCL)	• Gains caregiver's perception about child's competencies and behavior problems • Measures magnitude of social withdrawal, somatic complaints, anxiety/depression, social problems, thought problems, attention problems, delinquent behavior, and aggressive behavior • Yields individual scores for each symptom and total scores of externalizing and internalizing problems • Compares with the Teacher Report Form (TRF) and Youth Self-Report Form (YSR)
	The Behavior Assessment System for Children (BASC)	• Evaluates personality characteristics, behavior, emotional development, and self-perceptions of children and adolescents ages 4–18 • Allows clinicians to see how a child places on dimensions of behaviors associated with diagnostic criteria • Indicates the presence and magnitude of *Diagnostic and Statistical Manual (DSM)* symptoms for different disorders, including anxiety and depression
	NICHQ Vanderbilt Assessment Scale	• Reveals symptom and severity information for attention-deficit/hyperactivity disorder, anxiety/depression, conduct disorder, and oppositional defiant disorder • Parent and teacher scales used in conjunction have high sensitivity and specificity
	Child Health & Development Interactive System (CHADIS)	• Web-based program screening for health, emotional, and behavioral issues prior to primary care visits • Assessment tools for children ages 0–3, 4+ and adolescents • Parental questionnaires to assist care providers in addressing issues of child behavior and development • Avoids interference with routine visits
Anxiety disorders	Revised Children's Manifest Anxiety Scales (RCMAS)	• Assesses the level and nature of anxiety in children ages 6–19 • 37 yes/no questions divided into 4 scales • Successful in distinguishing youth with anxiety disorders from youth without
	Screen for Child Anxiety Related Emotional Disorders (SCARED)	• Correctly differentiates between anxiety disorder subtypes and anxiety disorders and disruptive disorders • 41-item parent and child symptom inventory using a 3-point scale to assess for *DSM-IV* anxiety disorders
	Multi-dimensional Anxiety Scale for Children (MASC)	• 39-item self-report scale for children and adolescents ages 8–19 • Subscales distinguish between physical symptoms, social anxiety, harm avoidance, and separation anxiety • Has the strongest supportive evidence for use as an anxiety screening tool

Table 18.1. Screening Tools for the Identification of General Behavior/ Emotional Functioning, Anxiety Disorders, and Mood Disorders *(continued)*

Area of Screening	Scale	Description
Mood disorders	Children's Depression Inventory (CDI)	• Assesses cognitive, affective, and behavioral signs of depression in children ages 6–17 • Designed for use in school setting, pediatric practice, child guidance clinics, and psychiatric settings • 27 items reveal total scores and scores for each individual scale: negative mood, interpersonal difficulties, negative self-esteem, ineffectiveness, and anhedonia • Only takes 5–10 minutes to complete and written at a first-grade reading level
	Beck Depression Inventory (BDI)	• Clinician-administered measure that monitors intensity, depth, and severity of depression in psychiatric patients • Also identifies depressive symptoms in a primary care setting • Short version is composed of only 7 questions (as opposed to 21) • Scoring varies between patients that have been diagnosed with depression and those in primary care settings
	Mood and Feelings Questionnaire (MFQ)	• Quick screening measure to determine extent of depressive symptoms • Items heavily weighted toward affective and cognitive components of depression • Separate child (13 questions) and parent (11 questions) versions • When administered consistently throughout treatment, it can be used to progress monitor depressive symptoms
	Center for Epidemiologic Studies Depression Scale for Children (CES-DC)	• 20-item self-report depression symptom inventory
	Children's Depression Rating Scale, Revised (CDRS-R)	• Brief rating scale (15- to 20-minute administration) based on a semi-structured interview with the child (or an adult informant) • Designed for 6- to 12-year-olds and has been used successfully with adolescents • Can be used in clinical, school, and pediatric settings Has been used in National Institute of Mental Health and other medication trials

2. The Behavior Assessment System for Children (BASC) is designed to evaluate the personality characteristics, behavior, emotional development, and self-perceptions of children and adolescents from 4 to 18 years of age. The BASC allows clinicians to see how a child places on dimensions of behaviors associated with diagnostic criteria.[21] Scores on the BASC indicate the presence and magnitude

of *DSM* symptoms for different disorders (ie, depression and anxiety). There are teacher and self (ages 8–18 years) versions of the BASC designed to aid in the comparison of collected information.

3. Parent and teacher versions of the National Initiative for Children's Healthcare Quality (NICHQ) Vanderbilt Assessment Scale are available that, when used in conjunction, have high sensitivity and specificity.[22,23] These scales reveal symptom and severity information for ADHD, anxiety/depression, conduct disorder, and oppositional defiant disorder.

Anxiety Disorders

Anxiety disorders are among the most common conditions in children and adolescents, and depending on the type of anxiety, affect between 2.8% to 27% of youth.[3,24,25] The subtypes of the different *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, Text Revision (DSM-IV-TR)* anxiety disorders have several features in common and some very different associated symptomatology. The following presents an overview of assessment and basic treatment guidelines. Additional reading and study is recommended for the professional frequently addressing these disorders. This chapter incorporates assessment and evidence-based treatments, when available, and otherwise presents well-accepted standards for good clinical care.

Epidemiology and Risk Factors

Anxiety disorder diagnosis rates in preadolescents range from 2.6% to 41.2%, and lifetime rates of anxiety have been reported to range from 8.3% to 27.0%.[25] Anxiety has been reported to be more prevalent than depression and conduct or behavioral disorders in young children.[25] Further, even with the most conservative estimates for anxiety prevalence, at least 1 child in the average elementary school class of 30 children will have an anxiety disorder.[25] Research suggests that these disorders are not likely to go away as the child ages and are often associated with other problems later, such as an increased risk of depression or substance abuse.[26] There is evidence of a bidirectional continuity from anxiety to depression and back to anxiety,[3] and symptoms of both disorders may overlap.

In preschool children, anxiety is considered from the perspective of developmental norms, since stages of fear and anxiety are expected as a child grows. However, approximately 15% of young children demonstrate excessive or persistent fear or anxiety, putting them at risk for later anxiety disorders. Research in preschoolers has indicated no significant gender bias, although age may play a role, as 4- to 5-year-olds are more likely than 2- to 3-year-olds to have an anxiety disorder.[25]

Definition and Core Characteristics

Anxiety disorders are often associated with psychosocial impairments, such as poor social relationships, decrease in academic performance, and low self-esteem.[27] If left untreated, these disorders may continue throughout adulthood. In general with all

anxiety disorders, the focus of the worry may shift, but the inability to control the intrusive thoughts about the worry persist. Children with anxiety disorders can often appear to have difficulty concentrating or processing information because they cannot get their mind off of their worries, which must be differentiated from ADHD, primarily inattentive subtype. Some of these youth need constant reassurance and others may be overly perfectionistic or critical of themselves or others.

The categories of anxiety disorders in the *DSM-IV-TR*[28] are generally based on adult criteria, except for separation anxiety disorder and selective mutism. Symptoms are considered to meet criteria for an anxiety disorder if they are severe enough to adversely affect social or occupational functioning. The *Classification of Child and Adolescent Mental Diagnoses in Primary Care: Diagnostic and Statistical Manual for Primary Care (DSM-PC) Child and Adolescent Version* is designed to assist primary care providers in determining the spectrum from normal variations to disordered behavior. This classification system can help the pediatric health care professional determine severity of symptoms by outlining differences between a normal developmental variation (anxious variation), anxiety problem, and anxiety disorder.[29] Hallmarks of these disorders are described below and in Table 18.2.

Generalized Anxiety Disorder (300.02)

Generalized anxiety disorder (GAD) was introduced in the *DSM-IV,* replacing the *DSM-III–R* diagnosis of overanxious disorder of childhood.[30] Generalized anxiety disorder is associated with persistent, excessive, and unrealistic worry that is not focused on a specific object or situation. Children with GAD worry more often about personal safety or the safety of other family members, and they worry more intensely than other children in the same situation. Diagnostic criteria include excessive and uncontrollable worry about multiple issues with at least one somatic complaint present for at least 6 months.

Separation Anxiety Disorder (309.21)

Separation anxiety disorder can only be diagnosed in childhood or adolescence. It is defined as developmentally inappropriate and excessive anxiety concerning separation from home or from those to whom the child is attached. Separation anxiety disorder must present for at least a 4-week duration in youth older than 6 years, since developmentally appropriate fears of separation can be present prior to that time.

Social Anxiety Disorder (300.23)

Two subtypes of social phobia or social anxiety disorder (SAD) are identified: generalized SAD (GSAD), which is characterized by avoidance of most social situations, and non-generalized SAD, characterized by avoidance of a few social situations. Social phobia involves intense distress in response to public situations and persistent fear of social or performance situations where embarrassment may occur, with symptoms persisting for more than 6 months. Children with this disorder may not

Table 18.2. Youth Anxiety Disorders and Potential Associated Behavioral and Cognitive Components[a]

Disorder	Associated Behaviors	Potential Associated Cognitions
GAD	Verbal reports of discomfort and worry about many situations; concerns and questions raised about the impact of the child's behavior on future outcomes	The youth reports many worried thoughts that he/she did something wrong that will result in reprimand or negative interactions; reports concern about safety; believes that negative consequences will result from her/his actions.
Separation anxiety disorder	The child refuses to separate from a parent or major attachment figure; experiences great distress when away from the parent or attachment figure; sometimes refuses to attend school; experiences somatic symptoms when separated—many headaches and stomachaches are described	My parent(s) will be harmed or become sick when I am separated from her/him; I will be harmed or will get sick if I am away from my parent(s); the world outside of my home is not safe.
SAD	The child avoids social situations, usually large group settings; is very quiet in social situations	The child believes that others are evaluating her/him and the evaluation is negative; eg, They think I am stupid; they don't like the way I look and act.
Selective mutism	The child is reluctant to speak to people outside of the family or refuses to speak to others outside of the family	They will not like what I say; they will laugh at the way I talk; they will think what I have to say is stupid.
Specific phobia	The child avoids situations where the phobic object may be encountered; avoids pictures, talk, or sounds associated with the phobic object	All dogs (or other phobic object) are dangerous; I will get hurt by the phobic object; something bad will happen if I encounter the phobic object.
Panic disorder	The child or teen experiences extreme levels of anxiety in a number of settings and begins to avoid situations or will not go out without being accompanied by a parent or another trusted person	I am going to have a heart attack; these feelings mean something is seriously wrong with me; my parent (or other trusted companion) will protect me and help me escape if something happens to me.
PTSD	Youth experiences flashbacks to the traumatic event; avoids situations associated with the event; has a heightened level of tension and bodily discomfort; acts out traumatic event in play over and over	The event is happening again; I will experience another trauma if I return to the scene of the event; I cannot handle anxiety associated with the event.
OCD	Youth repeats actions, washes hands, avoids touching objects, or describes worries about getting contaminated; seeks reassurance from parents; checks on locks, lights, and actions; counts items over and over; touches items in a ritualized pattern	I might have touched germs and I will die; I did not turn off the light, lock the door, or do that action the right way; If I count a certain number of times, bad things will not happen; I have to touch things evenly, once with the right side of my body and once with the left side to make sure bad things don't happen.

Abbreviations: GAD, generalized anxiety disorder; SAD, social anxiety disorder; PTSD, posttraumatic stress disorder; OCD, obsessive compulsive disorder.

[a] From: Silva RR, Gallagher R, Minami H. Cognitive-behavioral treatments for anxiety disorders in children and adolescents. *Prim Psychiatry*. 2006;13(5):68-76. Reprinted with permission.

understand their fear as excessive or unreasonable, while adolescents and adults do. Individuals with social phobia typically experience symptoms resembling panic during a social encounter, such as speaking in public, using public restrooms, eating with other people, or social contact in general. Patients fear being humiliated or embarrassed by their actions and may become intensely anxious, with increased heart rate, diaphoresis, and other signs of autonomic arousal. These physiological symptoms may cause additional anxiety, often leading to a conditioned fear response that reinforces the anxiety of public situations. Individuals with true social phobia go to great lengths to avoid social situations, usually to their own detriment. The fear of embarrassment is "ego-dystonic" (ie, patients with social phobia are disturbed by their symptoms).

Selective Mutism (313.23)

Selective mutism (formerly elective mutism) is a rare but severe syndrome that can occur early in the course of SAD.[31] Selective mutism symptoms include a consistent failure to speak in specific social situations. Diagnosis requires that the duration of the disturbance is at least 1 month (not limited to the first month of school).

Specific Phobia (300.29)

Specific phobia includes an extreme fear of a specific situation or object. Children often do not understand their fears as unreasonable or excessive, while adolescents and adults do. Symptoms must persist for at least 6 months in individuals younger than 18 years for diagnosis.

Panic Disorder With Agoraphobia (300.01) and Panic Disorder Without Agoraphobia (300.21)

Panic disorder includes unexpected panic attacks accompanied by worry about future attacks. Agoraphobia is diagnosed if the individual avoids places in which escape would be difficult or embarrassing.

Obsessive Compulsive Disorder (300.3)

Obsessive compulsive disorder (OCD) is defined by either obsessions or compulsions that are excessive or unreasonable and that cause marked distress and take up to 1 hour or more a day. Obsessive compulsive disorder should be considered in a child that has a high degree of compulsive and ritualistic behavior, such as repetitive hand washing; repeating words silently; and frequently checking, counting, or ordering items. The obsessions and compulsions may interfere with selective and sustained attention. Often these behaviors are ego-syntonic (make sense to the child). Children are more likely to engage in these behaviors at home and may be reluctant to discuss their rituals; therefore, parental report may be the most reliable history.

Post-traumatic Stress Disorder (309.89)

Post-traumatic stress disorder (PTSD) should be considered in any child known to have witnessed or experienced an event or events outside of usual human experience, including the witnessing or experiencing of violence, tragedy, natural disasters, or severe medical illness or emergency. Child abuse is an all too common problem; it results in many long-term physical and emotional effects, including adjustment, mood, and anxiety, including PTSD. Children who develop PTSD have typically faced trauma that threatened a loved one or their own integrity, safety, or even life. The loss of control, the unpredictability, and the extremely aversive nature of the event(s) are the main pathogenic elements of PTSD. The most common precipitant of PTSD is intrafamilial violence, yet this disorder can emerge after exposure to community violence, natural disasters, war, and motor vehicle accidents, as well as medical illness, tragedies, and treatments. Of the youth exposed to traumatic events, PTSD affects between 5% to 45%.[32–34] The essential features of PTSD include the following:

- A child is exposed to actual or threatened death or serious injury to himself or herself or another person, causing a reaction that includes intense fear, horror or, particularly in children, disorganized or agitated behavior.
- The child reexperiences the event (eg, through flashbacks or nightmares). In children, nightmares may have general frightening themes rather than one that specifically involves the abuse. Reexperiencing may also take on the form of repetitive play.
- The child avoids stimuli associated with the trauma, has a numbing of emotional responsiveness, and experiences diminished interest and a sense of a foreshortened future. Children may not report diminished interest, but caretakers may observe it. In children, a sense of a foreshortened future may manifest as a belief that they will never become adults.
- Children may also have somatic symptoms, such as stomachaches and headaches.
- The child has increased physical arousal with an exaggerated startle response.

Duration must be greater than 1 month. Acute stress disorder is a more appropriate diagnosis if the disturbance lasts between 2 days and 4 weeks. The hallmarks of PTSD are based on adult criteria and are often difficult to assess, especially in young children. It has been suggested that young child symptom criteria be modified developmentally. Scheeringa et al[35] suggest that criteria for PTSD be reduced to 4 categories (reexperiencing, numbing of responsiveness, increased arousal, and new fears and aggression) and require only one item to be met from each category for a diagnosis of PTSD. The UCLA PTSD index is a screen that a professional with expertise in childhood trauma could administer and is available online and in the public domain.[36]

Anxiety Disorder Not Otherwise Specified (300.00)

The anxiety disorder not otherwise specified (NOS) diagnosis includes disorders with prominent anxiety or phobia that do not meet criteria for any other specified anxiety disorder. Examples include mixed anxiety-depressive disorder and clinically significant social phobic symptoms.

Anxiety Disorder Due to a Medical Condition With Panic Attacks (293.84)

Anxiety disorders can be caused by medical conditions, such as hyperthyroidism or cardiac problems. The diagnosis is given if panic attacks are the predominant clinical representation of anxiety.

Adjustment Disorder With Anxiety (309.24)

Adjustment disorders with anxiety are defined by the development of excessive worry, jitteriness, or nervousness within 3 months of experiencing a stressor and lasting no more than 6 months. In children, there may be fears of separation from a loved one. Adjustment disorder with anxiety has also been identified as a maladaptive response to the stress associated with having a general medical condition.

Etiology/Pathophysiology

The causes and pathophysiology of anxiety are not fully understood. There are certain life events that may increase the risk of anxiety disorders, including childhood adversity, stress, certain personality types, poor social supports, and illness. There is strong evidence of genetic transmission for a vulnerability to anxiety disorders, especially OCD. Most researchers believe that anxiety and depression involve brain neurotransmitters, particularly serotonin and norepinephrine. Theories have arisen based on the efficacy of pharmacologic agents used to treat anxiety. Thus evidence of a serotonergic etiology exists, as selective serotonin reuptake inhibitors (SSRIs) help alleviate symptoms. Similarly, there seems to be an adrenergic etiology, given the success of propranolol therapy.[37] Serotonin (5-HT), dopamine, and glutamate appear to be involved in the pathophysiology of OCD.[38]

A controversial and poorly understood phenomenon, pediatric autoimmune psychiatric disorders associated with streptococcal infection (PANDAS) has been reported in pediatric patients. This disorder is marked by seemingly overnight onset of obsessions and compulsions, symptomatic of OCD, which then exhibit periods of abrupt relapse and remission. It has been reported that the periods of relapse coincide with streptococcal pharyngitis or scarlet fever.[39] Active streptococcal infections always need to be treated with a usual course of antibiotics. Experimental treatments, such as plasmapharesis, have not been adopted as a standard of care.

Screening for Anxiety

The use of standardized screening tools has shown to be effective in identifying psychological issues.[1] Given the internalizing nature of anxiety symptoms, a child

self-report is recommended when screening for anxiety disorders.[40] The CBCL,[41] BASC,[21] and NICHQ Vanderbilt Assessment Scale[22] are excellent instruments for screening overall behavioral/emotional functioning that contain subscales for anxiety and depression. Standardized screening instruments that have been designed to specifically identify risk for anxiety disorders include the following (see Table 18.1):

1. Revised Children's Manifest Anxiety Scales (RCMAS)[42] consist of 37 yes or no questions divided into 4 scales. This measure is designed to assess the level and nature of anxiety in children between the ages of 6 and 19 years. The RCMAS is successful in distinguishing youth with anxiety disorders from youth without, but it is unclear how useful it is for distinguishing between psychiatric disorders.[43]
2. Screen for Child Anxiety Related Emotional Disorders (SCARED)[44] is a 41-item parent and child symptom inventory using a 3-point scale to assess for *DSM-IV* anxiety disorders. This measure has been shown to correctly differentiate between anxiety disorder subtypes and anxiety disorders and disruptive disorders.[43]
3. Multidimensional Anxiety Scale for Children (MASC)[45] is a 39-item self-report scale for children and adolescents from age 8 to 19 years. Subscales distinguish between physical symptoms, social anxiety, harm avoidance, and separation anxiety. This measure has the strongest supportive evidence for use as an anxiety screening tool.[43]
4. Child Health & Development Interactive System (CHADIS)[46] is a Web-based program that screens for health, emotional, and behavioral issues prior to primary care visits. The CHADIS has assessment tools for children ages 0 to 3, 4+, and adolescents. Parents complete CHADIS questionnaires to assist care providers in addressing issues of child behavior and development while not interfering with routine visits.

Treatment of Anxiety

Effective and evidence-based treatments for pediatric anxiety that have emerged over the past 2 decades include psychopharmacologic management and cognitive-behavioral therapy (CBT).[47] Cognitive-behavioral therapy generally includes psychoeducation, somatic management, cognitive restructuring, problem-solving, exposure, and relapse prevention. Patients participating in CBT have been shown to have demonstrated, maintained, or enhanced treatment gains at follow-up.[48,49] This treatment has also been shown to be an effective prevention and treatment measure in school-based settings.[50] Some studies have indicated greater success rates in children undergoing CBT with an additional parent-based component. More cohesive families have been associated with greater treatment efficacy.[27]

Cognitive-behavioral therapy for children was adapted from adult versions to suit a child's developmental level. The interventions include cognitive restructuring, relaxation training, contingency management, and exposure to feared situations.[51] The

purpose of CBT is to teach the child coping skills so they may reduce anxious symptoms in anxiety-provoking situations. Family components that teach parents as well as the child how to effectively manage cognitions and emotions have been added to CBT.[48] For children with social phobias, group treatment settings are effective in reducing anxious symptoms because they provide more exposure to the feared situation.[49] Cognitive-behavioral therapy is an effective intervention with treatment gains shown to last 7 years posttreatment.[52]

The Child/Adolescent Anxiety Multimodal Study (CAMS) is the largest placebo-controlled study of anxiety disorders in youth and included subjects with one or more of the diagnoses of SAD, GAD, and social phobia.[53] Subjects were randomly assigned to 1 of 4 groups: sertraline only, CBT only, sertraline and CBT combined, and placebo. All 3 treatment arms were superior to placebo, with 81% in the combined condition, 60% in the CBT group, and 55% in the sertraline-only group responding, compared with 24% in the placebo group. These findings suggest that a multimodal approach is most likely to be successful when treating childhood anxiety disorders.

The Pediatric OCD Treatment Study tested the effectiveness of CBT alone, sertraline alone (mean dose 170 mg/day), and combined CBT and sertraline against a placebo group.[54] This study of 112 children demonstrated that combined therapy was most effective, with an efficacy rate of 54%, followed by CBT (39%), which was approximately equal in effectiveness to sertraline alone (21%). The response of the placebo group was only 4%.[54] In one study of 63 adolescents with comorbid anxiety and depressive disorders, the combination of imipramine and CBT (54%) demonstrated greater efficacy than the combination of placebo and CBT (17%).[55]

In the area of psychopharmacologic treatment alone, SSRIs have been shown to have short-term efficacy and safety in the treatment of childhood anxiety disorders. One of the seminal studies was an 8-week study of fluvoxamine for youth with a combination of anxiety disorders that showed a 76% response rate for the fluvoxamine group compared with 29% in a placebo group.[56] In the open-label follow-up study of continued medication treatment, 94% of the responders in the medication group showed sustained benefit 6 months after study completion. Of the nonresponders to fluvoxamine, 71% responded to fluoxetine.[57]

Differential Diagnosis

Distinguishing anxiety from developmentally appropriate fears and realistic worry is important. Throughout the pediatric years, children experience transitory fears while also learning to recognize and understand potential dangers in their environment. Typically there is a normal progression from immediate, tangible fears (eg, separation from caregiver or being around strangers) to anticipatory, less tangible fears (eg, bad dreams, getting hurt, or school failure). Children are expected to overcome and resolve these fears as part of the developmental process. Worry can be thought of as

feeling uneasy or concerned about something that is quite real. It can be an internal representation of a realistic threat. For example, a child that lives in an unsafe neighborhood may worry about the walk home, a child with a learning disability may worry about an upcoming examination, or a child with a medical condition may worry about an upcoming procedure. This kind of worry is expected to be specific to a situation, or to subside once the situation has passed. If the worry does subside, the temporal requirement for GAD diagnosis (6 months) is not met.[58]

If the symptoms of anxiety occur only during the course of an autism spectrum/pervasive developmental disorder, schizophrenia, or another psychotic disorder, an anxiety disorder diagnosis is not given. From a general medical standpoint, substance-induced anxiety disorder and anxiety disorder due to a generalized medical condition, such as thyroid problems,[59] cardiac arrhythmias, and the rarer pheochromocytoma, need to be considered. With anxiety due to a general medical condition, the anxiety must represent a physiologic, not emotional, consequence of a general medical condition. This must be evidenced from the history and physical examination or from radiologic or laboratory findings. Acute onset, lack of family history, and occurrence within the context of an acute medical illness is consistent with this diagnosis, and the severity of anxiety often correlates with the severity of the medical illness. The clinician should take appropriate steps to determine the presence of a medical condition and establish a direct relationship between symptoms of anxiety and the medical condition. It is important for the clinician to note the temporal relationship between the onset of anxiety symptoms and medical condition as well as any symptoms atypical of a primary anxiety disorder. The physical symptoms of anxiety due to a general medical condition, such as shortness of breath, tremor, and rapid heart rate, are often prominent.

Mood Disorders

Depression: Definition and Core Characteristics

Mood disorders are a serious health concern that can be a major cause of morbidity across the life span. They are a risk factor for suicide, as well as for poor academic and psychosocial functioning in childhood. Depressive disorders exist across a range of severity, with the least severe being an adjustment disorder with depressed mood and with major depression being the most severe (associated with the greatest level of functional impairment). Dysthymia is a chronic depression disorder lasting at least 1 year in youth. Depressive disorders in adolescents and children are often comorbid with other disorders, such as ADHD, anxiety, conduct disorders, and substance abuse.[60] In addition, rates of depression can be twice as high in medical populations as in those without chronic medical illness.

Adult criteria are used for the diagnosis of depression in pediatric populations, and this can be a challenge, as the presentation in children is not always the same as in

adults. One of the most important differences between age groups is that children and adolescents can present with irritability instead of depressed mood. They may also appear oppositional when having feelings of worthlessness and low self-esteem. Instead of weight loss, they may not gain weight as expected or may gain excessive weight when depressed. Fatigue may present in children and adolescents as boredom or lack of motivation. Hallmarks of these disorders follow.

Major Depression (296.2 or 296.3)

Major depression is the occurrence of at least one major depressive episode (depressed mood or lack of interest lasting at least 2 weeks). Differing from adult major depressive disorder (MDD), prepubertal boys and girls are equally affected. Major depression often co-occurs with OCD and other anxiety disorders.[4,61] The diagnosis of pediatric MDD includes depression or irritability with 5 additional symptoms for a duration of at least 2 weeks. As opposed to adults, children and adolescents are less likely to identify sadness, present with melancholic symptoms, or have delusions or auditory hallucinations. Often, children have greater irritability symptoms than adults.

Dysthymic Disorder (300.4)

Dysthymic disorder may present in children and adolescents as a persistent depressed or irritable mood for 1 year versus 2 years in adults. Major depression cannot be present for the first year, but it can exist after that period during dysthymia as so-called double depression.[28]

Depressive Disorder Not Otherwise Specified (311)

This diagnosis is applied in children and adolescents with symptoms of depression that do not meet criteria for either major depression or dysthymia. Some characteristics of depressed children and adolescents include maladaptive attributive styles, cognitive distortions, negative self-concept, social skills deficits, impaired problem-solving, and passive or avoidant coping strategies.

Adjustment Disorder With Depressed Mood (309.0)

Adjustment disorder with depressed mood develops within 3 months of a specified psychosocial stressor and must resolve within 6 months after the experience of the stressor to meet diagnostic criteria. Significant behavioral or psychological symptoms, as well as marked distress, are characteristic of adjustment disorders, while tearfulness or feelings of hopelessness allow for the specification of depressed mood.

Mood Disorder Not Otherwise Specified (296.9)

Mood disorder NOS includes disorders with mood symptoms that do not meet criteria for any specific mood disorder and that cannot be fully diagnosed by depressive disorder NOS or bipolar disorder NOS.

Mood Disorder Due to a General Medical Condition (293.83)

A mood disorder due to a general medical condition is a prominent and persistent disturbance in mood as a direct result of the physiological effects of a medical condition. This disorder may resemble MDD or manic, mixed, or hypomanic episodes and may cause clinically significant distress or impairment in functioning.

Etiology of Mood Disorders

As with anxiety disorders, the pathophysiology involved in mood disorders is not fully understood. Some biological features and genetic contributions have been elucidated, and knowledge regarding the role of brain circuits associated with the dysfunctions of depression is expanding. Brain structures involved in these circuits include the orbital and medial prefrontal cortex, amygdala, hippocampus, and basal ganglia.[62] Neurotransmitters, particularly norepinephrine, serotonin, and acetylcholine, play a role in the course of depression,[63] and an individual's stress responsiveness has been shown to be related to depression risk in studies implicating hypothalamic-pituitary-adrenal (HPA) axis reactivity.[64] Moreover, genetic and biological mechanisms both alter and, in turn, are altered by the environment.[65] Decreased sleep efficiency and delayed sleep onset are associated with depressive episodes and recurrence.[66] There is some evidence that children and adolescents with recurrent depression may have shorter REM latency during the period of depression as well as during periods of remission.[67]

Several neurotransmitter systems are thought to be involved in the emergence of depressive disorders, including the noradrenergic, serotonergic, cholinergic, and dopaminergic systems. Studies of children and adults with depression have shown serotonergic agents increase negative feedback of endogenous glucocorticoids with resultant blunted cortisol secretion.[68,69] Studies of adults have also shown that there are lower levels of the serotonin metabolite 5-hydroxyindoleacetic acid in the cerebrospinal fluid of both those who attempt and complete suicide. Studies have shown that children with depressive disorders may have blunted growth hormone responses when challenged with adrenergic agents such as clonidine. Stressful events in young children may lead to permanent HPA axis dysregulation, which correlates with depression in adulthood.[70]

Epidemiology of Mood Disorders

Current estimates suggest that the point prevalence of depression in prepubertal children is 1% to 2%, with that figure rising to 3% to 8% in adolescence.[3,71,72] Major depressive disorder occurs in older adolescents at rates comparable to adults. During adolescence, a 3:1 ratio of females to males with depressive disorders emerges, in contrast to the relatively equal ratio in prepubertal children.[73] By the end of adolescence, the lifetime prevalence of depressive disorders is estimated to be 20%, with higher rates for females, especially after puberty. Approximately 5.4% of Americans

older than 12 years (>1 in 20) are depressed during any given 2-week period,[74] and an estimated 20% of individuals will experience the onset of a depressive episode before the age of 18 years.[75] The risk of depression in first-degree relatives is great, with heritability in the range of 31% to 42%, and this will most likely be higher with more reliably diagnosed major depression.[76] Overall, about 1.3 million young people have depression between ages 5 and 19 years in the United States.[77,78]

In children and adolescents with depression, studies have shown that the various risk factors, which may include biological, psychosocial, familial, or environmental factors, interact in ways that are complex and poorly understood to determine likelihood of depression, length of depressive episodes, and likelihood of later relapse. Prepubertal depression is most commonly associated with adverse family environments, comorbid behavioral problems, and an increased risk of antisocial disorder. However, unlike adolescent depression, prepubertal depression is not likely to be associated with adult depression. Early-onset of puberty (for girls); sexual activity; experimenting with alcohol, tobacco, or drugs; decreased adult supervision and communication; low socioeconomic status; and exposure to abuse, suicide, substance use, and sexual activity are all risk factors for depression.[79] In addition, a greater need for sleep with a tendency to sleep less is associated with an increased risk for adolescent depression.[79] Studies have demonstrated a greater heritability of adolescent-onset depression than childhood depression; however, there is also evidence that environmental factors, such as family environment, neglect or abuse history, and social network, may play as large a role in depression as genetic factors.[79]

Comorbidity is common in children and adolescents with depression. Dysthymia may co-occur from 30% to 80% of the time. Disruptive behavior disorders can occur from 10% to 80% of the time with depression. Substance abuse, particularly for adolescents, can occur 20% to 30% of the time with depression.[80] Anxiety disorders coexist up to 80% of the time with depression. Comorbid anxiety has been linked to longer episodes of depression, but not to recurrence, while the presence of family conflicts has been linked to both longer episodes and increased chance of recurrence. Lifetime MDD is linked to comorbid GSAD.[4] Female gender predicts neither length nor chance of recurrence but does represent an increased risk of a depressive episode.[81]

In longitudinal studies, children diagnosed with MDD have a significant chance of relapse or recurrence. Early-onset depression predicts future depressive episodes during adolescence and adulthood. In addition, children with dysthymic disorder have an increased chance of developing MDD, often within 5 years of dysthymia diagnosis, and the onset of MDD is associated with increased risk for suicide; substance abuse; eating disorders; adolescent pregnancy; and academic, social, and family relationship problems later.[82] Adolescents diagnosed with MDD have been shown to be more likely to have a second episode later in adolescence and are more likely to be depressed or diagnosed with personality disorders as young adults.

Children and adolescents who are depressed may also be at risk for developing bipolar disorder later in life, especially if they present with psychosis, psychomotor retardation, pharmacologically induced hypomania, or mania and a family history of bipolar disorder.[81]

Primary pediatric health care professionals need to be aware that the physical symptoms of depression can be interpreted as a physical illness. Some patients tend to somatize and demonstrate their worries through physical complaints. In addition, the stress of having a medical disorder may lead to feelings of helplessness.

Suicide is the most alarming complication of depressive disorders. If there are any safety concerns, the clinician must assess and ensure a safety plan exists. Suicide is the fifth leading cause of death in children and the third leading cause of death in adolescents. Suicide is uncommon in childhood (ages 5–14 years), with rates of 0.5 for females and 0.9 for males per 100,000, but it tends to rise dramatically in adolescence, with rates of 12.0 for females and 14.2 for males per 100,000.[83] It has been suggested that cognitive development plays a role in carrying out the act of suicide, due to the increasing rate as children age and the fact that many younger children who commit suicide are reported to be early developers.[84] Psychosocial, familial, and psychiatric risk factors must be taken into account for the identification of heightened-risk individuals. Parental divorce is reported in nearly half of all childhood and adolescent suicides, while parental mental problems, history of suicide attempts, substance abuse, and weakened parental support are also prevalent.[6] A study by Brent et al[85] reported that suicide attempts, and not suicidal ideation, are positively associated with first-degree relative adolescent suicide, indicating suicide as a trait that is common within families. Some psychosocial risk factors include academic difficulty, disciplinary problems, individual loss, not being enrolled in school, and being unemployed.[6] It is estimated that more than 90% of youth who commit suicide have an associated psychiatric illness, most commonly a mood disorder.[86] Psychological autopsy has revealed that 81% to 95% of young people who commit suicide have a mental disorder, 50% to 75% of them being affected by a mood disorder, most commonly depression.[87] Health care professionals need to be aware that characteristics of oppositional conduct disorder, disruptive behavior disorders, and psychiatric comorbidity are also common risk factors for suicide.

Screening for Mood Disorders

Despite common reluctance to give psychiatric diagnoses to preschool-aged children, research in the 1980s pointed to the existence of clinically relevant *DSM-III* criteria for MDD in children.[88] However, the *DSM* criteria are based on adult populations and do not always account for variations in child symptoms due to developmental changes. To account for this, the *Diagnostic Classification of Mental Health and Developmental Disorders of Infancy and Early Childhood (DC:0-3)*[89] has been developed, with more young child–centered criteria (in addition to modified criteria from the *DSM-IV),* which include symptoms based on play observation. In research,

anhedonia has been proven to be a strong indicator of preschool depression, occurring almost exclusively in depressed children and not in control groups. In addition, irritability or sadness is observed in 98% of the depressed group, making it the most sensitive marker.[90]

It may be difficult to obtain accurate information for the diagnosis of anxiety and mood disorders in preschool children, as caregivers may give skewed information; however, self-report techniques, including a modified Children's Depression Inventory[91] or the Berkley Puppet Interview-Symptom Scales,[92] have been shown to be reliable screening techniques, as has observation of parent-child dyad playing. The Preschool Age Psychiatric Assessment (PAPA), a "semi-structured parent interview for assessing psychiatric symptoms in research studies," is a new tool for diagnosing depressed preschoolers.[90]

In diagnosing MDD in preschoolers, it is important to rule out PTSD and adjustment disorder with depressive features—both differential diagnoses are preceded by a significant stressful or traumatic experience more commonly than MDD. When concerns about depression have been identified through a direct interview or use of a screen, such as the PSC, then more focused self-report assessment tools may be indicated. Standardized screening instruments that can be used by primary pediatric health care professionals in their efforts to evaluate children and adolescents with potential mood disorders include the following (see Table 18.1):

1. The Children's Depression Inventory (CDI) is designed to assess cognitive, affective, and behavioral signs of depression in children from 6 to 17 years of age.[91] This instrument is designed for use in the school setting, pediatric practice, child guidance clinics, and psychiatric settings. The 27 items of the CDI reveal total scores, as well as scores for each of the individual scales: negative mood, interpersonal difficulties, negative self-esteem, ineffectiveness, and anhedonia.[91] The CDI takes only 5 to 10 minutes to complete and is written at a first-grade reading level.
2. The Beck Depression Inventory (BDI) is a clinician-administered measure that monitors the intensity, depth, and severity of depression in psychiatric patients, and it also identifies depressive symptoms in a primary care setting.[93] A short version composed of only 7 questions, as opposed to 21, is available, as well as a version designed to be administered by primary care professionals. Scoring on the BDI varies between patients that have been diagnosed with depression and those in primary care settings. For patients with a diagnosis, scores are rated as minimal, mild, moderate, and severe depression, while a cutoff score of 21 suggests a diagnosis of depression for primary care patients.[93]
3. The Mood and Feelings Questionnaire (MFQ) is designed to be used as a quick screening measure to determine the extent to which children and adolescents exhibit depressive symptoms. The items on the MFQ are heavily weighted toward the affective and cognitive components of depression; however, other

diagnostic criteria are also assessed (eg, restlessness, fatigue, and poor concentration). There are separate versions for the child (13 questions) and parent (11 questions), and the scale takes approximately 5 minutes or less to complete. Child scores at or above 8 and/or parent cumulative scores at or above 12 suggest a clinically significant problem with depressive symptoms. When administered consistently throughout treatment, it is possible to monitor symptoms and determine the positive benefits of treatment on depressive symptoms.[94,95] See http://devepi.mc.duke.edu/MFQ.html for more information.

4. The Center for Epidemiologic Studies Depression Scale for Children (CES-DC) is a self-report depression symptom inventory. Patients answer 20 items. Scores on the CES-DC range from 0 to 60, with 15 being the cutoff score suggestive of depression symptoms in children and adolescents.[19]
5. The Children's Depression Rating Scale, Revised (CDRS-R) is a brief rating scale based on a semi-structured interview with the child (or an adult informant). It is designed for 6- to 12-year-olds, and has been used successfully with adolescents. Administration time is estimated at 15 to 20 minutes. It can be used in clinical settings to diagnose depression and monitor treatment response. It can also be used in school and pediatric clinics as a screening tool to identify children who might benefit from professional intervention.[96] This rating scale has been used in National Institute of Mental Health (NIMH) and other medication trials.

Bipolar Disorder

Bipolar disorder in children and adolescents continues to be better understood as more research emerges. A meta-analysis by Kowatch and colleagues[97] indicated the following symptoms as prevalent: increased energy, distractability, grandiosity, pressured speech, irritability, racing thoughts, decreased need for sleep, and euphoria. Compared with adults, differential diagnosis can be even more challenging in children and adolescents with disruptive behavior and other mood disorders, such as an irritable depression. By carefully reviewing symptoms and particularly considering those mentioned above, physicians and other clinicians can more effectively identify and treat bipolar disorder. The AAP TFOMH recommends that bipolar disorder should be managed by a psychiatrist and not by most primary pediatric health care professionals. However, it is extremely beneficial for the primary pediatric health care professional to be familiar with bipolar disorder so he or she understands appropriate treatment and follow-up and also because in some isolated areas, he or she may be the only health care professional available to provide treatment.

In adolescents with depression, 20% (and possibly up to 40%) may convert to bipolar disorder. This means conversely that 60% to 80% do not. However, the rate of conversion in adolescents appears be higher than in adults. Risk factors for conversion include early-onset depression, psychomotor retardation or psychosis, family history of any mood disorder, and medication-induced hypomania or mania (the highest rate of conversion when treated with an antidepressant occurs at 10–14 years of age).[78,80]

Probably the greatest diagnostic and treatment challenges in the field of pediatric mental health are pediatric bipolar spectrum disorders. Some evidence-based treatment studies have been criticized for including youth with conduct disorders and trauma histories. Nearly 60% of bipolar sufferers presented their first bipolar episode before age 20 years.[98–100] Early onset of the disorder is often associated with increased recurrence and chronicity, as well as a higher risk of comorbidity and poor outcomes. Studies have indicated that depressed youth with comorbid ADHD are at a high risk of developing bipolar disorder later.[101] Bipolar depression, unlike mania, will often go untreated for long periods, due to its less noticeable symptoms. Perhaps due to the disorder's initial similarity to depression or other disorders, 70% of patients in a National Depressive and Manic-Depressive Association (now the Depression and Bipolar Support Alliance) survey reported at least one misdiagnosis, most commonly MDD.[100] On average, these patients waited 9 years before a correct diagnosis was made. In adolescents, this kind of delay of proper treatment may result in interpersonal and vocational consequences throughout adult life. In diagnosis, the *DSM-IV* makes a distinction between treatment-emergent affective switches (TEAS), which are often precipitated by medication, and spontaneously occurring manic episodes. A TEAS will not lead to a bipolar diagnosis according to the *DSM* standards. In addition, the less severe episodes of hypomania may often be overlooked, leading to prolonged misdiagnosis.[101] Psychotherapy techniques, such as CBT, psychoeducation, and family interventions, have demonstrated responses equal to or greater than that of pharmacotherapy for treating bipolar disorder, especially in relapse prevention. Family-focused therapies may prove especially effective in teenagers, due to their dependence on family support.[102]

The Young Mania Rating Scale (YMRS) screen may help with differentiating possible bipolar illness from some other area of concern and is available online.[102] This 11-item scale is able to assess manic symptoms, such as elevated mood and energy level, rate of speech, and sleeplessness,[102] with cutoff scores of 25 for mania and 20 for hypomania on a 1 to 60 scale. A score of 13 indicates a potential case for mania or hypomania, while a score of 21 or higher indicates a probable case.[103]

Treatment of Mood Disorders

Two types of psychological treatment strategies have demonstrated empirical support for the treatment of adolescent depression: CBT and interpersonal therapy (IPT).[104] However, pharmacotherapy may need to be considered if there is insufficient response to psychosocial and cognitive behavioral strategies.

The combination of fluoxetine and CBT was superior to either alone according to the results of the Treatment for Adolescents with Depression Study (TADS).[105,106] This study evaluated short- and long-term effectiveness of fluoxetine alone, CBT alone, combination of fluoxetine and CBT, and placebo. Four hundred thirty-nine

youth between the ages of 12 and 17 who met *DSM-IV* criteria for MDD participated. At 12 weeks the combination of fluoxetine and CBT was superior, with a response rate of 71%. Fluoxetine was more effective than CBT alone, with response rates of 61% and 43%, respectively; CBT was statistically equivalent to placebo (35% response rate). The participants were followed for a total of 36 weeks, and response rates at the end of the study were 86% for combination therapy, 81% for fluoxetine therapy, and 81% for CBT at week 36. Combination therapy achieved remission earlier than monotherapy with either fluoxetine or CBT and was overall more effective.

A 2006 review by Boylan et al[107] describes the challenges of treating pediatric MDD, including the likelihood of multiple comorbid disorders; psychosocial or academic problems; and increased risk of suicide attempts, self-harm, and substance abuse. If mild, MDD may be treated with supportive therapy like CBT, IPT, or psychoeducation. In moderate to severe cases, pharmacotherapy should be considered, although CBT, IPT, or psychotherapy may increase the rate of improvement.

Since 1992, the suicide rate has dropped in adolescents and children in the general population. That period has seen a 35% decrease in youth suicide in 15 countries.[108,109] There has been no corresponding decrease in substance abuse or firearms availability, or any corresponding increase in mental health screening.[110] However, there has been an increase in antidepressant medication treatment for young people during this period.[111] Unfortunately, as a result of the black box warning issued by the US Food and Drug Administration (FDA) in 2003 (which warns about the increased risk of suicidal ideation in association with the initiation of antidepressant medication treatment), the rate of prescriptions of antidepressant medication for children has gone down.[112] This decrease has been associated with an increase in suicide rates among children and teenagers.[113]

Tips for Determining Whether to Reassess, Actively Treat, or Refer

A key component for determining appropriate treatment, as prescribed by the *DSM-IV-TR*[28] and the *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, Primary Care Version*[114] is the extent of impairment or severity. Symptoms of anxiety or depression can vary from mild (can be easily managed in primary care) to severe (definitely need to be addressed by a very experienced clinician). For patients that require more comprehensive treatment, daily support through their school, day hospitalization, inpatient hospitalization, or psychopharmacologic treatment may be needed to help reduce symptoms. Early intervention is always best, as the preservation of a child's self-esteem is more likely if problems are identified before they worsen or become chronic.

The successful management of mental health disorders in children is often determined by the degree of complexity of the presenting problem, the availability of mental health care clinicians, and the interest and motivation of primary pediatric health care professionals to learn state-of-the-art assessment and treatment methods.

Some professionals may have little interest or experience in treating these disorders but may be the only available source for acute or ongoing psychiatric care for families. Some primary or specialty care clinicians may unknowingly convey a condescending message that these problems are not important or that they are "crazy." This can discourage follow-up for assessment or treatment.

The urgency of intervention and the type of triage or treatment should always be determined by concerns for current and future safety. The necessity of treatment should be based on an assessment of how much the disorder is impairing the functioning of the child. It is very important that health care professionals communicate to families that symptoms of anxiety or depression are not a sign of a personal weakness. It is often useful for the health care professional making a referral to state that they want to help and that they need help to provide the best care. Primary pediatric health care professionals also need to convey that they will continue to be involved in the child's care while the child is seen by a mental health professional. Periodic phone calls by the clinician's office to ensure that families have scheduled an appointment, have followed up, and have remained in treatment can often ensure that recommendations are followed.

Pharmacotherapy of Anxiety and Mood Disorders

Anxiety Disorders

Selective serotonin reuptake inhibitors, often prescribed when CBT is not effective or when anxiety symptoms are severe and interfere with everyday functioning, have repeatedly been shown to be safe and effective in treating anxiety disorders in adolescents and children.[53] However, the FDA recently added a black box warning about the potential risks for "suicidal thinking and behavior" with these medications. This places a greater burden on the prescribing physician to assess the severity of the child's anxiety, the degree of impairment caused by the symptoms, the level of family functioning, and the family's ability to monitor the medication and potential side effects before establishing treatment protocols.

Treatment of OCD has the largest body of supporting evidence in children. Medications with FDA approval for the treatment of OCD in children from 7 to 17 years of age include the SSRIs fluvoxamine, fluoxetine, and sertraline and the tricyclic antidepressant clomipramine. Placebo-controlled clinical trials have generally shown that approximately 50% of children or adolescents with anxiety respond positively to SSRIs versus approximately 25% of children who receive placebo.[54,115–117] Cognitive-behavioral therapy in conjunction with medication may increase drug effectiveness. Response and relapse may be worsened by comorbidity with other psychiatric illnesses.[118] Increased symptom severity is also related to lower treatment effectiveness.

Table 18.3. Antidepressant Medications

Type/ Class	Medication	Dosing Guidelines	Recommended Administration	FDA Approval (OCD) as of 2009	Evidence-based Medication Trials	FDA Approval (Depression) as of 2009	Evidence-based Medication Trials	Common Side Effects (note: all antidepressants have black box warning for increased suicidality)
SSRI	Fluoxetine	Initial daily dose 5–10 mg Target daily dose 20 mg Maximum daily dose 40–80 mg	AM Oral elixir preparation available	Yes, ages 7–17	Yes	Yes, ages 8–17	Yes	Nausea, headache, insomnia, sexual dysfunction, restlessness
SSRI	Citalopram	Initial daily dose 10–20 mg Target daily dose 20 mg Maximum daily dose 60 mg	AM Oral elixir preparation available	No	Yes	No	Yes	Nausea, headache, insomnia, sexual dysfunction, restlessness
SSRI	Escitalopram	Initial daily dose 5–10 mg Target daily dose 10 mg Maximum daily dose 20 mg	AM Oral elixir preparation available	No	Yes	Yes, ages 12–17	Yes	Nausea, insomnia, restlessness, sexual dysfunction is less common than other SSRIs
SSRI	Sertraline	Initial daily dose 25–50 mg Target daily dose 50–100 mg Maximum daily dose 150–200 mg	AM Oral elixir preparation available	Yes, ages 7–17	Yes	No, FDA withdrew approval	Yes	Nausea, headache, insomnia, sexual dysfunction, restlessness
SSRI	Paroxetine (CR)	Initial daily dose 10–20 mg (12.5–25 mg) Target daily dose 20 mg (25 mg) Maximum daily dose 40–60 mg (62.5–75 mg)	AM or HS Oral elixir preparation available	No	Yes	No	Limited	Nausea, drowsiness or insomnia, headache, sexual dysfunction, weight gain

Table 18.3. Antidepressant Medications *(continued)*

Type/ Class	Medication	Dosing Guidelines	Recommended Administration	FDA Approval (OCD) as of 2009	Evidence-based Medication Trials	FDA Approval (Depression) as of 2009	Evidence-based Medication Trials	Common Side Effects (note: all antidepressants have black box warning for increased suicidality)
SSRI	Fluvoxamine	Initial daily dose 25 mg Target daily dose 50 mg–200 mg Maximum daily dose 200–300 mg	HS	Yes, ages 8–17	Yes	No	Yes	Nausea, headache, insomnia, sexual dysfunction, restlessness
Other	Bupropion (SR/XL)	1–7 mg/kg/day Initial daily dose 100 mg (100–150 mg/150 mg) Target daily dose 225–300 mg (200–300 mg/300 mg) Maximum daily dose 225–300 mg (200–300 mg/300 mg)	TID ≤150 mg/ dose (BID ≤200mg per dose/daily)	No	No	No	Limited	Headache, insomnia. Increased risk of seizures with doses >450 mg/day. Not associated with sexual dysfunction or weight gain.
SNRI	Venlafaxine (XR)	Initial daily dose 37.5–75 mg (same) Target daily dose 150–225 mg (75–225 mg) Maximum daily dose 375 mg (225 mg)	BID (daily)	No	Yes	No	Limited	Nausea, headache, insomnia, sexual dysfunction, increased blood pressure

Table 18.3. Antidepressant Medications *(continued)*

Type/ Class	Medication	Dosing Guidelines	Recommended Administration	FDA Approval (OCD) as of 2009	Evidence-based Medication Trials	FDA Approval (Depression) as of 2009	Evidence-based Medication Trials	Common Side Effects (note: all antidepressants have black box warning for increased suicidality)
TCA	Imipramine	Initial daily dose 1.5 mg/kg or 25 mg Target daily dose 100 mg FDA upper dose 2.5 mg/kg but evidence shows maximum daily dose 5 mg/kg or 200 mg (serum level 150–250 ng/mL)	HS	No	Yes	No	No	Orthostatic hypotension, conduction abnormalities, Anticholinergic, drowsiness, nausea, headache, sexual dysfunction, weight gain
TCA	Clomiramine	Initial daily dose 25 mg Target daily dose 3 mg/kg or 100 mg Maximum daily dose 3 mg/kg or 200 mg	HS	Yes	Yes	No	IV route only	Anticholinergic, orthostatic hypotension, conduction abnormalities, drowsiness, sexual dysfunction, weight gain

Abbreviations: AM, morning; BID, twice a day; FDA, US Food and Drug Administration; HS, at bedtime; IV, intravenous; OCD, obsessive-compulsive disorder; SNRI, serotonin-norepinephrine reuptake inhibitor; SSRI, selective serotonin reuptake inhibitor; TCA, tricyclic antidepressant; TID, 3 times a day.

Table 18.4. Dosing Guidelines for Antidepressant Agents

Type/Class	Medication	Initial Starting Daily Dose	Initial Target Daily Dose (Serum Level)*	Maximum Daily Dose (Serum Level)	Recommended Administration
SSRI	Citalopram	10–20 mg	20 mg	60 mg	AM
	Escitalopram	5–10 mg	10 mg	20 mg	AM
	Fluoxetine	10–20 mg	20 mg	40–80 mg	AM
	Paroxetine (i)	10–20 mg	20–30 mg	40–60 mg (ii)	AM or HS
	Sertraline	25–50 mg	50–100 mg	150–200 mg	AM
SNRI	Duloxetine	20–30 mg	40–60 mg	120 mg (iii)	Daily or BID
	Venlafaxine	37.5–75 mg	150–225 mg	375 mg	BID
	Venlafaxine XR	37.5–75 mg	75–225 mg	225 mg	Daily
Other (iv)	Bupropion	75 mg	225–300 mg	450 mg	TID ≤150 mg/dose
	Bupropion SR	100–150 mg	200–300 mg	400 mg	BID ≤200 mg/dose
	Bupropion XL	150 mg	300 mg	450 mg	Daily
	Mirtazapine	7.5–15 mg	30 mg	60 mg (v)	HS
TCA	Amitriptyline	25–50 mg	150–200 mg	300 mg	HS
	Clomipramine	25 mg	100–150 mg	250 mg	HS
	Desipramine	25–50 mg	150 mg (>ng/ml)	300 mg	HS
	Imipramine	25–50 mg	150 mg (>ng/ml) (vi)	300 mg (200–400 ng/ml) (vi)	HS
	Nortriptyline	25–50 mg	75–100 mg (50–150 ng/ml)	150 mg (50–150 ng/ml)	HS

SSRI: selective serotonin reuptake inhibitor **SNRI:** serotonin-norepinephrine reuptake inhibitor **TCA:** tricyclic antidepressant

* Antidepressant dosage can be increased every 2 to 3 weeks as tolerated if remission has not occurred. (i) Paroxetine and paroxetine CR have similar side effect profiles, comparable half-lives, and reach steady state plasma concentrations at similar time intervals. (ii) Manufacturer recommended maximum dose for major depressive disorder (MDD) is 50 mg/day. (iii) Manufacturer recommended maximum dose for MDD is 60 mg/day. (iv) Trazodone is not included as a treatment option for MDD because therapeutic doses are hard to achieve due to excessive sedation (therapeutic dose 300–600 mg/day). Trazodone may be considered during the acute treatment phase as adjunctive therapy when sedation is desired. (v) Manufacturer recommended maximum dose is 45 mg/day. (vi) Serum level includes parent drug and active metabolite (imipramine and desipramine, respectively).

From: Evidence-Based Best Practices for the Treatment of Major Depressive Disorder in South Carolina.pdf. Published February 2008. Available at: http://www.sccp.sc.edu/centers/SCORxE

Table 18.5. Common Side Effects (SEs) of Antidepressant Medication **0 = Absent or rare to 4+ = common**

Class	Antidepressant	Anticholinergic	Conduction Abnormalities	Drowsiness	Gastrointestinal Distress	Headache	Insomnia	Orthostatic Hypotension	Sexual Disturbances	Weight Gain	Comments
SSRI	Citalopram	0	0	0	3+	2+	2+	0	4+	1+	Activation, agitation and restlessness reported with all SSRIs
	Escitalopram	0	0	0	3+	0	2+	0	2+	1+	
	Fluoxetine	0	0	0	3+	2+	2+	0	4+	1+	
	Paroxetine	1+	0	1+	3+	2+	2+	0	4+	2+	
	Sertraline	0	0	0	3+	2+	2+	0	4+	1+	
SNRI	Duloxetine	1+	1+	1+	3+	2+	2+	0	4+	0	
	Venlafaxine	1+	1+	1+	3+	2+	2+	0	4+	0	Higher rate of hypertension with doses >225 mg/day
Other	Bupropion	0	0/1 +	0	1+	2+	2+	0	0	0	Increased risk of seizures with doses >450 mg/day
	Mirtazapine	1+	1+	3+	0+	1+	1+	1+	2+	3+	Less sedation with doses >15 mg/day
TCA	Amitriptyline	4+	3+	4+	1+	1+	1+	3+	1+	4+	
	Clomipramine	4+	3+	4+	1+	1+	2+	2+	4+	4+	
	Desipramine	1+	2+	2+	0	0	1+	2+	1+	1+	
	Imipramine	3+	3+	3+	1+	2+	2+	4+	4+	4+	
	Nortriptyline	2+	2+	2+	0	0	0	1+	0	1+	

SSRI: selective serotonin reuptake inhibitor **SNRI:** serotonin-norepinephrine reuptake inhibitor **TCA:** tricyclic antidepressant

References: *Drug Information Handbook for Psychiatry*. Fuller MA, Sajatovic M, editors. Hudson, OH. Lexi-Comp 2007: *Clinical Handbook of Psychotropic Drugs*. Bezchlibnyk-Butler KZ, Jeffries JJ, editors. Toronto, ON. Hogrefe & Huber, 2006.

There is less research on other pediatric anxiety disorders (like social phobia, GAD, and separation anxiety disorder), which are often treated similarly due to clinical overlap. Selective serotonin reuptake inhibitors have been shown to be an effective treatment for social phobia, GAD, SAD, selective mutism, and PTSD.[119–122] The most important study to date, the CAMS, showed that in 488 subjects aged 7 to 17 years with separation anxiety disorder, SAD, and GAD, a combination of CBT and sertraline was the most effective treatment, followed by CBT alone and sertraline alone, which were equally effective compared with placebo.[123] The treatment was well tolerated in general. Benzodiazepines are generally not recommended for treatment of anxiety in the pediatric population given lack of evidence of efficacy and high potential for abuse. In anxiety comorbid with ADHD, while older studies had reported findings that indicated up to 25% of children and adolescents' anxiety worsened with stimulant treatment, the recent NIMH Multimodal Treatment Study of ADHD (MTA) showed that stimulants did not worsen anxiety.[124]

The course of treatment may depend on the disorder. For example, GSAD is associated with greater impairment because of its earlier onset, negative effect on social skills, and increased odds ratio for MDD later in life.[4] A striking 25% to 31% of adolescents and adults with SAD have comorbid depressive disorder[4] and 50% of those who suffer from SAD as adults experience onset of the disorder before age 13 years. In children, SAD presents with symptoms similar to adults, but may also manifest a characteristic such as selective mutism. Any medications that are helpful with anxiety are also helpful in the treatment of depression.

Mood Disorders

In a review of controlled pharmacotherapy trials by Singh and colleagues,[125] it is suggested that for moderate to severe cases of pediatric mood disorders, the first-line treatment is usually pharmacotherapy either by itself or with CBT or IPT. Currently fluoxetine and escitalopram are the only FDA-approved drugs for the treatment of MDD in children older than 7 years. However, a recently generated algorithm for the treatment of pediatric MDD has fueled a large number of short-term placebo-controlled studies of antidepressants in children, comparing the safety and efficacy of varying medications. The review by Singh et al showed that SSRIs have been proven effective for treating children and adolescents with MDD, but there are no published studies concerning the treatment of mild depression or dysthymia. When prescribing SSRIs, there is a risk that the side effects may include an increased risk of suicidal thoughts and behaviors. When treatment begins, it is important to monitor the patient's environment, and psychoeducation for the family may be beneficial.[107] Tricyclic and heterocyclic antidepressants have not been demonstrated to have a greater rate of effectiveness than placebo, and there have been no placebo-controlled trials of monoamine oxidase inhibitors in adolescents. In addition, studies concerning the use of lithium have been inconclusive.[126] Decisions about the treatment of depressive disorders with comorbid ADHD should depend on the severity of each

condition. The treatment of ADHD with atomoxetine may decrease both ADHD and depressive symptoms,[127,128] and bupropion has been shown to be effective for both disorders.[129]

According to Singh et al,[125] there are few double-blind, placebo-controlled studies of the efficacy of drug treatment on pediatric bipolar disorder and related conditions; however, the American Academy of Child and Adolescent Psychiatry has recently endorsed lithium, the anticonvulsant divalproex sodium, and atypical antipsychotics as first-line treatment agents and has recommended the use of drugs approved by the FDA for adults.[130] The continuing effectiveness of lithium for long-term treatment has been called into question by recent research, which indicates that patients on lithium randomly assigned to either a placebo group or a group continuing treatment did not experience a significantly different incidence of mania.[131] In addition, the side effects of lithium (including cognitive dulling, tremor, exacerbation of acne, weight gain, and long-term kidney and thyroid problems) cause it to be a less acceptable treatment for younger patients. Anticonvulsant therapies have also been tested, with mixed efficacy, but valproate is now the most commonly prescribed mood stabilizer in children and adolescents, due in part to its greater tolerability.[101] Studies have shown risperidone and aripiprazole (a partial dopamine agonist) to be effective in the treatment of pediatric bipolar disorder if the other drugs are not effective, and both are FDA approved for youth ages 10 to 17 years with bipolar mixed or manic episodes.[132–136]

Advances have been made in both the understanding of and treatment for childhood and adolescent bipolar disorder. Research suggests that if treatment is initiated soon after a mood disorder (particularly bipolar disorder) becomes evident, both the severity and chronicity may be reduced. The clinician must balance emerging evidence that supports early treatment with the fact that research has still yet to provide a full picture of this disorder among youth, leaving the diagnosis in a very controversial area.

Conclusion

Anxiety and mood disorders commonly exist in patients who are seen in pediatric general practice. The primary pediatric health care professional must be able to screen for and identify these disorders, as well as recommend appropriate treatment with medication as part of the primary care medical home. Families and children presenting to primary pediatric health care professionals with anxiety and mood disorders rely on the medical home to provide the same quality of evidence-based treatments that would be expected when receiving treatment for any other medical condition. At first glance, treatment of anxiety and mood disorders may seem daunting, yet the primary pediatric health care professional holds a great advantage given his or her ability to work with patients from birth through late adolescence and is subsumed under the primary care medical home. Many patients demonstrating symptoms of more complex disorders, particularly bipolar disorder and comorbid

anxiety and depression, are ideally referred to specialists. However, given the shortage of mental health professionals, the primary pediatric health care professional should be as knowledgeable as possible about assessment and treatment of the spectrum of anxiety and depressive disorders.

Useful Web Sites and Resources

Screening tool
http://www.massgeneral.org/schoolpsychiatry/screeningtools_table.asp

Professional journal article
Wagner AP. Cognitive-behavioral therapy for children and adolescents with obsessive-compulsive disorder. *Brief Treat Crisis Interv.* 2003;3(2):291–306

Treatment manual and kit for professionals
Wagner AP. *Treatment of OCD in Children and Adolescents: A Professional's Kit.* 2nd ed. Rochester, NY: Lighthouse Press; 2007

Professional magazine article
Wagner AP. The Worry Hill: A child-friendly approach to OCD. *Psychotherapy Netw.* 2008;May/June:63–68

Book on anxiety and CBT for parents, schools, and clinicians
Wagner AP. *Worried No More: Help and Hope for Anxious Children.* 2nd ed. Rochester, NY: Lighthouse Press; 2005

Professional kit for use with anxious kids
Wagner AP. *Worried No More: Teaching Tools and Forms on CD.* 2nd ed. Rochester, NY: Lighthouse Press; 2005

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Chapter 19

Evidence-based Psychological and Behavioral Interventions

Michael W. Mellon, PhD, ABPP

Introduction

The limited amount of time available to complete a health maintenance visit requires the primary pediatric health care professional to have a specific method in place to cover a wide range of topics in a time-efficient manner. Except for physical trauma and acute infections, developmental and behavioral issues are the most common presenting problems in primary pediatric care.[1] As much as one-half of primary care pediatricians' time is devoted to assessing and managing childrearing, discipline, developmental, educational, and behavioral problems.[2] It has been shown that pediatricians feel responsible for identifying and managing child behavioral/mental health problems.[3] Eighty percent of surveyed pediatricians felt it was their responsibility to identify disorders such as attention-deficit/hyperactivity disorder (ADHD), eating disorders, depression, substance abuse, and behavior problems; 59% agreed they were responsible for the identification of learning disabilities. However, excluding ADHD, less than a third felt responsible for treating/managing mental health issues. More than 85% indicated they would refer these patients to other mental health professionals for care. However, a dearth of mental health professionals are available, and the primary care medical home is expected to encompass care for behavioral/mental health disorders in children and adolescents. The American Academy of Pediatrics Task Force on Mental Health has recommended that primary pediatric health care professionals achieve the following competencies: (1) select appropriate tools to screen for mental health problems and functional impairment in infants, children, and adolescents and for family issues adversely affecting their behavioral and mental health in a health maintenance visit; (2) establish and use mechanisms to monitor progress toward functional goals for children/adolescents with behavioral and mental health challenges; and (3) select appropriate tools for gathering behavioral and mental health diagnostic and/or functional information from collateral sources.

Given this need to manage mental health disorders within the medical home, it is essential for primary pediatric health care professionals to be knowledgeable about the most effective, evidence-based psychological and behavioral interventions available to their patients.

Given the plethora of unproven and disproven therapies, many of which are widely available and marketed specifically for the treatment of children with developmental-behavioral problems, such as sensory integration therapy, visual training, auditory integration training, music therapy, facilitated communication, play therapy, and others,[4] the purpose of this chapter is to assist primary pediatric health care professionals in confidently guiding families to choose effective evidence-based psychological and behavioral treatments for their children with developmental and behavioral problems (Table 19.1).

Evidence-based Interventions for Internalizing, Externalizing, and Developmental Disorders

Chapter 4 in this manual reviews common behavioral problems that are identified by primary care providers and that can be effectively managed within the medical home. However, children will also present with emotional and behavioral problems that warrant referral to mental health professionals. The important decisions for primary pediatric health care professionals to make are to properly conceptualize the problem and identify an evidence-based intervention to which to refer their patients and their families. The reader should recognize an emphasis on evidence-based psychological interventions that are primarily delivered through a "parent training" model or, at the very least, a model in which parents play an instrumental role in the

Table 19.1. Evidence-based Psychological/Behavioral Interventions for Children and Adolescents

Indication	Evidence-based Intervention
Anxiety/depression	Cognitive-behavioral therapy (CBT)
Attention-deficit/hyperactivity disorder	Behavior therapy
Oppositional, disruptive behavior	Parent-child interaction therapy (PCIT), Incredible Years, Triple P-Positive Parenting Program
Conduct disorder/substance abuse	Multisystemic therapy
Autism spectrum disorders/intellectual disability	Applied behavior analysis (ABA)
Learning disabilities	Direct instruction
Enuresis	Urine alarm
Encopresis	Behavior therapy (following bowel clean out)
Weight loss, smoking cessation	Motivational interviewing
Medical procedure-related and chronic pain	Hypnosis, biofeedback

treatment, rather than an emphasis on direct psychotherapy provided individually to the child. This emphasis is made due to the unquestionable influence parents have on children, and children on parents, and the behavioral and developmental outcomes that result.[5] The goal of parent training approaches is to teach parents to apply operant and social learning methods, with an understanding of child development, to assist their children in enhancing positive and adaptive behavior while extinguishing disruptive or maladaptive behavior.

A common categorization of broad areas of concern that may warrant referral for evidence-based psychological/behavioral interventions in children includes internalizing disorders, externalizing disorders, and developmental disorders. The following section of this chapter will be dedicated to specifying examples of specific diagnostic entities within these broad categories and the types of evidence-based interventions that appear to be most effective in treating these disorders.

Internalizing Disorders

Anxiety and depression are usually referred to as internalizing disorders, as the manifestations of these problems include poorly regulated feelings that may interfere with the child's environmental adaptability. Anxiety disorders of childhood come in many versions (ie, phobias, separation anxiety, generalized anxiety, school avoidance, etc) and are common, with prevalence estimates of 10% to 20% of the population included.[6] Although somewhat less common than anxiety, depression in childhood will also present to primary pediatric health care professionals, with prevalence rates estimated to be 10% to 15% of children and adolescents showing some symptoms of depression[6] (see Chapter 18).

The treatment known as cognitive-behavioral therapy (CBT) has been employed with both anxiety and depressive disorders. This treatment includes components designed to address maladaptive or inaccurate thoughts that elicit maladaptive emotional functioning that further leads to maladaptive behavior. Essentially, the intervention attempts to change the thoughts that disrupt a child's productive engagement of the environment. These components include direct intervention with the child and parent as a dyad. An example of this approach for anxiety is the Coping Cat program developed by Kendall.[7] The Coping Cat program, combined with parental involvement, is considered to be a "probably efficacious" treatment for anxiety disorders in children.[8]

The use of CBT for depression does not have as much evidence compared with that for anxiety disorders in children. Cognitive-behavioral therapy for depression typically includes social skills training, assertiveness training, relaxation training, and imagery and cognitive restructuring, all of which are designed to help the child reengage in behaviors that previously brought satisfaction. The intervention developed by Lewinsohn et al,[9] the Coping with Depression Course, contributed to fewer self-reported feelings of depression, cognitive clarity, and increased activity levels in adolescents, and is considered a probably efficacious treatment.

Externalizing Disorders

Much more is known about externalizing disorders, perhaps because these disorders may be associated with adverse outcomes that affect not only the individual child or adolescent but also those around them, such as those relationships in the family, school, and community. As such, children thought to exhibit an externalizing disorder are more likely to be treated.[10] These disorders include ADHD, oppositional defiant disorder (ODD), and conduct disorder. The reported prevalence rates of these disorders are 7% of the school-aged population for ADHD,[11] and from 2% to 16% for the disruptive behavior disorders of ODD and conduct disorder.[12] These disorders also evidence significant overlap as comorbidities. A recent epidemiologic study reported that optimal treatment of ADHD is associated with a significant reduction in lifetime costs of treatment and psychosocial burden on the child.[13] The treatments that have the best evidence for effectively treating externalizing disorders are multimodal treatment for ADHD (which includes various parent training methods), parent-child interaction therapy (PCIT), Incredible Years, Triple P-Positive Parenting Program, and the Adolescent Transition Program. Each of these approaches will be briefly described with target populations and evidence for their effectiveness.

With regard to the optimal treatment of ADHD, much has been learned from the study funded by the National Institutes of Mental Health known as the Multimodal Treatment of ADHD (MTA) study.[14] The MTA study used a well-controlled scientific method to compare the effectiveness of stimulant medications alone (MED), behavior therapy alone (BT), or the combination of the two (MED+BT) in reducing the symptoms of ADHD and improving the general functioning of the children and their relationships. Results demonstrated clinically significant improvements for all treatments in multiple domains.[14] However, the combined treatment (MED+BT) showed greater improvement in reducing core ADHD symptoms in most functional domains, reduction of comorbid problems, and greater parental satisfaction with treatment. The inclusion of BT was associated with lower dosages of medication (25%–70%) needed compared to the MED condition alone. The BT treatment used contingency management programs that taught standard behavioral techniques such as time-out, point systems, contingent attention, teacher consultation with a daily report card for behavior, response-cost, and social skills training with the children.

For externalizing disorders, such as ODD or disruptive behavior disorder, including emerging conduct disorders, there are a few evidence-based behavioral interventions that deserve mentioning. Interventions that approach the problem through parent training include PCIT[15] and the Incredible Years.[16] Parent-child interaction therapy involves direct and individual coaching of parenting behaviors to improve the quality of the parent-child relationship through play, and improving a parent's child management skills through effective communication and the appropriate use of time-out. Clinically significant improvements in the quality of the parent-child relationship, child cooperation, and parental well-being have been documented. The Incredible Years is delivered in a group format in which parents master positive child manage-

ment skills by viewing videotaped vignettes followed by discussion and role-playing practice by the parents. The children also participate in social skills training. The Incredible Years has demonstrated clinically significant reductions in child disruptive behaviors at home and school.

A treatment that incorporates a multilevel approach with a primary focus on prevention of disruptive behavior problems is known as the Triple P-Positive Parenting Program.[17] Five levels of intervention are offered from parent education and awareness of resources to direct treatment of a child's behavior problems through a clinician. The unique aspect of the Triple-P approach is that it is coordinated within a community, spanning the primary care office to the home and family. There is evidence that parents report less frequent child behavior problems and greater parent competence and satisfaction than wait-list control families.

For adolescents diagnosed with conduct disorder and involved with substance abuse, multisystemic therapy (MST) is considered an empirically supported treatment.[18] This is a manualized intervention that provides therapy to the family and within multiple environments or systems in which the adolescent evidences maladaptive behaviors. The systems include the family, school, peer groups, neighborhood, family social supports, and juvenile justice and child welfare systems. Therapeutic change is focused on the adolescent within each of these psychosocial systems by enhancing contingency management parenting skills, communication skills, and effective discipline techniques. Multiple researchers have demonstrated significantly improved family functioning, decreased antisocial behaviors, and decreased psychiatric symptoms.[18]

For those primary pediatric health care professionals with an interest in learning more about evidence-based behavioral interventions, many professional psychological associations have promoted knowledge of evidence-based practice in the treatment of mental health disorders. The Society of Pediatric Psychology (Division 54 of the American Psychological Association) is one. Its flagship journal has published a special series regarding empirically supported treatments in pediatric psychology (www.jpepsy.oxfordjournals.org).

Developmental Disorders

Pervasive Developmental Disorders and Intellectual Disability/ Mental Retardation

Pervasive developmental disorders, such as autism, and the cognitive disabilities, such as intellectual disability (mental retardation), have been monitored and measured in the population. The Centers for Disease Control and Prevention (CDC) reports an overall average prevalence of 9.0 per 1,000 population (95% confidence interval = 8.6–9.3), or 1 child in every 110 meeting the criteria for autism, and this figure appears to be rising.[19] The CDC reports that 1 in 83 children (12 per 1,000 8-year-olds) meet the criteria for intellectual disability (IQ <70), and this figure

appears to be falling.[20] Barbaresi et al[21] explain these shifts in prevalence due to the influence of refinements in the diagnostic process and changes in educational entitlements, especially for autism.

Although autism and intellectual disability are defined by specific criteria and can co-occur, both are generally associated with significant impairments in cognitive, communicative, self-help, and social functioning. These impairments place these children at a profound disadvantage when it comes to naturally learning from the familial and social milieus within which they are developing. Further, there is no psychopharmacologic intervention that directly treats the core symptoms of autism or intellectual disability.[22] Therefore, evidence-based behavioral and educational interventions are the primary treatment interventions to support children with autism and intellectual disability in achieving their optimal developmental potential.

Over the past 40 years a large body of literature has shown the successful use of applied behavior analysis (ABA) to reduce problem behavior, increase appropriate skills, and enhance learning in individuals with intellectual disability, autism, and related disorders.[23] Applied behavior analysis treatment is based on clearly defining the deficits and excesses of the disorders in observable behaviors, systematically manipulating the immediate learning environment to determine the effect on the target behaviors (both deficits and excesses), and applying the empirically supported laws of learning to improve the adaptive functioning of children with autism and/or intellectual disability in school or clinic settings. Since the earliest efforts to use the ABA approach in the 1960s, there has been a tremendous contribution to the lives of children who were previously thought to be relatively unteachable. Although ABA has become a widely recognized and effective treatment for autism,[24] the same behavioral methods were developed and have been demonstrated to be effective for children who have intellectual disability without an autism spectrum disorder.[25–27] The criteria for effective and appropriate ABA treatment of children with autism and/or intellectual disability include a behavioral emphasis, family participation, one-to-one instruction, integration with typical learning environments, comprehensiveness, intensity, recognition of individual differences, and close quality control.[28]

Children who meet the criteria for intellectual disability show significant variability in functioning, from children in the mild range, who may be fully integrated into regular classrooms with appropriate levels of support, to children in the profound range of impairment, who require total care and support just to obtain the most basic necessities of life. The individualized and evidence-based approach of ABA is quite useful for children with intellectual disability and their optimal learning and development. However, there has been a greater emphasis on children with intellectual disabilities more fully participating in typical educational environments, as that peer group provides ample opportunities for appropriate modeling of desired behaviors. The multiple reauthorizations of the Individuals with Disabilities Education Act (PL 105-17) have stressed the importance of inclusion

and collaboration in the educational setting for children with intellectual disability. Providing these children the chance to participate in the *least restrictive environment* requires educators to implement Individualized Education Programs (IEPs) in the regular classroom, directly to the child, and while collaborating with all other professionals needed to support the IEP (special education teacher, speech therapist, school psychologist, physical and occupational therapist, etc). Further, the standard for an effective, individualized education requires documentation that the stated educational goals are being achieved. The ABA approach, which emphasizes data-driven intervention, becomes an important tool in which to support the philosophic goals of inclusion and collaboration.

In addition to advocating for evidence-based ABA methods in special education settings, primary pediatric health care professionals should identify professionals in the community that have familiarity with evidence-based ABA approaches to treating children with autism and/or intellectual disability. A professional organization that recently emerged to provide public education and certification of professionals who proclaim to have expertise in applied behavior analysis is that of the Behavior Analyst Certification Board. This organization maintains a registry of certified behavior analysts that can be accessed at www.bacb.com. The primary professional organization for mental health professionals interested in the applied behavioral approach is the Association for Behavior Analysis International. This organization maintains a directory of members and can be accessed at www.abainternational.org.

Learning Disorders

During the health maintenance visit, primary pediatric health care professionals should always inquire about a child's progress in school. Simply asking to review report cards, teacher comments, and any group achievement test scores with the parent can highlight the necessity for further detailed assessment by a psychologist or educational specialist. Often, children who are struggling in school will not be referred for evaluation of possible learning disorders until they actually fail. The primary pediatric health care professional plays an important role in early detection and referral for effective treatments. Learning disorders are defined by the US Office of Education[29] as "…a severe discrepancy between achievement and learning ability in one or more areas, including oral expression, written expression, listening comprehension or reading comprehension, basic reading skills, mathematic calculation, mathematic reasoning, or spelling" (see Chapter 15). Noticing emerging evidence for learning inefficiencies in one or more of these domains should prompt the primary pediatric health care professional to guide the parents through the referral process in the public schools for a learning assessment. All children are entitled to a fair and appropriate education, and when satisfactory learning progress is not occurring, a free assessment by the public school may be needed.

Whether a specific learning disability is documented, or whether the problem is just a mild learning weakness, the primary pediatric health care professional should be

aware of effective educational interventions for learning disorders that are available in the school and community settings. This becomes a difficult task, as the field of education has a long and disappointing history of promoting instructional methods and curricula that are without evidence and simply do not work.[30,31] However, scientific reviews of the educational literature support one type of instructional method that has demonstrated efficacy in an evidence-based manner. This method is known as direct instruction (DI).[32]

Direct instruction is characterized by features such as teacher-directed, well-organized instructional materials that are individualized for the student; brisk pacing with verbal responding by the student; and the use of social reinforcement. A meta-analysis that reviewed all of the randomized controlled trials of DI over a 25-year period for both reading and math instruction definitively supports DI as an effective teaching strategy.[32] Results indicated effect sizes of 0.68 for reading and 1.11 for math. Effect sizes of 0.25 are considered statistically significant, and effect sizes of the magnitude produced by DI are rare in educational research. Primary pediatric health care professionals are not directly involved in educational interventions for children with learning problems, but they play an important role in providing information to parents regarding what types of educational interventions are effective, so that they can be requested of the public schools or from private tutoring agencies. A wonderful resource for parents and teachers about effective teaching strategies, such as DI, can be found at www.ldonline.org.

Evidence-based Psychological/Behavioral Interventions for Use in Primary Care Offices

Primary care management of behavioral problems in the office setting is reviewed extensively in Chapter 4. The following section of this chapter reviews evidence-based behavioral interventions that can be most easily incorporated into primary care practice.

Behavioral Treatment of Elimination Disorders

Toileting difficulties, including enuresis and encopresis, are common presenting concerns encountered by primary pediatric health care professionals during health maintenance visits. In fact, prevalence studies report that 8% to 10% of school-aged children will meet the criteria for enuresis, and 1% to 3% of children will present with encopresis.[33] Years of research have demonstrated the effectiveness of the urine alarm as an evidence-based behavioral intervention for treating simple nocturnal enuresis or bedwetting, with cure rates consistently reported in the 70% to 80% range.[34] Treatment will usually take 16 to 20 weeks to complete.

Encopresis treatment frequently requires bowel clean-out and prevention of recurrence of constipation, followed by behavioral interventions aimed at reestablishment of regular toileting and taking responsibility for personal hygiene. Behavioral inter-

ventions can promote the necessary lifestyle changes for successful recovery, improve cooperation between parent and child, and encourage total independence in toileting.[35]

Motivational Interviewing

Motivational interviewing (MI) is a patient-centered counseling style that helps patients recognize and do something about problems by building on intrinsic motivation.[36,37] It is especially useful for patients who are reluctant to change. Motivational interviewing creates a partnership with the patient to explore and resolve ambivalence about behavioral change, and get them moving on the path to change. Although most well studied for substance use and obesity, MI has been applied to other behavioral goals. It is practical in focus, with the goal of increasing the patient's intrinsic motivation to change from within rather than change being imposed extrinsically. It can be contrasted with CBT, which is a directive approach emphasizing skill training that assumes the patient is already motivated to change and in the "action" stage. Therefore, CBT emphasizes teaching how to change, rather than why to change.[38]

Motivational interviewing is based on a transtheoretical model developed 25 years ago by Prochaska and DiClemente, and involves what they termed "stages of change," which is a continuum of readiness to change behavior. The stages range from those who are not interested in change (precontemplation stage), those who are committed to change (determination/preparation stage), and those whose behavior change has already started (action stage) to those whose behavior change has been well-established (maintenance). The primary pediatric health care professional matches counseling strategy to patient readiness.[39] Motivational interviewing consists of the following 5 broad principles:

1. Express empathy.
2. Develop discrepancy.
3. Avoid argumentation.
4. Roll with resistance.
5. Support self-efficacy.

Accepting patient beliefs and behaviors leads to the ability to express empathy with the patient and therefore facilitate behavior change. Following this, develop discrepancy by recognizing inconsistencies between current behavior and potential goals. Avoid argumentation and head-to-head confrontations by using a gently persuasive style to increase awareness of problems and the need to do something about these problems. Roll with resistance (eg, arguing, denying there is a problem, etc) by recognizing ambivalence about behavior change is a normal feeling. When the patient recognizes a problem and believes he or she can do something about it, support self-efficacy by expressing optimism and identifying effective alternatives for the patient to reach his or her goal.[38]

Motivational interviewing employs open-ended questions, reflective listening, affirmations, and summaries to build rapport. Reflective listening consists of nonjudgmental restatement, clarifications, or expansion of what the patient has said. Patients are encouraged to talk about their thoughts when open-ended questions are used. Affirmations are used to express appreciation with the patient (eg, "Thank you for being so honest with me."). Summaries help outline how a patient's expression of thoughts or feelings fit together.[39]

A behavior change plan is appropriate when the patient is in the preparation for change stage, or close to ready for an immediate change. Sample components might include the following:

The changes I want to make are:
The reasons why I want to make these changes are:
The steps I plan to take in changing are:
The ways other people can help me are:
Some things that could interfere with my plan and possible solutions are:

Comprehensive MI can be labor intensive and time consuming, but variations including the key strategies described above can be adapted for use in primary care offices.

Hypnosis and Biofeedback

One final brief comment on the treatments known as hypnosis and biofeedback is warranted. These treatments possess some empiric support and are considered to be promising interventions primarily for treating chronic pain and pain associated with medical procedures and may be considered as adjunctive treatments for sleep and elimination problems.[40,41]

Conclusion

Primary pediatric health care professionals play an important role in the behavioral and mental health care of children, as they are typically the first professionals to recognize developmental-behavioral problems in the pediatric population. Further, they are responsible for accurately assessing and referring children with problems of development and behavior for effective evidence-based treatments. As there is certainly no shortage of unproven and potentially harmful (medically or financially) therapies that are widely marketed specifically for the treatment of children with developmental-behavioral problems, it is critical for primary pediatric health care professionals to be knowledgeable about those relatively few psychological/behavioral interventions to which to guide families for evidence-based treatment of their children with developmental-behavioral problems.

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Chapter 20

Principles of Psychopharmacologic Management

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Basics of Psychotropic Medication Management

This chapter addresses psychopharmacologic management for primary pediatric health care professionals who are increasingly called on to provide assessment and treatment for children and adolescents with disorders that require psychopharmacologic intervention. There are far too few physicians with expertise in severe psychopathology, such as child and adolescent psychiatrists or neurodevelopmental or developmental-behavioral pediatricians. It is often left to primary care clinicians to provide care for children and adolescents with complex neurodevelopmental and behavioral disorders.

The past 2 decades have seen a dramatic increase in both the absolute numbers of prescriptions written for psychotropic medication in children and adolescents and also a widening range of medications used to treat behavioral and emotional problems in children and adolescents.[1–4] There are extraordinary differences in the rates of prescription prevalence between the United States and other developed countries, with a much higher rate in the United States.

The rate of growth of new information regarding pediatric psychopharmacology is rapid. It is a challenge, particularly for the primary care clinician, to differentiate well done studies with meaningful results from other investigations. Federally funded investigations may have had more valid conclusions compared with studies funded by the pharmaceutical industry. The task for clinicians is to follow the growing evidence base, to accurately diagnose and treat children, putting those who would likely benefit from medications on carefully administered trials and not substituting psychopharmacologic interventions for problems that more appropriately require individual or family psychotherapy, or no treatment at all.

Most commonly prescribed are the stimulant medications.[5] There also have been sizable increases in the prescribing of antidepressant medications, particularly selective serotonin reuptake inhibitors (SSRIs), and for atypical antipsychotics. Embedded in these overall figures, however, are more subtle observations. The rise in stimulant use has been steady, at about 10% per year for the last several years, but the growth in the last 5 years has been principally in medications prescribed in adults. As to the SSRIs, there had been a steady increase for several years in the number of prescriptions written per year in the United States. This occurred in the context of substantial advertising regarding the use of antidepressants to both health care providers and consumers. Between 1998 and 2002 there was an annual adjusted rate of increase of 9.2%. However, there has been a 20% reduction since the recent controversy regarding potential increases in suicidal thinking and behavior (suicidality) associated with antidepressant use, and the subsequent US Food and Drug Administration (FDA) black box warning issued in October 2004.[6]

Regarding the atypical antipsychotic medications, there has been a 6-fold increase in the last 10 years.[7] Whether rising concerns regarding their side effects will affect changes in prevalence over time remains to be seen.

In consideration of the increased prescriptions, a corollary question is whether this reflects best practice. A more specific question is whether too many or too few children are being treated with psychotropic medications. There is evidence for both sides of the argument. For example, in a study of children sampled across the country, three-quarters of the children who met criteria or a diagnosis of attention-deficit/hyperactivity disorder (ADHD) were not on stimulant medication.[8] In the Great Smokey Mountain Project study, of those children who were being treated with stimulant medication, only one-quarter had met criteria for ADHD.[9]

Clinical practice has at times exceeded the existing evidence base. There is a very limited evidence base to support the use of even 2 medications at once, with some support for combinations of stimulant medications and alpha II agonists, stimulant medications and atomoxetine, stimulant medications and SSRIs, and atypical antipsychotic medications and mood stabilizing anticonvulsant medications.[10] There is no study that supports the use of more than 2 medications at a time. Clearly, it is difficult to ascertain relative benefit and unique contributions of medications when a complex psychopharmacologic array is being prescribed.

Polypharmacy is an increasingly common practice, with children and adolescents being treated with several psychopharmacologic medications at one time.[11,12] Surveys have indicated that it is common practice to have children on 4 or 5 medications at one time. An unfortunate reality is that clinicians are more likely to add medications rather than reevaluate the diagnosis and treatment when other than a robust response to treatment has been obtained. Most importantly, when a child is on multiple psychotropic medications there must be careful attention to costs and benefits.

Psychotropic medication treatment should be based on a psychiatric diagnosis, or on a specific behavioral pharmacologic hypothesis that results from a diagnostic and functional assessment (target symptom approach). That assessment should address the following:

- Medical pathology
- Psychosocial and environmental conditions
- Health status (with particular attention to ruling out pain)
- Current medications (including over-the-counter medications)
- Presence of any psychiatric diagnosis
- Response to previous intervention
- A functional analysis of behavior

In a pediatric population, initial dosing should be low and titrated slowly. There should be periodic efforts to test the effects of dosage. Frequent drug and/or dose changes should be avoided if at all possible. It is important to collect baseline data for initiation of treatment, identify specific behavioral indices, and track those behavioral indices using sound behavioral principles (eg, using frequency counts, time sampling, and rating scales). Outcome data should be collected, and treatment objectives need to be regularly evaluated.

Any psychopharmacologic approach should be part of an overall and integrated care plan with careful communication between all of the relevant parties, including parents, teachers, primary care medical home, and consultants. There is evidence of synergy of effects of medication with other psychosocial interventions, as demonstrated in the Multimodal Treatment of ADHD (MTA) study in which the combined form of treatment was expert psychopharmacologic management coupled with intensive behavior management.[13] While there were comparable levels of improvement in ADHD symptoms with medication alone and combined treatment, a reduction in the dosages of stimulant medication was obtained for those children treated with a combined treatment of medication and behavior management compared with medication alone. In addition, specific symptom areas were improved with combined treatment, including social skill development, anxiety regulation, and aggression. Similarly, in the Treatment of Adolescents with Depression Study (TADS), the combined treatment arm of cognitive-behavioral therapy (CBT) with medication (fluoxetine) compared with medication alone was associated with a numerically superior (albeit not statistically significant) difference in response rate and also a lower rate of emergence of suicidal ideation and attempts.[14] While there are barriers, adding psychosocial interventions to the care plan is important to achieve the best outcomes.

This chapter specifically addresses psychopharmacology. Other chapters in this manual focus on assisting primary pediatric health care professionals in the recognition, diagnosis, and broader management of developmental and behavioral disorders. While the focus in this chapter is on medication, it is critical to understand

that medication is most helpful when the comprehensive treatment needs of the child or adolescent and family are being addressed. The overall goal of drug therapy for mental health conditions is not curative, but to control symptoms. In most cases non-pharmacologic treatment will also be needed.

There are other key principles that must be considered when developing an evidence-based management plan for psychotropic medication use. Developmental factors must be taken into account. This includes use of appropriate dosage by weight, but also includes consideration of the effect of development on medication responsiveness. For example, the evidence with regard to ADHD is that dosage is lower on a milligram per kilogram basis for preschoolers compared with older children.[15] There was also a higher discontinuation rate in young children as well as more irritability and emotional outbursts. It is also essential to recognize that when a psychotropic medication prescription is provided, there is the potential for complicated dynamics that involve at least the child and their parents, but may also involve extended family members, who can have strong opinions regarding medications. Many medications are often prescribed in the context of school and must involve the perspectives of teachers and school authorities. General principles of psychopharmacologic management also include[16]

- First, do no harm.
- Establish a therapeutic relationship.
- Know the disorder and use drugs when indicated.
- Choose the best drug.
- Understand the drug and its properties.
- To the extent possible, minimize medication usage and dose.
- Try to keep things simple.
- Avoid polypharmacy if possible.
- Don't follow fads.
- Take special care with children.
- Ensure adherence to treatment with family and school/early education centers.

A number of psychiatric disorders have well-established indications for use of psychotropic medications. This is particularly true for ADHD, for which there have been many studies with large numbers of children reported in the scientific literature. There also has been considerable research in the area of depression in children and adolescents with some support, but also with questions regarding the cost–benefit ratio of medications in that population. There is research evidence supporting benefit of psychotropic medication use in anxiety disorders, especially for obsessive compulsive disorder (OCD) but also for other anxiety disorders.

While no psychotropic medication has a precisely known mechanism of action, they are thought to exercise their effects on modulation of one or more neurotransmitters, particularly dopamine, norepinephrine, serotonin, and the inhibitory neurotransmitter gamma-aminobutyric acid. Whereas some medications, such as stimulants, are

indicated specifically for the treatment of a single disorder (ADHD), other medication classes, such as SSRIs and atypical antipsychotic medications, have demonstrated efficacy for a number of disorders. Also, a number of disorders have multiple medication classes that have been used effectively. Medications are often considered for symptoms that do not aggregate into specific diagnoses, such as aggression, self-injury, mood dysregulation, and sleep difficulties.

There is evidence for effectiveness of psychotropic medication in children and adolescents for a number of diagnoses, for which there are formal FDA indications and substantial evidence-based support. There are also disorders and symptoms that have yet to secure FDA indications, but for which there is evidence from randomized, controlled trials with robust numbers of subjects in the investigations. Clinical practice in youth, however, extends to other medications for which there is either only open label support or no data whatsoever in this age group. Results are often extrapolated from studies with adults, which at times may not apply to children.

In this chapter there is an effort to delineate best practice. The evidence base for safe and effective treatment with psychotropic medications is reviewed, examining several psychiatric disorders and symptom clusters, including ADHD, depression, anxiety disorder, tic disorders, bipolar disorder, sleep problems, aggression, and psychoses. Multiple medication groups, including stimulants, atomoxetine, alpha-2 agonists, SSRIs, tricyclic antidepressants, benzodiazepines, atypical antipsychotics, and other mood stabilizers are reviewed. Longitudinal considerations regarding monitoring of dosage and side effects of psychotropic medications are discussed.

Attention-Deficit/Hyperactivity Disorder

Stimulant Medications

Attention-deficit/hyperactivity disorder is by far the childhood psychiatric disorder most extensively studied regarding response to medication. Most widely studied are the stimulant medications (methylphenidate, amphetamines, and dexmethylphenidate). Atomoxetine and the long-acting preparation of the alpha-2 agonist guanfacine also have FDA indications for ADHD. Atomoxetine is generally considered a second-line agent, except possibly first line when comorbid substance abuse, anxiety, or tics are present. Guanfacine is also a second-line medication for ADHD. Other medications with evidence for effectiveness include the alpha-2 agonist clonidine, bupropion, tricyclic antidepressants, and modafinil.

The best evidence is for the use of stimulant medications, which are the agents of first choice, unless there are specific reasons to consider alternative medications. In general, there is about the same response rate with amphetamines and methylphenidate. Most children with ADHD respond favorably to both methylphenidate and amphetamines. Some children respond to one stimulant class and not the other.[17] Thus either may be initiated, and if a patient does not respond or has unacceptable side effects to one class, the patient should be switched to the other class.

In the past 3 decades, there have been thousands of children studied in many randomized, double-blind, placebo-controlled trials (RDBPCTs) with stimulant medication. In a review of multiple studies of stimulant medications there was a significant improvement on active medication: 65% to 75% compared with 4% to 30% of patients treated with placebo. The effect sizes average about 1.0.[18]

Dosing limit guidelines have been articulated by the FDA, but it is common practice that some patients are treated with higher doses. This should be done with caution and only in children in whom there is relative freedom from side effects and for whom lower dosages have resulted in partial but not full response.

Prescription medications should always be part of an overall, comprehensive treatment plan that requires a positive therapeutic alliance with the patients and their parents, assent from young children, and consent from older children and parents/guardians, as dictated by state regulations. Issues with noncompliance are common in the treatment of ADHD and must be taken into account in assessment of treatment effect.[19]

Medications used to treat ADHD are generally very well tolerated. The discontinuation rate because of side effects for stimulant medications, for atomoxetine, and for alternative non-FDA approved medications is very low. That said, a significant percentage of patients treated with stimulants, particularly early in treatment, will have side effects. The most common are headaches and stomachaches. Also, potentially problematic are reduced appetite and possible accompanying growth velocity deceleration, sleep initiation problems, and irritability. Some children will have exacerbation of preexisting tics or development of tics that had not been previously present. Whereas FDA guidelines indicate that stimulants are contraindicated in the presence of tics, there is no evidence that the longitudinal course of tics is affected by treatment with medications for ADHD.[20] The consensus of experienced clinicians is to cautiously initiate stimulants in the presence of tics and continue treatment with a cost–benefit analysis as treatment proceeds, should tics occur or worsen during the course of treatment. Irritability is the most problematic adverse behavioral effect of stimulants and may require shifting to alternative medications. Dysphoria and frank sadness may also occur and should be mentioned to parents.

It is important to have trials with adequate dosage and with alternative medications. It is not uncommon for clinicians to begin at a relatively low dose and not to advance medication throughout the typical range in which response can reasonably be expected, or to not switch medications if no benefit is obtained from the first medication even with maximum dosage. An adequate titration should be done; however, dosage should not be advanced up to the point of generating prominent side effects.

Attention-deficit/hyperactivity disorder is a chronic disorder, with most individuals continuing to have signs and symptoms over time.[21] Follow-up studies have been as long as 15 years after initial diagnosis, with over half having enduring significant

symptoms. Some children will demonstrate continued need for and benefit from medication treatment over several years. Others may be able to function satisfactorily without medication sometime after treatment initiation.

After initiation of medication, children should have follow-up appointments approximately once a month until they are stable on their medication as to therapeutic benefit, dosing, and side-effect management. There should be a facile means of accessing health care professionals as needed as doses are titrated between clinic appointments. Most children will stabilize in terms of their medication, both as to specific medication and dose, within 3 months. Once stable, patients should be seen on an ongoing basis every 3 to 4 months. In addition to procuring information from the patient and family regarding medication response and overall functioning, there should be regular contact with the school system in order to help monitor the therapeutic response and report results for the follow-up visits at 3 to 4 months. Height, weight, and vital signs should be obtained at each visit. Laboratory tests and electrocardiograms (ECGs) are not routinely required, but should be done if there is a clinical indication (eg, complete blood count [CBC] if malnutrition is suspected; ECG if any personal or family history of cardiovascular symptoms [eg, syncope] or sudden unexplained cardiac death). The persistent need for medication treatment for ADHD should be systematically reevaluated at least every 2 years, either through analysis of unintended breaks in use of medication, or in those instances in which there has been steady compliance, with a trial off medication to determine whether the child continues to require medication.

The goal with medications is to optimize adaptive functioning, striking a balance between a therapeutic response and relative freedom from side effects. Most children treated with medications will not normalize, but will have at least 30% reduction in level of symptomatology from baseline, as measured by change from baseline on a standardized ADHD rating scale such as the Vanderbilt or ADHD Rating Scale-IV.[22] In children in whom less than 30% symptom reduction is achieved, or in whom prohibitive side effects occur, it is appropriate to switch to alternative medications. There is a preference by families and patients for the longer-acting stimulant medication preparations, given greater convenience and improved adherence to treatment. As such, they are the agents of first choice. It is also preferable to have a single dosing per day if at all possible. That said, the long-acting medications commonly do not provide full treatment effect for the desired durations of action of 8, 10, 12, or even 16 hours. In the past decade, there have been a number of large, well-designed studies that have confirmed the benefit of long-acting forms of methylphenidate, mixed salts of amphetamine, and dexmethylphenidate in both short-term controlled trials and long-term open-label trials.[23–27]

More caution is indicated in young children, as demonstrated in the Preschool ADHD Treatment Study (PATS).[15] It is preferable to initiate a trial of parent training and behavior management prior to the institution of medication. Given the research

studies in children younger than 6 years, methylphenidate is the medication of first choice. Dosage should begin very conservatively, and typically not at all with children younger than 3 years. For those between 3 and 6 years of age, dosage should begin with 2.5 mg twice a day. Dosage may be titrated upward every 4 to 7 days, with a maximum dose of methylphenidate in this population of 30 mg/day. Maximum benefit should occur within 2 weeks after maximum dose.

Methylphenidate is equally efficacious, but half as potent, as dexmethylphenidate and amphetamine preparations. If no benefit is obtained, and prominent ADHD symptoms are still present, it is appropriate to switch to an amphetamine-based product, beginning at a dose of 2.5 mg/day and titrating in 4- to 7-day intervals to a maximum of 15 mg/day.

There is a general relationship between dose response and weight, but it is not exact. For children 6 years of age and older, amphetamines, methylphenidate, or dexmethylphenidate are all appropriate initial medications. Initial dose of methylphenidate for children 6 years of age and older should be 5 mg 2 or 3 times a day. Long-acting preparations of methylphenidate products may be initiated at the outset of treatment at an equivalent dosage, including Concerta, Ritalin LA, or Metadate CD. Dosing titration should move forward in a 4- to 7-day interval to typically no higher than 60 mg/day, not to exceed 2 mg/kg/day. If an amphetamine is initiated, it may be done with an immediate-release preparation, such as dextroamphetamine or mixed salts of amphetamine tablets, or with longer-acting agents, including Adderall XR and Vyvanse. Dexmethylphenidate (Focalin or Focalin XR) may be initiated at dose levels similar to the amphetamines. The maximum dose for amphetamines and dexmethylphenidate is typically 30 mg/day, not to exceed 1 mg/kg/day. As mentioned, in some children, dosage may exceed these ceilings, especially in older and larger adolescents, but should very rarely exceed 120 mg/day of methylphenidate or 60 mg/day of amphetamine or dexmethylphenidate. In instances of use of higher dosages, there must be carefully documented evidence of added efficacy, and relative freedom from side effects must be present.

Best practice is to attempt to treat with a single dose of a single medication per day. While the long-acting preparations offer the benefit of requiring a single dose per day, they are much more expensive than generic preparations. The only currently available (as of 2010) generic truly long-acting preparation is the XR form of mixed salts of amphetamine. Some children require supplementation with the same stimulant medication in the afternoon. For example, if a long-acting methylphenidate preparation, such as Concerta, Metadate CD, or Ritalin LA, is used, and there is not sustained benefit up until the evening, a dose of immediate-release methylphenidate may be helpful when administered in the afternoon. Similarly, immediate-release amphetamine may be added in the afternoon to a morning dose of Adderall XR or Vyvanse if duration of effect is inadequate (Table 20.1).

Table 20.1. Medications Approved by FDA for ADHD (Alphabetical by Class) 1*

Generic Class *Brand Name*	Dosage Form	Typical Starting Dose	FDA Max/Day	Off-Label Max/Day	Comments
AMPHETAMINE PREPARATIONS					
Short-acting					
Mixed amphetamine salts *Adderall***	5, 7.5, 10, 12.5, 15, 20, 30 mg	3–5 yr: 2.5 mg qd ≥6 yr: 5 mg qd–bid	40 mg	>50 kg: 60 mg	>Short-acting stimulants often used as initial treatment in small children (<16 kg) but have disadvantage of bid–tid dosing to control symptoms throughout day
Dextroamphetamine *Dexedrine*** *DextroStat*	5 mg 5, 10mg	3–5 yr: 2.5 mg qd ≥6 yr: 5 mg qd–bid			
Long-acting					
Dextroamphetamine *Dexedrine Spansule*	5, 10, 15 mg	≥6 yr: 5–10 mg qd–bid	40 mg	>50 kg: 60 mg	>Longer-acting stimulants offer greater convenience, confidentiality, and compliance with single daily dosing but may have greater problematic effects on evening appetite and sleep
Mixed amphetamine salts *Adderall XR*	5, 10, 15, 20, 25, 30 mg	≥6 yr: 10 mg qd	30 mg	>59 kg: 60 mg	
Lisdexamfetamine *Vyvanse*	10, 20, 30, 40, 50, 60, 70 mg	30 mg qd	70 mg	Not yet known	

Table 20.1. Medications Approved by FDA for ADHD (Alphabetical by Class) 1* *(continued)*

Generic Class *Brand Name*	Dosage Form	Typical Starting Dose	FDA Max/Day	Off-Label Max/Day	Comments
METHYLPHENIDATE PREPARATIONS					
Short-acting					
Dexmethylphenidate *Focalin*	2.5, 5, 10 mg	2.5 mg bid	20 mg	50 mg	>Short-acting stimulants often used as initial treatment in small children (<16 kg) but have disadvantage of bid–tid dosing to control symptoms throughout day
Methylphenidate *Methylin*** *Ritalin***	5, 10, 20 mg	5 mg bid	60 mg	>50 kg: 100 mg	>Methylin is available as a liquid preparation
Intermediate-acting					
Metadate ER	10, 20 mg	10 mg qam	60 mg	>50 kg: 100 mg	>Longer-acting stimulants offer greater convenience, confidentiality, and compliance with single daily dosing but may have greater problematic effects on evening appetite and sleep
Methylin ER	10, 20 mg				
*Ritalin SR***	20 mg				
Metadate CD	10, 20, 30, 40, 50, 60 mg	20 mg qam	60 mg	>50 kg: 100 mg	>Metadate CD and Ritalin LA caps may be opened and sprinkled on soft food
Ritalin LA	10, 20 ,30, 40 mg				

Table 20.1. Medications Approved by FDA for ADHD (Alphabetical by Class) 1* *(continued)*

Generic Class *Brand Name*	Dosage Form	Typical Starting Dose	FDA Max/Day	Off-Label Max/Day	Comments
Long-acting					
Concerta	18, 27, 36, 54 mg	18 mg qam	72 mg	108 mg	>Swallow whole with liquids >Nonabsorbable tablet; shell may be seen in stool
Daytrana Patch	10, 15, 20, 30 mg patches	Begin with 10mg patch qd, then titrate up by patch strength	30 mg	Not yet known	
Focalin XR	5, 10, 15, 20 mg	5 mg qam	30 mg	50 mg	>Capsules may be opened and sprinkled on soft food
SELECTIVE NOREPINEPHRINE REUPTAKE INHIBITOR					
Atomoxetine					>Not a Schedule II medication, can be refilled
Strattera	10, 18, 25, 40, 60, 80, 100 mg	Children and adolescents <70 kg: 0.5 mg/kg/day for 4 days; then 1 mg/kg/day for 4 days; then 1.2 mg/kg/day, given in a single qam or hs dose or bid	Lesser of 1.4 mg/kg or 100 mg	Lesser of 1.8 mg/kg or 100 mg	>Consider if active substance abuse or severe side effects of stimulants (mood lability, tics) >Give qam or divided doses bid (effects on late evening behavior) >Do not open capsule >Monitor closely for suicidal thinking and behavior, clinical worsening, or unusual changes in behavior
Guanfacine *Intuniv*	1, 2, 3, 4 mg	1 mg titrate weekly to effectiveness	4 mg	Not yet known	>Monitor BP weekly during titration phase >Taper when discontinuing to avoid rebound hypertension

* Modified from: Practice Parameter for the Assessment and Treatment of Children and Adolescents with Attention-Deficit/Hyperactivity Disorder. *J Am Acad Child Adolesc Psychiatry*. 2007:46(7):894-921. Reprinted by permission of Lippincott Williams & Wilkins.

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Atomoxetine

For those children who do not respond to stimulant medications, an FDA-approved agent for use in ADHD is atomoxetine (Strattera), which is a noradrenergic reuptake inhibitor. Randomized, double-blind, placebo-controlled trials have shown benefit.[28] Direct comparisons of atomoxetine to stimulants have shown a greater treatment effect for methylphenidate.[29] In a meta-analysis of atomoxetine and stimulant studies, the effect size for atomoxetine was 0.62 compared with 0.91 and 0.95 for immediate-release and long-acting stimulants respectively.[30] It may be an alternative, particularly for those children who do not respond to a stimulant, develop irritability or severe appetite suppression while on stimulant medication, or have a history of substance abuse or anxiety symptoms.[31] There is no published experience with this agent in children younger than 6 years, and use in that age group should be cautiously implemented. For children older than 5 years, dosage begins at approximately 0.5 mg/kg/day and can be titrated up every 4 to 7 days up to a ceiling dose of the lesser of 100 mg or 1.4 mg/kg/day. Many children will not respond immediately to atomoxetine, and it may be necessary to keep them on a maximum dose for at least 4 to 6 weeks to ascertain full benefit, much longer than the time to observe benefit with stimulant medication.[31] It is usually effective given once a day, in the morning or at bedtime. Dosage can be given twice a day, which may reduce side effects.

Side effects with atomoxetine include appetite suppression, headaches, nausea, irritability, and possibly tics. In contrast to stimulants, however, it does not tend to exacerbate sleep problems, and some children are sedated. It is important to note that the FDA warning regarding possible emergence of mood-related side effects relevant to antidepressant medications extends to this agent, though mood-related side effects are uncommon with atomoxetine (0.4% vs 0 on placebo). Atomoxetine has shown benefit in patients with co-occurring ADHD and anxiety disorders.[31]

Alpha-2 Receptor Agonists and Antidepressants

There are other medications with evidence of effectiveness and safety in treatment of ADHD but without an FDA indication (Table 20.2). Two noradrenergic alpha-2 receptor agonists, clonidine and guanfacine, may be used alone for the treatment of ADHD, with evidence, especially for guanfacine, of reduction of impulsivity and hyperactivity.[32,33] Extended-release guanfacine (Intuniv) is now available with a specific indication for the treatment of ADHD. Clonidine and guanfacine are typically used as adjuncts to stimulant medication or atomoxetine for persistent hyperactivity, impulsivity, or aggression not responsive to other medication treatment, for sleep induction, or to address exacerbation of tics associated with alternative ADHD medications.

Dosing with clonidine (which does not have an FDA indication for ADHD) for children weighing less than 45 kg, should begin at 0.05 mg by mouth at bedtime and can be titrated every 4 to 7 days to a maximum of typically no more than 0.2 mg/day. Dosing for children weighing more than 45 kg begins at 0.1 mg by mouth at bedtime

and can be titrated in 0.1 mg increments up to a maximum typically of 0.4 mg/day given 2 to 4 times per day. The potency of guanfacine is one-tenth of clonidine. Therefore, children weighing less than 45 kg should begin guanfacine at 0.5 mg by mouth at bedtime and increase by 0.5 mg increments up to a 4 times per day dosing, to a maximum daily dose of typically no more than 2 mg/day. For children weighing more than 45 kg, begin at 1 mg by mouth at bedtime and titrate up to a 4 times per day dosing of typically no more than 4 mg/day given 2 to 4 times per day. The new extended-release guanfacine preparation (Intuniv) is available in doses ranging from 1 mg to 4 mg once daily. Side effects include sedation (especially with clonidine), depression, and hypotension. Care also must be given to avoid rebound hypertension by tapering slowly if a patient has been on an alpha-2 agonist for more than 2 weeks.

In the event that stimulant medications and atomoxetine are not successful, some antidepressant medications have also been successfully employed with the treatment of ADHD. Bupropion (Wellbutrin) is a noradrenergic antidepressant, which comes in a number of preparations. Bupropion can lower the seizure threshold. There is less evidence of lowering the seizure threshold in the long-acting XL preparation of this medication, and therefore the long-acting XL form, though more costly, is the agent of choice. Dosage should begin with the lesser of 3 mg/kg or 150 mg/day and titrated every 4 to 7 days to the lesser of 6 mg/kg/day or a total daily dose of 450 mg, with no single dose greater than 150 mg.

There is some evidence of benefit with tricyclic antidepressants in the treatment of ADHD, including desipramine, imipramine, and nortriptyline. Concerns regarding possible cardiovascular toxicity, including sudden death, make these agents less desirable in this era and should be reserved for extraordinary cases.[34] It is essential to obtain a baseline ECG and serial monitoring of ECG and blood levels, being mindful of the development of a prolonged QTc interval and with blood levels of imipramine not to exceed 400 ng/mL, or in the case of nortriptyline, 200 ng/mL. The beginning dose with imipramine is 1 mg/kg/day, advanced to a maximum dose of 5 mg/kg/day or not to exceed 200 mg. For nortriptyline the dosing is half that for imipramine.

Recent Developments

There have been several recent developments regarding ADHD. An FDA warning was issued in August 2006 regarding all stimulant medications and possible cardiovascular risks and possible psychiatric side effects, including psychosis.[35] While this will hopefully lead to increased monitoring of vital sign parameters as well as growth issues, it is not clear that there are significant cardiovascular risks in otherwise healthy children, and the development of psychosis is very rare.

Recently there has been an appreciation that ADHD symptomatology present in children with other diagnoses, such as autism spectrum disorders (ASDs), may obtain some benefit from the agents traditionally used to treat ADHD, including

Table 20.2. Medications Used for ADHD, Not Approved by FDA*

Generic Class *Brand Name*	Dosage Form	Typical Starting Dose	FDA Max/Day	Comments
Bupropion				>Lowers seizure threshold; contraindicated if current seizure disorder >Usually given in divided doses, bid for children, tid for adolescents, for both safety and effectiveness >Obtain baseline ECG before starting imipramine and nortriptyline
*Wellbutrin***	75, 100 mg tab	Lesser of 3 mg/kg/day or 150 mg/day	Lesser of 6 mg/kg or 600 mg, with no single dose >150 mg	
Wellbutrin SR	100, 150, 200 mg tab			
Wellbutrin XL	150, 300 mg tab			
Imipramine				
*Tofranil***	10, 25, 50, 75 mg tab	1 mg/kg/day	Lesser of 4 mg/kg or 200 mg	
Nortriptyline				
*Pamelor*** *Aventil***	10, 25, 50, 75 mg tab	0.5 mg/kg/day	Lesser of 2 mg/kg or 100 mg	
ALPHA-2-ADRENERGIC AGONISTS				
Clonidine				
*Catapres***	0.1, 0.2, 0.3 mg tab	<45 kg: 0.05 mg qhs: titrate in 0.05 mg increments bid, tid, qid >45 kg: 0.1 mg qhs; titrate in 0.1 mg increments bid, tid, qid	27–40.5 kg: 0.2 mg 40.5–45 kg: 0.3 mg >45 kg: 0.4 mg	>May be used alone or as adjuvant to another medication for ADHD >Effective for impulsivity and hyperactivity; modulating mood level; tics worsenin from stimulants; sleep disturbances >May not see effects for 4–6 weeks >Review personal and family cardiovascular history >Taper off to avoid rebound hypertension
Guanfacine				
*Tenex***	1, 2 mg tab	<45 kg: 0.5 mg qhs; titrate in 0.5 mg increments bid, tid, qid >45 kg: 1 mg qhs; titrate in 1 mg increments bid, tid, qid	27–40.5 kg: 2 mg 40.5–45 kg: 3 mg >45 kg: 4 mg	

* From: Practice Parameter for the Assessment and Treatment of Children and Adolescents with Attention-Deficit/Hyperactivity Disorder. *J Am Acad Child Adolesc Psychiatry.* 2007;46(7):894–921. Reprinted by permission of Lippincott Williams & Wilkins

**generic formulation available

methylphenidate, guanfacine, and atomoxetine.[36] Under *Diagnostic and Statistical Manual, Fourth Edition, Text Revision* criteria, if ADHD symptoms occur only in the context of an ASD, a diagnosis of ADHD cannot be made. Previous reports suggested that stimulant medications were contraindicated in this population. However, more recent investigations have demonstrated that there is benefit with these agents, although less robust evidence, in this highly complex population.

There has been extension of the database to young children with the PATS.[15] This is the largest study to date of children younger than 6 years with ADHD treated with stimulant medication (methylphenidate). There was a rigorous treatment protocol that required children and their families to go through behavior management protocols prior to the initiation of medication treatment. As a result, there was considerable patient attrition. However, in the children who did complete the study, there was evidence of positive response of methylphenidate compared with placebo. Several differences were noted in terms of outcomes compared with studies of older children with ADHD treated with stimulants. The doses that were optimally beneficial were substantially lower than in trials of older children, with the mean optimal dose of 14 mg/day. There was a higher rate of side effects with stimulant medications. Finally, while the response was positive, the rate of response or degree of benefit obtained in these investigations was less robust than studies in older populations, with an effect size ranging from 0.4 to 0.8.

There is emerging evidence regarding the potential benefit of the combination of atomoxetine with stimulant medications.[10] This should be reserved for instances in which there is a partial response to one or the other of these agents and should be done judiciously.

An alternative medication to treat ADHD is modafinil, which has an FDA indication for narcolepsy and arousal issues associated with sleep apnea. This medication has shown benefit in controlled trials for ADHD, with an effect size similar to atomoxetine. It had been submitted for FDA approval for treatment of ADHD in children and adolescents but was withdrawn following emergence of a possible Stevens-Johnson–type rash, though no cause and effect was established. Use should be reserved for patients with debilitating ADHD who are unresponsive to the medications described previously.

There has been a continuation of the development of a number of delivery systems of medications used to treat ADHD, including the transdermal methylphenidate patch (Daytrana) released in June 2006.[37] This preparation provides steady methylphenidate release while the patch is applied, with therapeutic effect for up to 3 hours after removal of the patch. It has an indication for benefit up to 12 hours with the instruction to leave the patch on for 3 hours less than the desired effect, up to 12 hours. However, at the ninth hour, 41% of the methylphenidate remains in the patch that could still diffuse across the membrane, thus allowing for a potential duration of

action of longer than 12 hours, though the side effect burden may be higher. It is helpful for those children who are unable to swallow medication, or who have significant difficulties with an up and down course of response to medications over the day or do not have a long enough duration of response. Dosing equivalence between the transdermal patch and oral methylphenidate is not exactly established, but the patch is roughly twice as potent.

A novel product, lisdexamfetamine (Vyvanse), an amphetamine prodrug in which amphetamine is bound to the amino acid lysine, has been created.[38] This preparation is not biologically active until lysine has been cleaved. There is little biological effect if the medication is inhaled or injected intravenously. There had been hope that this would have a lower abuse potential and the likeability indices with this agent were lower; however, it was released in June 2007 under the same Schedule II status as all of the other stimulant medications.

Another longer-acting medication in the developmental pipeline is a triphasic amphetamine capsule with a 3-bead system, such that one-third of the medication is immediately released, one-third at 4 hours, and one-third at 8 hours.[39] The duration of effect is up to 16 hours in clinical trials, but it has not yet received approval by the FDA. Nicotinic agonists are also being studied.

Depressive Disorders

Do antidepressant medications benefit children and adolescents with major depression? While there have been a number of studies, there still remain significant questions. There is much yet to be learned about the fundamental nature of depression and of the role of antidepressants in the treatment of depression in youth. Studies of 4 SSRIs (fluoxetine, sertraline, citalopram, and escitalopram) have demonstrated benefit, albeit not all robust.[14,40–45] Although in each of these studies there was a significantly greater number of responders on active drug compared with placebo, there still were a number of patients who did not respond. No other antidepressants have been shown to have efficacy to treat depression in youth in published controlled trials, and there have been several trials with negative results. In addition, no studies have been done in children and adolescents who suffer from other forms of depression, such as dysthymia. This contrasts with an extensive literature in adults demonstrating the benefit of not only SSRIs, but also many other antidepressants.

In the TADS at 12 weeks, fluoxetine alone and fluoxetine combined with CBT were superior to CBT.[14] By 18 weeks, CBT had "caught up" in terms of the percentage of responders.[42] At 36 weeks, 80% of subjects in each of the 3 treatment groups (fluoxetine CBT, fluoxetine alone, and CBT alone) had responded by the study's definition. Suicidal events (either thinking or behavior) were more common in those patients treated with fluoxetine alone (14.7%) compared with the combined treatment (8.4%) and CBT alone (6.3%).

Table 20.3. Published Double-Blind, Placebo-Controlled Trials of Antidepressants for the Treatment of Major Depression in Children and Adolescents, With Statistically Significant Positive Results

Medication	Duration of Treatment, wks	No. of Patients	Mean Dosage, mg/day	Responders Active Drug, %	Responders Placebo, %
Fluoxetine					
Emslie et al[40] 1997	8	96	20	56	33
Emslie et al[41] 2002	9	219	20	65	53
TADS[14] 2004	12	439	33 [28[a]]	61 [71[a]]	35
Other SSRIs					
Sertraline					
Wagner et al[43] 2003	10	376	131	69	59
Citalopram					
Wagner et al[44] 2004	8	174	24	36	24

Abbreviations: SSRI, selective serotonin reuptake inhibitor; TADS, Treatment for Adolescents with Depression Study.
[a] Fluoxetine plus cognitive-behavioral therapy.

Given the evidence base, it is reasonable to consider fluoxetine as the SSRI of first choice in the case of major depression in a child or adolescent. There are FDA indications for fluoxetine and escitalopram for depression in youth. If the patient is not tolerating a trial of fluoxetine or if there is a family history of a positive response to an alternative SSRI, the first recommendation is an SSRI other than fluoxetine. If there has been an adequate trial of fluoxetine, defined by adequate dosage of at least 20 mg/day for at least a month, the secondary choice is an alternative SSRI (Tables 20.3 and 20.4).

Support for this approach comes from the Treatment of Resistant Depression in Adolescents (TORDIA) study.[46] In a randomized controlled trial, 334 patients from 12 to 18 years of age with a primary diagnosis of major depression who had not responded to a 2-month trial with an SSRI were studied. Patients were assigned to 1 of 4 interventions: switch to a second SSRI, either paroxetine, citalopram, or fluoxetine 20 to 40 mg/day; switch to a second SSRI plus CBT; switch to venlafaxine 150 to 250 mg/day; or switch to venlafaxine plus CBT. The main outcome measures were a Clinical Global Impression-Improvement (CGI-I) of 2 or less and a decrease of at least 50% on the Childhood Depression Rating Scale-Revised. Results were significant and best for a switch to either medication with CBT (with a response rate of 54.8%); with a medication switch alone 40.5% responded. There was no difference in response between venlafaxine and a second SSRI, but there were more side effects on venlafaxine, including increased diastolic blood pressure and pulse and a rash. There were no differences in any of the 4 treatment arms in the rate of suicidal thinking or behavior.

Table 20.4. Selective Serotonin Reuptake Inhibitor Dose Guidelines

Medication	Initial Daily Dose, mg	Dose Range, mg
Fluoxetine	5–10	10–60
Sertraline	25–50	50–200
Paroxetine	5–10	10–60
Fluvoxamine	25–50	100–300 twice daily
Citalopram	5–10	10–60
Escitalopram	5–10	10–30

Second-line agents for depression include bupropion, mirtazapine, and venlafaxine. It should be noted, however, that there have been negative trials with venlafaxine and mirtazapine in youth.

There is considerable use of augmentation strategies in adults with complex depression, with the use of bupropion as well as mood stabilizers, including lithium. There is no experience in the controlled literature with regard to children and adolescents about augmentation strategies. Experts in pediatric psychopharmacology responded in a psychopharmacology perspectives review that they would add in an augmenting strategy—medications such as lithium, bupropion, antiepileptic medications, or atypical antipsychotics—to improve a partial response to fluoxetine.[47] If a patient is unresponsive to several single medication trials, these may be options, but there are no controlled data in youth to support these practices.

In general, the SSRIs are well tolerated. Children with anxiety, however, are more vulnerable to side effects and consequently they often need to start at lower doses and advance the dose more slowly. Selective serotonin reuptake inhibitor side effects include gastrointestinal upset, diarrhea, headaches, sexual dysfunction, insomnia, sedation and/or agitation, sweating, and vivid dreams. Manic episodes can also be induced in those individuals predisposed to bipolar disorder. Additionally, an increased risk of suicidal thinking and behavior has been noted in youth taking antidepressants and has resulted in FDA black box warnings for these medications, whether prescribed for depression or anxiety.[48,49] Food and Drug Administration findings were a prevalence of suicidal thinking and behavior in 4 out of 100 on active medication versus 2 out of 100 on placebo.[50] A subsequent review concluded that a risk was present, but somewhat lower.[51] There were no completed suicides in the more than 4,400 youths younger than 18 years who have participated in controlled trials.

Education must be provided to patients and families regarding the data supporting the use of SSRIs and the potential side effects. This education must include informing the patient and family about the black box warning regarding the potential for the development of suicidal ideation in children and adolescents. The risk

of increased suicidal thinking and behavior also applies to youth taking SSRIs for anxiety (ie, it is not limited to youth with depression).

Close monitoring, especially early in treatment with antidepressant medications (whether used for depression or anxiety) is indicated. It is also essential to have clear communication with the child and family when antidepressants are being used, with special attention to the course of mood symptoms over time, particularly regarding self-harm, suicidal ideation, and suicide attempts. Whether close monitoring will mitigate risk remains to be determined. There is likely a small subset of children for whom risk of suicidal thinking or behavior is heightened. It has not been demonstrated that completed suicide is associated with use of SSRIs in youth.

The use of rating scales can assist in the assessment of treatment response. If available and allowed by the patient and family, monitoring of signs and symptoms by school personnel can be helpful. Since children and adolescents in treatment studies for most mental health disorders have a high rate of placebo response, it is sometimes difficult to evaluate whether improvement in symptoms is due to placebo response, medication effect, or concurrent therapy effects.[52–54]

Evidence-based long-term treatment guidelines for youth do not currently exist. Given our knowledge of the course of depression in data in adults, there is general agreement that ongoing pharmacologic treatment of major depression in youth associated with clinical improvement should be continued for a customary duration of at least 3 to 6 months after remission is achieved, with efforts to taper medication after a single episode of depression. There are no clear guidelines in youth in the event of recurrence, but possibly with one and probably with 2 recurrences, there should be indefinite use of an antidepressant.

Best practice combines treatment with both medication and psychotherapy, especially CBT and interpersonal psychotherapy, and requires that important safety issues must be monitored carefully when children and adolescents are treated for depression. On balance, there seems to be evidence of the benefit for antidepressant medications for treatment of depression in children and adolescents, which outweigh safety concerns and likely with overall reduced risk of suicidality.

Anxiety Disorders

Medication interventions often provide relief to anxiety symptoms but may not be adequate to fully improve a child's functioning. Psychotherapeutic approaches should be combined with medication treatment to ensure an optimal effect. Patients and families need to be alerted to the black box warning regarding the risk of increased mood-related side effects in youth being treated with antidepressant medications, whether prescribed for depression or anxiety.

It is important to note that the medications for anxiety, apart from OCD, are not FDA approved for this indication in children and adolescents. Their use in these

circumstances, thus, is off-label. However, several of the commonly used medications have been approved for use in children for other purposes. Effective pharmacotherapy of anxiety requires collaboration with the family. The family should be clearly advised of the off-label use of medication and the potential benefits and risks.

The most extensive evidence for medication efficacy in child and adolescent anxiety disorders exists for OCD. While the first controlled trial demonstrating medication efficacy in OCD was with clomipramine, the first-line agents for treating OCD are the SSRIs due to their superior risk and safety profiles.[55–60] Clomipramine and 3 SSRIs (fluoxetine, fluvoxamine, and sertraline) have been FDA approved for the treatment of OCD in youth. There have been positive trials with other SSRIs in youth with OCD, but they do not have FDA approval. There is no evidence that any particular SSRI is superior in the treatment of OCD.[61] The SSRIs have also been well studied in adults with OCD.

Factors such as family history of response to medication, side effects, drug interactions, half-life, cost/formulary limitations, and dosing forms (eg, availability of liquid) are often relevant in selecting a specific SSRI. The effective treatment of OCD may require higher doses of SSRIs compared with the dosages typically used to treat depression or other anxiety disorders. It is necessary to start with low doses and proceed slowly in advancing the dose in order to avoid adverse side effects. It is safe to increase the dose weekly, as long as no prominent side effects are reported. However, it may be necessary to advance more slowly in children who are sensitive to adverse effects.

A medication trial for OCD should be continued for at least 8 weeks and preferably 12 to 16 weeks. Some studies have even suggested that continued benefits may accrue for months beyond this initial period, so patience should be encouraged. Pharmacologic treatments for OCD are generally only partially effective in relieving symptoms. The goal of treatment is focused on symptom reduction and improvement in functioning. Once a medication has shown some benefit in addressing OCD symptoms, it can be difficult to decide how long to continue treatment, and there are no studies to guide length of treatment. Many individuals with OCD experience their illness as a chronic condition and find medication treatment helpful indefinitely. At least a year of treatment is generally necessary. It is best to continue medication until symptoms are under good control and remain in good control for several months before considering a careful and slow tapering of several weeks. Additionally, relapse-prevention and psychotherapeutic techniques for symptom management should be emphasized prior to medication discontinuation.

Clomipramine (Anafranil), a tricyclic antidepressant with serotonin reuptake inhibition effects, is a second-line agent for the treatment of OCD. It is usually considered after a patient has failed adequate trials of at least 2 SSRIs. It has been FDA approved for the treatment of OCD in youth, but it is generally reserved for patients who have

not responded to alternative medication interventions due to the risk of serious adverse effects. Most of clomipramine's side effects are relatively benign, such as dry mouth, sedation, dizziness, constipation, and urinary hesitancy. More serious adverse effects can occur, however, such as tachycardia and cardiac conduction problems. Clomipramine, like other tricyclic antidepressants, can be lethal in overdose, and the therapeutic window is relatively narrow. Other toxic effects in overdose include seizures and coma. Consequently, ECGs must be checked before and during the course of a tricyclic antidepressant trial, with special attention to evidence of QTc prolongation, which may be a precursor to ventricular tachyarrthymias. Monitoring of clomipramine and norclomipramine levels may help to avoid toxicity. As a result, clomipramine is typically prescribed after consultation with a psychiatrist.

Atypical antipsychotics are another class of medications sometimes used to augment treatment for severe and treatment-resistant OCD based on evidence of augmentation effects in adults.

As to anxiety disorders other than OCD, while there are no medications with FDA indications there is a substantial evidence base of positive controlled trials with SSRIs in adults with anxiety disorders, and there are now some studies showing efficacy and safety in youth for anxiety disorders, including RDBPCTs.[62–69] There is no evidence that any medication in this class is more effective than another. Typical benefits from medication treatment can include an overall decrease in anxiety, improvement in somatic concerns related to anxiety, and an increased ability to tolerate distress. The goal is to restore function, but not necessarily to resolve all symptoms.

The most important study of anxiety other than OCD to date was the Child/Adolescent Anxiety Multimodal Study (CAMS), a large multicenter 12-week RDBPCT of sertraline alone up to 200 mg/day (mean dose 150 mg/day), CBT alone, and the combination.[64] The results were best for the combination, but all active treatment groups were far superior to placebo. In a study of fluvoxamine in 128 children with anxiety disorders, including social phobia, separation anxiety disorder, and generalized anxiety disorder, 76% were improved in the acute phase of the trial with maintenance of response in a 6-month open-label extension.[65,66] The mean final dose was 131 +/- 86 mg/day. Venlafaxine was studied in an 8-week RDBPCT in children with generalized anxiety disorder that documented significant reduction in ratings on anxiety scales at doses of 37.5 to 225 mg/day.[68] In an RDBPCT in adolescents with comorbid anxiety and depression who also were refusing school, imipramine (mean dose 185 mg/day), in combination with CBT, was found to be significantly more efficacious than placebo with CBT, as measured by improved school attendance.[62]

Anxious children can be particularly sensitive to somatic concerns, and sometimes anxiety can be paradoxically increased when medications are increased too quickly. A cautious titration can also ultimately improve compliance, since some children

may refuse medications if they experience side effects. As with OCD, gradual dose increases can be done weekly, as long as no significant side effects are reported. If a child has a positive response to a very low dose of an SSRI, the medication does not have to be titrated higher. The medication trial should be continued for at least a month at a therapeutic level to ensure adequate time for a response to emerge. Additional benefits can occur over the course of 8 to 12 weeks. The time of day of the medication administration can vary. Some children experience mild sedation with an SSRI, and they tolerate their medication better at bedtime. Other children experience an increase in energy and alertness and find a morning dose to be preferable. If daytime sedation or a sleep disturbance emerges when using an SSRI, modifying the time of medication administration can help.

There are inadequate data with regard to appropriate duration of treatment. In general, if a patient has found an SSRI to be substantially helpful and their symptoms have been stable for 4 to 6 months, it may be appropriate to consider gradually tapering off the medication, especially at a time when the environment is relatively stable and limited stressors exist. Some children may be candidates for chronic administration of medication, especially if they have a relapse of symptoms during or after a discontinuation trial. The SSRIs vary significantly in terms of half-life, and this can be relevant in deciding how rapidly to taper the medication. For example the half-life of fluoxetine is quite long in comparison to the other medications in the class. Abrupt discontinuation of SSRIs with shorter half-lives can lead to muscle stiffness, runny nose, and general malaise. Additionally, there is some evidence that children may metabolize these medications more rapidly than adults and, consequently, they may be more vulnerable to withdrawal symptoms.

Second-line agents for treating anxiety in youth include benzodiazepines. While these medications are known to be effective in adults with anxiety, they have been inadequately studied in children. The scant evidence to date shows mixed results in studies with children. There are several different benzodiazepines, and they vary largely according to duration of action. Among the most commonly prescribed agents are the long-lasting medication clonazepam (Klonopin) and the intermediate-acting lorazepam (Ativan) and alprazolam (Xanax). Benzodiazepines can be a desirable intervention at times due to their potential for almost immediate relief. They typically are used in conjunction with an SSRI at initiation of treatment when severe symptoms are present, since they can provide some benefit in the early weeks of treatment while the antidepressant medication is still starting to take effect. Unfortunately, children are more likely to experience some adverse effects of benzodiazepines than adults, particularly disinhibition, paradoxical activation, and agitation. Benzodiazepines can also impair memory and learning, making them less appealing to many families. When benzodiazepines are used, they should generally be considered cautiously as a short-term intervention, and consultation with a child psychiatrist should be pursued if their use continues to seem necessary over the long term (Table 20.5).

Table 20.5. Benzodiazepine Dose Guidelines

Medication	Initial Dose, mg	Dose Range, mg
Clonazepam	0.25 once or twice daily	0.25–1 twice daily
Lorazepam	0.5 once or twice daily	0.5–2 two to three times daily
Alprazolam	0.25 once or twice daily	0.25–1 three times daily

Other medications to consider as second-line agents include mirtazapine, venlafaxine, and buspirone. If venlafaxine is used, provide education regarding the potential side effect of increased blood pressure and tachycardia. The sedative and appetite-stimulating properties of mirtazapine make it an option for anxious patients with insomnia and prominent appetite suppression. However, there are no randomized, controlled trials of mirtazapine in youth with anxiety. There are no published randomized, controlled trials to support the use of buspirone in youth with anxiety. This medication generally requires dosing at least twice and sometimes 3 times per day, and the side-effect profile is generally benign.

Rating scales can assist in the monitoring of symptoms. Instruments such as the Multidimensional Anxiety Scale for Children (MASC), the Scale for Childhood Anxiety and Related Emotional Disorders (SCARED), or the Children's Yale-Brown Obsessive Compulsive Scale (C-YBOCS) should be administered at least once every 3 months to evaluate the effectiveness of treatment.[70–72]

Sleep Disorders

Sleep problems are common in children and adolescents. They are especially prominent in children who meet criteria for specific psychiatric diagnoses, including ADHD, depression, and mania, and in children with developmental disorders, such as ASDs. A careful evaluation is essential, which needs to consider specific sleep disorders, general medical conditions (such as iron deficiency anemia), and psychiatric disorders associated with sleep impairment. Referral to a sleep specialist may be necessary to clarify diagnosis. Patient and parent education are the first steps with attention to sleep hygiene. This is often sufficient.

While these problems are common, there is essentially no controlled evidence to guide health care professionals in the management of these problems, particularly as to psychopharmacologic interventions. No medication has a specific FDA indication to treat insomnia in youth. Medications, however, are commonly prescribed for insomnia, with 75% of practitioners recommending nonprescription medications and 50% having prescribed a sleep medication.[73] Medications recommended included diphenhydramine, melatonin, trazodone, and clonidine.

If medications are prescribed, use in most children and adolescents should be short term, typically for no longer than 2 weeks at a time. Children with ADHD, autistic disorder, or intellectual disabilities/mental retardation may have more chronic sleep

problems and be candidates for longer-term treatment. Even in these instances, treatment needs should be reassessed every 3 to 4 months. If a primary psychiatric diagnosis can be identified, it may be appropriate to entertain medication specifically treating that disorder. For example, if ADHD is present, the most commonly prescribed medication for sleep is the use of the alpha-2 agonist clonidine. Similarly, if depression is present, it is preferable to assess the benefits of an antidepressant medication on sleep architecture and quality of sleep before resorting to a separate medication for sleep. Often, however, the problems do not fit nicely into a diagnosis or even a group of diagnoses.

Melatonin administered orally mimics the effects of endogenous melatonin. There is probably the best empiric support for the use of melatonin for sleep problems.[74] It has both hypnotic effects and chronobiotic effects. For use in induction of sleep in a hypnotic capacity, dosage is typically of the order of 3 to 5 mg orally to be taken 30 minutes prior to the anticipated time of sleep. When used for its chronobiotic effects with its potential to shift circadian rhythms, the dose is significantly lower at 0.5 mg administered 6 or 7 hours before the anticipated time for sleep. The specific dose necessary for both hypnotic and chronobiotic effects is not predictable and must be titrated. A melatonin receptor agonist, ramelteon (Rozerem), is also available. While having a selective effect on melatonin receptors, its clinical efficacy is modest.

Trazodone is a 5-HT agonist with a short-term effect. Despite its widespread use, there have been 2 failed trials in terms of its effect on sleep in nondepressed adults and no trials in children. With clonidine, abrupt discontinuation can lead not only to rebound hypertension, but also to rebound nightmares. There is also clinical experience with antihistamines, particularly diphenhydramine, but no controlled trials. It is important to be aware of the potential for habituation over time. Long-term effectiveness for sleep has not been demonstrated for any medication.

There are a number of novel non-benzodiazepine receptor agonists used for the treatment of sleep disorders in adults, including agents such as zolpidem (Ambien), zaleplon (Sonata), and eszopiclone (Lunesta). There is now a longer-acting form of zolpidem (Ambien CR), with reports in adults of benefit for up to 8 hours of uninterrupted sleep per night and safety over a 6-month period.

On the other side of the sleep spectrum are conditions in which there is excessive sleepiness. To the extent that there is a specific psychiatric disorder, such as an atypical depression, it is appropriate to proceed with treatment directed at that presentation. Narcolepsy is an uncommon illness in children and adolescents, defined by rapid entry into REM sleep and excessive daytime sleepiness. Daytime sleepiness is also associated with sleep apnea syndromes. Benefit has been demonstrated in both of these conditions with stimulant medications, including methylphenidate and dextroamphetamine, and modafinil, and there has been a positive trial with mazindol.

Disorders of Impulse Control, Mood Dysregulation, and Aggression

This section of the chapter addresses management of aggression, agitation, severe disinhibition, and emotional lability and considers a number of diagnoses, including ASDs, bipolar disorder, and psychosis.

Acute Management of Agitation

If a child presents with acute and severe agitation, the first steps are to clarify diagnosis and implement strategies to de-escalate the patient. Anti-agitation medications are to be considered if other calming measures are deemed ineffective or inappropriate. Oral agents are generally preferred over intramuscular (IM) medications.

If the patient is already on behavioral medications, consider giving his or her usual dose as scheduled (unless toxicity is suspected). For example, to treat agitation, consider giving one-half to one-quarter of the total daily dose of the patient's prescribed benzodiazepine or antipsychotic medication ONE TIME, if between scheduled doses.

For use of NEW medications, determine the type of agitation and CHOOSE ONE appropriate medication from the options listed below.

Sedative/Anxiolytic Medications

These agents are first-line therapy for patients with anxious agitation or unknown type agitation. (NOTE: These medications may cause disinhibition, especially with very young or cognitively impaired patients. Oral route preferred, but consider IM or intravenous if necessary.)

Diphenhydramine 1 mg/kg by mouth One Time Unscheduled; maximum dose = 50 mg
Lorazepam 0.05 mg/kg by mouth One Time Unscheduled; maximum dose = 2mg
Diazepam 0.1 mg/kg by mouth One Time Unscheduled; maximum dose = 10 mg

Risperidone

This drug is first-line therapy for patients with psychosis, mania, aggression, delirium, or unknown type agitation. (NOTE: Risperidone may cause dystonia [muscle stiffness or restlessness] that should be treated with diphenhydramine or benztropine. It can also cause hypotension, lowered seizure threshold, or prolonged QTc interval. Do not use if history of adverse response.)

For patients 4 to 6 years old
    Risperidone 0.25 mg by mouth One Time
For patients 6 to 10 years old
    Risperidone 0.5 mg by mouth One Time

For patients 11 to 16 years old
Risperidone 1 mg by mouth One Time
For patients 17 years and older
Risperidone 2 mg by mouth One Time

Haloperidol

This drug is second-line therapy for patients with psychosis, mania, maladaptive aggression, or delirium type agitation. (NOTE: Haloperidol may cause dystonia [muscle stiffness or restlessness] that should be treated with diphenhydramine. It can also cause hypotension or prolonged QT interval, and has a warning regarding possible development of torsades de pointes. Do not use if history of adverse response. Lower 0.025 mg/kg dose preferred for preadolescents.)

For preadolescents
Haloperidol 0.025 mg/kg IM One Time
For adolescents
Haloperidol 0.05 mg/kg IM One Time

Emergency/Antidote Medications

For treatment of antipsychotic medication-induced dystonia (muscle stiffness or restlessness).

Diphenhydramine 1 mg/kg by mouth ONE TIME ONLY as needed for dystonic reaction; maximum dose = 50mg

Atypical Antipsychotic Medications

An area of increasing investigation has been atypical antipsychotic medications. This class of medications has demonstrated effectiveness in the management of extreme mood and behavior dysregulation in ASDs, developmental disorders, conduct disorder, and psychotic disorders, including childhood bipolar disorder.

Atypical antipsychotic medications include clozapine (Clozaril), risperidone (Risperdal), olanzapine (Zyprexa), ziprasidone (Geodon), quetiapine (Seroquel), aripiprazole (Abilify), and paliperidone (Invega). There has been a dramatic recent increase in the prescription of atypical antipsychotic medications in youth. Atypical antipsychotics that carry an FDA indication are risperidone for the treatment of autistic disorder, schizophrenia, and bipolar disorder type I; aripiprazole for the treatment of agitation associated with autistic disorder, schizophrenia, and bipolar disorder, type I; quetiapine for schizophrenia and bipolar disorder; olanzapine for schizophrenia and bipolar disorder; and ziprasidone for schizophrenia and bipolar disorder.

The use of atypical antipsychotics in treatment of labile disruptive behavior symptomatology is complicated in at least 2 respects. First, there is considerable controversy and disagreement in the field regarding bipolar disorder in children and

adolescents. In general, children with a bipolar disorder diagnosis are behaviorally dysregulated and irritable with mood lability. Whether they ultimately will go on to have a condition consistent with what is known about the adult form of bipolar disorder remains to be seen. Second, there is evidence that a number of psychiatric disorders and symptom domains in children and adolescents are responsive to atypical antipsychotic treatment, suggesting a more nonspecific response. There are, however, significant numbers of children who are mood dysregulated, with episodic and/or severe reactive aggression problems who may require medication.

The first medication to achieve an FDA indication in the atypical antipsychotic group in youth was for risperidone, following 2 short-term RDBPCTs of risperidone in individuals who had ASDs, with evidence of reduction in aggression, emotional lability, self-injurious behavior, and agitation.[75,76] A long-term follow-up study also demonstrated continued effectiveness at 6 months.[77]

In addition, risperidone has been studied in children with subaverage[78,79] and average intelligence[80] with disruptive behavior disorders in RDBPCTs. In an RDBPCT of 50 children with normal intelligence, there was evidence of significant improvement in terms of aggressive disruptive oppositional behavior, with a mean dose of 1.23 mg/day of risperidone, and also in an open-label follow-up of 50 subjects for 48 weeks.[80,81] As would be expected, weight gain and sedation were ongoing and there were persistent side effects.

There have been several controlled trials and open-label studies documenting efficacy of other atypical antipsychotic medications in children and adolescents diagnosed with bipolar disorder, but none reported to date with paloperidone.[82–88] As with other psychotropic medications, dosage must be carefully titrated. Table 20.6 provides dosing guidelines. In the management of mood dysregulation and aggression, duration of treatment should be 6 to 12 months with reevaluation. Rating scales, such as the Aberrant Behavior Checklist and the Young Mania Rating Scale, can complement clinical interviews with the patient and parents and information from schools.[89,90]

Table 20.6. Dosing Guidelines for Atypical Psychotic Medication

Medication	Initial, mg/day	Daily Dose, mg/day	Not to Exceed, mg/day
Risperidone	0.5–1	1–3	6
Olanzapine	2.5–5	5–20	30
Quetiapine	25–50	50–400	800
Ziprasidone	5–20	20–80	160
Aripiprazole	5	5–10	30

Side Effects of Atypical Antipsychotic Medications

There are a number of potential side effects associated with atypical antipsychotic medications. Whereas the risk of serious movement disorders, including extrapyramidal symptoms and tardive dyskinesia, associated with traditional antipsychotic medications is substantially less with the atypicals, they do occur. Other potentially serious side effects include weight gain, hyperlipidemia, diabetes, and galactorrhea. Thus, while there is evidence of benefit in using these medications, they are not benign agents.

Table 20.7 depicts the side-effect monitoring recommendations regarding weight, glucose, and lipid levels. More frequent monitoring may be necessary if abnormalities arise. The side-effect profiles of medications in this class are not identical. For example, there is relatively less weight gain with ziprasidone and aripiprazole. Galactorrhea is more common with risperidone. Nonetheless, if patients are treated with any atypical antipsychotic, monitoring guidelines must be followed.

Table 20.7. Monitoring Protocol for Atypical Antipsychotics

	Baseline	1 Month	2 Months	3 Months	And Then After the First 3 Months as Follows	Quarterly	Annually
Personal and family history regarding diabetes, cardiac disease, and lipid profile	X						X
Weight	X	X	X	X		X	
Blood pressure	X			X			X
Fasting glucose	X			X			X
Hemoglobin H1c	X			X			X
Fasting lipid profile	X			X			X

Mood Stabilizers

Mood stabilizers include one agent, lithium, for which there is an FDA indication for bipolar disorder for individuals 12 years and older, and also a number of anticonvulsant medications that have been identified to have potential mood stabilizing effects, including divalproex sodium, carbamazepine, lamotrigine, oxcarbazepine, gabapentin, topiramate, and clonazepam. Whereas mood stabilizing agents have been extensively studied in adults, with several medications having an FDA indication for adults with bipolar disorder, there is considerably less controlled evidence with children and adolescents. The medication for which there are best data in adults is lithium. While it does have an FDA indication for children 12 years and older for bipolar disorder, there have been comparatively few trials in children and adolescents.[91–94] A large, multicenter National Institute of Child Health and Human Development–funded study investigating lithium in the treatment of pediatric mania is in progress.

Lithium requires close monitoring. The therapeutic-to-toxicity ratio is quite close, with significant risk of toxicity at doses close to the therapeutic dose range (<2 times above the upper limit of normal). In the absence of pediatric data, the approach to treatment in children and adolescents follows adult guidelines. In acute manic episodes, lithium is quickly titrated to achieve a trough blood level of 0.8 to 1.4 mEq/L. There are no studies in any age group that indicate benefit at doses higher than this. Once stabilization has occurred, the dose range targets are typically between 0.6 and 1.0 mEq/L, dependent on clinical response.

Serum monitoring of lithium levels should be done every 1 to 2 weeks at the initiation of a titration until stable and then every 1 to 2 months during continuation. In addition, there are a number of side effects that need to be carefully monitored. The common side effects include the development of tremor, acne, hair loss, weight gain, polyuria, polydipsia, and hypothyroidism. Therefore, in addition to lithium monitoring, it is essential to get a baseline thyroid profile, renal function tests (including blood urea nitrogen and creatinine), urinalysis, and an ECG. Thyroid and renal function tests and urinalyses should be repeated every 3 to 6 months.

There are no guidelines regarding the duration of treatment in children or adolescents. For patients with a manic episode successfully treated with lithium, it should be continued for at least 4 to 6 months after stabilization. With a single episode, it would be appropriate to consider careful tapering; possibly after 1 or probably after 2 recurrences of mania, indefinite treatment is indicated.

Divalproex sodium is also commonly prescribed in children and adolescents. There is an FDA indication for treatment of acute mania in adults. It may be more effective for mania associated with rapid cycling or with mixed episodes. The picture in children and adolescents is mixed, with some open-label support for use for acute mania and maintenance, but also negative trials.[95–98]

There also are significant monitoring issues regarding use of divalproex sodium. As with lithium, serum levels should be monitored. There are no standards that guide this in children and adolescents, but in adult studies, benefit has been identified with serum levels between 50 and 150 mcg/mL. There is no evidence of added benefit at higher serum levels.

There are a number of side effects with this medication, including sedation, weight gain, polycystic ovary syndrome, hair loss, thrombocytopenia, pancreatitis, and hepatitis. At baseline, a CBC (with platelet count) and liver function tests should be obtained. Thorough serum valproate level, CBC, and liver function tests should be obtained 5 to 7 days after dose changes during titration and then approximately every 3 to 6 months in the continuation phase. Typical dosage to achieve targeted blood levels is 10 to 20 mg/kg/day.

There are a number of other anticonvulsant medications with evidence of mood stabilization in adults. For example, lamotrigine has been identified in adult studies as effective in the prophylaxis against bipolar depression, resulting in an FDA indication in adults but not in youth. There was an open-label investigation of adolescents with bipolar depression that suggested benefit with this agent.[99] Titration must be started at the lower of 0.3 mg/kg/day or 25 mg/day, and increase no more rapidly than the lower of 0.3 mg/kg/day or 25 mg every 2 weeks, not to exceed 200 mg/day to avoid a potentially life-threatening rash. Additional care and half the dosage are required if the patient is on valproic acid. Studies have been done with carbamazepine and oxcarbazepine in adults, with findings that are not as strong as they are for divalproex sodium, with few studies in adults showing efficacy and a negative trial in youth. For carbamazepine, the target serum level is 4 to 14 mcg/mL typically associated with dosing of 10 to 20 mg/kg/day (200–1,200 mg). Side effects include dizziness, headaches, leukopenia, and rashes. At baseline, a CBC and liver function tests should be obtained. Serum blood level and CBC should be done every 1 to 2 weeks during the titration stabilization phase and then approximately every 3 months in the maintenance phase. There are rare instances of agranulocytosis associated with carbamazepine that may not be captured by periodic monitoring, so families must be advised of this potential rare but serious side effect and to watch for signs, including easy bruising or unexplained fever. A distinction must be made between nonprogressive leukopenia, which is relatively common (10%–15%), compared with profound agranulocytosis or aplastic anemia. Topiramate has been associated with negative trials in adults. While the drug was tolerated, there was no evidence of efficacy in a study with adolescents.[100] Gabapentin has been suggested at times as a potential augmenting agent with regard to bipolar disorder, but a large controlled trial was negative in adults and there are no pediatric RDBPCTs.

Tic Disorders

Tics are very common in children, with as many as 15% of children having at least transient tics by age 15 years. If they are severe and are associated with significant impairment, such as occurs in Tourette disorder, medication may be required.

There is evidence of efficacy with psychopharmacologic treatment for tic disorders. As with many other psychiatric conditions, medications can reduce symptoms, but do not necessarily completely eliminate tics. The decision to treat tics pharmacologically is complicated by several factors. There is an inherent waxing and waning of the course of tics over time. Not all tics require pharmacologic intervention. A judgment has to be made that there is a significant adaptive problem, such as impairment of self-esteem or interference with daily activities, before medications are warranted.

Two typical antipsychotic medications, haloperidol and pimozide, have an FDA indication for treatment of tics. However, both of these medications have a number of significant side effects associated with them. Extrapyramidal symptoms are common with haloperidol. Pimozide has the potential to affect cardiac rhythm, and ECG monitoring is required if this medication is used.

There is evidence of benefit of the alpha-2 agonists clonidine and guanfacine in the reduction of tic severity, though less robust than for antipsychotic medication.[32] Given their relative benign side-effect profile, they are the medications of first choice, particularly if tics are mild.

For prominent tics, however, first choice is an atypical antipsychotic medication. Several newer atypical antipsychotics have demonstrated efficacy in RDBPCTs but do not have an FDA indication.[101–103] These newer agents, however, are generally better tolerated and likely are more effective; thus they should be considered among the first agents of choice. Best evidence is for risperidone and ziprasidone, with a 30% to 50% decrease in tics. Dosages in these trials have been relatively low, with attention to the usual constellation of side effects associated with atypical antipsychotics. Dose guidelines are lower than those noted in Table 20.6. Monitoring parameters mentioned in Table 20.7 also apply in the treatment of tic disorders.

Conclusion

This chapter endeavors to provide practical information regarding the psychopharmacologic treatment of children and adolescents with psychiatric disorders who are being treated in a primary care setting. As noted, this is a rapidly changing field and doubtless much information will be added to our knowledge base over time. A clinician must have a means of staying in touch with the literature and the field as knowledge grows. It is essential that a team be put together to appropriately treat children who are candidates for psychotropic medications, which ideally includes the child; family; and community team, including school, therapist, and pediatrician. While it may be difficult at times to access a child and adolescent psychiatrist, it is useful for a

primary pediatric health care professional to have consultative access to subspecialists in this area. It is also important to make clear decisions about what the scope of one's practice is, as to what appropriately can be managed within a practice and what needs to be referred out for subspecialist care. There is little doubt that children with mental health disorders can be significantly helped with medications embedded in an evidenced-based comprehensive care plan.

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Chapter 21

Complementary and Alternative Medicine in Developmental and Behavioral Pediatrics

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James is a 2-year-old boy who presents for a developmental evaluation. His parents report that at 12 months of age, he was able to say "mama" and "dada," as well as what sounded like "tickle." In the months following his first birthday, he stopped using these words, and his eye contact and overall social responsiveness decreased as well. While James' parents did not have specific concerns about his development prior to 12 months of age, communicative pointing was never observed, and it is questionable whether he ever waved "bye-bye" in his first year of life. At the age of 2, James was nonverbal, had significantly diminished social reciprocity, and exhibited a variety of repetitive behaviors. His parents, after investigating various treatment options they found on the Internet and consulting with a physician who promotes a "biomedical" approach for the treatment of autism, elected to start a number of nonstandard therapies: multiple vitamins (including methyl B_{12} injections), various other supplements (coenzyme Q10, dimethylglycine, L-glutamine, N-acetyl cysteine, cod liver oil, colostrum, transfer factor, probiotics), topical agents (lithium cream, zinc cream), 2 different oral chelators, several homeopathic remedies, a special diet (gluten-free, casein-free), and hyperbaric oxygen therapy.

This story raises a number of important questions that any rational person should ask. What is the scientific rationale and evidence regarding the use of these therapies individually and/or in combination for children with developmental/behavioral concerns? What are the potential risks of these therapies? Is it even plausible that these agents should have efficacy in improving the symptoms of a developmental disorder, based on what we already know about neuroscience and how the body works? How does one balance the desire of a parent to pursue a particular treatment with the right of a child not to be subjected to unproven therapies that may be ineffective or even harmful? And importantly, how do we help caregivers learn the skills they need to critically evaluate the morass of remedies promoted in the media and on the Internet? While there may not be clear answers to all these questions, they all require careful consideration.

Complementary and Alternative Medicine: Definition and Background

Various definitions of complementary and alternative medicine (CAM) have been offered. Complementary and alternative medicine has been defined as "a broad domain of healing resources that encompasses all health systems, modalities, and practices and their accompanying theories and beliefs, other than those intrinsic to the politically dominant health systems of a particular society or culture in a given historical period,"[1] and as practices "not presently considered an integral part of conventional medicine."[2] More recent terminology includes "holistic" and "integrative" medicine, the latter term defined as care that uses both mainstream and complementary therapies to promote health for the whole person in the context of his or her family and community.[3] The National Center for Complementary and Alternative Medicine (NCCAM), a National Institutes of Health (NIH) center, categorizes CAM practices into domains of mind–body medicine, manipulative and body-based practices, energy medicine, biologically based practices, and alternative medical systems. The diversity of practices included under this umbrella highlight the importance of evaluating the specific merits of individual therapies and methods (some of which, such as vitamin supplements, may be considered either CAM or conventional depending on the context in which they are being used).

The use of CAM therapies is common among adults,[4,5] and trends of increasing use have been noted over recent years.[6–8] Complementary and alternative medicine use is more common in children of parents who use these therapies themselves and is associated with higher levels of parental education.[9–12] Population-based data suggest that the use of these therapies is not particularly common among the general pediatric population,[12,13] although significant variability has been reported among selected outpatient and inpatient pediatric groups.[10,11,14–20] Complementary and alternative medicine use does seem to be widespread among children with chronic medical conditions.[21–26] High rates of CAM use have also been reported among children with developmental and behavioral disorders, including autism spectrum disorders (ASDs), attention-deficit/hyperactivity disorder (ADHD), Down syndrome, cerebral palsy, and spina bifida.[27–38]

An increase in research into CAM practices has occurred over the past decade. The NCCAM was created in 1999 for the purpose of investigating complementary and alternative practices using the methods of rigorous science, training CAM researchers, and disseminating authoritative information regarding CAM to the public and professionals. Since its inception, NCCAM has been the recipient of more than $1 billion in federal appropriations.[39] Total NIH funding for CAM research over the past decade may be 2 to 3 times this amount. Criticism has been leveled at the quality and appropriateness of studies funded by NCCAM.[40] Unfortunately, few NCCAM-funded projects have been directed at issues relevant to pediatrics, and as yet limited information has been developed proving or disproving the value of any specific CAM therapy.

How to Evaluate Therapies

Families of children with developmental and behavioral disorders, and the clinicians caring for these children, should ask certain questions to help them identify therapies that have an insufficient evidence base or are fundamentally implausible. These questions can be grouped into 3 main categories pertaining to the theoretical basis of the therapy, the evidence base of the therapy, and the tactics used to promote the therapy (Box 21.1).

Evaluating the Theoretical Basis of a CAM Therapy

Therapies considered complementary and alternative have diverse origins and arise from a variety of theoretical frameworks. The following questions can help identify therapies that have a weak theoretical foundation. Is the treatment based on a theory that is overly simplistic? Is the treatment based on proposed forces or principles that are inconsistent with accumulated knowledge from other scientific disciplines? Has the treatment changed little over a very long period?

Many of the CAM therapies used in children with developmental disorders are based on hypotheses that do not account for much of what we already know about the neurobiology of these disorders. For example, the belief that autism is caused by a discrete environmental insult (such as mercury exposure) has gained traction on the Internet and among some parent groups. While environmental factors that modify

Box 21.1. Twelve Questions to Ask About a Complementary or Alternative Therapy[a]

Questions related to the underlying theoretical basis for the therapy

1. Is the treatment based on a theory that is overly simplistic?
2. Is the treatment based on proposed forces or principles that are inconsistent with accumulated knowledge from other scientific disciplines?
3. Has the treatment changed little over a very long period?

Questions related to the scientific evaluation of the therapy

4. Is it possible to test the treatment claim?
5. Have well-designed studies of the treatment been published in the peer-reviewed medical literature?
6. Do proponents of the treatment "cherry pick" data that support the value of the treatment while ignoring contradictory evidence?
7. Do proponents of the treatment assume a treatment is effective until there is sufficient evidence to the contrary?
8. Do proponents claim that a particular treatment cannot be studied in isolation, only in combination with a package of other interventions or practices?

Questions related to the promotion and marketing of the therapy

9. Is the treatment promoted as being free of adverse effects?
10. Is the treatment promoted primarily through the use of anecdotes?
11. Do proponents of the treatment use scientific-sounding but nonsensical terminology to describe the treatment?
12. Is the treatment promoted for a wide range of physiologically diverse conditions?

[a] Adapted from: Lilienfeld et al 2003[41] and Nickel 1996.[42]

disease expression certainly should be explored, even in disorders that have a strong genetic basis, therapies (such as chelation therapy in autism) based on a belief about the role of some environmental "trigger" are unjustified in the absence of good evidence that the particular environmental factor is actually etiologically related to the disorder. There should also not be a blind leap to link associated medical issues to the etiology of a particular developmental disorder. For example, even if medical issues such as gastrointestinal dysfunction occur more commonly in children with autism, it does not necessarily follow that these issues are causally related to the core neurobehavioral features—to contend otherwise necessitates that a body of evidence about the neurobiology of autism be disregarded. While treating a gastrointestinal problem, or any other associated medical problem, is certainly important for the overall health and comfort of a child with a developmental disability, the claim that treatment of these associated medical issues should ameliorate core neurobehavioral features is currently not supported by available evidence. One has to look no further than Down syndrome to find a disorder in which various gastrointestinal and immune abnormalities are common but not related in a cause-effect manner to the fundamental neurodevelopmental issues.

Certain CAM therapies also seem to be quite disconnected from what we already understand about how the natural world works. Therapeutic touch, for example, is based on the belief that an energy "biofield" exists in proximity to the human body and that imbalances in this energy field are responsible for human disease (including developmental disturbances). Practitioners of therapeutic touch believe that this energy field can be manipulated manually and can result in objective improvements in some aspect of physical functioning. This theory is fundamentally inconsistent with much of the accumulated knowledge in biology and physics. While people may certainly experience subjective improvement in some symptom after undergoing therapeutic touch, the mechanism for this improvement is likely based on placebo effects and not on the adjustment of "energy" imbalances. Another energy-based practice, acupuncture, also illustrates the principle that therapies remaining unchanged for many years (or centuries) may not be undergoing the error correction that is a necessary element of scientific practices.

While the scientific investigation of novel therapies is an important pursuit, and is a primary mission of NCCAM, some therapies do not merit any further study, given that their underlying theoretical basis is so implausible or at odds with an accumulation of reliable, reproducible knowledge from other scientific disciplines. The appropriateness of considering plausibility in determining which novel therapies are worthy of formal investigation is especially relevant when research resources are limited.

Evaluating the Evidence Base for a CAM Therapy

The question of whether a particular treatment can actually do what it is claimed to do is a fundamental issue that should concern clinicians who care for children with

developmental disorders. Several questions can shed light on whether there is an adequate evidence base to support the use of a specific therapy. Is it possible to test the treatment claim? Have well-designed studies of the treatment been published in the peer-reviewed medical literature? Do proponents of the treatment "cherry pick" data that support the value of the treatment, while ignoring contradictory evidence? Do proponents of the treatment assume a treatment is effective until there is sufficient evidence to the contrary? Do proponents claim that a particular treatment cannot be studied in isolation, but only in combination with a package of other interventions or practices?

The evidence base for CAM therapies in developmental disorders is quite limited, and some of the most widely used therapies are not supported by any published studies. Summary recommendations for the use of various CAM therapies in developmental disorders are outlined in Box 21.2. There is insufficient evidence to indicate any value for the various alternative medical systems (traditional Chinese medicine, ayurvedic medicine, naturopathy, and homeopathy) in the treatment of developmental disorders. There have been efforts to test traditional Chinese medicine methods in developmental disorders including ADHD, cerebral palsy, and intellectual disabilities (mental retardation), although significant methodological deficiencies limit any conclusions that can be drawn from these studies. Likewise, there are no published controlled trials indicating efficacy for any ayurvedic practice in the treatment of developmental-behavioral disorders. Homeopathy, which has a highly questionable scientific basis, has not been shown to be beneficial in the treatment of ADHD.[43]

Mind–body medicine techniques have a similar lack of supporting evidence. Electroencephalographic biofeedback has been promoted for the treatment of children with ADHD and remains under investigation. However, it is not currently a well-validated treatment approach in this population. Meditation and relaxation training have been investigated in children with ADHD, cerebral palsy, and intellectual disabilities, and short-term improvements in certain behavioral measures have been observed in some studies.

Manipulative and body-based practices include such approaches as sensory integration therapy, massage, auditory integration training, and chiropractic manipulation. Although sensory integration therapies enjoy widespread use among children with developmental disorders, there is limited evidence supporting the therapeutic value of these methods.[44] A number of studies of massage in infants have been published, but there is no convincing evidence of measurable developmental benefits in preterm or low birth weight infants.[45] Similarly, there have been several trials of auditory integration training (which is based on the hypothesis that abnormal auditory perception contributes to various developmental and behavioral symptoms) in children with autism, but there is no sufficient evidence to support its use.[46] Other manipulative or body-based practices, including optometric visual training,

Box 21.2. Summary Recommendations for the Use of Selected Complementary and Alternative Therapies in Developmental Disorders Based on Available Evidence

Not recommended: insufficient or absent empiric support (or strong evidence of inefficacy), low plausibility

- Homeopathy
- Chiropractic
- Auditory integration therapy
- Vestibular stimulation
- Vision therapy, visual perceptual training
- Reflexology
- Craniosacral therapy
- Patterning, Doman-Delacato method
- Acupuncture
- Therapeutic touch
- Magnet therapy
- Reiki, Qi gong
- Spiritual therapies
- Hypnosis
- Pharmacologic doses of vitamins (except in known metabolic disorders)
- Herbal remedies
- Chelation therapy
- Secretin in autism
- Hyperbaric oxygen for cerebral palsy
- Antifungal agents
- Antiviral agents
- Antioxidants

More research needed: limited empiric support, limited plausibility

- Biofeedback, electroencephalogram/electromyogram biofeedback
- Meditation, relaxation techniques
- Music therapy
- Art therapy
- Massage
- Sensory integration therapy
- Gluten-free, casein-free diet in autism
- Omega-3 fatty acids

Adequate empiric evidence to support use

- Melatonin for prolonged sleep latency

craniosacral therapy, and chiropractic manipulation, lack scientific plausibility and do not have any current role in the treatment of children with developmental disabilities. Children with Down syndrome and atlantoaxial instability may be particularly susceptible to injury from chiropractic manipulation.

Energy therapies, including therapeutic touch and acupuncture, are based in the belief that energy fields are present around all living organisms and that a disturbance in these fields is a cause of disease. There is no convincing evidence that acupuncture is beneficial in the treatment of any developmental disorder, and most published studies in these populations have been uncontrolled or had

unreliable outcome measures. There are no published studies of therapeutic touch in children with developmental disorders, and the theoretical basis of this particular therapy remains highly questionable.

Biologically based therapies include special diets, vitamins, supplements, and similar interventions. While these approaches can potentially have greater biological plausibility than many other CAM therapies, they also have weak or nonexistent evidence of efficacy in the treatment of developmental and behavioral disorders. Dietary manipulations have long been used in attempts to effect a positive change in the behavior or developmental functioning of children with various disorders. The Feingold diet (based on the hypothesis that various food additives are the cause of hyperactive behavior) gained popularity in the 1970s, and early trials did not show any consistently positive effect.[47] More recent data suggest the possibility that certain food additives may exert negative behavioral effects in subgroups of young children.[48] Oligoantigenic diets arose from the belief that food allergies may play a role in the etiology of hyperactive behavior. There are 6 controlled trials of oligoantigenic diets in children with ADHD between 1985 and 1997 listed in the Cochrane Central Register of Controlled Trials, data which have been reviewed both positively and negatively.[49,50] Effect sizes in these studies were not large. A more recent study indicated a beneficial effect of a carefully monitored elimination diet in a group of children with ADHD,[51] although a clear role for such an intervention in ADHD management remains undetermined.

The gluten- and casein-free (GF-CF) diet has gained widespread popularity in children with autism. There have been several trials of this diet, which is based on the unproved hypothesis that peptides derived from gluten and casein cross into the bloodstream from the gastrointestinal tract and subsequently exert a central nervous system effect that leads to behavioral abnormalities. Despite its widespread use, the existing scientific literature does not support the GF-CF diet as a treatment for children with autism.[52]

Various vitamins, minerals, and other supplements are commonly used in autism and other developmental and behavioral disorders.[28,30] Vitamin B_6 and magnesium are among the most extensively studied supplements in children with ASDs. While some investigations have shown apparent benefits, many of these studies had methodological deficiencies, and the best data currently do not support the combined use of these supplements in the treatment of autism.[53] Sleep disturbances are extremely common in children with developmental disorders, and melatonin has been used as a treatment to shorten prolonged sleep latency and increase total sleep time. There is published evidence that supports the use of melatonin for this purpose.[54,55] Interest in omega-3 fatty acids as a potential therapy has also been strong. At the present time, there is insufficient evidence to support the use of omega-3 fatty acids in children with developmental disorders or in the treatment of specific psychiatric conditions.[56] A pilot study in 2007 was reported as showing a beneficial effect of omega-3

fatty acids in children with autism.[57] However, significant inequality in baseline behavioral characteristics between treatment and control groups in this study raises questions about this conclusion. A larger NIH-funded study is underway.

More invasive nonstandard biomedical interventions in developmental disorders include heavy metal chelation and hyperbaric oxygen therapy (HBOT). Chelation therapy has potentially serious adverse effects, including death,[58] and there is no published evidence supporting the efficacy of this treatment for autism or any other developmental or behavioral disorder. In 2008 an NIH-funded study of chelation in autism was suspended over safety concerns. Chelation should never be performed in the absence of an appropriate indication (such as lead poisoning). Hyperbaric oxygen therapy remains popular in some areas for children with cerebral palsy and other developmental disorders, despite a lack of replicated experimental evidence showing any value in these conditions. A review in 2003 concluded that there is insufficient evidence to determine whether hyperbaric oxygen treatment improves functional outcome in children with cerebral palsy.[59] Hyperbaric oxygen therapy has also been promoted for the treatment of autism. There is a single controlled trial in the medical literature. This study suggested benefit of HBOT (primarily as measured by Clinical Global Impression–Improvement) in a group of children with autism.[60] However, certain concerns about this trial have been raised (including the modest oxygen partial pressures used, which could have been achieved using ~30% oxygen delivered via simple face mask),[61] and replication of these findings will be necessary before the use of HBOT in autism can be endorsed.

Instead of following a logical, forward-moving process in which credible therapies are studied and either verified or discarded, the scientific community has commonly been forced to disprove the value of untenable therapies already in wide use. The use of secretin in autism is one such example. A small case series published in 1998 reported improvement in language and social features in 3 children with autism who had undergone diagnostic endoscopy.[62] The gastrointestinal hormone secretin, which had been used during these endoscopy procedures, subsequently was widely touted (on the Internet and in the mainstream media) as a therapy for autism, and thousands of children were treated. Numerous subsequent controlled trials indicated that secretin is no more effective than placebo.[63]

Certain CAM therapies are framed in such a way that objective analysis of efficacy is discouraged. For example, some therapies (such as therapeutic touch) have been described as being highly dependent on the "skill" of individual practitioners. This creates an environment in which experiments that do not show benefit for the therapy can be more easily dismissed by believers. Another common ploy used to obscure evidence of inefficacy is for a claim to be made that a specific therapy cannot be studied in isolation—rather, a large package of interventions, or entire systems of care, must be evaluated. This makes the creation of an appropriately controlled study difficult and may serve to hide the fact that a particular therapy has no measurable benefit.

While it is appropriate to recognize that there are different levels of scientific evidence, not all evidence should be viewed as carrying equal weight. The evidence at the top of this hierarchy should most strongly inform clinical decision-making. Much of the evidence (for instance, individual anecdotes) that supports the value of CAM therapies in developmental and behavioral disorders does not even reach Level 5 in the Oxford Centre for Evidence-Based Medicine classification of levels of evidence (Table 21.1)[64] and is woefully inadequate to support any positive recommendation for the use of these therapies. Unfortunately, even the peer-reviewed medical literature contains studies in which the conclusion of a positive effect of a particular treatment for children with a developmental or behavioral disorder is undercut by analytical flaws.[57] It is crucial for health care professionals attempting to practice evidence-based medicine to read the peer-reviewed literature with an appropriately critical eye.

As of December 2008, only a single NIH-sponsored clinical trial of a CAM intervention for a developmental disorder was recruiting patients.[65] Between 2006 and 2008 the NCCAM funded only 2 clinical trials studying a CAM therapy in children with a developmental disorder.[66] Despite a great deal of talk to the contrary, there is not much action currently underway to expand the evidence base, and at the present time, no intervention that falls under the NCCAM definition of CAM that has an evidence base that justifies its use in children with autism[67] or other developmental disorders.

Table 21.1. Oxford Centre for Evidence-based Medicine Levels of Evidence[a,b]

Level	Evidence
1a	Systematic review (SR) (with homogeneity) of randomized controlled trials (RCT)
1b	Individual RCT (with narrow confidence interval)
1c	All or none[c]
2a	SR (with homogeneity) of cohort studies
2b	Individual cohort study (including low-quality RCT; eg, <80% follow-up)
2c	"Outcomes" research; ecological studies
3a	SR (with homogeneity) of case-control studies
3b	Individual case-control study
4	Case series (and poor-quality cohort and case-control studies)
5	Expert opinion without explicit critical appraisal, or based on physiology, bench research, or "first principles"

[a] Adapted from: Oxford Centre for Evidence-Based Medicine[64] (www.cebm.net/?o=1025).
[b] For definitions of terms used, see glossary at http://www.cebm.net/?o=1116.
[c] Met when all patients died before the therapy became available, but some now survive on it, or when some patients died before the therapy became available, but none now die on it.

Evaluating the Strategies Used to Promote a CAM Therapy

Additional useful information about a therapy can be gleaned from an analysis of the methods by which the therapy is promoted and marketed. Is the treatment promoted as being free of adverse effects? Is the treatment promoted primarily through the use of anecdotes? Does advertising for a therapy use scientific-sounding but nonsensical terminology? Is it promoted for a wide range of physiologically diverse conditions?

Certain treatments, particularly herbal remedies, may be advertised as "natural," with the unspoken implication being that the therapy is safer than conventional medicines. This, of course, is not necessarily the case, and there are a variety of potentially serious risks associated with herbs and supplements.[68] Advocates for a particular CAM therapy, when challenged about the safety of their therapy, may respond by pointing out the risks of conventional medical approaches—a *tu quoque* logical error that merely serves to divert attention from the original question.

Complementary and alternative medicine therapies are often promoted through the use of anecdotes and testimonials. The Internet has been a major driving force in the popularization of many of these therapies. Because of the various biases that inevitably color our perceptions of what we observe, however, anecdotes can never take the place of more objective methods of analysis (such as controlled trials) that serve to minimize these biases. Any treatment promoted exclusively through testimonials should be viewed with great caution.

Therapies that are advertised as being effective for a wide range of diverse conditions (eg, treatment for candidal infection[69]) also warrant increased scrutiny. It is often possible to construct a convoluted narrative that links multiple different health problems back to a single purported cause. Such narratives may seem to have a scientific veneer, but on closer inspection have serious fundamental flaws. We should rightfully question whether disorders with quite different underlying physiological bases should be treatable with a single "one size fits all" biomedical intervention. Any benefit observed from such a treatment is very likely nonspecific (ie, a placebo response) and not related to the proposed mechanism of action of the therapy.

Counseling Families of Children With Developmental Disorders About CAM

Virtually every article written about the widespread use of CAM therapies in children with chronic medical conditions concludes with the recommendation that medical providers need to discuss the use of these therapies with the caregivers of these children. What, then, should be said? The American Academy of Pediatrics has suggested that providers increase their knowledge about nonstandard practices, be able to critically analyze the merits of specific therapies, identify potential safety risks, provide families with information about all therapeutic options, and emphasize the importance of investigating nonstandard practices using rigorous scientific

methodology.[70] However, it is challenging to put such recommendations into practice when there is little or no reliable evidence of safety or efficacy to draw on.

Even for CAM therapies that have been tested in an appropriately rigorous manner, an all too common occurrence is that proponents develop such a strong belief in the therapy itself that they are blinded to the reality that all available evidence indicates the therapy has no greater benefit than a placebo. When this occurs, believers in the therapy often propose further investigation (to study an ever-smaller component of an herb, a different patient population, etc) in an effort to continue the search for evidence that the therapy has value. Caregivers of children with developmental and behavioral disorders may also consider evidence of inefficacy in an idiosyncratic manner. One of the first controlled trials of secretin in children with autism showed that secretin was no more effective than placebo in improving behavioral and communication features.[71] Even after the parents of the participants were informed of the negative results, however, a large percentage expressed interest in continuing the treatments. This suggests that caregivers of children with developmental disabilities may be considerably influenced by factors other than what the scientific community considers acceptable evidence in their decisions about which treatments to pursue for their children. One potential explanation is that when anxiety is high as a consequence of having a child with a condition that has unclear etiology and uncertain outcome, a caregiver may exhibit a greater likelihood of accepting a belief that might be viewed as unscientific. Such beliefs can be powerfully reinforced by the social milieu (such as support groups or Internet-based organizations) in which parents may find themselves immersed. A major challenge for the scientific community is to find new ways to actively influence these networks to accept and disseminate accurate, scientifically supported information. It remains to be seen whether the most vocal CAM proponents are willing to change their practices in response to scientific evidence—the fact that CAM therapies with good evidence of inefficacy are still commonly used in children with developmental and behavioral disorders suggests otherwise.

The need for health care professionals to be accepting of diverse healing traditions is commonly invoked as a reason why CAM therapies should not be marginalized by the medical community.[72] However, this issue may not be particularly relevant when considering CAM use among children with disabilities. For instance, only a minority of caregivers of children with ASDs cite cultural reasons as important in their decision to pursue a particular CAM therapy.[28] Analysis of other surveys of CAM practices in children with autism illustrates that most of the therapies that are being commonly used have their origins in scientifically implausible beliefs about the etiology of the disorder, as opposed to being associated with any particular cultural or religious tradition.[27,29–31]

Health care professionals need to use several different approaches when discussing CAM therapies with families. Families should be strongly advised to avoid those

therapies that are clearly risky and have insufficient evidence of efficacy (eg, chelation therapy in autism). Therapies for which there is adequate evidence of safety and efficacy in children with developmental and behavioral disorders should be recommended; however, few (if any) CAM therapies currently fall into this category. A larger group of therapies may have some degree of potential plausibility but have not been adequately tested. These therapies should not yet be promoted, but should undergo more rigorous clinical investigation. Finally, the small number of practices that are clearly part of cultural or religious traditions should usually be tolerated, assuming that there are no major anticipated safety concerns. Health care professionals should not be afraid to use reasoned argument to dissuade families from pursuing inappropriate CAM therapies—no one else may be willing or able to confront the health fads that have the potential to create risks for children with developmental and behavioral disorders.

Ethical Considerations

The story of James at the beginning of this chapter highlights some of the complexities inherent in evaluating the ethics of CAM use in children with disabilities. Principles that need to be considered within an ethical framework include beneficence, non-maleficence, autonomy, justice, and truthfulness. In the case of CAM use in children, these principles may come into direct conflict with one another, and clinicians must give careful attention to each when rendering advice about nonstandard medical practices.

A health care professional has the responsibility to administer therapies that are beneficial to the patient and society. The importance of basing treatment decisions on the best available evidence is paramount—as is the need for the scientific community to continue to develop reliable and valid evidence. It is challenging to know, however, whether the principle of beneficence is truly being upheld in situations where there are no reliable data supporting the value of a therapy, or if all we have are indirect or weak surrogate outcome measures (such as parental perceptions of the effectiveness of the therapy). An equally difficult situation arises when the safety of a CAM therapy is considered. Again, safety data for many of the commonly used CAM therapies are lacking. While each of the treatments prescribed to James can probably be broadly viewed as "safe," how should we consider the aggregate burden of subjecting a child to nearly 30 different treatments (some administered multiple times per day)? At some point, this approach could be viewed as primarily fulfilling a need to "do something" as opposed to actually contributing to the well-being of the child. While courts have typically upheld the right of a parent to pursue a CAM therapy for their child (except in cases of a life-threatening disease), health care professionals need to continue to carefully balance the principles of beneficence, non-maleficence, and autonomy in deciding whether it is justifiable to recommend (or tacitly support) an ineffective, albeit probably safe, therapy.

The ethical principle of justice often relates to the fair allocation of medical resources and treatments. This principle has been invoked in discussions regarding the lack of access to CAM therapies.[73] Consideration should also be given, however, to the flip side of this argument. In some parts of the world, therapies that fall under the rubric of CAM are provided to populations as cheap substitutes for medical practices with a much stronger evidence base, with little concern for therapeutic equivalence. In this sense, CAM therapies may serve as agents that contribute to the unjust distribution of health services. This is also true in situations in which CAM proponents contribute to attitudes that decrease the use of medical practices (such as immunizations[74]) that have clear individual and public health value, or when the time spent in an ineffective therapy directly detracts from the pursuit of other interventions known to be effective (particularly if the cost of the ineffective therapy, which for some CAM therapies can be excessive, creates undue financial burden for the family).

Placebo and other expectancy effects may account for most or all of the benefits appreciated by users of many CAM therapies. Some CAM proponents would argue that the mechanism of efficacy is less important than the fact that benefit occurs. In fact, the use of de facto placebos may be quite common in conventional medical practice as well.[75] The deception that occurs, however, when a provider prescribes an innocuous but ineffective therapy purely to achieve some degree of placebo effect runs counter to the ethical principle of truthfulness. Although a potential role for the clinical use of placebos in developmental pediatrics has been suggested,[76] a consensus has not been reached on the appropriateness of this notion.

Conclusion

Although the use of CAM therapies is widespread among children with developmental disorders, there is a paucity of reliable data supporting the value of virtually any of these approaches. Some treatments that have their origin as a complementary or alternative therapy may ultimately develop a sufficient evidence base that supports their widespread use. Until this occurs, health care professionals should remain appropriately skeptical, and the rational scientific community needs to remain on guard against therapies, whether viewed as CAM or conventional, whose use is not justified by the available evidence. Pediatricians in the medical home benefit from a continuous, longitudinal relationship with the family and patient. When pediatricians build therapeutic relationships with families through trusting, open relationships, the family is likely to be more willing to disclose CAM use and to consider physician advice regarding these therapies. Truly family-centered care should include efforts to teach families of children with developmental and behavioral disorders to evaluate therapeutic claims critically and to clearly understand the potential hazards (physical, emotional, and financial) that can accompany the use of unproved therapies.

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Chapter 22

Sensory Impairments: Hearing and Vision

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Hearing Impairment

Hearing loss is a potentially hidden impairment that can have a devastating impact on language, social, and learning abilities if not detected and treated. The adoption of universal screening of hearing in newborns in the United States has significantly lowered the average age of identification of hearing loss, but many children with delayed onset or progressive hearing loss are still diagnosed only after crucial developmental opportunities have been missed.[1] Comprehensive treatment with amplification of hearing and educational interventions to promote communication and potential surgical interventions, such as cochlear implants, facilitate optimal outcome, even in children with profound hearing loss. Primary pediatric health care professionals must therefore be knowledgeable regarding early identification and diagnosis of hearing impairment and their role in addressing the multiple factors that can influence successful functioning in these children.

Epidemiology and Etiology

The prevalence of hearing loss varies depending on the population studied, but the overall rate of congenital severe-to-profound bilateral sensorineural hearing loss is reported to be 1 to 2 per 1,000 live births.[2] A further 2 to 3 per 1,000 subsequently acquire severe loss. Many more children suffer milder degrees of hearing impairment or unilateral hearing loss. The broad categories of hearing impairment include *sensorineural* (dysfunction of the cochlea and/or its neural connections to the cortex) and *conductive* hearing loss (interruption along the conductive pathways: the pinna, external auditory canal, tympanic membrane, and middle ear structures). The clinician should also be alert to mixed hearing loss, where a conductive component is superimposed on an underlying sensorineural hearing loss.

Sensorineural hearing loss results from genetic causes in 30% to 50% of cases. Of these, 70% to 80% have an autosomal recessive pattern of inheritance, frequently

with no family history of hearing loss or external physical manifestations of the disorder. More than 60 loci for genes associated with non-syndromic hearing impairment have been identified.[2] Mutations in the gap junction proteins beta 2 and beta 6 (GJB2 and GJB6) are a common cause of hearing impairment, and a mutation of GJB2, which encodes the Connexin protein 26 (one of the main factors in potassium homeostasis in the cochlea), is responsible for up to 50% of the hearing loss in certain populations.[3] In addition, more than 500 syndromic forms of deafness have been identified, along with many mitochondrial genes for syndromic and non-syndromic hearing impairment. Table 22.1 outlines some of the more common genetic syndromes associated with hearing loss.

Prenatal acquired causes of hearing impairment include the congenital infections toxoplasmosis, rubella, cytomegalovirus, and herpes simplex, which can be associated with progressive hearing loss. Prenatal exposure to toxins, such as alcohol and mercury, can also lead to subsequent hearing loss.[4] Extremely premature infants are at increased risk of hearing loss due to a variety of factors, including hypoxia, acidosis, hypoglycemia, hyperbilirubinemia, high levels of ambient noise, and ototoxic drugs, such as aminoglycosides.[4] Bacterial meningitis is associated with sensorineural hearing loss in up to 10% of cases.[5] Audiological follow-up of these children is essential, as the hearing loss can be progressive. Prolonged exposure to loud noise, either environmental or recreational (especially related to the use of audio headphones), is an increasingly common cause of high-frequency hearing loss (Table 22.2).

Table 22.1. Genetic Syndromes That Are Associated With Hearing Impairment

Syndrome	Associated Clinical Features
Autosomal Dominant	
Waardenburg	White forelock; heterochromia iridis Lateral displacement of inner canthus of each eye
Alport	Nephritis
Branchio-oto-renal	Branchial arch anomalies; renal anomalies; mixed hearing loss; can have temporal bone abnormalities
Stickler	Flat facies; myopia; spondoepiphyseal dysplasia; hypotonia
Treacher-Collins	Malar hypoplasia; downslanting palpebral fissures; malformation of external ear (conductive hearing loss)
Autosomal Recessive	
Usher	Retinitis pigmentosa; vestibular dysfunction (3 types with varying degrees of hearing loss—progressive)
Pendred	Enlarged vestibular aqueduct Goiter; can have normal thyroid function
Jervell and Lange-Nielsen	Cardiac conduction problems (prolonged Q-T interval)

Table 22.2. Degree and Effects of Hearing Loss[a]

Degree of Hearing Loss	Hearing Level (dB)	Effects
Normal	0–15	• Can detect all aspects of speech
Minimal	16–25	• May miss up to 10% of speech • May respond inappropriately • Peer social interaction affected
Mild	26–40	• May miss up to 50% of speech • May be labeled as "behavior problem" and "poor listener"
Moderate	41–55	• May miss 50% to 100% of speech • Speech quality likely to be poor • Vocabulary is limited • Compromised communication ability • Low self-esteem possible
Moderate/ Severe	56–70	• 100% of normal volume speech lost • Delayed speech and poor intelligibility • Social isolation likely
Severe	71–90	• Loud voices only heard within 12 inches of ear • Delayed speech and language if loss is prelingual • Declining speech abilities and atonal voice if loss is postlingual
Profound	90+	• Sound vibrations felt rather than heard • Visual cues primary for communication • Peer group of hearing impaired children preferred

[a] Adapted from: Bachmann KR, Arvedson JC. Early identification and intervention for children who are hearing impaired. *Pediatr Rev.* 1998;19:155–165.

Identification

The key to an optimal outcome for the child with a hearing impairment is early identification and intervention. In 2000 the Joint Committee on Infant Hearing, which included representatives from the American Academy of Pediatrics (AAP), issued a position statement recommending universal screening of newborns for hearing loss, and this practice has been adopted by most states.[1] It is also recommended that all infants who have risk indicators for delayed onset or progressive hearing loss should have regular assessment of hearing every 6 months until age 3 years. Box 22.1 lists risk factors indicating the need for follow-up hearing evaluations.

The Centers for Disease Control and Prevention (CDC) Early Hearing Detection and Intervention program has set "1-3-6" goals, namely, hearing screening before 1 month of age, diagnostic audiological evaluation for children with hearing loss before 3 months of age, and early intervention for these children before 6 months of age.[6] The AAP, in partnership with the National Center for Hearing Assessment and Management and the CDC, has developed an algorithm to assist physicians in the process of early hearing detection and intervention.[7]

Box 22.1. Risk Factors for Delayed Onset of Hearing Loss[a]

History
- Parental or caregiver concern regarding hearing, speech, language, and/or developmental delay
- Family history of permanent childhood hearing loss

Neonatal Indicators
- In utero infections—cytomegalovirus, rubella, syphilis, herpes, or toxoplasmosis
- Hyperbilirubinemia at a serum level requiring exchange transfusion
- Persistent pulmonary hypertension
- Use of extracorporeal membrane oxygenation
- Use of ototoxic medications

Postnatal Indicators
- Syndrome associated with sensorineural and/or conductive hearing loss or eustachian tube dysfunction
- Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and Usher syndrome
- Neurodegenerative disorders such as Hunter syndrome
- Sensory motor neuropathies such as Friedreich ataxia and Charcot-Marie-Tooth syndrome
- Postnatal infections associated with sensorineural hearing loss including bacterial meningitis
- Head trauma
- Recurrent or persistent otitis media with effusion for at least 3 months

[a] Modified from: Joint Committee on Infant Hearing, American Academy of Audiology, American Academy of Pediatrics, American Speech-Language-Hearing Association, Directors of Speech and Hearing Programs in State Health and Welfare Agencies. Year 2000 position statement: principles and guidelines for early hearing detection and intervention programs. *Pediatrics*. 2000;106:798–817.

Primary pediatric health care professionals thus should not rely on behavioral symptoms to identify hearing loss, although such symptoms might lead parents to raise concerns. The obvious manifestations of hearing loss include failure of an infant to startle to loud noises or to turn to localize a sound. Toddlers might not respond to environmental sounds or might appear to ignore requests or instructions or request a higher volume on electronic sound sources. A key clinical sign in children with severe to profound hearing loss is the failure to develop "canonical babbling" (use of discrete syllables such as "ba" "da" "na") by 11 months. Delayed development of speech is a universal symptom of hearing impairment, and even milder degrees of hearing impairment can cause difficulties with language development, especially the processing and production of the softer, higher frequency sounds such as some consonants (eg, "s" and "t"). It should be noted that youngsters with even a profound hearing loss will begin to vocalize before 6 months of age, although further language development is impeded.

Clinicians must be alert to parental concerns regarding a child's hearing, delays in language development, or significant articulation deficits. Such concerns should be identified as part of standardized developmental screening that is performed in accordance with the recent algorithm recommended by the AAP (see Chapter 6).[8]

In children with otitis media with persistent middle ear effusion, the level of hearing loss should be documented and monitored closely.

Assessment of Hearing

Screening of hearing in the clinical office setting using automated otoacoustic emission measuring devices or audiometers can be a useful first step. There should not be reliance on response to environmental sounds, such as rustling paper, as this does not rule out milder levels of hearing loss or differentiate thresholds of hearing at various frequencies. If there is any question of hearing impairment, the child should be referred for formal audiological evaluation.[9]

Objective measures are most reliable in infants and younger children who cannot provide consistent behavioral responses. *Auditory evoked potentials* are electrophysiologic responses that assess auditory function and neurologic integrity. A click is introduced by an inserted earphone or a headphone at the external canal and the transmission of the low energy evoked potential through the brain stem pathways to the auditory cortex is recorded by means of scalp electrodes. It is important to note that auditory brainstem response (ABR) testing does not measure how the sound is being interpreted and processed, and it should be used in conjunction with behavioral audiometry whenever possible. Automated ABR tests (where responses are interpreted by computer and reported as "pass" or "fail") are used frequently in newborn hearing programs.

Otoacoustic emissions (OAE) testing measures the integrity and sensitivity of the cochlea, as well as indirectly reflecting middle ear status. The OAE are a form of acoustic energy produced by active movements of the outer hair cells of the cochlea in response to sound. Testing entails the introduction of a click via a probe in the ear canal with measurement of the emissions from the inner ear by a microphone. This test is relatively simple and highly sensitive, but it is less specific than ABR testing and can be affected by outer ear canal obstruction and middle ear effusion and can be inaccurate in specific circumstances. For instance, OAE testing will be falsely normal (false negative) in the infant who has pure sensorineural hearing loss (eg, from hyperbilirubinemia).

In newborn screening, if OAE testing indicates hearing loss, follow-up with ABR is recommended. Another helpful application of OAE is in the identification of infants and children with auditory neuropathy, where cochlear function is normal (normal OAE) but auditory nerve conduction is distorted (as measured by ABR).

Hearing tests that elicit *behavioral responses* allow for more frequency-specific testing and confirmation that sound is being perceived by the child. *Behavioral observation audiometry* can be used in very young infants (birth–6 months) to establish estimated levels of hearing. This technique entails controlled observation of an infant's behavioral response to sound stimulation under controlled conditions. These

responses include the auro-palpebral reflex, startle and arousal responses, and rudimentary head turning. This type of testing is prone to relatively high false-negative and false-positive responses. *Visual reinforcement audiometry* can be used in children by 6 months of age, and it is particularly helpful in the 1- to 3- to 4-year age range. This technique uses systematic reinforcement of behavioral responses. The typical application is to have the child seated on the parent's lap in the sound booth with animated lighted toys placed such that when the child turns in response to sound from the speaker, the toy at that speaker is lit to reinforce the response. After conditioning, the sound is presented before the toy lights up, and thus the child's response to the varying auditory stimuli can be measured.

Play audiometry can be used in children 2 years of age and older as attention spans increase. The child responds to sound by performing tasks such as dropping or stacking blocks or placing rings on pegs. *Pure tone and speech audiometry* provides more accurate measurement of response to pure tones or speech, where older children are asked to respond to signals generated by a calibrated audiometer. Use of speakers has the limitation of only reflecting hearing "in the better-hearing ear," and once children accept the use of headphones, more accurate assessments can be made in each ear. The results of hearing tests are represented graphically on an audiogram, which displays auditory threshold in decibels as a function of frequency in hertz. This form of evaluation relies on the attention and cooperation of the child (eg, a young child with autistic disorder might have normal hearing but fail behavioral audiometry testing and require ABR for accurate assessment).

Medical Evaluation

When hearing loss has been identified, further medical assessment is necessary.[9] A thorough history can establish risk factors and potential etiologies. In children with sensorineural hearing loss, it is essential to rule out any associated conductive component. A detailed general physical examination should include pneumatic otoscopy and tests of vestibular function. Comprehensive evaluation is important to look for associated disabilities. For example, unexplained fainting spells in a deaf child might signal a cardiac conduction defect (long QT interval) of Jervell and Lange-Nielsen syndrome. Ophthalmologic evaluation is also essential to rule out conditions such as retinitis pigmentosa with progressive loss of vision, which occurs in children with Usher syndrome. Chorioretinitis accompanies some of the congenital infections, and this finding might help establish an etiologic diagnosis. Routine evaluation for refractive errors is important to ensure optimal vision for these children, who are more reliant on visual input for communication and learning.

The extent of special investigations to be performed depends partly on clinical presentation. It is currently recommended that all children with sensorineural hearing loss should have a high-resolution computed tomography (CT) scan of the temporal bone to rule out conditions such as an enlarged vestibular aqueduct (associated with progressive hearing loss) or abnormalities of the cochlea and semicircular canals.

Magnetic resonance imaging scans do not have as high of a yield but can also identify anatomic abnormalities or neoplasms, and a CT brain scan might also reveal calcifications indicating congenital infection. Genetic testing should include DNA testing for genes, including Connexin 26 and 30, and mitochondrial mutations, including A1555G (increased susceptibility to aminoglycoside toxicity) and PDS (pendrin—present in Pendred syndrome).

Other special investigations could include tests of renal function or metabolic function, immunologic testing, or an electrocardiogram depending on clinical findings. If congenital or acquired infection is suspected, consultation by a pediatric infectious disease specialist is helpful in ordering and interpreting immunologic tests (cytomegalovirus, toxoplasmosis, rubella, herpes, syphilis).

Developmental Assessment

Formal assessment of cognitive, language, and social abilities should be carried out by professionals who have experience in testing children with hearing impairment. Tests of cognition include the Hiskey-Nebraska Test of Learning Aptitude[10] (specifically developed for children with hearing impairment), the performance subtests of the Weschsler Intelligence Scales,[11] (WISC-IV), or the Leiter International Performance Scale.[12] Visual-motor tasks, including the Bender-Gestalt and Developmental Test of Visual Motor Integration, are also used frequently.

Treatment

Comprehensive management should include attention to medical conditions, interventions to promote language development, educational interventions, use of assistive devices, and support and advocacy.[9] This is best accomplished by a team of professionals working in partnership with families, including the primary care clinician, otolaryngologist, audiologist, speech-language pathologist, and an educator of children who are deaf or hard of hearing.

The primary care clinician, working with the parents and other health care professionals, provides the medical home to facilitate and coordinate many of these interventions.[13] Audiologists confirm the existence and degree of hearing loss and provide recommendations for amplification and assistive technology. Otolaryngologists will be able to assess middle ear function and to evaluate for any surgically correctable causes of hearing loss, such as cholesteatoma, ossicular abnormalities, or other anomalies of the conductive system. They can also provide consultation regarding candidacy for cochlear implantation.

When a significant hearing loss has been discovered, the child should be fitted with a hearing aid as soon as possible. Hearing aids can be fitted in infants based on estimates of hearing thresholds from ABR measurements. Once a child is old enough to participate in behavioral hearing tests, these results can be incorporated into more precise calibration of hearing aids. The goal of amplification is to make speech and

other environmental sounds audible while avoiding high-intensity sound levels that are aversive or could damage residual hearing. A variety of forms of amplification are available, including behind-the-ear or ear-level hearing aids that fit behind the pinna with amplified sound transmitted to the ear canal via the custom-fit ear mold. These devices can also be used with telephones and with direct input from FM auditory trainers, where the primary speaker (usually the classroom teacher) wears a lapel-type microphone that transmits the speaker's voice directly to the hearing aid. Bone conduction devices are used for children with certain types of conductive hearing loss, such as atresia of the external auditory canal.

While hearing aids are effective for children with moderate to severe hearing loss, cochlear implants are revolutionizing the management of children with profound hearing loss.[14] A cochlear implant is an electronic device, part of which is surgically implanted into the cochlea and the remaining part worn externally. The external speech processor consists of a microphone, usually worn behind the ear, that transmits sounds to a speech-processing computer, that in turn converts the sound into an electric code. An external coil then transmits the signal across the skin to the internal receiver system implanted within the temporal bone and connected to multichannel electrodes placed within the cochlea. The electrodes are located at different sites to use the tonotopic organization of the spiral ganglion cells within the cochlea. Clinical trials have indicated positive outcomes, with significant improvement in appreciation of sound in everyday situations, speech recognition and understanding, and expressive language abilities. Recent studies have demonstrated lack of significant surgical complications and positive functional outcomes, even in children who receive their implants before 12 months of age.[14] Implantation before 2 years of age has been found to provide the most advantage with regard to speech perception and language development, with studies suggesting that most children with profound deafness who receive implants between 12 and 24 months of age will enter school with near-normal language skills. Children who use any form of amplification device, and especially those who have cochlear implants, need auditory training to help them understand the meaning of the newly amplified sounds.

A number of assistive devices are available, including telecommunication devices for the deaf, closed captioning of television, and adapted warning devices, such as vibrating devices or flickering lights to indicate a ringing alarm or telephone. Advances in information technology have, of course, enabled enormously increased opportunities for communication for individuals with hearing impairment. The Internet and e-mail, as well as text messaging by cellular telephones and voice to text technology, have broken down barriers at many levels.

Early intervention to promote language development remains the most critical management challenge for children with hearing impairment.[15] The child with profound hearing loss and his or her parents and other caregivers should receive professional assistance in establishing a functional system of communication as soon as possible.

There are many differing opinions regarding the most appropriate communication and instructional techniques. Options include sign language (manual communication), lip reading and use of speech (oral communication), or a combination (total communication). Children with profound hearing loss who have not received cochlear implants usually experience great difficulty learning to read lips and speaking fluently; they are best served by early exposure to visual and manual forms of communication, such as sign language. However, children with milder degrees of loss, and those who have received cochlear implants, are better able to communicate with those of normal hearing by developing their oral language skills.

The advent of universal newborn hearing screening has provided a unique opportunity to study the effects of early intervention on child development, particularly as related to hearing impairment. Studies involving children in the Colorado Home Intervention Program have definitively established that early intervention services for families with infants with hearing loss identified in the first few months of life results in significantly better language, speech, and social-emotional development.[15] Children who were diagnosed and received services before 6 months of life did significantly better than those diagnosed later, where intervention kept language delays from increasing, but it did not enable them to catch up with regard to delays that were already present at the time of diagnosis.[15]

For school-aged children, educational interventions should be tailored to the individual needs of each student. These services are mandated through the Individuals with Disabilities Education Act (IDEA). Options for those whose hearing loss has not been fully corrected range from use of interpreters in a regular school and classroom to special programs in a regular school or enrollment in a school for the deaf. Children with hearing impairment must have the opportunity for full participation in academic and social activities. The optimal school setting to achieve this goal depends on individual characteristics of the child and the educational system in that geographic region.

Parents of the child with newly diagnosed hearing loss are dealing with significant grief, while at the same time being faced with enormous amounts of information and the need to make decisions regarding treatment approaches.[16] Counseling can be helpful in assisting the parents to work through their feelings and adapt to their new roles. The primary pediatric health care professional can be a vital source of information and support for parents who often receive conflicting advice regarding both medical and educational interventions deemed necessary for the child. Parents face numerous stresses, including adjustment to the diagnosis and the need to learn new forms of communication, and they need their clinician to assist them in accessing the most appropriate therapies and interventions for their child.

Hearing Impairment: Conclusion

Preventive efforts are crucial, including rubella immunization and other measures to decrease the risk of prenatal infections and minimize the other risk factors previously listed. Immunization against *Haemophilus influenzae* type b infection has significantly decreased the incidence of this form of bacterial meningitis and the deafness associated with it. Primary pediatric health care professionals should also counsel families regarding risks of exposure to loud noises, such as the use of headphones or earphones to listen to loud music. As the consistent provider of longitudinal care for children with hearing impairment, the primary pediatric health care professional thus has a crucial role in their lives, from early identification of problems to coordination of the subsequent evaluation and management interventions, to support and advocacy for the child and family as he or she progresses through the educational system, to success as an adult.

Vision Impairment

In humans, of the 5 primary senses and their respective central nervous system (CNS) networks for processing information, vision is unique. Unlike touch, smell, and taste, vision enables reception of environmental information that is both near and distant. While it shares with hearing an important role in processing discontinuous *sequential* bits of information (particularly critical for use in receptive language, including verbal/auditory and nonverbal/visual elements), vision also provides *simultaneous* and *continuous* reception of immense chunks of contextual information in a gestalt. Because of this power, vision assumes a unique role in enabling rapid synthesis of information pouring in from all of the senses, automatically boosting the efficiency of the brain's associative and executive functions. For infants and toddlers, it also serves as a powerful motivator for further exploration, which in turn stimulates increasingly precise reaching and moving through space. Vision therefore facilitates safe movement through space and bodily coordination. For young, normally developing children, most learning is not taught by others; rather, it is through incidental acquisition of information through experience within their environments. It has been estimated that as much as 80% of this learning is visually mediated.

Visual impairment is a relatively low-incidence disability, often associated with neurologic comorbidities, which can have profound influence on how children learn, move, and experience the world. Some vision-impairing medical conditions, when detected early enough, can be treated and prevented from causing permanent and significant loss of visual functions. For other ophthalmologic and vision-related CNS disorders for which there is no medical or surgical treatment available, early detection followed promptly by tailored educational interventions, environmental modifications (eg, enlarging print size, reducing glare, increasing contrast), "low vision" devices to augment any residual visual function, and support services for families can make a major difference in the child's ultimate learning, functional independence, and social/emotional health.

Definitions of Blindness and Visual Impairment

The term *legal blindness* refers to best corrected distance visual acuity in the better eye of 20/200 or less or a visual field restriction in the better eye of less than 20 degrees, or both. An individual with visual acuity of 20/200 must be 20 feet or closer from the target in order to see visual detail that someone with normal vision could discriminate at a distance of 200 feet. Approximately 75% of legally blind children have some useful vision. Traditionally, corrected visual acuity between 20/70 and 20/200 in the better eye is considered *low vision,* and in many states, establishes a child's eligibility for special education services based on visual impairment. However, because visual function in children is determined by many factors besides distance visual acuity, planning medical and educational interventions for the child with any significant visual impairment should ideally be based on a thorough evaluation of visual function that includes such parameters as distance and near acuity, visual fields, sensitivity to contrast, depth perception, preferred head and eye positions, variability of visual function from day to day, response to illumination, color vision, eye motility, and responses to stationary versus moving targets.

Epidemiology and Etiology

The prevalence of visual impairment and blindness in children varies depending on definitions of severity, methods of ascertainment/surveillance, and populations' classification regarding economic wealth/poverty. Of the approximately 1.5 million children and adolescents worldwide who are blind (visual acuity <20/400), less than 4% live in developed countries.[17] In the United States and other industrialized countries, the prevalence of childhood blindness is approximately 3 per 10,000 children. In these countries, the combined prevalence of visual impairment, severe visual impairment, and blindness is approximately 10 to 22 per 10,000 children younger than 16 years. In contrast, the combined prevalence in some impoverished, developing countries is nearly 40 per 10,000.[18]

The epidemiology of visual impairment also depends largely on the relative balance between genetic and environmental risk factors on the one hand and access to health services, ophthalmologic care, and advanced medical technology on the other. In the most highly developed countries, the more prevalent causes of visual impairment are related to underlying CNS conditions, sequelae of extreme prematurity following technologically advanced care in neonatal intensive care units, and genetic conditions. In contrast, most children with visual impairments in developing countries, where poverty is widespread, have eye conditions caused by infections (eg, trachoma, toxoplasmosis, onchocerciasis), nutritional deficiencies (eg, vitamin A deficiency), and inadequate access to ophthalmologic treatments (eg, amblyopia due to lack of or delayed treatment of infantile cataracts or untreated strabismus).

Causes of visual impairment in children can be classified by primary anatomical site, etiological process (eg, infectious, genetic, metabolic, traumatic, etc), or timing of

onset. Prenatal causes of visual impairment include genetic conditions, other types of fetal malformations, prenatal infections, and hypoxia. Congenital abnormalities of the brain and/or eye may be limited to one specific structure in the eye or brain (eg, isolated coloboma of the iris or diminished number of rod cells in the retina), might affect multiple parts of the visual system (eg, both retinal dysfunction and anomalous routing of optic nerve fibers in oculocutaneous albinism), or be part of a multisystem syndrome that is either chromosomal (eg, Down syndrome), metabolic, or of unknown origin (eg, CHARGE anomalad, which includes coloboma, heart disease, atresia choanae, mental retardation, genital anomalies, and ear anomalies/hearing loss). In the perinatal period, visual impairments may result from hypoxia/ischemia, retinopathy of prematurity, and infection. Postnatal etiologies include tumors, nutritional deficiencies, trauma (accidental and non-accidental), infection, increased intracranial pressure, and systemic conditions. In industrialized countries, at least half of all children with visual impairment have multiple disabilities, and hereditary conditions account for roughly half of all childhood blindness.[19] However, in economically poor countries, where infectious and nutritional diseases affecting the eyes continue to play a major role, genetic conditions cause a significantly smaller percentage of blindness. In these developing countries, it is estimated that up to 40% of children who are blind have conditions that are preventable.

In the United States, the most common causes of severe visual impairment among preschoolers, in decreasing order of frequency, are cortical/cerebral visual impairment, retinopathy of prematurity, optic nerve hypoplasia, structural abnormalities of the eye, and albinism.[20,21] See Table 22.3 for features of these and other common causes of childhood blindness and visual impairment. For a more complete listing, please refer to the table in Teplin et al.[22]

Identification

One of the most important responsibilities for primary pediatric health care professionals is early detection of major conditions that interfere with normal vision, particularly during the newborn period, infancy, and through the preschool years (2–5 years). Recent guidelines issued jointly by the AAP and the American Academy of Ophthalmology recommend that appropriate screening for eye disorders, including red reflex examination, be performed at birth and with each well-child visit.[23] They recommend that the child's first screening for visual acuity occur "at the earliest possible age that is practical (usually at approximately 3 years of age),"[24] preferably using a Snellen chart or its equivalent (eg, HOTV cards, Allen Cards, or LEA symbols) for preschool-aged children. Photoscreening and photo-retinoscopy are new technologies that may allow rapid, reliable screening of preschoolers for conditions that can cause amblyopia.[25] Detection by the primary pediatric health care professional of possible abnormalities of the ocular structures, abnormal visual acuity screening results, or persisting parental concerns regarding a child's visual abilities warrant a referral to a pediatric ophthalmologist or other specialist trained and experienced in diagnosis and treatment of children's eye disorders. The clinician should also

Table 22.3. Common Disorders Causing Childhood Blindness or Visual Impairment

Condition	Pathophysiology and Exam Findings	Commonly Associated Eye, Neurologic, and Systemic Disorders	Common Developmental/Behavioral Features and Resources
Cortical/ cerebral visual impairment	Injury to or maldevelopment of optic radiations and/or visual processing areas of the brain. Normal ocular and optic nerve structure and function. Origin may be prenatal (eg, genetic/metabolic disorder), perinatal (eg, hypoxic-ischemic brain damage, periventricular leukomalacia, etc), or postnatal (eg, non-accidental brain trauma, meningitis, etc). Exam: normal eye exam, but poor central visual fixation and tracking; usually without nystagmus (unless comorbid ocular disorder).	Extremely heterogeneous, depending on locations, extent, timing of brain abnormalities. Often associated with one or more neurologic problems (eg, seizures, cerebral palsy, motor or verbal dyspraxia, intellectual disabilities, learning disabilities, hearing loss, autism). Visual function often fluctuates from hour to hour and day to day. Visual function often improves over first 1–3 years.	Often developmentally delayed beyond what would be expected based solely on the visual impairment. Difficulty maintaining focused attention. Easy fatigability. Tendency to avoid loud noise or visual clutter; difficulty with transitions between activities; may have prosopagnosia (difficulty recognizing faces even when visual acuity is adequate). Tendency to turn head and eyes away from object during reach for it. Prognosis is probably worse when cause is PVL vs occipital lobe damage.[a]

Abbreviations: AR, autosomal recessive; BW, birth weight; CNS, central nervous system; GA, gestational age; IGF-I, insulin-like growth factor-I; MRI, magnetic resonance imaging; PVL, periventricular leukomalacia; VEGF, vascular endothelial growth factor; VI, visual impairment; XR, X-linked recessive.

[a] Good WV. Cortical visual impairment. In: Dennison E, Hall Lueck A eds. *Proceedings—Summit on Cerebral/Cortical Visual Impairment, Educational, Family, and Medical Perspectives*. New York, NY: AFB Press; 2006:282.

[b] Termote J, Schalij-Delfos NE, Donders RT, Cats BP. The incidence of visually impaired children with retinopathy of prematurity and their concomitant disabilities. *J Am Assoc Pediatr Ophthalmol Strabismus*. 2003;7:131–136.

[c] Msall ME, Phelps DL, DiGaudio KM, et al. Severity of neonatal retinopathy of prematurity is predictive of neurodevelopmental functional outcome at age 5.5 years. *Pediatrics*. 2000;106:998–1005.

[d] Fiedler AR, Quinn GE. Retinopathy of prematurity. In: Taylor D, Hoyt CS eds. *Pediatric Ophthalmology and Strabismus*. 3rd ed. Edinburgh, UK: Elsevier Saunders: 2005:516517.

Table 22.3. Common Disorders Causing Childhood Blindness or Visual Impairment *(continued)*

Condition	Pathophysiology and Exam Findings	Commonly Associated Eye, Neurologic, and Systemic Disorders	Common Developmental/Behavioral Features and Resources
Retinopathy of prematurity (ROP)	Disorder of retinal neovascularization affecting immature retinal vessels in premature infants. Retinal oxygen levels regulate secretion of VEGF. In addition to other factors, ROP is believed to result from overproduction of VEGF and insufficient IGF-I. ROP occurs in most extremely low birth weight premature infants (BW <1,000g), but in the US, spontaneously resolves in 90%. However, even years later, even children with regressed ROP remain at some risk of late ocular complications (glaucoma, retinal detachment) and require regular ophthalmologic monitoring.	Extremely premature infants also have a higher incidence of strabismus, nystagmus, and high myopia. High prevalence of multiple disabilities in children with advanced stages of ROP; higher risk of other disabilities associated with worsening visual function.[c] Timely cryo- or laser-ablative therapy of avascular retina for "threshold" ROP disease can prevent retinal detachment and increase chances of better functional vision. Laser treatment is currently procedure of choice. In wealthier, developed countries, VI due to ROP primarily affects babies born at GA ≤31 weeks, with BW <1,250 g, contributing 3%–8% of prevalence of childhood blindness. In middle income countries, premature babies with higher GAs and BWs are affected, accounting for about 40% of childhood blindness.[d]	Sometimes associated with greater than expected difficulties with spatial awareness and motor coordination. Increased risk of behavior problems.[b] Parent support: Association for Retinopathy of Prematurity and Related Disorders: www.ropard.org

Abbreviations: AR, autosomal recessive; BW, birth weight; CNS, central nervous system; GA, gestational age; IGF-I, insulin-like growth factor-I; MRI, magnetic resonance imaging; PVL, periventricular leukomalacia; VEGF, vascular endothelial growth factor; VI, visual impairment; XR, X-linked recessive.

[a] Good WV. Cortical visual impairment. In: Dennison E, Hall Lueck A eds. *Proceedings—Summit on Cerebral/Cortical Visual Impairment, Educational, Family, and Medical Perspectives*. New York, NY: AFB Press; 2006:282.

[b] Termote J, Schalij-Delfos NE, Donders RT, Cats BP. The incidence of visually impaired children with retinopathy of prematurity and their concomitant disabilities. *J Am Assoc Pediatr Ophthalmol Strabismus*. 2003;7:131–136.

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Table 22.3. Common Disorders Causing Childhood Blindness or Visual Impairment *(continued)*

Condition	Pathophysiology and Exam Findings	Commonly Associated Eye, Neurologic, and Systemic Disorders	Common Developmental/Behavioral Features and Resources
Optic nerve hypoplasia (ONH)	Abnormal fetal development (diminished number of nerve fibers) of optic nerve. Etiology is variable (eg, maternal diabetes, fetal alcohol exposure, chromosomal abnormalities, Goldenhar syndrome, osteogenesis imperfecta, Apert syndrome, etc). May be unilateral or bilateral. Eye exam: small optic nerve head appearing gray or pale, surrounded by yellow margin and darker ring ("double ring sign"). Retinal vessels are tortuous; nystagmus present if significant visual impairment. Variable peripheral field cuts. Important to rule out constellation of ONH, absent septum pellucidum, and endocrinopathies due to various degrees of hypopituitarism. These comprise the elements of septo-optic dysplasia. Important to obtain brain MRI and endocrine consultation. May present in newborn with jaundice (hypothyroidism), temperature instability, and/or hypoglycemic seizures (adrenal insufficiency).	Visual acuity highly variable, ranging from normal to no light perception; visual acuity is usually stable over time. Often associated with variety of CNS anomalies, each of which may have its own neurologic characteristics. Monitor for emerging signs of hormonal deficiencies (eg, short stature, hospitalizations for dehydration or hypoglycemia). Risk of sudden death during febrile illness or anesthesia (inadequate adrenal response to stress).	Often associated with feeding problems, food refusals. Some children have sensory integration problems with hypersensitivity to certain textures or sounds. Cognitively, there is wide variability, with many having learning problems; some fall into the range of autism spectrum disorders. Some, however, are gifted in certain domains (eg, music). Family support: http://www.focusfamilies.org

Abbreviations: AR, autosomal recessive; BW, birth weight; CNS, central nervous system; GA, gestational age; IGF-I, insulin-like growth factor-I; MRI, magnetic resonance imaging; PVL, periventricular leukomalacia; VEGF, vascular endothelial growth factor; VI, visual impairment; XR, X-linked recessive.

[a] Good WV. Cortical visual impairment. In: Dennison E, Hall Lueck A eds. *Proceedings—Summit on Cerebral/Cortical Visual Impairment, Educational, Family, and Medical Perspectives*. New York, NY: AFB Press; 2006:282.

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Table 22.3. Common Disorders Causing Childhood Blindness or Visual Impairment *(continued)*

Condition	Pathophysiology and Exam Findings	Commonly Associated Eye, Neurologic, and Systemic Disorders	Common Developmental/Behavioral Features and Resources
Albinism	Two main types: oculocutaneous (AR) and ocular (XR), each of which has several subtypes, according to combination of 13 genes, which influence melanin metabolism. Reduced or absent melanin pigment production in eyes (retina, iris) and/or skin/hair. Visual acuity is reduced. Other eye findings: nystagmus, iris translucency, macular hypoplasia, and abnormal routing of optic nerve fibers through chiasm.	Visual acuity may actually improve after the first 4–6 months, and nystagmus may spontaneously improve later in childhood. Strabismus is common. Corrected visual acuity is variable, but typically is in the range of "low vision" (ie, between 20/70 to 20/200). A significant subgroup may have vision in the "legally blind" range (worse than 20/200).	No other associated disabilities. May have behavioral difficulties because of others' misinterpretation or misunderstanding of child's apparent aloofness (difficulty with recognizing people at a distance due to nystagmus or poor acuity). There may be transient fine motor delays due to problems with visual tracking and depth perception. May be some emotional difficulties secondary to stigma of "differentness," especially for children whose family members have dark skin. Benefit from consistent support from family, peers, and teachers. Low-vision interventions may include magnification aids, high-contrast materials, sunglasses, telescopes for distance viewing. Advocacy and parent support: National Organization for Albinism and Hypopigmentation: www.albinism.org

Abbreviations: AR, autosomal recessive; BW, birth weight; CNS, central nervous system; GA, gestational age; IGF-I, insulin-like growth factor-I; MRI, magnetic resonance imaging; PVL, periventricular leukomalacia; VEGF, vascular endothelial growth factor; VI, visual impairment; XR, X-linked recessive.

[a] Good WV. Cortical visual impairment. In: Dennison E, Hall Lueck A eds. *Proceedings—Summit on Cerebral/Cortical Visual Impairment, Educational, Family, and Medical Perspectives*. New York, NY: AFB Press; 2006:282.

[b] Termote J, Schalij-Delfos NE, Donders RT, Cats BP. The incidence of visually impaired children with retinopathy of prematurity and their concomitant disabilities. *J Am Assoc Pediatr Ophthalmol Strabismus*. 2003;7:131–136.

[c] Msall ME, Phelps DL, DiGaudio KM, et al. Severity of neonatal retinopathy of prematurity is predictive of neurodevelopmental functional outcome at age 5.5 years. *Pediatrics*. 2000;106:998–1005.

[d] Fiedler AR, Quinn GE. Retinopathy of prematurity. In: Taylor D, Hoyt CS eds. *Pediatric Ophthalmology and Strabismus*. 3rd ed. Edinburgh, UK: Elsevier Saunders: 2005:516517.

maintain a high index of suspicion for subtle visual dysfunction in any child with previously diagnosed neurologic impairments, particularly cerebral palsy, degenerative CNS disorders, hearing loss, and intellectual disabilities (mental retardation). Earlier detection of possible visual problems in children with known developmental disabilities can allow for thorough ophthalmologic evaluation. This in turn may identify treatments to ameliorate the visual problem and improve long-term outcome.[26] On the other hand, although vision dysfunction can contribute to learning problems, vision problems are not the underlying basis of dyslexia or other specific learning disabilities.[27]

Development

Motor

Wide variability is noted in the acquisition of motor skills by young children with visual impairment. Important determinants include the severity and type of visual impairment, comorbid disabilities (particularly those involving the motor system), and the degrees to which early environments are enriched with opportunities and encouragement for movement. For children with visual impairment but without other disabilities, early motor milestones that involve only balance, but no movement (eg, sitting and standing unaided), often occur on roughly the same timetable as for infants with normal vision. However, movement through space is typically delayed (eg, walking may not occur until 18–24 months). To prevent a tendency to remain passive and relatively immobile, it is important for these children to have active engagement with their environments. Invaluable services are provided by certified orientation and mobility specialists (COMS), whose job is to help children with visual impairments learn about the spatial arrangement of their environments and use that knowledge to move about efficiently and safely.[28]

Cognitive

For sighted infants, vision serves as a powerful motivator for further exploration, which in turn stimulates increasingly precise reach/grasp and movement through space in order to get to a desired person, place, or object. As they move and observe objects and other people move, young children with normal vision discover and repeatedly verify basic cognitive concepts (eg, object permanence and cause and effect). For infants with congenital blindness or severe visual impairment, understanding of these principles often takes longer, requiring learning via alternative sequences. Most severely visually impaired children have additional disabilities; for these children, greater fragmentation of incoming information creates more formidable obstacles to learning these fundamental concepts, causing greater delays in their acquisition. It is crucial that the young child with severe visual impairment be actively introduced to a variety of experiences, objects, qualities (eg, hot/cold, loud/soft, above/below, curved/straight, etc), and people to allow the child to use any available sensory functions to take in and store information. For example, instead of

simply telling the young blind child about a tree, it is more helpful in consolidating his concept of "tree" if he can simultaneously feel its bark and leaves, smell the blossoms, and hear the branches swaying in the wind. Parents of blind or severely visually impaired young children often need help in this teaching process. Parents also need support in resisting the temptation to anticipate the young child's every need or desire, so that they learn not to inadvertently restrict the child's learning opportunities by, for example, automatically retrieving objects the child has dropped or carrying the child around because of concern about inevitable but harmless collisions with nearby furniture. Caregivers need to understand how important it is for the child to experience the environment *actively* (ie, being given many opportunities to use developing language skills to verbally express needs and desires, listening skills and hands to search for dropped objects, and body and legs to move through space, even when this might be temporarily frustrating).[29]

Learning abilities and therefore required instructional methods may vary according to whether a child's visual loss was congenital versus adventitious because of the latter's advantage in having had prior visual experience.[30] This is especially evident regarding spatial orientation and spatial behaviors as well as acquisition of motor skills[31(pp54–55,106–107,110–111,117)]; however, studies have shown no correlation between age of vision loss and overall cognitive abilities.[31(pp88–89)] Teaching strategies for children whose blindness occurred prior to the establishment of visual memory (estimated to be approximately 5 years of age[30]) must rely exclusively on their nonvisual senses.

Language

As receptive and expressive vocabulary develop, language becomes an increasingly powerful tool for the child with blindness to learn about the environment. For many children with severe visual impairment or blindness and normal cognitive development, the pace of single-word acquisition is often comparable to that seen in children with normal vision. Some differences, however, are notable[32]—compared with sighted children, children with severe visual impairments are slower to acquire adjectives and verbs, have more prolonged phases of echolalic speech, are more likely to choose more egocentric topics to discuss, and have longer periods of pronoun confusion and reversals (eg, saying "You want a cookie?" when what they mean is: "I want a cookie."). As the child gets older, there may be gaps in understanding of words that have more visual referents (eg, "sky," "red") or that represent very large objects that would be difficult for the child to experience as a whole (eg, a building).

Play

Compared with young children with normal vision, preschoolers who are severely visually impaired but otherwise developmentally intact tend to engage in more solitary play and play with adults more than with peers. Compared with sighted preschoolers, there is about a 2-year delay across all categories of play. The level of visual functioning directly correlates with the sophistication of play. Vision plays a critical

role in acquisition of higher levels of play behavior among sighted children. For young children with severe congenital visual impairments, most of their spontaneous play is exploratory and focuses on simple manipulation, with much less functional play and even less symbolic/pretend play (eg, imaginative doll play). They are not likely to learn how to play without some guidance from others. When with peers, they have difficulty sustaining their social connection when there is a communication breakdown. These delays are likely to be even more substantial for children with visual impairment plus additional disabilities. Such difficulties also correlate with deficiencies in social skills.[33]

Strategies that can promote more interactive and educational play include providing toys and activities that are interesting to the child (eg, through tactile and auditory sensations), choosing play partners who have similar interests, and facilitating the child's awareness of the environment by use of verbal descriptions. Sighted peers can be shown how to interact with the child who is blind, with gradual fading of adult reinforcement and support.[34]

Social and Emotional

Infants with severe visual impairment often have facial expressions that are passive-appearing or somewhat serious; their social smiles are more subtle and fleeting, making it difficult for caregivers to detect and read the baby's social cues. The important early phase of pre-lingual, mutual social interaction may be jeopardized by possible misinterpretations by the caregiver. For example, the mother of an infant who is blind might notice the infant's lack of smiling on hearing her voice and mistakenly assume that he does not recognize her voice or does not want to be with her. A teacher of the visually impaired (TVI) specializing in young children could help this mother to understand that the infant's sudden stillness or a particular hand positioning may be subtle cues that he is responding to her. The child with severe visual impairment remains at risk for emotional and behavioral problems, largely related to difficulties in adapting to change and not having the abilities of sighted peers to read others' social cues or see their own impact on others. These potential difficulties in social expression, pretend play, and emotionally meaningful interactions with parents can often be prevented or at least ameliorated when parents and other early caregivers have access to services from TVIs for infants, toddlers, and preschoolers who can help them read the child's cues, respond reciprocally, and provide explicit modeling to promote the child's pretend play.[33] Often such TVIs are itinerant and work closely with the local early intervention (EI) teams.

Through a recent surge in research interest and new emphasis in both medical literature and popular media outlets, clinicians, teachers, and parents have become increasingly aware of clinical features often associated with autism spectrum disorders (ASDs) among sighted children (see Chapter 13). Children with visual impairments are no less likely than sighted peers to have ASDs, with higher risk among visually impaired children with evidence of other CNS damage or dysfunction (eg,

children with cortical visual impairment, CHARGE anomalad, and periventricular leukomalacia).[35] Studies have been conflicting as to whether children with other etiologies of blindness (eg, retinopathy of prematurity, Leber's congenital amaurosis, and septo-optic dysplasia) are at higher risk for ASDs. However, great caution must be exercised when considering the behaviors of children with significant visual impairment. Several common characteristics of an otherwise developmentally normal child with significant visual impairment could lead to erroneous identification of a comorbid ASD, including difficulties in establishing typical social relationships with peers, stereotypical/self-stimulatory behaviors (eg, hand flapping, rocking, eye-poking, light-gazing) when excited or upset, prolonged phase of echolalic language during the toddler and preschool periods, and what seem to be sensory processing "problems" (eg, oversensitivity to ambient noise, "inappropriate" touching or sniffing of objects and people). An autism diagnosis is therefore quite difficult to make in this setting and is best addressed by an interdisciplinary team of developmental specialists with input from a TVI or other professional who has had extensive experience working with children with visual impairments. Also of note is the observation that among children with severe visual impairment and stereotypical behaviors, these socially offensive behaviors often spontaneously decrease as the children pass through adolescence.[36]

Role of the Primary Pediatric Health Care Professional and Additional Interventions

Diagnostic and Medical Issues

As noted, the primary pediatric health care professional, as the medical home for the child and in partnership with the child's parents/caregivers, plays a key role in helping the child toward optimal functioning. The clinician should routinely provide surveillance and screening for possible ocular and CNS disorders and to identify visual problems as early as possible. In addition to making appropriate referrals to an eye care specialist for definitive diagnostic assessment of the eye problem, other pediatric subspecialty referrals may also be indicated (eg, neurologist, geneticist, and/or developmental-behavioral or neurodevelopmental pediatrician). An interdisciplinary developmental assessment team that includes a TVI and COMS can provide comprehensive evaluation of the child's strengths as well as the nature of his or her limitations related to the visual impairment. Other medical problems that are more frequently seen in children with visual impairments include sleep difficulties,[37] feeding problems in young children, and other sensory processing difficulties. The clinician may need to devote additional time to exploring these issues with parents or refer to a developmental consultant as needed.

Infants, Toddlers, and Preschoolers

The primary pediatric health care professional can make sure that the young child is being served by appropriate EI services, in collaboration with TVI and COMS professionals. The young child's parents may also benefit from contact with local,

regional, and national organizations that are dedicated to supporting parents of children with visual impairments, often related to specific medical diagnoses (see Table 22.1 and resources on page 489).

School-aged Children

Most students in the United States with blindness or low vision currently attend their own local public schools, receiving a range of special education services according to their needs. However, some school districts have specialized programs, TVIs who are on staff, and adaptive books and equipment, while others must scramble to find resources when a new child with visual impairment enters the system. Advanced planning can help considerably. Primary pediatric health care professionals, often in collaboration with the local EI program, can help parents of a toddler with visual impairment to advocate for current and future services (eg, establishing an early relationship with the school system's coordinator of special education, giving the system plenty of time to prepare for the child's entry at age 3 years). Often the intensity and format of these services varies over time. For example, during the early elementary years, a child may spend more time in a specialized resource setting with a TVI, where he or she can focus on learning necessary compensatory skills unique to this disability, such as reading braille, learning safe and independent mobility with a COMS, learning to use residual vision more efficiently, building listening skills, and using adaptive low vision devices (eg, magnifiers, closed circuit TV, etc). Depending on a student's abilities and unique educational needs, many can be successfully included into regular education classes for at least part of the day if appropriate support or consultation services from the TVI and COMS are available. For some students, intense skill-building courses, for example during the summer at a state's residential school for the blind, may provide unique opportunities unavailable in the local school to learn about specialized adaptive technologies (eg, computer software and devices that can interconvert braille, written text, and synthetic speech, etc) while also offering socialization opportunities for students from distant schools who have similar disabilities and interests.

In addition to coping with unique developmental and academic hurdles, students with visual impairments may also require specific help in learning appropriate social skills, many of which can rely heavily on learning about picking up on verbal and other nonvisual cues. They also may need guidance in self-regulation of common self-stimulatory behaviors, such as eye poking or rubbing, rocking, head-swaying, and conversational skills, such as staying on topic, taking turns, and determining when to interrupt.[38] These skills are among the unique learning needs specified in the American Foundation for the Blind's Expanded Core Curriculum[39] as requiring explicit, systematic, and sequential instruction to students with visual impairments. Other topics on this list include independent living skills, recreation and leisure skills, use of assistive technology, visual efficiency skills, and self-determination. Clinicians can help parents advocate for the inclusion of these instructional goals within their child's individualized education plan.

Adolescents

Adolescents with significant visual impairment must cope with the same upheavals and issues as youth with normal vision. However, as with most youth with chronic illness or other developmental disabilities, they also struggle with more basic issues of independent identity. Their success in activities leading to greater independence (eg, dating, exploration of job opportunities, higher skill levels) hinge to some extent on whether they have already mastered more basic self-help skills, such as personal grooming, food preparation, money management, travel, etc. They are at risk for distorted concepts about sexuality due to having missed typical visual cues during childhood regarding appropriate and inappropriate sexual behavior. Sex education needs to begin early. Through use of anatomically correct dolls and ongoing factual discussions, they can acquire knowledge of fundamental facts and vocabulary. As with sighted youth, sex education is most effective when embedded within learning about the emotional context of sexual relationships.

Deaf-Blindness

Children who have significant impairments of both vision and hearing constitute a special group because the educational interventions and support services required are different than those typically needed for children with either single disability. Deaf-blindness encompasses a range of severity of each disability. Approximately 17% are totally blind or have light perception only, 24% are legally blind, and 21% have low vision. Approximately 39%, 13%, and 14% have severe to profound, moderate, and mild hearing loss, respectively.[40] The most common etiologies include a variety of hereditary conditions, complications of prematurity, hypoxic-ischemic encephalopathies, CHARGE anomalad, post-infectious complications, and Usher syndrome. Approximately 85% of children with this dual sensory impairment have additional disabilities, particularly intellectual disability (mental retardation), speech/language problems, and orthopedic conditions.[41] The most important developmental issues include social interaction, mobility, communication, and cognitive grasp of concepts.[42] For individuals who are deaf-blind, their hands must be carefully, patiently, and consistently trained to become the primary sensory organs as well as the main means of communication with others.[43]

Vision Impairment: Conclusion

During recent decades, research interest and our knowledge base about developmental implications of blindness and visual impairment in children and adolescents has expanded, both supporting and challenging clinicians and teachers of the visually impaired to meet the unique developmental, educational, and social needs of these children. However, compared with other disabling conditions, blindness/visual impairment still represents a relatively small percentage of all children with special needs. As a result, much of the expanding knowledge base fails to get transferred and integrated into medical and educational practice where it is most needed. Primary

pediatric health care professionals can help close this gap by maintaining awareness of professionals and specialized agencies in their community or region who have expertise and experience in working with infants/preschoolers, school-aged students, and adolescents who are visually impaired and making prompt referrals when encountered as patients. Referral to a pediatric eye care specialist is mandatory, but that is only the beginning. It will usually be the primary care clinician's medical home, not the ophthalmologist's expertise, that will continue reinforcing the family's resilience, monitoring the child's developmental status and obstacles, and smoothing the path toward educational and behavioral consultation in order to optimize the child's progress. As devastating as it may seem initially for the family (and clinician) to confront the reality of a child's blindness or severe visual impairment, the health care provider's practical and positive attitude and commitment to helping parents advocate for necessary services and supports will prove extremely valuable.

Resources and Publications for Physicians and Parents of Children With Visual Impairment

Agencies and Web Sites

American Association for Pediatric Ophthalmology and Strabismus
http://www.aapos.org
Information about latest research efforts; concise overviews of common conditions causing visual impairment in children and treatment approaches.

American Foundation for the Blind (AFB)
www.afb.org
Resources, materials, educational devices and toys, useful suggestions for parents and teachers.

American Printing House for the Blind (APH)
www.aph.org
APH is a source of publications and adapted educational equipment for children and youth who are visually impaired.

Assist the Development of Visually Impaired Students through Online Resources
www.e-advisor.us
Useful information for primary care physicians as well as families about diagnostic and therapeutic procedures regarding a variety of diagnoses affecting visual impairments.

Blind Babies Foundation
http://blindbabies.typepad.com/resources/ http://blindbabies.typepad.com/resources
Excellent fact sheets summarizing major conditions of visual impairment in children.

National Association of Parents of Children with Visual Impairments (NAPVI)
http://www.spedex.com/napvi/index.html
A nonprofit organization of, by, and for parents committed to providing support to the parents of children who have visual impairments.

National Federation of the Blind (NFB)
www.nfb.org
A membership organization of blind people in the United States. NFB has numerous publications for parents and professionals on blindness and visual impairment and includes a national parent organization.

Texas School for the Blind and Visually Impaired
http://www.tsbvi.edu
Comprehensive Web site with many instructive articles on topics related to visual impairment in children, medical and educational approaches, and glossaries of terms.

Suggested Reading

Holbrook MC. *Children with Visual Impairments: A Parent's Guide.* Bethesda, MD: Woodbine House; 2006
This book is authored by an expert team of parents and professionals. It offers jargon-free advice to parents of young children with visual impairments from birth to age 7.

Lueck AH, Chen D, Kekelis L. *Developmental Guidelines for Infants with Visual Impairment: A Manual for Early Intervention.* Louisville, KY: American Printing House for the Blind; 1997
This manual was written for professionals working with children with visual impairments from birth through 2 years of age. The manual includes narrative chapters and developmental charts in the domains of social-emotional, communication, cognitive, and motor development.

Pogrund RL, Fazzi DL. *Early Focus: Working with Young Children who are Blind or Visually Impaired and Their Families.* 2nd ed. New York, NY: American Foundation for the Blind Press; 2002
This book addresses young children with visual impairments, including those with additional disabilities. It addresses challenges and intervention recommendations in all developmental domains including early literacy, daily living skills, and orientation and mobility.

Adapted with permission from: Teplin SW, Greeley J, Anthony TL. Blindness and visual impairment. In: Carey WB, Crocker AC, Coleman WL, Elias ER, Feldman HM, eds. *Developmental-Behavioral Pediatrics.* 4th ed. Philadelphia, PA: Elsevier; 2009:698–716.

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Chapter 23

Social and Community Services

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Introduction

Developmental disabilities and many behavioral disorders are life-long chronic conditions with no medical cures. Even acute behavioral disorders that seem to resolve are often rooted in the child's biophysiology, from which other behavior problems arise over a lifetime. While modern medicine provides pharmacologic treatments to decrease symptoms of these conditions, patients and their families continue to present to the primary pediatric health care professional with problems that are not amenable to medication alone. While not always easy to find, there are programs, including many governmental and nonprofit ones, available to support children with chronic developmental and behavioral conditions, and their families. The American Academy of Pediatrics (AAP) emphasizes that pediatricians should provide a medical home for these patients, linking them and their families with appropriate services and resources, even nonmedical ones, in a coordinated effort to achieve optimal outcome.[1] Recent studies indicate that families of children with these conditions want their pediatricians to help coordinate their children's care in the manner advised by the AAP, including putting them in touch with parents of children with similar conditions.[2] For these reasons, primary pediatric health care professionals need to be familiar with all social and community services available to children with developmental disabilities and behavioral disorders, and their families.

Care Coordination and Case Management

From the time of the initial diagnosis of a developmental disability or behavioral disorder, primary pediatric health care professionals make multiple recommendations for their patients that involve accessing services provided by a third party not under the clinician's or parent's control. For example, the clinician often writes orders or recommendations for therapies, school services, and evaluations in other clinics, some of which may require the patient's parent to be reliant on the third party with regard to schedule and payment. The need for additional third parties to address

new situations continues for life given the chronic nature of many developmental and behavioral disorders. This can cause patients, their caretakers, and their physicians ongoing stress due to lack of control. Both primary care physicians and parents of individuals with developmental disabilities and/or behavioral disorders report frustration over wait times for appointments with these third parties, confusion over the exact services needed from each of the third parties, and inconsistency regarding insurance/Medicaid coverage for the services.[3,4]

Clinicians' designation of a care coordinator and provision of care coordination services within their practices help caregivers navigate the health care system and access needed services for all of their patients, none the least being those with developmental disabilities or behavioral disorders. Building such care coordination services within a pediatric practice is one of the emphasized cornerstones of the AAP family-centered medical home model, so much so that the AAP provides the tools for such coordination through a Web site (http://www.medicalhomeinfo.org/tools/coordinating%20care.html).[5] This Web site even gives information regarding Title V Maternal and Child Health Bureau/children with special health care needs programs and other resources and services available in individual states if the user clicks "state pages" at the top of the homepage, plus information regarding evaluating managed care plans for children with special needs/Medicaid coverage if the user clicks "insurance/managed care" at the side of the homepage.

Various agencies, insurance plans, and even some states' Medicaid programs offer case managers who can serve as care coordinators for eligible patients. This care coordination is limited to patients meeting inclusion criteria for having a case manager. In the cases of patients eligible for such a service through a managed care organization, the case manager may be limited to seeking only the services aimed at disease management with no coordination for educational or social support or secondary behavioral health needs. In light of these possible limitations with use of outside sources, care coordination within the pediatric practice is the preferred practice since this would be available to all patients, addressing all condition-related needs and not simply those considered a medical treatment.[6(p5)] At times, however, use of an outside case manager might be needed. Helpful Web sites for review of case management services by states are the AAP site mentioned previously and http://www.hrsa.gov/reimbursement/, which will allow the user to put "case management by state" in the search bar to yield the information.

Effective case coordinators from inside a practice (or case managers from outside sources) do the following: (1) help caregivers understand the health needs of their child and maintain their child's health history; (2) provide information regarding their child's condition; (3) identify the medical, social, educational, and mental health services their child may need; (4) assist in applying for services for which their child is eligible; (5) develop a written comprehensive plan of care; (6) arrange for services to be provided and monitor the provision of services; (7) follow up on referrals; (8) arrange for transportation; and (9) coordinate among service

providers.[7] The staff member chosen to be the case coordinator can vary, although there are indications that the most effective choice with regard to cost and family satisfaction would be using a nurse.[8]

Addressing specific nonmedical needs of individuals with developmental disabilities or behavioral disorders or those of their families has various benefits. One study showed that case management–based behavioral intervention can decrease returns to the psychiatric emergency department by individuals with anxiety, thus reducing the overall cost of care.[9] Another study showed that having an individual provide case management and counseling for grandparents raising grandchildren with developmental disabilities increases the grandparents' sense of empowerment and caregiving mastery, while decreasing their depressive symptoms.[10] Case managers also seem to be the key to successful vocational programs for children with developmental disabilities transitioning to adulthood, and they offer hope for reducing the maltreatment of children with disabilities, estimated to be around 3 to 4 times higher than the maltreatment of children without disabilities.[11,12] Care coordination seems to improve continuity of care and decrease delayed or missed services.[13–15] Such coordination also seems to reduce fragmented or duplicative care, length of hospital stays, charges during hospital stays, and caretaker stress.[6]

Children's Mental Health Services

The 2001 National Survey of Children with Special Health Care Needs showed that of the approximately 10,200 children in the survey with a chronic emotional, behavioral, or developmental problem, 67% of their parents reported that their child needed mental health services, with 18% of mental health service needs not met.[16] These data are consistent with findings over the past 30 years.[17] Even among young children ages 0 to 3 years with developmental disabilities, a significant number had behavior problems consistent with a psychiatric disorder, including tantrums, aggression, oppositional behaviors, hyperactivity, and self-injury.[18] Caring for a child with mental health care needs affects financial well-being more than caring for a child with other special health care needs.[19] Undoubtedly, clinicians and their office staff who are aware of and can access children's mental health services while counseling to alleviate some of the stigma of referral to a mental health provider can offer hope to the numbers of families projected to have such needs in their practices. Primary pediatric health care professionals can search network affiliates for state by state mental health services at the following Web sites: www.nami.org and www.nmha.org.

Supplemental Medical Insurance and Other Financial Assistance Programs

Raising a child with a developmental disability and/or behavioral disorder costs 4 times more on average than does raising a child without one of these conditions.[20] The Medical Expenditure Panel Surveys of 1999 and 2000 targeting families of youth

with disabilities younger than 18 years revealed that these families paid approximately 11% of health care bills out of pocket, with the financial burden worse on families having the most severely affected children, the lowest incomes, and/or the least health insurance.[20] The 2001 National Survey of Children with Special Health Care Needs reported that 21% of families of the youth with chronic physical, developmental, behavioral, or emotional conditions on the survey list reported that their child's health care caused financial problems, and 30% of these surveyed families had to cut back or quit work because of their child's condition.[21] In addition, 17.7% of the surveyed families reported their children experienced unmet health care needs.[21] Despite the availability of Medicaid and the State Children's Health Insurance Program (SCHIP) to low-income parents with uninsured children with developmental disabilities and/or behavioral disorders, many parents who qualify for these programs report not using them due to the belief that they are not eligible for the programs or because they feel that the application process is too difficult.[22] Given these data, it is critical for primary pediatric health care professionals or their office staff to know how to assist families in accessing and applying for supplemental medical insurance and other financial assistance programs (eg, supplemental security income [SSI]) available to children with developmental disabilities and their families. Primary pediatric health care professionals can also assist families by providing a clear, concise summary of medical records that document the child's disability to the SSI physician. A resource for clinicians to provide information about financial assistance programs for their patients with developmental disabilities, including the Medicaid waiver plan, SSI, SCHIP, and Temporary Assistance for Needy Families (TANF), can be found at the following Web site, listed under Financial Assistance and US Department of Health and Human Services (HHS) Agencies: www.hhs.gov/children/index.html#income.

Educational Advocacy Services

Since primary pediatric health care professionals are often the first individuals to express a concern regarding a child's development or behavior or the first with whom parents share a concern, they are in a unique position to be able to access appropriate early childhood and special education services for their patients. Children with developmental disabilities and behavioral disorders may need special education and related services to be successful in school. Strock-Lynskey and Keller[23] detail the history of special education laws beginning with Section 504 of the Rehabilitation Act of 1973, the impetus for subsequent laws that provide civil rights protections to individuals with special needs, require rehabilitation services in schools, and prohibit discrimination in institutions receiving federal funds. The law governing the provision of special education services was passed in 1975 with the Education of All Handicapped Children Act (EHA, PL 94-142) and established the right to a free and appropriate education for all children with disabilities from 3 to 21 years of age in the *least restrictive environment,* regardless of the functioning level of the child's abilities. In 1986 an amendment to the EHA extended the provision of special

educational services to include early childhood intervention for children from birth to 3 years of age. In 1990 PL 94-142 was reauthorized and renamed the Individuals with Disabilities Education Act (IDEA, PL 101-476). In 1997 amendments were made to IDEA that "expanded the role of parents in the educational system and promoted a family-centered approach to service."[23] The most recent reauthorization of IDEA was in 2004.[24] Over the years, these laws have been expanded to include more children with special health care needs living in their local communities, including children in residential settings, in hospitals, or homebound. Partnerships, family choice, and the provision of information needed for parents to become advocates are supported by these laws. In addition, advocates for children with disabilities and/or behavioral disorders can also depend on the Americans with Disabilities Act of 1990, which "prohibits the denial of education services, programs or activities to students with disabilities and prohibits discrimination against all such students," and Section 504 of the Rehabilitation Act (details at http://www.ed.gov/about/offices/list/ocr/docs/edlite-FAPE504.html).[25] Finally, the No Child Left Behind law, passed in 2002, made among other things "academic achievement of students with disabilities a national priority."[26]

Infants and young children with a suspected developmental delay or behavior disorder, as well as children with a medically diagnosed physical or mental condition having a high probability of resulting in delays, are eligible for early childhood intervention referral.[27] Disabilities that qualify school-aged students for special education services under IDEA are as follows: orthopedic impairment, other health impaired (OHI), auditory impairment, visual impairment, deaf-blindness, mental retardation, emotional disturbance, learning disability, speech-language impaired, autism, traumatic brain injury, multiple disabilities, and non-categorical.[28]

Families of children with developmental disabilities or behavioral disorders have become increasingly knowledgeable of the laws governing their child's educational rights and have developed advocacy skills to ensure provision of services. Educational advocacy has become a recognized need and service of many disability groups. Across the country, there are seminars, trainings, and conferences for parents and professionals devoted to ensuring the educational rights of children with disabilities.

Primary pediatric health care professionals can assist families whose children have a developmental disability and/or a behavior disorder with a referral for early childhood intervention, and they can support a parent's request for a school district special education and related services (such as occupational therapy, physical therapy, and speech/language therapy) evaluation. Pediatricians and other physicians are the only individuals who can identify children whose health conditions affect their strength, vitality, or alertness and allow them to access special education services and modifications through the OHI eligibility, which requires a licensed physician's signature. The OHI eligibility covers attention-deficit/hyperactivity disorder (ADHD), developmental coordination disorders, and other medical conditions that negatively

impact the child's education. While not within the realm of special education, Section 504 of the Rehabilitation Act also allows for special accommodations, such as preferential seating, which the clinician can advise parents to seek as needed for their children.[24]

A primary pediatric health care professional does a great service for the child and parents by listening to parent concerns in a time-limited clinic visit and guiding them to the appropriate early intervention or school district evaluations, even when the clinician does not observe the stated behavior or delay. While primary pediatric health care professionals do not have a direct role in remediation, they are able to initiate interventions aimed at helping the child meet his or her full potential. They can monitor the child's progress, help parents identify their child's strengths, and advocate for needed educational and therapeutic programs from year to year. In some cases they can also help families accept realistic goals for a child. Community agencies, such as The Arc (www.thearc.org), can also provide educational advocates to accompany parents to their children's Individualized Education Program plan meetings at school to assist them in accessing the most appropriate special educational and therapeutic services for their children. A "one-stop shop" for resources related to IDEA and state contacts can be accessed at http://idea.ed.gov.

Support Groups

Parent-to-parent support is a resource of great value that clinicians can offer to parents as they adjust and cope with their child's new diagnosis of a developmental disability or behavioral disorder. These support groups help decrease the feelings of stress and social isolation that often follow the initial diagnosis.[29] When a child is diagnosed with a developmental disability or a behavioral disorder, the impact can be overwhelming and affect family relationships. The time immediately following the diagnosis, often described as "a critical period" for families, is a particularly apt time for referral to parent support resources.[30] During this critical period, parents can be confused about where to obtain appropriate early educational and therapeutic services, which services are most effective and most evidence-based, and how to pay for them. It can greatly help for a parent to speak to another parent whose child has the same or similar diagnosis who has been through the same process of adjustment and with whom they can share their feelings and gain answers in how to navigate the systems of care.

As time passes, parents of children with developmental disabilities and/or behavioral disorders run the gamut of emotions from joy to despair as they navigate the systems of care—therapeutic, educational, and psychological—to help their child and family. Clinicians may feel unprepared in how to advise and advocate for these primarily nonmedical interventions. Parent support groups and parent-to-parent networks offer information, advice, and often an experienced hand to help with finding educational, social service, and other needed community resources. As children age, or as

families move to other cities, these support groups provide needed information regarding local schools, therapists, and extracurricular activities.

There are many support group options, both for specific disabilities and behavioral disorders as well as for disabilities of unknown etiology. Formalized meetings with special topics, gatherings at houses of worship, and many times an informal network of parents give opportunity to those who want to band together to energize their spirits, decrease their sense of isolation, and enhance their abilities to meet their children's needs. These groups offer daytime and nighttime meetings, so as to include both mothers and fathers, as they often differ in adaptation to their child's diagnosis.[31] They also offer sibling groups for brothers and sisters of children with developmental and/or behavioral disorders, where they can develop friendships and gain support from those in similar circumstances. Grandparent support groups allow for information sharing as well. In such a mobile society, support groups help those whose families live elsewhere. The Web has become an excellent aid for allowing parents to bridge miles and even countries to network with each other, especially in cases involving children with rare disorders. Family Voices is a national advocacy and support group of families of children with special health care needs and has offices in each state and territory of the United States (www.familyvoices.org).

Research demonstrates the benefits of parent-to-parent support, showing that parents are "uniquely qualified to help each other."[32] Clinicians can help by keeping a close eye on parental well-being, supporting and encouraging parents' efforts to seek out other parents of children with developmental disabilities and/or behavioral disorders. They and their staff should supply support group information at the onset of diagnosis and thereafter as needed. Primary pediatric health care professionals can access parent group listings by state at www.yellowpagesforkids.com.

Functional Behavioral Analysis/In-Home Behavior Management Counseling Services

Mothers of children with disabilities exhibit high rates of depressive symptoms associated with child behavior problems, maternal stress, coping style, and support.[33] Interventions that provide parents behavioral training together while addressing their well-being reduce parent distress.[34] Parents can better implement child behavioral management with specific behavioral counseling.

A practical and evidence-based method for helping parents manage their child's behavior problems is to align them with individuals who can provide a home-based functional behavioral assessment (FBA), and then provide in-home behavior management counseling services to the parents. Similarly, schools also use FBAs and behavioral plans to help teachers and other staff optimally work with a child with behavioral problems in their setting. The behavior therapist performing the FBA observes and precisely describes the specific maladaptive behaviors, including when they happen and when they stop. The behavior analyst uses the information

from the observation to understand the factors influencing the behaviors and the triggers and consequences of the behaviors to determine the driving forces behind each behavior. Then the behavior analyst provides the parents on-the-job training on ways to bring about appropriate changes in the behavior of their child. Resources for FBA can be found at http://www.nichcy.org/Pages/behavassess.aspx.

Respite Care

Raising a child with a developmental disability and/or behavioral disorder presents many challenges, affecting all family relationships. Respite is short-term temporary care that provides parents and caregivers with important time off from the constancy of their child care responsibilities. This family support service is often critical for the long-term stability of the family and child at home. Caregivers take pride in their ability to provide good care for their child; however, there is often a lack of time and energy for personal, marital, and family activities, which results in social isolation. Many hours are spent on therapy appointments, obtaining medical supplies, and/or modifying the home to ensure safety. The physical, emotional, and financial demands may be overwhelming, adversely affecting the health and well-being of all family members.[35] Since the family is the child's best resource, families need respite services to avoid the burnout that often accompanies the provision of 24-hour care. Respite helps to maintain family stability and to enable children with developmental disabilities and behavioral disorders to remain in a nurturing home.

In-home and out-of-home respite are options that are based on the parent's needs, funding, and availability. A few hours, a few days, or weeks at a time helps to replenish a parent's or caregiver's energy. It has been reported that respite is the most requested family support service for children with developmental disabilities and behavioral disorders.[36] This report is not surprising, since respite allows parents and caregivers a chance to spend time with a spouse, friends, or their nondisabled children, who may often be overshadowed by their sibling's needs. Respite care helps to increase positive family interactions and enables families and caregivers to cope with chronic and potentially stressful situations. By decreasing parent and caregiver stress, respite reduces the possibility of maltreatment, known to be increased among children with developmental and behavioral disorders compared to children without such disorders in the general population. Sullivan and Knutson[37] estimated that the rate of maltreatment for children with disabilities was 31%. A recent AAP clinical report noted that children with behavioral disorders are at highest risk of maltreatment and that neglect is the most common type of maltreatment across all disability types.[38]

Respite also provides opportunities for children with developmental disabilities and behavioral disorders to meet new people and friends and to enjoy new experiences outside the home. Their siblings may be included as well. These activities can increase the child's self-esteem while improving parent and caregiver relationships.

Clinicians can help by asking parents about their needs for supportive care and by monitoring for burnout. Such inquiries acknowledge the extra care that these parents provide, a recognition parents complain is often missing.[39] They can encourage and support families in this endeavor by filling out necessary paperwork for respite care services and by having information regarding community respite services available in their offices. Because they know firsthand the impact of care, they can also be advocates in the community for more available and diverse respite options to meet the needs of these parents and caregivers. A state-by-state listing of respite care services can be found at http://chtop.org/ARCH/National-Respite-Locator.html.

Personal Care Attendant Services

Caring for a child with a developmental disability or behavioral disorder who also has self-care limitations can require additional assistance at home. Clinicians should be aware that personal care attendant services are a health care benefit provided under the Medicaid state plan of each state as an optional, not required, service that states may select so that children with developmental disabilities and behavioral disorders can receive services to assist in day-to-day living in their community. Children with severe physical and medical disabilities can receive assistance to help with mobility and self-care skills, allowing these children to enjoy and participate in their home and community life. Children with severe behavioral disorders can have supervision to help perform daily tasks, communicate, and learn new skills. Personal care attendants can assist with activities of daily living, such as eating, bathing, toileting, and dressing, as well as with positioning, lifting, transferring, exercising, and adaptive skill building.[40]

Personal care attendant services allow children with developmental and behavioral disorders to live at home in a nurturing environment with parents, siblings, or other extended family members or caregivers. Families are often able to choose and supervise service providers for their children through Consumer Directed Services, a potentially more effective, less costly approach to care.[41] Regardless of the severity of their disability or behavioral difficulties, with personal care services children are able to function as a member of a family, attend school, and participate more fully in social and recreational activities in their community, with an improved quality of life for the child, family, and community.

Clinicians can assist families in accessing personal care attendant services by completing required forms that document the severity of the disability and define the services and supports that are needed. This is a time-consuming task, not often reimbursable, but well appreciated by families and service providers. Clinicians can also help by advocating for these services with the appropriate state officials and groups.

Information regarding personal care attendant services for children with disabilities can be found by going to http://www.hhs.gov/, which will allow the user to put "personal care attendant services for children with disabilities" in the search bar to yield the information.

Counseling Regarding Long-term Legal and Financial Planning Issues

Just as parents of children without disabilities need advice about designing a will that addresses care of their children younger than 18 years in case of parental demise, the parents of children with developmental disabilities and/or behavioral disorders need to do the same, although often in a more detailed fashion to specifically address disability/behavior, financial, and guardianship issues. In addition, parents of children with developmental disabilities and/or behavioral disorders need advice and legal help to plan for their children's adulthood and to write specific instructions for their adult children's care, if they as parents are no longer able to give care or to be available to make decisions regarding this care.

Even parents of children with milder disabilities like dyslexia or ADHD need to consider special adult provisions for their children if the parents anticipate that the income of their adult children with milder disabilities may be less than that of their unaffected adult children. Parents of children with certain behavioral disorders also need to address ways to protect against these children unwisely spending inherited money as adults. Parents whose children have disabilities and/or disorders limiting their independence, whose children's conditions prevent adult decision-making, and whose children's conditions require long-term medical treatment need to deal with the financial and legal aspects of these situations well before their children reach 18 years of age.

Finding appropriate counseling regarding long-term legal and financial planning issues is of paramount importance to families of children with developmental disabilities and/or behavioral disorders. Laws limiting the amount of assets or income an individual can have and still receive such aids as SSI and Medicaid are confusing and often changing. Therefore, parents need professionals who listen to them, and who have experience in estate planning, the use of special needs trusts, guardianship, power of attorney, tax laws, regulations governing Medicaid/social security disability income/SSI/other governmental benefits available, and addressing the issues of families like their family. Fortunately, well-respected national, state, and local organizations serving individuals with specific disabilities/disorders offer help to families with regard to knowing what they need to do and where to find appropriate professionals to help them.

Primary pediatric health care professionals and their office staff are in a unique position to assist parents of their patients with developmental disabilities and/or behavioral disorders with regard to long-term planning for their children and finding

appropriate financial/legal counseling for this planning to take place. Because the clinician is usually one of the first professionals to talk with the parents regarding their children's diagnoses and usually follows the children on a longitudinal basis, the clinician and his or her staff can advise parents to begin planning for their children's future as soon as possible, as long-term care lists like those for Medicaid waivers in some states take up to 7 to 10 years of being on the list before services are available. Primary pediatric health care professionals should advise all parents, including those with limited financial resources, to begin estate planning and establishing special needs trusts.[42] While this information is difficult for parents to hear, it should be given in a sensitive manner soon after a diagnosis is given to avoid causing a loss of eligibility for certain government programs while still seeing to the child's supplemental needs. At each clinic visit, health care providers can update anticipatory guidance for what the children will need as adults and remind parents that at 22 years of age even individuals with severe disabilities will lose certain previously guaranteed care, such as that through public school services and specific government programs. They can also guide parents toward reputable Web sites/organizations offering appropriate financial and legal counseling to address their children's needs. While the benefit of helping families with appropriate long-term planning for their children with disabilities and/or behavior disorders is intuitive, there are some studies that document this benefit.[43,44] Resources to assist families in obtaining information about long-term legal and financial planning can be found at http://specialchildren.about.com/od/longtermplanning/LongTerm Planning_ Finances_and_the_Future.htm.

Extracurricular Activities and Summer Camps

Extracurricular activities, such as clubs, sports, scouting, arts, crafts, theater, and music, are an important part of the educational experience of all students, including children with developmental disabilities and behavioral disorders. Children with developmental disabilities and behavioral disorders may experience low self-esteem related to their inability to keep up academically and socially with peers. Their need for special education and related services often sets them apart from the general student population, and often siblings reinforce their feelings of being different and less capable than other children.[45]

Participation in athletic and creative activities provides children with developmental disabilities and behavioral disorders with opportunities to develop self-confidence and experience success in fun and non-stressful ways. Participation in these activities may also help the child and his or her family identify a child's special talent that can be a huge boost to that child's self-esteem.

Federal laws governing special education services recognize the importance of extracurricular activities. The IDEA amendments of 1997 include a "statement of program modifications or supports for school personnel that will be provided for

students with disabilities to participate in extracurricular and other nonacademic activities. School districts shall take steps to provide nonacademic and extracurricular services in such manner as necessary to afford children with disabilities an equal opportunity for participation in those services and activities."[46]

Nonschool-sponsored extracurricular activities are available through Special Olympics, special needs scouting, YMCA programs, disability groups (eg, The Arc or United Cerebral Palsy [UCP]), as well as city parks and recreation departments. Specialty programs, such as hippotherapy, offer recreational activities specifically for children with disabilities.

Summer camps are an additional extracurricular activity. Not only do they provide children with developmental disabilities and behavioral disorders with the opportunity to experience success outside the classroom, they also allow these children to be like their siblings and same-age peers who go to camp. Both day and overnight camps support the development of independence and self-esteem. Specialty camps help children to understand and cope with a specific disability or chronic illness, while also enjoying horseback riding, swimming, and other fun activities. Children with even the most severe disabilities can enjoy the opportunity to be away from home, while their parents may enjoy the temporary respite from child care duties.[47]

Primary pediatric health care professionals can help parents with selecting extracurricular activities, summer programs, and camps appropriate for their child's developmental level. As clinicians, their letters of support and medical input regarding level of participation and care needs will aid activity and camp directors in planning and programming for children with developmental disabilities and behavioral disorders. In addition, they can address their medical treatment of these children toward maximizing their ability to participate in these activities.[48] The clinician's support for extracurricular activities and summer camps emphasizes their value for children with developmental disabilities and behavioral disorders. Information about extracurricular activities and summer camps for children with special health care needs can be found at the following Web sites:

- www.familyvillage.wisc.edu/leisure/
- www.specialolympics.org
- www.bestbuddies.org
- www.scouting.org/scoutsource/boyscouts/thebuildingblocksofscouting/specneeds.aspx
- www.acacamps.org
- www.kidscamps.com

Assistive Technology

Assistive technology resources now provide children with developmental disabilities and behavioral disorders with an extensive array of tools to communicate their needs and to demonstrate their ability to learn. Access to these resources in the educational

setting is mandated by IDEA. Assistive technology can help children with disabilities to more fully participate with typical peers in both school and extracurricular settings. These resources can also help children better connect and communicate with family, friends, and the world outside. Children with even the most severe conditions may benefit from assistive technology. Clinicians should request an assistive technology evaluation for even their youngest patients with developmental disabilities.[49] Such evaluations can be requested through early childhood education programs for children ages 0 to 3 years, special education programs and services for children ages 3 to 21 years, and agencies such as UCP. Library-related resources to connect children to technology tools and toys can be accessed at www.ataccess.org/resources.

Conclusion

Laws and resources will change over time. However, when primary pediatric health care professionals partner with parents to identify and access all community and social services available to children with developmental disabilities and/or behavioral disorders and their families, they can develop the most comprehensive plan for ensuring that these children reach their full potential.

Summary of Web Sites for Accessing Social and Community Services for Children With Developmental and/or Behavioral Disorders and Their Families

Due to each state and local government having their own services for individuals with developmental disabilities and behavioral disorders, clinicians and caregivers should also use Internet search engines to look for specific services in their state/local area. In addition, clinicians and caregivers should use Internet search engines to look for specific diagnosis-related national/state/local organizations, as Web sites for such organizations often aid in finding needed services.

Case Management
http://www.hrsa.gov/reimbursement/
Allows the user to put "case management by state" in the search bar to yield information.

Children's Mental Health Services
www.nami.org
www.nmha.org
Search network affiliates for state-by-state services at these sites.

Supplemental Medical Insurance and Other Financial Assistance Programs
www.hhs.gov/children/index.html#income
Various Web sites regarding financial assistance listed under Financial Assistance and HHS Agencies, including Medicaid, Medicaid waiver plan, SCHIP, TANF, SSI, etc.

Educational Advocacy Services
http://idea.ed.gov
"One-stop shop" for resources related to IDEA and state contacts and information.

www.thearc.org
The Arc is the world's largest community-based organization for people with intellectual and developmental disabilities. The Arc is devoted to promoting and improving supports and services for all people with intellectual and developmental disabilities and their families.

Support Groups
www.yellowpagesforkids.com
Parent group listings state by state.

Functional Behavioral Analysis/In-Home Behavior Management Counseling Services
http://www.nichcy.org/Pages/behavassess.aspx
Provides multiple Web sites.

Respite
http://chtop.org/ARCH/National-Respite-Locator.html
Respite services, state by state, in local area to match specific needs.

Personal Attendant Care
http://www.hhs.gov/
Allows the user to put "personal care attendant services for children with disabilities" in the search bar to yield information.

Counseling Regarding Long-term Legal and Financial Planning Issues
http://specialchildren.about.com/od/longtermplanning/LongTerm_Planning_Finances_and_the_Future.htm
Provides multiple Web sites.

Extracurricular Activities/Summer Camps
www.familyvillage.wisc.edu/leisure/
www.specialolympics.org
www.bestbuddies.org
www.scouting.org/scoutsource/boyscouts/thebuildingblocksofscouting/specneeds.aspx
www.acacamps.org
www.kidscamps.com
Includes special needs leisure/camps.

Assistive Technology
www.ataccess.org/resources
Resources, library-related resources connecting children to technology tools and toys.

General

www.TheArcLink.org
Resources and services for individuals with disabilities and their families on a state-by-state basis.

www.thearc.org
Voluntary organization committed to the welfare of all children and adults with mental retardation and their families (state chapters and resources).

www.autismsocietyofamerica.org
Official site of Autism Society of America with state-by-state information.

http://clerccenter.gallaudet.edu/infotogo/184.html
Access Directory of National Organizations of/for Deaf and Hard of Hearing People.

www.developmentaldisabilitiescouncil.org
State chapters of the Developmental Disabilities Council.

www.DisABILITIESBOOKS.com
Books of interest to parents of children with disabilities, family members, and professionals who serve children and families.

www.disabilityresources.org
A guide to Disability Resources' publications and services, with access to state listings.

www.easterseals.org
Services to ensure that people living with autism and other disabilities have equal opportunities to live, learn, work, and play (state chapters).

www.epilepsyfoundation.org/aboutus/AfilliateLookup.cfm
Find services for individuals with epilepsy.

www.familyconnect.org
Check for state-to-state support groups/toys/services for the visually impaired.

www.familyvillage.wisc.edu/index.html
A global community of disability-related resources.

www.familyvoices.org
Family-to-family health and education information (state resources).

http://www.medicalhomeinfo.org/publications/eNews/announce.html
This is an e-newsletter dedicated to providing medical home information and resources for children with special needs.

http://www.medicalhomeinfo.org/tools/coordinating%20care.html
National center of medical home initiatives for children with special health care needs (tools for coordinating care).

http://www.medicalhomeinfo.org/states/index.html
National center of medical home initiatives for children with special health care needs (state resources).

www.nichcy.org
Look under state resources/topics A–Z.

http://www.pacer.org
Parents helping parents, training and information center, state resources.

http://www.spinabifidaassociation.org/site/c.liKWL7PLLrF/b.4861757/k.BEF1/Resource_Directory.htm
Ideas for helping individuals with spina bifida that gives ideas that can extend to helping individuals with other conditions.

www.ucp.org
"One-stop resource guide" for every US state and territory.

http://www.wrightslaw.com
Information about special education law, education law, and advocacy.

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Chapter 24

Transition to Adult Medical Care

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Transitions are part of normal, healthy development and occur throughout life. As survival for youth with developmental disabilities now is approaching that of the general population, socioeconomic factors are being recognized as important determinants of health, and social inclusion and quality of life are the focus of outcome, not just survival.[1] Youth with developmental disabilities experience the same transitions in life as those without disabilities. Primary pediatric health care professionals have an essential role in this transition process because of their longitudinal contact with many of these young people and the close relationships that they develop with children with disabilities and their families.

Assessing long-term outcome in youth with disabilities no longer consists primarily of collecting mortality data; it is now important to assess the youth's level of participation in adult society. The stage is now set for dramatic improvement in the services for all youth to prepare them for their working future. In the business world that is geared for change, business people would say that the ingredients needed to create a significant change in an area are (1) available knowledge, (2) gap between knowledge and practice, (3) examples where the gap has been closed, and (4) societal pressure to make the needed improvement. Transition for youth with developmental disabilities is ready for such a transformation. While transition outcomes for youth with chronic health issues and disabilities have shown some improvement and promising practices are beginning to be implemented, many young people continue to face barriers that prevent their full participation in school, community, and work, thus impairing their maximal independence and quality of life.

Transition to adulthood can be a most challenging time, as it is a time of biological, emotional, and social change for the young person. Health and wellness are often not the primary concerns of youth in their transition years, but good health is central to their being successful in all the other areas of transition: school to work, home to community, dependence through independence to interdependence, and pediatric to adult health care.

This chapter focuses on the key issues of health care transition that primary pediatric health care professionals need to know in order to plan, provide, and create services, policies, and support systems within a medical home for children and youth with developmental disabilities who need them most, who use them often, and who may struggle as they move from pediatric to adult health systems and services. This chapter also describes the essential components of transition in health care; the wishes of youth with disabilities and their families for their transition; and the actions needed to meet the critical first steps outlined in a consensus statement created by health care professionals, families, and youth with special health care needs (SHCN) to maximize the young person's lifelong functioning.[2] With each critical step, tools are outlined to assist the primary pediatric health care professional to create a successful transition process for those youth with disabilities in their practice.

In planning for the ultimate outcome of living successfully as an adult, it is important to acknowledge that children age normally but mature depending on their unique circumstances that may define, but do not necessarily limit, their possible outcomes. Youth with developmental disabilities have a variety of ways to interact with the adult health care system depending on their cognitive levels and supports. Some are interdependent and manage their own health and health care like most adults without disabilities, while others need support full time through legal avenues, and many have arrangements in between these 2 extremes. Timing and tempo of the transition process is key to a successful transition that allows the youth to assume adult roles. For some, it means transferring their care from a pediatric health care professional to an adult-trained health care professional, and for others, it means a reorientation of the clinical interaction within the same practice, such as that seen in family medicine. As health status impacts productivity and quality of life, children and youth with developmental disabilities need their childhood years to

- Learn and practice the skills to optimize their outcomes.
- Become more knowledgeable about staying well.
- Become able to access systems of care.
- Learn how to be heard and how to be more informed decision-makers.

All of the above prepare adolescents with disabilities to transition to adulthood systems and services. The adults in their lives, whether medical professionals, families, and/or caregivers, need to provide opportunities for practice. This is critical for youth with disabilities to gain skills and expertise that are essential for survival in navigating care and staying healthy to be able to be ready to live and work to their full capacity.

What Is Transition?

Transition is defined as "the purposeful, planned movement of youth from child-centered to adult-oriented systems."[2] Successful transition planning is the result of partnerships among the individual, his or her family, school personnel, the health care system, local community and adult service organization representatives, and

interested others. The goal is to maximize lifelong functioning, social participation, and human potential. Transition is the deliberate, coordinated provision of developmentally appropriate and culturally competent health assessments, counseling, and referrals. Many have chronicled the hurdles that families, youth, and health care professionals have to overcome in creating a successful transition (Table 24.1).

Components of successful transition include
- Self-determination
- Person-centered planning
- Preparation for adult health care
- Work/independence
- Inclusion in community life

Table 24.1. Hurdles to Successful Medical Transition[a]

Young Person	• Lack of familiarity with adult clinic and team • Reluctance to leave pediatric team especially if long disease duration • Immaturity—lack of social skills • Dependence on parents • Non-adherence • Not yet independent in managing their own health care • Prevention and management of comorbidities
Parents Family Caregivers	• Lack of familiarity with adult clinic and/or team • Dislike of individual approach of adult care • Overprotectiveness • Negative preconceptions of adult care • Awareness of lack of confidence of pediatric team in adult service
Pediatric Health Professional	• Reluctance to let go long-term patients • Lack of confidence in adult system • No pediatric-adult interface • Lack of knowledge of transition issues/resources
Adult Health Professional	• Lack of confidence in pediatric illnesses • Lack of training in adolescent health care • Individual rather than family-centered approach • Negative attitudes of paternalistic style of pediatric care • Lack of knowledge of transition issues/resources
Delivery System	• Lack of planning • Lack of preparation of young person, their family and the adult team • Suitable clinic space • Difficulties in administrative transfer of patient notes and x-rays • Consensus regarding management guidelines between pediatric and adult specialty services • Lack of health care insurance availability
Time and Money	• Lack of available time for the full spectrum of transitional care • Lack of funding

[a] Reprinted by permission. HRTW National Resource Center.

What Is Health Care Transition?

The optimal goal of health care transition is to provide health care that is family-centered, continuous, comprehensive, coordinated, compassionate, and culturally competent (ie, a medical home that is as developmentally appropriate as it is technically sophisticated).[3] Several general principles of successful transition are summarized below.

1. *Continuum:* Transition is a process, not an event. Planning should begin as early as possible on a flexible schedule that recognizes the young person's increasing autonomy and capacity for making choices. Transfer refers to the event when the youth with SHCN moves from pediatric-oriented to adult-oriented health care systems. Transition to adult services should occur prospectively, rather than during a crisis, and when the young person's medical problems are under good control.
2. *Start Early:* The transition process should begin in earnest by the age of 11 to 13 years[4] and include long-term sequential planning toward goals of independence and self-management.
3. *Coordination:* Coordination between health care, educational, vocational, and social service systems is essential. It is particularly important to recognize the complex interplay between health and social outcomes as young people age into employment and, in the United States, into an employment-based health insurance system.
4. *Changing Roles:* As the role of the young person changes in transition to adult systems, the families' and the health care professionals' role in the life and health care of the young person also should change. Primary pediatric health care professionals and the family should appreciate the young person's change in status as they move from adolescence to adulthood.
5. *Self-determination:* Self-determination skills should be fostered throughout the transition process. Best practices for transition services call for a young person–centered and asset-oriented approach that involves young people as decision-makers during the entire transition process.

Why Transition?

Children and youth with SHCN, like all individuals, deserve to be cared for in a developmentally appropriate environment. Having an SHCN can adversely affect the physical, mental, financial, and social well-being of children and their families. Functional limitation is especially associated with increased health care needs, and the demands on the health care system grow as the youth becomes more physically and cognitively limited.[5]

When Should Transition Occur?

There is much discussion as to when preparation for health care transition should begin and transfer should occur. Age has been a central way to discuss these transition milestones. In a large study by Geenen et al,[6] parents and health care professionals agreed that around the average age of 17 years, the youth should be transitioned to an adult health care professional. In contrast, health care professionals thought that they should begin teaching youth to manage their own health at approximately 9.5 years, and the parents reported an age of 12 years. Similarly, health care professionals thought they should begin seeing the youth independently for part of the visit at around age 12 years, but parents felt that this should occur around age 14 years. The literature today suggests the process of transition planning should begin at the latest between the ages of 11 to 13 years, as that is when the youth is most engaged in the process, before other issues of adolescence may become more interesting, such as their peer group and social activities.[4]

What Do Youth and Families Want for Their Transition?

Youth

In the past decade many surveys have been done that have asked youth with disabilities/SHCN what they believe are the best ways to build a successful adult life (employment, college, health, and independent living). The following is a representative list[7,8] of the health transition issues reported by youth with SHCN:

- Learning to stay healthy
- Getting a good doctor who treats adults
- Having family members that expect me to be a successful adult
- Figuring out what accommodations I need and how to ask for them
- What could happen if my condition gets worse
- What to do in an emergency
- How to get health insurance

Families

After the diagnosis, and as families begin "living" with their children with disabilities, many wonder what the future is going to look like. They want to know what they need to do to prepare their child for adulthood and how they can help serve as a navigator as they go through the maze of medical, educational, therapeutic, legal, insurance, social, and community systems.

In 2003 Geenen and colleagues[6] surveyed parents of youth with SHCN and following is a list of transition activities that parents most desired:

1. Take care of my child's health and my child's disability.
2. Coordinate my child's care with other health care professionals.
3. Help my child keep health insurance and find an adult health care professional for my child.

4. Teach my child to manage their own health to be successful at work.
5. Work with school to coordinate care.
6. Connect my child with other services in the community.
7. Screen my child for other mental health problems.

National Surveys of US Families on Transition Services: How Successful Are Current Efforts?

In 2005–2006 a national telephone survey of 40,804 families with youth with SHCN younger than 18 years of age was completed.[9] This survey reported that 48.8% of families with youth with SHCN aged 12 to 17 years felt that their youth received the services necessary to make appropriate transitions to adult health care, work, and independence but only 10% reported receiving discussion on how their youth could keep health insurance.

A recent study by Lotstein et al[10] confirmed the lack of transition services and reported that of the youth with disabilities who aged out of the Maternal and Child Health Bureau (MCHB) Title V programs (the only federal program that focuses solely on improving the health of all mothers and children), 24% had no usual source of care, 27% had forgone medical needs, 39% had delayed care, and 40% had insurance gaps since turning age 21. Thus fewer than half of families with youth with SHCN were receiving only a few transition services and had many gaps in their health care.

Transition Consensus Statement and 6 Critical Steps for a Successful Transition

To move the transition concept forward, policies have been devised by many groups. In 2001 an MCHB-invited group of experts drafted a Consensus Statement on Health Care Transition for Young Adults with Special Health Care Needs that outlined 6 critical first steps "that the medical profession needs to take to realize the vision of a family-centered, continuous, comprehensive, coordinated, compassionate, and culturally competent health care system that is as developmentally appropriate as it is technically sophisticated."[2] This was endorsed in 2002 by the American Academy of Pediatrics (AAP), American Academy of Family Physicians, and American College of Physicians-American Society of Internal Medicine.

The 6 critical steps of the consensus statement are outlined below, with suggested steps that pediatric health care professionals and pediatric practices can consider adopting to improve the quality of services addressing transition for youth with developmental disabilities in their practices.

Critical Step 1. *Medical Home Primary Care:* Ensure that all young people with SHCN have an identified health care professional who attends to the unique challenges of transition and assumes responsibility for current health care, care

coordination, and future health care planning. This responsibility is executed in partnership with other child and adult health care professionals, the young person, and his or her family.[2]

In ensuring that health care transition is included in the medical home concept, a lead person should be identified within the practice who is familiar with transition issues and processes. This person should guide the transition efforts of the practice and partner with families and youth in developing the needed processes and procedures. It is usually better for continuity of care to transition only one service at a time if multiple services are involved in the care of the youth. If the transition process is started between the ages of 11 and 13 years, the health care professional has the time to prepare for the young person's transition and allows the youth time to practice their skills in self-advocacy in the primary care setting. Pediatric health care professionals who are looking to transition their patients to adult health care professionals should develop a referral list of physicians willing to accept these referrals. Comfortable relationships with the adult health care professionals dramatically facilitate transition. A recent survey of internal medicine specialists[11] revealed the lack of training in childhood chronic illnesses, adolescent medicine, and adolescent behavior as most concerning to adult health care professionals.

In addition, adult health care professionals stated receiving a letter of referral, a medical record, and an ability to discuss the patient with the sending physician would be key and occurred only half of the time. In a survey of pediatricians in a medial home program, only 33% had a transition policy, 31% had created transportable medical records for their youth with SHCN, 19% discussed legal issues on medical decision-making before age 18, and 5% had developed individual transition plans.[12] A practical way to assist in this transition process is to suggest an observational visit to interview potential adult health care professionals. Talk with the youth and family about what they want to learn about the adult health care professional and also talk with the adult health care professional about how prepared the youth was to interact with the adult practice. Thus both pediatric and adult health care professionals can benefit from working together to create the continuum of care needed as the youth transitions from one medical home to the other.

Critical Step 2: *Core Knowledge and Skills:* Identify the core knowledge and skills required to provide developmentally appropriate health care transition services to children and youth with SHCN and make them part of training and certification requirements for primary care residents and physicians in practice.[2]

To understand areas that a practice should incorporate into their practice processes and procedures, see Appendix A for Healthy & Ready to Work National Resource Center (HRTW) Tool—Core Knowledge & Skills for Pediatric Practices checklist. This checklist is divided into 5 areas: policy, medical home, family/youth involvement, health care insurance, and screening.

Another key issue that requires a flexible approach by the practice is that some youth, depending on cognitive level, decision-making ability, and/or level of medication treatment that may impair informed decision-making, require a more formal support for partnering around their medical care. This may be the family or another trusted adult and, when needed, can be legalized through a power of attorney for medical decisions. It is critical to address this issue of a power of attorney before the youth reaches 18 years of age. Many of these issues are discussed on the National Disability Rights Network Web Site (http://napas.org/aboutus/default.htm). Long before the age of 18 years, pediatric health care professionals should discuss with those youth who can make their own medical decisions that they will have control of all such decisions at the age of 18 and give them opportunities to test out this new role.

Critical Step 3: *Portable Medical Summary:* Prepare and maintain an up-to-date medical summary that is portable and accessible. This information is critical for successful health care transition and provides the common knowledge base for collaboration among health care professionals.[2]

An essential component of transition is the communication of information from one set of health care professionals to another. The consensus statement suggests that the practice develop a 1- to 2-page medical summary that can be kept up to date. This can be placed in the electronic medical record, given to the youth and families on a flash drive, or live on the youth's/family's computer. It is always updated and available as a communication tool. Suggested components include

Youth

Medical

- Family health portrait: https://familyhistory.hhs.gov/
- Critical problems that are life-threatening
- List of inpatient history, treatment protocols
- List of surgeries
- Medications (prescribed, over-the-counter, and alternative medicines/herbs)
- Prevention actions (general: nutrition, physical activity guidelines, routine screenings, tests according to age; specific actions/screenings required due to the family health portrait, eg, heart disease, diabetes)
- Do not resuscitate status
- Power of attorney, health surrogate, guardianship
- List of medical care professionals, including dentists and appropriate diagnostic codes if unusual

Supports

- Levels of supports needed for care (family, grandparents, home health, others)
- Cultural practices to consider when developing a treatment plan
- Established emergency plan for vacations, hospitalization, or unexpected disasters
- Place ICE (in case of emergency) in the youth's phones (circle of support)

Youth needs

- Method of learning for the youth: verbal, verbal reinforced with written materials, verbal/written with demonstration using a manipulative, etc
- Communication supports: picture cards, sign language, gestures, verbal cues, or assistive technology devices that provide verbal communication for the youth

For an example of a blank template and a filled in medical record from HRTW, see Appendix B and C.

Families

Many families have learned how to organize medical records, tests, and other documents into a 3-ring binder. For children and youth who have multiple issues, these binders, while an impressive collection of historical medical treatments, are not well received by incoming health care professionals, who are often time-compressed and need to focus on today's preventive screening and maintaining and improving wellness. Creating a concise 1-page medical summary enables the family who is acting as an agent of the minor child to quickly communicate with medical professionals and home health services. Such summaries should include health care professional contact information, medications, and diagnoses (including *International Classification of Diseases, Ninth Revision* codes). Including patient data such as height, date of birth, and cultural and communication strategies is also helpful. It is suggested to give copies of the medical summary to key members of the family, support circle, and youth to carry with them and have immediately on hand.

Critical Step 4: *Health Care Transition Plan:* Create a written health care transition plan by age 14 together with the young person and family. At a minimum, this plan should include what services need to be provided, who will provide them, and how they will be financed. This plan should be reviewed and updated annually and whenever there is a transfer of care.[2]

Steps for the Preparation for Transition to Adult Health Care

1. **Develop a practice transition policy; put it in writing and make it visible!** One of the most important first steps to start the transition process for a practice is to develop a transition policy that is agreed on by all of the key players to ensure consensus and mutual understanding of the processes involved and to provide a structure for evaluation. The policy should be posted for the youth and families to see,[13] yet many practices have not developed such a written policy that is endorsed by the entire practice.[14] A key component of such a policy is a preparation for transition of health care timeline. As mentioned earlier, starting this process early (at the latest around ages 11–13 years) allows the youth to practice taking on responsibility as soon as the family and health care professional feel the youth is capable of beginning to take responsibility for their health and health care. Starting early allows time for the youth to practice the skills required for their changing role, time for parents to let their youth

have increased independence and learn who they need to ask for help (develop adult interdependent relationships), and time for exploration of options and obstacles with regard to continuing health care insurance coverage and applying/maintaining/leaving government benefits (Supplemental Security Income, Vocational Rehabilitation).

2. **Discuss differences in the cultures of the pediatric and adult health care systems.**
   Preparation for transition prior to the transfer of care is central to success. Youth with developmental disabilities and their families need support, encouragement, education, and time to practice essential skills to learn how to prepare for maintaining health and wellness and accessing care in the adult health care system. Youth and their families find that they have to negotiate 2 contrasting cultures and systems of health care: the pediatric and adult health care systems.[3] Table 24.2 outlines some of the major differences in the 2 systems. The different approaches in the adult health care system and the timing for moving to the adult-oriented system must be discussed with the young person and family, so the young person can be knowledgeable about what to expect and be an integral part of the process. Adult-oriented systems focus on the individual, not the family unit, and are rarely structured to provide multidisciplinary care. The demands of treating young adults with chronic illnesses may outweigh the benefits in the eyes of some adult health care professionals, especially if the young adult with SHCN was not prepared to be their own health advocate by the pediatric system and is non-adherent. In addition, systems of care (availability of case managers,

Table 24.2. Cultural Differences between Pediatric and Adult Health Care[a]

	Pediatric	Adult
Age-related	Growth & development, future focused	Maintenance/decline: Optimize the present
Focus	Family	Individual
Approach	Paternalistic Proactive	Collaborative Reactive
Shared decision-making	With parent	With patient
Services	Entitlement	Qualify/eligibility
Non-adherence	Greater assistance	Less tolerance
Procedural pain	Lower threshold of active input	Higher threshold for active input
Tolerance of immaturity	Higher	Lower
Coordination with federal systems	Greater interface with education	Greater interface with employment
Care provision	Interdisciplinary	Multidisciplinary
No. of patients with special needs in practice	Fewer	Greater

[a] Reprinted by permission. HRTW National Resource Center.

social workers, and mental health services) and financial reimbursement practices have not caught up with the numbers of young adults with SHCN. In contrast, pediatric health care professionals tend to focus on the family, and many are reluctant to "let go" of their youth with SHCN. Last but not least, youth with SHCN make up a smaller percentage of patients in pediatric practices, as most children in pediatric practices come for well-child care. This is a contrast to the adult system, where most of the individuals in an adult practice have chronic illnesses. Thus the youth with SHCN and their families may find that they do not stand out as being as different from others in the adult practice as they had in the pediatric practice, and they need to become excellent advocates to have their needs addressed in the new adult setting. Plans to discuss these differences and strategies as to how the youth and family negotiate these differences and add the skills required to address these differences need to be components of the youth transition plan.

3. **Develop a transition plan that includes the changing roles and skills to be acquired by the youth and family and coordinate the skill development with activities in the youth's Individualized Education Program (IEP) at school.** Coordination of care is a cornerstone of the medical home and is an essential component of a successful transition process. Developing a common medical transition plan among all health care professionals can assist in this process. A more detailed discussion of the pediatrician's role can be reviewed in the AAP policy statements on care coordination.[15–17] In light of the differences in the 2 approaches of pediatric and adult health care outlined previously and understanding the capacity of the youth involved, it is recommended that a transition plan be developed that matches the demands of the new system to the skills needed. It is thus critical that the new skills needed be included as goals on the youth's IEP at school to prepare the youth for the skills needed to negotiate the added demands of the adult health care system. If the demands match the skills, there will be far less anxiety on behalf of the youth and family as they move through the transition process.

Areas to include in a transition plan

Delineate the needed youth and family skills: Developing a health care transition plan is a key component of the consensus statement. This should be developed in conjunction with the youth and family in consultation with other health care professionals and related services (school, work, community). Discussing the components necessary for the youth to be able to care for their health, learn/practice to be a more informed decision-maker, and formulate a timeline to practice and acquire those skills are core components of a transition plan. See Appendix D and E for an HRTW tool that lists skills for inclusion in a transition plan. This can be used as a screening tool to decide which skills need to be acquired for each youth with SHCN.

Discuss legal health care decision-making before age 18, including the issue of guardianship. Ask the youth to give assent before they are required to give consent.

Assist youth to provide a concise medical report. Central to receiving appropriate health care is the ability to clearly describe medical symptoms and to ask questions about the process, outcome, and therapy. If the youth is dependent, the health surrogate should learn to communicate this information as well. The 1-page medical summary discussed previously is an essential part of the interaction and must be updated after each appointment. It is essential to include the acquisition of medical reporting skills in the transition plan. It is recommended that the child/youth be encouraged to come up with 5 questions to ask at each visit. Another HRTW tool that can assist the youth is contained in Appendix F. As part of the preparation for the visit, the family or teacher can assist the student in inserting their questions under the correct body system. Note that the listing of systems follows the routine encounter questionnaire often used by health care professionals. Using this approach, the youth receives practice asking their questions to their health care professional, the health care professional has a glimpse of what is important to their patient, and the youth is in sync with the order of what will be asked and learns that these recorded questions will be acknowledged.

Plan when the youth will start seeing their health care professional alone. An important correlation in transition studies is that youth who were seen alone during their health care visit were more likely to take charge of taking their own medications.[18] See Appendix G.

Create a portable medical record (see critical step #3). Another essential component to coordinated care is communication between all health care professionals and the youth and family. Key to this is the development of a portable medical record that is easily understood and compact. In Appendix B (medically complex) and C (cognitive support needs) are examples of a medical record. It also includes *Current Procedural Terminology* codes to assist the next health care professional to bill easily for the services offered. Overall the portable medical summary

- *Expedites accurate medical information reporting* in emergency situations, new medical encounters, and intake for service care professionals
- Provides *quick scan to document disability* often required for program eligibility
- *Increases patient knowledge of diagnosis and medical history* (over time with repeated review, the patient will increase recall of accurate dates, names/dosage of medications, and contact information of health care professionals and payers)

Create an emergency plan. Youth and their families should be prepared for a medical emergency. The youth should use the ICE (in case of emergency) designation in their cell phone with the appropriate numbers and medications/allergies listed after the ICE designation. Most youth carry a cell phone, and this is the first place the emergency personnel look for important numbers and information. Youth are interested in knowing what to do if their condition worsens and in knowing the signs that their health is deteriorating. Clarifying these issues (including knowing their baseline information, such as temperature, blood pressure, and respiratory rate), and having a plan for how the youth can access their health care professional in a timely manner, improves the quality of their care.

Outline steps for health promotion. Regular exercise and good nutrition are essentials to good health. Youth with developmental disabilities have similar rates of obesity and risky behaviors (tobacco and alcohol use) as people without disabilities and are less likely to participate in prevention screening programs.[19] Using prevention schedules for adolescent health, such as Bright Futures,[20] is important for the youth's general health. Participation in community-based programs helps the youth maintain healthy lifestyles after leaving school. Leisure activities are especially important for youth with developmental disabilities, as they often become socially isolated. Therapeutic recreation programs that incorporate skills training assist individuals with developmental disabilities to develop social skills and teach healthy and fun behaviors. The Special Olympics is a good example of such a program, and the National Center on Physical Activity and Disability (http://www.ncpad.org) is a clearinghouse to promote physical activity for people with disabilities.

Outline educational goals. Pediatricians should assist youth with developmental disabilities attain the best education possible, as level of education is related to health outcome.[21] Attending postsecondary education increases the likelihood of attaining adult social roles among young adults with childhood disabilities.[22] It is recommended that the AAP policy statements concerning the roles of the pediatrician in postsecondary education and work for youth with disabilities be reviewed.[23,24] In preparation for postsecondary education, see Appendix H for a list of questions to elucidate the accommodations needed.

Outline career/work goals. Regular school attendance and volunteer and paid jobs during high school improve future success in the workplace.[25] The importance of being on time for school and appointments should be emphasized, as this translates to employment skills needed to successfully maintain jobs.

Discuss independence and transportation issues. A handout with a list of community resources, such as summer camps and travel training, should be provided. See Selected National Web Resources on Transition on page 528 for a list of national Web sites for additional information.

Plan a visit to an adult practice or plan a joint visit with an adult health care professional. To assist the youth in learning what the adult health care system is like, an observational visit a year or more before the actual date of transfer should be planned. This allows the youth and family to see what the adult system requires. The youth and family can then return to the pediatric health care professional to assess the level of skills still needed to prepare for the transition to adult medical care. This also affords the youth and family the opportunity to select the adult health care professional of their choosing when the youth is medically stable. Alternatively, a joint visit with an adult health care professional can be planned, so that there can be verbal transfer of medical information, written medical record transfer, and discussion of remaining skills that are needed to be accomplished from the original transition plan.[26]

Critical Step 5: *Screening:* Apply the same guidelines for primary and preventive care for all adolescents and young adults, including those with SHCN, recognizing that young people with SHCN may require more resources and services than do other young people to optimize their health. Examples of such guidelines include the American Medical Association's *Guidelines for Adolescent Preventive Service;* the American Academy of Pediatrics' *Bright Futures: Guidelines for Health Supervision of Infants, Children, and Adolescents;* and the US Public Health Service's *Guide to Clinical Preventive Services.*[2]

In addition to the preventive health screening mentioned above, it is also essential to screen for mental health issues. Individuals with developmental disabilities are approximately 4 times more likely to develop mental health problems than the general population, and they report inadequate emotional support compared with the general population.[27] Behavioral concerns should be attended to and not be attributed to the developmental disability. Altered sleep and appetite can be signs of anxiety and depression.[28]

Critical Step 6: *Health Insurance:* Ensure affordable, continuous health insurance coverage for all young people with SHCN throughout adolescence and adulthood. This insurance should cover appropriate compensation for (1) health care transition planning for all young people with SHCN and (2) care coordination for those who have complex medical conditions.[2]

Key issues that a health care professional should consider when discussing the options for continuous health insurance coverage include the following:

Maintaining Insurance: Generally there are 3 situations when a parent's/family's health insurance coverage may be extended to cover adult children after they turn 19: he/she (1) is a student (usually covered until 22–25 years of age), (2) is an adult-dependent disabled child (no age limit as long as parent does not change jobs), or (3) opts to continue coverage through the Consolidated Omnibus Budget Reconciliation Act (COBRA) due to a change of employment status of the insured family member.

Student Dependent Children (age 19 to usually 22–25 years of age): Coverage may be continued for unmarried children if they are full-time students and dependent on their parents/family for support and maintenance. Some plans have higher age limits. In the past, if a student had to reduce classes or withdraw a semester due to illness, he or she would lose their health insurance. With the passage of "Michelle's Law" (federal mandate) a student who has to leave college due to illness can remain on the family plan if he or she needs to take a medical leave. The extension of eligibility is intended to protect group health benefit coverage of a sick or injured dependent child for up to 1 year. The new law affects all health benefit plans including ERISA, according to the Internal Revenue Code and the Public Health Services Act. (http://www.jdsupra.com/documents/15e5b928-d695-47bd-ae98-d93c04f56f6e.pdf.)

Adult Disabled Dependent Children (no age limit): In general, a dependent (child or disabled adult child) is a person for whom the parent/guardian provides more than half (50%) of yearly living expenses and declares that person as a dependent on his or her federal income tax return or as specified in a divorce decree. Once approved, insurance companies request annual recertification. This is routine paperwork, although it is easier to have documents prepared and updated yearly. Currently, 37 states mandate this approach via their state health insurance commissions, but 13 states do not (AL, AK, CO, DE, DC, IA, KS, KY, OK, OR, PA, RI, WV).

Court-Ordered Health Care Benefits: If an adult disabled dependent child is enrolled in health insurance under a parent's family health care insurance plan as part of a court-ordered child support decree, it is important that a time frame is specified (until age 18 years, lifelong, etc). This may require consulting a lawyer who specializes in family law to avoid having to go back to court again.

New Trend Emerging: Since 2006 many states have been exploring extending the age of family plan coverage for young adults with and without a disability to ages 25 to 30. New Jersey was the first state to pass this provision and allows coverage up to age 30. As of 2010, 28 states have adopted this approach and enacted legislation (CO, CT, DE, FL, IA, ID, IN, IL, KY, ME, MD, MA, MI, MN, MT, NH, NJ, NM, NY, RI, SD, TN, TX, VT, UT, VA, WA, WV).

Conclusion

Youth with developmental disabilities and SHCN want a full range of opportunities like everyone else and require a coordinated system of care. Although federal and state mandates exist and more inclusive opportunities are available, outcomes for youth with SHCN continue to be less than optimal. Coordination of care is the cornerstone of the medical home, and much needs to be done to make the transition from pediatric to adult health care a seamless one. To improve the process for all involved, preparation is central. Primary pediatric health care professionals should begin by developing a transition policy for their practice and post it for all to see. This allows everyone involved to plan for the important next steps. The pediatric practice should start the transition process around the age of 11 to 13 years, identify the core knowledge and skills needed by the youth and family for transition and allow them to practice these skills, create and use a transition plan and a portable medical record with the youth and family, and assist the family in identifying continuous health insurance options. Another challenge for all primary pediatric health care professionals is to simultaneously improve the system that serves youth with SHCN and prepare youth and their families with the knowledge and skills necessary to promote self-determination and successful navigation of adult systems of care, so that youth can be…

Healthy…Living…Learning…Working…Playing...
And have the ultimate of all outcomes: a successful transition to adulthood.

Selected National Web Resources on Transition

Youth
Family Voices KASA—Kids As Self Advocates
www.fvkasa.org

National Youth Leadership Network (NYLN)
www.nyln.org

Family
Family Village
www.familyvillage.wisc.edu

Family Voices
www.familyvoices.org

Cultural Competence
National Center for Cultural Competence (NCCC)
http://nccc.georgetown.edu

Education
National Center on Secondary Education and Transition (NCSET)
www.ncset.org

Independent Living
Centers for Independent Living (CILs)
www.ncil.org/about/CentersforIndependentLiving.html

National Council on Independent Living (NCIL)
www.ncil.org

National Directories of Centers for Independent Living
www.ncil.org/directory.html

Government Benefits
Government Benefits: Online
www.govbenefits.gov

Social Security Administration (SSA)
www.ssa.gov

SSA Benefit Eligibility Screening Tool (BEST)
http://connections.govbenefits.gov

Employment
SSA Youth With Disabilities
http://www.socialsecurity.gov/work/youth.html

Health Care Transition
Consensus Statement on Health Care Transitions for Youth and Young Adults With Special Health Care Needs
http://aappolicy.aappublications.org/cgi/content/full/pediatrics;110/6/S1/1304

Healthy & Ready to Work (HRTW) National Center
www.hrtw.org

National Center for Medical Home Implementation
www.medicalhomeinfo.org

American Academy of Pediatrics Illinois Chapter
"Preparing for the Future: Transition to Adulthood"
internet.dscc.uic.edu/forms/transition.pdf

Physical Activity and Disability
National Center on Physical Activity and Disability (NCPAD)
www.ncpad.org

Parents With Disabilities
Through the Looking Glass
www.lookingglass.org

Self-Determination
National Center for Self-Determination
www.self-determination.com

Sexuality
Quality Mall.org—Person Centered Services Supporting People With Developmental Disabilities
http://www.qualitymall.org/directory/dept1.asp?deptid=16

Disability Solutions
Sexuality Education: Building a Foundation for Healthy Attitudes
www.disabilitysolutions.org/pdf/4-5.pdf (Part I)
www.disabilitysolutions.org/pdf/4-6.pdf (Part II)

Technology
AbleData
www.abledata.com

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Chapter 25

Billing and Coding for Developmental and Behavioral Problems in Outpatient Primary Care

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Evolving and increasing interest by parents and professionals in areas of children's development and behavioral regulation has resulted in more medical care for children in these areas. Parents and professionals want early identification and ongoing supportive management to ensure optimal cognitive and behavioral development and social adaptation. Getting paid for this care can be problematic, as payers often view visits focusing on preventing problems and ongoing non–face-to-face services as difficult to manage from a cost-containment perspective. Satisfying professional needs for complete care, parental requests for comprehensive management, and third-party payer insistence on cost-containment can result in many needed services being provided gratis by the primary pediatric health care professional. Planning can ameliorate this. Published resources are available to inform medical providers and help them better understand the codes describing services and conditions.[1,2]

International Classification of Diseases

The *International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM)*[3] is used to classify diseases and operations, originally only for morbidity and mortality statistical purposes but now universally used for claims submission. Although a tenth edition was published in 1993 by the World Health Organization, *ICD-9* is still used in the United States except for the coding of hospital mortality statistics. (The rest of the world has already started using *ICD-10,* but this change has not occurred in the United States as of this book's publication.) The *ICD-9* diagnostic codes are meant to completely and accurately describe the patient's clinical picture. These codes describe *why* the service was performed. These codes are often used by payers as part of risk management (eg, to determining preexisting conditions and refusing payment for diagnoses). It is beyond the scope of this chapter to discuss *ICD-9-CM* diagnostic codes in great detail; for that the reader is referred to one of several published editions of *ICD-9-CM* appearing both online and in print.[4–6]

Germane to this discussion, however, are those diagnostic codes in the *ICD-9-CM* series **290–319** labeled the "mental health" codes. These codes are also described in the *Diagnostic Statistical Manual, 4th Edition, Text Revision (DSM-IV-TR)*[7] as well as in the *DSM for Primary Care Child and Adolescent Version (DSM-PC)*.[8] The *ICD-9-CM* series **290–319** codes are often referred to during discussions of parity. Just as payers may choose not to pay for some *Current Procedural Terminology (CPT®)* services (such as non–face-to-face care services, eg, telephone calls, e-mail messages, and extended reports), payers also may exclude (in their contracts) care for certain *ICD-9* diagnostic codes, either individually or by groups. Mental health codes *(ICD-9-CM* **290–319** series) often fall in this excluded code designation, as diagnoses restricted to "mental health providers."

Pediatricians providing a significant amount of developmental care may consider being paneled as a mental health provider by payers. However, this may be a case of "Be careful what you wish for!" Typically, physicians identified by payers as mental health providers are expected to describe their services through the **908XX** *CPT* procedure codes (see next section for more description of *CPT* coding), restricting these providers from using evaluation and management (E/M) codes when providing services for conditions in the **290–319** *ICD-9-CM* Chapter 5 series. The problem is that the **908XX** codes may not accurately describe the extensive counseling and care management both general pediatricians and developmental-behavioral or neurodevelopmental specialists provide to their patients. This counseling and care coordination is not covered in the relative value units (RVUs) allotted in the code series **9908XX.** This results in uncompensated work! This is a true conundrum in the health care system—one that will only be corrected when mental health care achieves parity with other medical conditions.

Another problematic group of diagnostic codes are the V codes. These secondary codes are located in a separate section at the end of Volume 1 of *ICD-9-CM* and also comprise many of the codes described in the *DSM-PC.* Generalists often refer to these codes as the "well-child" diagnostic codes and may be leery of using them as the primary diagnosis when providing non-preventive E/M services, as denials for payment may occur. (V code diagnoses usually may be the primary diagnosis if the patient was not seen for an active illness/injury.) If payers do permit V code use, they may require supporting documentation. Providers also must be aware that payers also may consider "V-code visits" as checkup visits and this may eventually result in nonpayment for a well-child visit if the patient has used up their allotted number of checkup visits. Providers should be cautious when using V codes as second or third diagnoses because, as Medicare automatically denies these claims, many other payers who adopt Medicare policies may also deny the claim. This may be especially true for clinics located in facilities that primarily serve adults. They often adopt Centers for Medicare & Medicaid Services (CMS) policies rather than *CPT* policy.

CPT: Background

As another part of intelligent practice planning, it is important to understand how services for developmental/behavioral conditions are described in *CPT 2010*.[9] These procedure codes are Level I codes of the broader-based Healthcare Common Procedure Coding System (HCPCS).[10] Level II HCPCS codes, developed by CMS, are 5-digit alphanumeric codes with a letter as the first character and are used to report services and supplies not identified in *CPT. Current Procedural Terminology 2010* is the current iteration of a process developed in 1966 to "provide a uniform language that will accurately describe medical, surgical, and diagnostic services, and will thereby provide an effective means for reliable nationwide communication among physicians, patients and third parties." *Current Procedural Terminology* allows providers to concisely describe services provided at a patient encounter. If the provider understands the *CPT* system, it is possible to accurately "capture" most, if not all, of the work done. If the provider is not knowledgeable or chooses to ignore published codes and modifiers, it is quite likely significant work will go unpaid by providers! It must be remembered, however, that payers are free to set their individual payment policies. Even when there are published RVUs for a procedure code, payers may refuse to pay for the services.

The CMS has a process to assign values (RVUs) to medical service codes. The assumption is that service value is quantifiable, and each service has a relative value based on the resource costs needed to provide each service. Each code value comprises 3 elements: physician work, practice expense (direct and indirect costs), and medical liability expense. These 3 factors (each assigned an individual RVU or part thereof) are summed to get the total RVU. This total RVU is multiplied by a Medicare conversion factor ($ per RVU) and a geographic conversion factor (reflecting that expenses vary from community to community) to give the Medicare payment for that service. Medicare conversion factors are updated every 6 months (January 1 and July 1: 1/1–6/30; 7/1–12/31.)

Relative value units (either work RVU or total RVU) are often used by larger group practices and larger institutions, such as university centers, to appropriately divide the group's income for determining individual salaries and to establish benchmarks for physician work. Relative value units are used in managed care contracts to determine the costs involved in providing the services. When *CPT* codes are examined, it is often noted that codes describing procedures have higher RVUs per time unit than codes describing cognitive services.[11] This is a particularly relevant concern for physicians providing developmental-behavioral services, as most of this work does not involve procedures. The non-proceduralist spends considerable time delivering cognitive services and may not be able to generate the same number of RVUs on a given day when compared with a provider who engages in more procedures. Developmental-behavioral specialists and generalists providing developmental and behavioral care in university-based and private multispecialty clinics can influence charges by

advocating for more realistic RVU benchmarks by their administrators. Advocating stronger payment for cognitive services with payers during contract renewal discussions also can help level the playing field.

CPT for Developmental-Behavioral Services

Outpatient face-to-face codes pertinent to developmental-behavioral services describe new, established, consultation, and preventive E/M services. Appropriate inpatient hospital *CPT* codes may also be correctly used to describe inpatient services for developmental and behavioral conditions. Codes for developmental screening and objective developmental testing also are available. If a service was modified in some manner, modifiers may be appended to the basic service code to inform the payers that both outpatient and inpatient services were modified, which may result in a change in charge or payment. Non–face-to-face services are also used when providing developmental and behavioral/mental health care: codes are available for those services as well as for telephone care, online medical evaluation, care plan oversight, extensive reports, and team conferences.

While clinicians may rail against the current payment system, believing that it undervalues cognitive work, the system may be used more effectively if providers understand "the system" and appropriately create clinical services in synchrony with the current reimbursement system. This does not mean abandoning *quality* care, but it does mean understanding *efficient* care. The following code descriptions are provided to improve code awareness and to link codes to efficient service delivery.

Outpatient E/M Codes

Outpatient developmental and behavioral care is provided using E/M services. These services include preventive health care **(99381–99384; 99391–99394),** new **(99201–99205)** and established **(99211–99215)** patient office visits, and consultation **(99241–99245)** services on request of another appropriate professional. A patient is considered new if the provider or any other provider from the same specialty in the same medical group practice either has never provided care or has not seen the patient in the past 3 years in any clinical setting. All E/M services, with the exception of preventive visits, may be coded on the basis of either the complexity of the visit or on the amount of time required for counseling and case management on the date of service. Non-preventive E/M visit components are defined by 4 levels of service: problem focused, expanded problem focused, detailed problem focused, and comprehensive. Complexity entails consideration of the 3 key components: history, physical examination, and the complexity of the medical decision-making required by the provider. Tables 25.1, 25.2, and 25.3 illustrate the 5 levels for consultation and new and established patient visits.

Note the sole difference between the detailed problem focused and the comprehensive levels is the difficulty of medical decision-making! Determining difficulty takes into consideration (1) the number of possible diagnoses and/or potential treatments;

Table 25.1. New Office Consultation[a]

Component	99241	99242	99243	99244	99245
History	Problem focused	Expanded problem focused	Detailed	Comprehensive	Comprehensive
Exam	Problem focused	Expanded problem focused	Detailed	Comprehensive	Comprehensive
Decision-making	Straightforward	Straightforward	Low complexity	Moderate complexity	High complexity
Time (minutes)	15	30	40	60	80

[a]Documentation of all components (history, examination, and decision-making) is required for all levels of care.

Table 25.2. New Patient Office Visit[a]

Component	99201	99202	99203	99204	99205
History	Problem focused	Expanded problem focused	Detailed	Comprehensive	Comprehensive
Exam	Problem focused	Expanded problem focused	Detailed	Comprehensive	Comprehensive
Decision-making	Straightforward	Straightforward	Low complexity	Moderate complexity	High complexity
Time (minutes)	10	20	30	45	60

[a]Documentation of all components (history, examination, and decision-making) is required for all levels of care.

Table 25.3. Established Patient Office Visit

Component	99211	99212	99213	99214	99215
History	Not required	Problem focused	Expanded problem focused	Detailed	Comprehensive
Exam	Not required	Problem focused	Expanded problem focused	Detailed	Comprehensive
Decision-making	Not required	Straightforward	Low complexity	Moderate complexity	High complexity
Time (minutes)	5	10	15	25	40

(2) the amount and/or complexity of ancillary information (medical records, diagnostic tests, other information); and (3) risk of complications, morbidity, and/or mortality associated with the patient's presenting problem.

The medical record documentation should always support both the *ICD-9* and *CPT* codes selected. If the provider spends at least half of the visit in counseling or care coordination, then time may be used to determine the *CPT* code (see below). A statement such as, "The visit took XXX minutes, and more than 50% of time was spent in counseling (or coordination of care)" should be included in the note. The clinician should code the diagnosis to the highest level of diagnostic *certainty* (the words in the descriptor) and *complexity* (the numbers in the *ICD-9-CM* codes). The first diagnosis listed on the billing sheet, however, should be the condition being actively managed on that date of service. A chronic condition, such as attention-deficit/hyperactivity disorder or depression, managed on an ongoing basis, may be coded and reported as many times as applicable to the patient's treatment. There is no provision for "rule out" in *ICD-9,* but there is a code **(799.99** deferred diagnosis). This could be properly used; however, payers may not choose to pay if **799.99** is listed first.

Preventive Service E/M Codes and Developmental-Behavioral Care

Health supervision (well-child checkups) and preventive medicine visits are properly described by the preventive medicine services codes (Table 25.4). These codes, like the other E/M codes, are subcategorized as new patient or established patient. Codes in these series are selected based on the age of the patient and not on the basis of time spent on the date of service.

Both new and established visits, however, assume counseling, anticipatory guidance, and risk-factor reduction interventions were offered during the session. If the provider wants to efficiently cover developmental and behavioral concerns in the typically busy primary care setting, developmental screening instruments and behavioral rating scales can be key procedures. These procedures may properly accompany the

Table 25.4. Preventive Services for New and Established Patients (99381-4; 99391-4)

Code	Description
New Patient	
99381	Initial comprehensive preventive medicine evaluation and management; infant (<1 y of age)
99382	Early childhood (age 1–4 y)
99383	Late childhood (age 5–11 y)
99384	Adolescent (age 12–17 y)
Established Patient	
99391	Periodic comprehensive preventive medicine evaluation and management; infant (<1 y of age)
99392	Early childhood (age 1–4 y)
99393	Late childhood (age 5–11 y)
99394	Adolescent (age 12–17 y)

preventive service and be described to the payer by appending an appropriate modifier to either the preventive service code, the procedure code or, in some circumstances, both.

Modifiers

Modifiers are 2-digit suffixes appended to the end of a *CPT* code to tell the payer that "this visit was different." The service may have been altered by a specific situation described by the modifier. Some modifiers may only be used with E/M codes, and others only may accompany procedures. Modifier use is extremely important, as they enable the payer software to permit these special circumstances. When modifiers are used, however, the provider must take care to clearly document the circumstances supporting their use. Despite correct modifier use and supportive documentation in the medical record, not all payers recognize all modifiers. Listing modifiers on the billing sheet will encourage their use!

The following modifiers may correctly be appended to E/M codes in the following circumstances:

25: Significant, Separately Identifiable E/M Service by the Same Physician on the Same Day as the Procedure or Other Service—This modifier tells the payer that the documentation requirements were met for a separate E/M service or procedure on that date. For example, the discussion of current concerns, medication side-effects, efficacy, etc, would contribute to the level of the separate E/M service selected. Modifier **25** would be properly appended to the E/M code to address the change in medication and not the preventive medical care visit. The medical record should have 2 separate sections documenting the 2 separate services.

Another example of correct modifier **25** use would be the situation when the parent completes a developmental screening instrument as part of a preventive medical care visit. In that situation, the procedure is described by code **96110** (developmental testing, limited) and modifier **25** would be properly appended to the selected **993XX** code, and **96110** also would be included as a separate service charge. Again, there must be separate sections in the medical record for the 2 provided services.

32: Mandated Services—This modifier is used when a third party requires the service being provided. For example, if a second opinion of a mental health or behavioral diagnosis is required before a child may receive treatment, modifier **32** would be appended to the E/M code describing the visit providing that evaluation service.

Modifiers used with procedures and not E/M codes include

52: Reduced Services—This is used when a procedure is started, but then prematurely stopped at the physician's discretion. This might occur during developmental testing if the child became oppositional and refused to respond to the testing. In that

case, **52** would be appended to **96111** (developmental testing, extended) to describe the premature termination of the testing session. Note, however, that the diagnosis code used at this visit should explain why the procedure was reduced. The physician is not required to reduce their charges for the reduced service, as the payer will make that decision.

59: Distinct Procedural Service—This modifier is used when procedures not usually reported together do occur appropriately on the same date of service. It indicates the procedure was distinct from the other procedures performed on that same date of service. Coders refer to this modifier as the "modifier of last resort," and the physician should be satisfied that no other modifier would be more appropriate. Clear documentation is almost always required to support payment of both procedures. An example of the use of modifier **59** would be a second office visit for developmental cognitive testing **(96125)** during which the parents also returned rating scales completed by the patient's child care provider for scoring and interpretation **(96110).** Modifier **59** would be appended to the second procedure marked on the billing sheet.

76: Multiple Procedures—Repeat procedure or service performed by the same physician on the same date of service. This code is used when the physician repeats a procedure on the same date. This modifier is appended to the second unit of the procedure to tell the payer more than one of that same procedure was performed. For instance, using **96110** (developmental testing, limited) as an example: if Pediatric Evaluation of Developmental Status (PEDS) forms were completed, scored, and interpreted for a child's mother (custodial parent), father (has weekly visitation), and daily babysitter, then the 3 forms could be described as

96110 (modifier not put on the first unit)
96110 76
96110 76

Alternatively, as some payers do not want the modifier as they prefer units, the same example would be put on the billing sheet as

(3) 96110
Always follow the payer guidelines!

Developmental and Behavioral Procedures: Screening and Objective Testing

Screening: 96110

Few procedure codes apply to physician provision of developmental-behavioral services, but those published and valued codes should not be ignored! These procedures include looking for possible areas of concern (screening or developmental testing:

limited), as well as formally assessing performance in one or more areas of development (developmental testing: extended; neurobehavioral status examination, standardized cognitive performance testing, computerized neuropsychological testing) (Box 25.1).

The identification process for developmental and behavioral differences begins with questions asked and provider observations made at every infant and young child's preventive service visit. This developmental surveillance is considered part of the well-child evaluation services. Eliciting parental observations of the child's communication efforts, emerging social reciprocity, and developing symbolic play may suggest closer monitoring of those children who are not developing in a typical manner. Developmental surveillance is augmented with periodic formal developmental screening. According to guidelines published in 2006 by the American Academy of Pediatrics (AAP),[12,13] formal developmental screening instruments should be administered regularly at the 9-, 18-, and 30-month preventive health visits and at other visits if any concerns arise during the developmental surveillance process. Standardized developmental screening instruments are available (eg, PEDS,[14] Ages and Stages Questionnaires [ASQ][15]) to assess the child's language, fine and gross motor, problem-solving, and social-emotional skills. These standardized measures meet the stated instrument criteria in the *CPT* descriptor for developmental screening. Developmental testing: limited **(96110)** may be properly coded in addition to the preventive service code to describe the services provided at the visit. When standardized developmental screening instruments are administered, scored, and interpreted as part of the preventive service visit, each screening instrument form is individually coded. For example, if an established 9-month-old patient is being cared for most of the time by the maternal grandmother while the child's mother is in school and working part-time, information from both the mother and grandmother may be needed for this child. Two units of **96110** would then be coded

2 x **96110**
(2) **96110**

It may be necessary to append modifier **25** (separate and identifiable service) to the preventive service code to alert the payer that the preventive service was a separate and identifiable service from the procedure **(96110)** also coded for at that visit. For the 9-month-old infant described, all services at that visit would thus be coded as

99391 25
(2) **96110**

Screening for autism spectrum disorders in younger children may include a formal measure, such as the Modified Checklist for Autism in Toddlers (M-CHAT).[16] Administration of such a formal autism screening instrument is recommended by

the AAP at the 18- and 24-month visits. Administering, scoring, and interpreting this instrument also fulfills the description for the **96110** service and may be properly coded along with other standardized developmental screening instruments at a health visit. For example, if the child described previously was 18 months of age, and a PEDS and an M-CHAT both were given to the child's mother and grandmother to complete, these services would accurately be coded as

99391 25
(4) **96110**

Screening for development, autism, and behavioral regulation may occur during non-preventive medical visits. If children miss their well-child checkups, office visits for other reasons can serve as opportunities for reviewing developmental, social, and communication status through caretaker reporting and completion of standardized screening instruments. If, for example, a child misses the 24-month anticipatory guidance visit and is not seen until a visit for otalgia and upper respiratory signs, the parent(s) may properly be asked to complete both a general developmental screening scale and an M-CHAT. The scored results would be reviewed by the medical provider and discussed with the family. If further action is needed, another visit could be scheduled or if a referral for additional consultation by other professionals seems indicated, this could be made. Coding for this visit would include

99214 (or 5) **25**
(2) **96110** (assuming one parent completed one developmental screening scale and one parent completed an M-CHAT)

It is important to note that the **96110** payment is based on the practice expense of the screening instrument administration (cost of the protocol, clinic overhead, nonphysician salary) and malpractice expense. There is no physician work in this code. The payment is based on someone other than the physician administering and scoring the instrument. The physician interpretation of the score, discussing the results with the family, acting on the results, and writing a brief summary of the results and action are included in the payment for the basic preventive or E/M service. If, however, the physician administers the screening instrument to the parent and scores and interprets the results, then this additional time and/or complexity of medical decision-making may be considered when selecting the proper code describing an E/M service. If more than 50% of the total visit time is devoted to counseling and/or care coordination, then the additional time needed to administer, score, and discuss the findings may add to the total time in this "counseling" service. Documentation of time counseling can be expressed as a fraction: time spent counseling/total time of visit. If, however, the visit is being coded on the basis of complexity of the service, then the interpretation of the results of the rating scale and the integration of these findings into the medical decision-making may increase the medical decision-

making complexity. Both examples show how developmental rating scales may contribute to a higher-level code (eg, **992X4→992X5).**

In late childhood and adolescence, rating scales are available to help identify those children and teens who need further evaluation for emotional conditions. One scale is the Screen for Child Anxiety Related Disorders (SCARED).[17] The physician often asks these questions directly during the visit and scores the results. In this instance, the separate code **(96110)** would not be appended to the visit, but the level of the service might be increased on either time or complexity of medical decision-making (with appropriate documentation). If, however, a nurse (or other trained, nonphysician) administers and scores the screening instrument and the physician interprets the results and reviews with the family, then **96110** can be used, and the time and effort of the interpretation and discussion by the physician can be factored into the E/M code.

Objective Testing: 96111, 96116, 96125

Physicians who have been properly trained in the selection, administration, scoring, and interpretation of standardized tests assessing varying areas of developmental attainment may quite properly perform this service. The *CPT* codes describing testing fall in the **96XXX** series. While all "psychological testing" codes (eg, **96101,** psychological testing; **96118,** neuropsychological testing) allow submission by physicians, the physician who uses these codes should be prepared to show very specific specialty training in the administration and interpretation of these tests. These codes

Box 25.1. Standardized Test Examples for 96111, 96116, 96125[20–29]

Code/Test
96111: Developmental Testing: Extended
Autism Diagnostic Observation Schedule (ADOS)
Beery-Buktenica Test of Visual-Motor Integration (VMI)
Peabody Developmental Motor Scales–Second Edition (PDMS-2)
Bruininks-Oseretsky Test of Motor Proficiency–Second Edition (BOT-2)
Clinical Evaluation of Language Fundamentals–Fourth Edition (CELF-4)
Bayley Scales of Infant and Toddler Development–Third Edition
96116: Neurobehavioral Status Exam
Woodcock-Johnson Tests of Cognitive Abilities–Third Edition (WJ-TCA-III)
Autism Diagnostic Observation Scale (ADOS)
Psychoeducational Evaluation Profile (PEP)
96125: Standardized Cognitive Performance Testing
Comprehensive Test of Nonverbal Intelligence (C-TONI)
Tests of Nonverbal Intelligence–Third Edition (TONI-3)
Woodcock-Johnson Tests of Cognitive Abilities–Third Edition: Thinking Ability Index

may be coded in multiple units to account not only for face-to-face administration of the tests, but also for scoring and report writing. In addition, 2 other newer codes—**96116** (neurobehavioral status examination) and **96125** (standardized cognitive performance testing) also allow multiple unit coding to cover test administration, scoring and interpretation, and report writing.

96111: Developmental Testing, Extended

This code was developed to include standardized testing for motor, language, social, adaptive, and/or cognitive functioning. It may be coded only once on each day of service and allows no additional units for test scoring, interpretation, or report writing. This code is applicable for testing sessions as brief as that needed for verbal fluency testing (eg, controlled oral word association test),[18] as well as testing lasting more than an hour. It may also be properly used for administering objective developmental screening instruments such as the Bayley Infant Neurodevelopmental Screening (BINS).[19]

Some payers may not permit the codes **96110** and **96111** to be charged together (developmental testing-limited and developmental testing-extended) as they interpret these services to be in conflict. They ask, "How can you bill for 'limited' developmental testing at the same time you are billing for 'extended' developmental testing? Isn't that 'unbundling'?" Addressing this will take additional clarification and advocacy with the individual payers.

96116: Neurobehavioral Status Examination

This code may be used for "clinical assessment" of thinking, reasoning, and judgment (eg, acquired knowledge, attention, language, memory, planning and problem-solving, and visual spatial abilities). Multiple units may be coded on the day of service to cover scoring and interpretation and report writing. While the descriptor does not specify use of standardized instruments, the physician who does not use a standardized test must be careful not to use this code to describe those tasks that would more customarily be included in the neurologic or mental status examination (eg, serial sevens). When documenting activity, the actual time spent for both face to face with the patient and the time interpreting test results and preparing the report should be reported—it should exceed 31 minutes and be less than 60 (for one unit of the code).

This service may properly be provided—and billed—in combination with **96110** (developmental testing—limited) and/or **96111** (developmental testing—extended). For example, if rating scales are completed, scored, and interpreted at a visit during which the neurobehavioral status examination is performed, both codes may be documented on the billing sheet. For example, if a level 4 E/M visit occurs with rating scales administration and completion, this could be coded as

962X4 25
(x units) **96110**
(x units) **96116 59**

Alternatively, if a level 4 E/M visit occurs with rating scales and separate and identifiable developmental cognitive testing by a non-psychologist, then this could be appropriate coding for that visit

992X4 25
(x units) **96110**
(x units) **96125**

96125: Standardized Cognitive Performance Testing

This code was developed for other professionals who also do standardized testing, but who were not included in the descriptors for **96101–96103** and **96118–96120.** As the descriptor says "qualified health professional's time," this code could quite properly be used by physicians. This code permits multiple unit coding on the day of service to allow payment for scoring, interpretation, and report writing.

Prolonged Services

Many times the visits for children with complex behavioral and developmental needs take longer than the time expected for the highest level of encounter. The average expected times are noted in Table 25.5, but the time may be more or less than noted below, depending on the clinical circumstances.

If the time exceeds the time for the appropriate highest-level service by more than 29 minutes, prolonged service codes may be appended. This may describe both face-to-face and non–face-to-face prolonged time spent in care on the same day as the appointment occurred.

Table 25.6 shows time guidelines for prolonged outpatient services. To correctly account for prolonged time beyond that specified in the selected E/M code, the first 30 minutes exceeding the E/M time is still included in the initial E/M payment. Therefore, a prolonged service code may not be used until after the first 30 minutes of the prolonged services have been provided. The medical provider has to spend

Table 25.5. Current Procedural Terminology Survey Time Guidelines for Highest-Level New and Established Outpatient Services

Code	Descriptor	Intra-Service Time (minutes)
99245	Consultation new patient	80
99205	New patient	60
99215	Established patient	45

Table 25.6. Guidelines for Prolonged Outpatient Services (99354-55–99358-5)

Total Duration of Prolonged Services (minutes)	Code(s)
Face to face, <30	Not reported separately
Face to face, 31–74	**99354**
Face to -face, 75–104	**99355,** each additional 30 minutes, past 74 minutes, use with **99354**
Face to face, 105–134	**99354** X 1 plus **99355** X 2
Before or after face to face, <30	Not reported separately
Before or after face to face, 31–74	**99358**
Before or after face to face, 75–104	**99359,** each additional 30 minutes, past 74 minutes, use with **99358**
Before or after face to face, 105–134	**99358** X 1 plus **99359** X 2

31 to 74 minutes with the patient and/or family in prolonged services before it is possible to correctly code for 31 minutes using code **99354.** What if the total prolonged services exceed 74 minutes? Code **99355** applies for each additional 30 minutes spent in face-to-face contact beyond the initial prolonged face-to-face 74 service minutes. To correctly use **99355,** however, the medical provider must spend at least 15 minutes beyond the initial 74 minutes of prolonged time.

It is essential to document the time and the services provided to substantiate the use of these prolonged services codes. Becoming familiar with these codes can help avoid the frequent (and disappointing!) situation of providing uncompensated care. Be aware, however, that even correct coding and documentation in the medical record may not result in payment. This is a payer issue. If request for payment is denied, ask for a written explanation of denials by the insurer. If the patient is on a capitated plan, you may request authorization to bill the family directly for these non-covered services.

Non–Face-to-Face Services: Telephone Care (99441–99443), Care Plan Oversight (993338–99339), and Extended Reports (99080)

Providing a medical home for patients with complex medical and behavioral conditions necessitates many non–face-to-face services. Primary care clinicians often develop "virtual teams" of community-based specialists to create comprehensive care plans for their patients with complex developmental and behavioral conditions. Reviewing interim reports occurs regularly. Telephone and written communications between medical providers (both primary and specialty), professionals in other areas, and family members are a routine and expected aspect of ongoing care. One option for requesting payment for these needed services is through the use of the care plan oversight codes **99338** (15–29 minutes/month) and **99339** (>29 minutes/month): recurrent physician supervision of a complex patient or a patient who

requires multidisciplinary care and ongoing physician involvement. These non–face-to-face services are reported separately from E/M services by the physician who has the supervisory role in the patient's care or is the sole provider. These 2 codes reflect the time required to supervise the care of the patient and are reported based on the amount of time spent/calendar month. Tasks appropriately included in this service would include reviewing previous medical records, reviewing laboratory results as part of medication monitoring, assessing progress of therapies (eg, speech/language, occupational, physical, or behavioral therapies), or documenting behavioral changes in psychodynamic therapy through telephone calls or written communication with the child's mental/behavioral health therapist. These codes are reported only once monthly and reflect the total time spent by the primary provider as care manager. To ensure accuracy in time calculation, a care log (Table 25.7) on which time and services are entered can be very helpful. A copy of this log can be attached when the billing sheet is submitted to the insurer, especially if there are denials for this service.

Table 25.7. Example of Service and Time Log for Care Management

	01/11/08	01/22/08	01/23/08	...	01/31/08
Telephone Call					
Child psychiatrist	10 minutes: discussed current Risperdal dose				
Lead school teacher		5 minutes: discussed not withholding recess			
School guidance counselor					15 minutes: discussed modifying Individualized Education Program (IEP)

If care plan oversight codes are regularly submitted, it would not be appropriate to submit bills for individual telephone calls. Telephone care, however, is often a significant part of providing care for children with complex developmental and behavioral disorders. Communication between the primary medical clinician and therapists, teachers, school counselors, and family members can be important to optimal management. While telephone calls to non–family members might correctly be included in care management services, telephone care to the child's family can be billed very appropriately as a separate and justifiable service. There are 3 published telephone care codes for use when the physician is part of the call. Relative value units for these 3 telephone care codes were published in 2008 (Table 25.8).

Table 25.8. Telephone Care (99441–99443)[a]

Code	Time of Medical Discussion (minutes)
99441	5–10
99442	11–20
99443	21–30

[a] Telephone evaluation and management (E/M) service provided by a physician to an established patient, parent, or guardian not originating from a related E/M service provided within the previous 7 days nor leading to an E/M service or procedure within the next 24 hours or soonest available appointment.

Again, remember that even correct coding and documentation of the telephone call in the medical record may not result in payment. This is another payer issue. If request for payment is denied, the provider may ask for a written explanation of the denial by the insurer. If the patient is on a capitated plan, you may request authorization to bill the family directly for these non-covered services. Before you bill families for non-covered activities like telephone calls, it would be wise to let them know that your practice may be doing so, for example, by sending all families a letter explaining that payment will be expected from them for these services and providing a fee schedule.

Finally, when parents purchase insurance coverage for their children with developmental or emotional/behavioral regulation conditions, insurers may require extensive documentation of need for this care on special forms. Self-contained schools and camps for children with significant developmental and behavioral conditions may require the primary clinician to complete extensive forms for the child's admission. Procedure code **99080** (special reports such as insurance forms, more than the information conveyed in the usual medical communication, or standard reporting form) is published and may be used for these services. There are no RVUs assigned to this code, and payers may not pay for this service. Clinicians should develop an office policy, similar to the decision for billing for telephone care, and advise families in advance if they will be expected to pay for this service.

Conclusion

Primary care physicians have an important role in providing children's developmental and behavioral health care. Developmental and behavioral differences should be identified as early as possible, and the medical home can provide appropriate care and care coordination. Understanding the current codes to describe medical services and the documentation needed to support them, using efficient methods of care, and modifying practice habits to minimize services not currently supported by procedural codes will permit the primary clinician to be paid in a more appropriate way for this medical care. Current medical procedure codes can legitimately be used to bill for care related to developmental and behavioral health needs and consistent use of these codes will help address payment barriers.

Resources

Commercial Pediatric Coding Newsletters

American Academy of Pediatrics. *Pediatric Coding Companion.* Elk Grove Village, IL: American Academy of Pediatrics

The Coding Institute. *Pediatric Coding Alert.* Durham, NC: Eli Research

Web Sites

www.dbpeds.org
Web site of the American Academy of Pediatrics Section on Developmental and Behavioral Pediatrics with coding information specific to developmental and behavioral health care for children.

www.aacap.org
Web site of the American Academy of Child and Adolescent Psychiatry with coding information specific to mental and behavioral health care for children.

www.cms.hhs.gov/MLNProducts
Documentation guideline revisions by CMS and the American Medical Association.

www.coding.aap.org
AAP updates on procedural and diagnostic coding.

www.thecodinginstitute.com
Commercial site for coding information.

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Appendices*

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Appendix A
Checklist for Transition: Core Knowledge & Skills for Pediatric Practices

Core Knowledge & Skills Checklist for Practices		
POLICY	**YES**	**NO**
1. Dedicated staff position coordinates transition activities		
2. Office forms are developed to support transition processes		
3. *CPT* coding is used to maximize reimbursement for transition services		
4. Legal health care decision-making is discussed prior to youth turning 18		
5. Prior to age 18, youth sign assent forms for treatments, whenever possible		
6. Transition policy states age youth should no longer see a pediatrician is posted		
MEDICAL HOME		
1. Practice provides care coordination for youth with complex conditions		
2. Practice creates an individualized health transition plan before age 14		
3. Practice refers youth to specific family or internal medicine physicians		
4. Practice provides support and confers with adult providers post-transfer		
5. Practice actively recruits adult primary care /specialty providers for referral		
FAMILY/YOUTH INVOLVEMENT		
1. Practice discusses transition after diagnosis, and planning with families/youth begins before age 10		
2. Practice provides educational packet or handouts on transition		
3. Youth participate in shared care management and self-care (call for appt/Rx refills)		
4. Practice assists families/youth to develop an emergency plan (health crisis and weather or other environmental disasters)		
5. Practice assists youth/family in creating a portable medical summary		
6. Practice assists with planning for school and/or work accommodations		
7. Practice assists with medical documentation for program eligibility (SSI, VR, college)		
8. Practice refers family/youth to resources that support skill-building: mentoring, camps, recreation, activities of daily living, volunteer/paid work experiences		
HEALTH CARE INSURANCE		
1. Practice is knowledgeable about state mandated and other insurance benefits for youth after age 18		
2. Practice provides medical documentation when needed to maintain benefits		
SCREENING		
1. Exams include routine screening for risk taking and prevention of secondary disabilities		
2. Practice teaches youth lifelong preventive care, how to identify health baseline and report problems early; youth know wellness routines, diet/exercise, etc.		

The HRTW National Resource Center believes these skills apply to all youth with and without a diagnosis.

The HRTW National Resource Center is headquartered at the Maine State Title V CSHN Program and is funded through a cooperative agreement (U39MC06899-01-00) from the Integrated Services Branch, Division of Services for Children with Special Health Needs (DSCSHN) in the Federal Maternal and Child Health Bureau (MCHB), Health Resources and Services Administration (HRSA), Department of Health and Human Services (DHHS). Activities are coordinated through the Center for Self-Determination, Health and Policy at the Maine Support Network. The Center enjoys working partnerships with the Shriners Hospitals for Children and the KY Commission for CSHCN.

NAME Address, Home Phone, Cell Phone, Email			
DOB	SS#	**Allergy**	**DNR SIGNED: N/ Y – ADD DATE**
Learns best by:			
Supports Needed:			
Legal Decision-Maker: __Self		**Guardianship: __ Limited __ Full**	
NAME: ADDRESS:		PHONE:	
Legal Health Surrogate:			
NAME:		PHONE:	

PRIMARY DIAGNOSIS/ICD-9 CODES	**AGE: XX HEIGHT X'X" (XX inches) WEIGHT XX lbs**
1. 2. 3. 4. 5.	

MEDICAL	
DOCTORS	**HOSPITAL**
MEDICINES Rx DAILY Rx MONTHLY Rx PRN	**IMMUNIZATIONS**
ADD NAME OF INSURANCE COMPANY *Primary Subscriber:* ADD NAME ADD Plan Code # ADD Subscriber # Customer service: ADD PHONE #	ADD NAME OF INSURANCE COMPANY *Subscriber:* ADD NAME ADD Plan Code # ADD Subscriber # Customer service: ADD PHONE #

Health Care/ Case Manager	ADD NAME	ADD PHONE #	ext. xx
Health Vendor	ADD COMPANY NAME/CONTACT	ADD PHONE #	ADD acc't. #
Home Nursing Agency	ADD COMPANY NAME/CONTACT	ADD PHONE #	ADD acc't. #
Pharmacy	ADD COMPANY NAME	ADD PHONE #	ADD RX #s
Dentist	ADD NAME	ADD PHONE #	

ADD NAME

Add Mailing Address Email, home phone, cell phone

INSURANCE	BlueCross BlueShield of Mass	Primary	Subscriber Name and # XXX-XX-XXXX BC/BS PPO Plan Code 200 Customer Service: 800-296-xxxx
		Secondary	Subscriber Name and # XXX-XX-XXXX BC/BS Blue Choice Plan 2, POS Code 200, Customer service: 800-222-xxxx

Legal	Name	Relationship	C XXX-XXX-XXXX W XXX-XXX-XXXX H XXX-XXX-XXXX
Health POA *	Name	Name	C XXX-XXX-XXXX H XXX-XXX-XXXX W XXX-XXX-XXXX

DOB 05-24-1973 **HEIGHT/WEIGHT**: 4' 3", 80 lbs **DNR SIGNED**: No **ORGAN DONOR:** No
SS# XXX-XX-XXXX **BLOOD TYPE**: A positive **ADVANCED DIRECTIVES:** Yes

---

- **High intelligence** (130 IQ), compliant patient, high tolerance to pain
- Incomplete Quad (has sensation), only movement left index finger 10 cm
- Need to explain EVERY procedure, when possible, ask for consent prior to doing
- **If unable to talk => one blink = yes / two blinks = no** - Read his lips - OR - letter/word board to direct his care.

ALLERGY: Sulfa, Adhesive Tape

HEALTH ISSUES		
Neuro Muscular	ICD-9 359 MD 335.1 SMA	Spinal Muscular Atrophy Type 2, dx age 9mos, 3/74 (Severe Anterior Horn Cell disease/Werdnig-Hoffman) Incomplete quad (has full sensation), no functional movement
Pulmonary	ICD-9 V44 Trach 518.81_Resp Failure 486 Pneumo Org NOS	Respiratory failure - trach and vent (9/01), Chronic RLL Atelectasis Recurrent pneumonia, Respiratory insufficiency, poor residual functions/reserved capacities, Elective Trach 3/82 for Ortho Surgery

MEDICATIONS	HERBS / DROPS	VENT - Pulmonetic LTV 900
Rx DAILY 1. Alprazolam (xanax) 0.5 mg QID anxiety 2. Aspirin-Child 81 mg 1 x prevent clots 3. Temazepam 15 mg H S sleeping pill 4. DuoNeb 1 vial QID nebulizer (Ipratropium, Bromide & Albunterol) Rx MONTHLY 1. Thiamine 100 mg monthly vitamin 2. Cyanocobalamin 1000 mcg/ml monthly (B12) Rx PRN 1. Darvocet-N pain 2. Zithromax SUS PFIZ 200/5ml 45ml antibiotic 3. Diphnoxylate/atropine 1-2 tablets diarrhea	1. Lymphatic 5 x2 2. Flu Balancing 10 x2 3. Respiratory 7 x2 4. Allertox –airborne 5 x2 5. " " Aleer-Total 3 x3 6. " " Allerdrain 10 x4 7. Immune 6 x2 8. Acute Rescue 5 x2 9. Urinary 8 x2 10. Digestive 3 x2 11. Mucous 5 x2 12. Cell 7 x2 13. Muscular 4 x2 14. Integumentary 8 x2 15. Er Cheng Tang 1 tsp x2	Breaths 05 Tidal Volume 310 Inspiration 1.1 Pressure Support 13 Sensitivity 02 High 40 Low 02 TRACH: Shiley 6 cuffed (deflated) SPEAKING VALVE: Passy-Muir PMV007 OXYGEN 1.5 liters

MEDICAL HISTORY		
GI		Decreased esophageal motility, s/p feeding
	SURGERY	
	ICD-9 V44.1	Gastrostomy tube, Foley 24Fr Age 10, 7/83, Cincinnati Children's Hospital: Dr. Martin
Ortho	ICD-9 737.4 ICD-9 754.89 ICD-9 754.81	Severe deformities: thoracic, pelvic obliquity, bilateral dislocated hips, flexion contractures, pectus excavatum
	SURGERY	
	ICD9- 81.0	Spinal fusion /Lueke Rod Age 9, 3/82, Cincinnati Children's Hospital: Dr. Alvin Crawford
Urological	ICD-9 752.51 753.3, V13.02	Undescended L testicle (since birth), Kidney Stones (3/79, 6/90); IVP (6/90) Intermittent cath: Age 28, 10/01, cath: 10 Fr
	SURGERY	
	ICD9- 752.51	Cystoscopy/left ureteral stent, Age 28, 10/01
Other	***SURGERY***	IV**:** Porta Cath (10/24/01) RIS right clavicle (PC 0603880 - lot 36HI124)

IMMUNIZATIONS						
Tetanus '85	TB '78, '87	Pneumococcal vaccine '79, '01	Flu '02	Mumps '74	Measles '74	DPT '73, '79

PHYSICIANS			
INTENSIVIST	Melvin XXXX, MD	XXX-XXX-XXXX	XXXXXXX, Ocala, FL 34482
ACUPUNCTURE	Barbara XXXXX RN, AP	XXX-XXX-XXXX	XXXXXXX, Ocala, FL 34482
PULMONOLOGIST	Robert xxx XXXXX MD	XXX-XXX-XXXX	XXXXXXX, Gainesville, FL 32608
OTHER			
BC/BS Case Manager	Debra XXXXXXX	800-392-xxxx	XXXXXXX, Ocala, FL 34482
Rx –Pharmacy	Bitting's	352-732-xxxx	XXXXXXX, Ocala, FL 34482
Dental	Yvette Gaya , DMD	352-xxx-xxxx	XXXXXXX, Ocala, FL 34482

Appendix D
Transitions—Changing Role for Youth

Health & Wellness 101 The Basics	Yes I do this	I want to do this	I need to learn how	Someone else will have to do this—Who?
1. I understand my health care needs, and disability and can explain my needs to others.				
2. I can explain to others how our family's customs and beliefs might affect health care decisions and medical treatments.				
3. I carry my health insurance card every day.				
4. I know my health and wellness baseline (pulse, respiration rate, elimination habits).				
5. I track my own appointments and prescription refill expiration dates.				
6. I call for my own doctor appointments.				
7. I call in my own prescription refills.				
8. Before a doctor's appointment I prepare written questions to ask.				
9. I know I have an option to see my doctor by myself.				
10. I carry my important health information with me everyday (i.e.: medical summary, including medical diagnosis, list of medications, allergy info., doctor's numbers, drug store number, etc.).				
11. I have a part in filing my medical records and receipts at home.				
12. I pay my co-pays for medical visits.				
13. I co-sign the "permission for medical treatment" form (with or without signature stamp, or can direct others to do so).				
14. I know my symptoms that need quick medical attention.				
15. I know what to do in case I have have a medical emergency.				
16. I help monitor my medical equipment so it's in good working condition (daily and routine maintenance).				
17. My family and I have a plan so I can keep my health care insurance after I turn 18.				

Appendix E
Transitions—Changing Role for Families

Health & Wellness 101 The Basics	Yes my child/ youth can do this	I want my child/ youth to do this	I need to learn how to teach my child/ youth	Someone else will have to do this for my child/ youth—Who?
1. My child/youth understands his/her health care needs and disability and can explain needs to others.				
2. My child/youth can explain to others how our family's customs and beliefs might affect health care decisions and medical treatments.				
3. My child/youth carries his/her health insurance card with him/her.				
4. My child/youth knows his/her health and wellness baseline (pulse, respiration rate, elimination habits).				
5. My child/youth tracks appointments and prescription refill expiration dates.				
6. My child/youth call to make his/her own doctor appointments.				
7. My child/youth calls in his/her prescription refills.				
8. Before a doctor's appointment my child/ youth prepares written questions to ask.				
9. My child/youth is prepared to see the doctor by him/herself.				
10. My child/youth carries his/her important health information everyday (i.e.: medical summary, including medical diagnosis, list of medications, allergy info., doctor's /drug store numbers, etc.).				
11. My child/youth helps file medical records and receipts at home.				
12. My child/youth pays co-pays for his/her medical visits.				
13. My child/youth co-signs treatment form (with or without signature stamp, or can direct others to do so).				
14. My child/youth knows his/her symptoms that need quick medical attention.				

Appendix E
Transitions—Changing Role for Families *(continued)*

Health & Wellness 101 The Basics	Yes my child/ youth can do this	I want my child/ youth to do this	I need to learn how to teach my child/ youth	Someone else will have to do this for my child/ youth—Who?
15. My child/youth knows what to do if they have a medical emergency.				
16. My child/youth knows how to monitor medical equipment so it's in good working condition (daily and routine maintenance).				
17. My child/youth and I have discussed a plan to be able to continue health care insurance after they turn 18.				

Appendix F
Prep for the Doctor Visit

GOALS: To improve patient reporting of health issues and symptoms
To improve skills in health care management/self-monitoring

Teach children & youth (and family) how to:

1. Take charge in providing symptoms and concerns
2. Report by body systems; learn which ones are critical to monitor health status, report improvement or change
3. Report comparing "norm" baselines; when to call to doctor increase/decrease rates
4. Prepare 5 questions prior to visit. Provides practice talking to the doctor (enter in the body system).

PREP FOR DOCTOR VISIT—5 Qs—GENERAL MULTISYSTEM EXAMINATION

REASON FOR TODAY'S VISIT

_____ Routine Checkup

_____ Specific Health Issue or Need: What is it? _____

		RECENT TESTS	REFERRAL NEEDED
TESTS: BLOOD WORK TESTS:			

BODY SYSTEMS—HISTORY, UPDATES & QS	CHANGE + / -	RECENT TESTS	REFERRAL NEEDED
CONSTITUTIONAL			
EYES			
EARS, NOSE, MOUTH AND THROAT			
NECK			
RESPIRATORY			
CARDIOVASCULAR			
CHEST (BREASTS)			
GI (ABDOMEN)			
GENITOURINARY			
LYMPHATIC			
MUSCULOSKELETAL			
SKIN			
NEUROLOGIC			
PSYCHIATRIC (MENTAL HEALTH)			
OTHER ISSUES:			

Developed by Patti Hackett, MEd-HRTW National Resource Center

Appendix G
Getting What You Need at Your Doctor Visit

When you make an appointment to see your doctor, whether if it's for a routine checkup or for a new health issue, there is more to do than just call the office to schedule the appointment. Planning ahead and being prepared can help lower your stress and make your visit more productive, both for you and your doctor.

BASIC IDENTIFICATION (ID)—ALWAYS CARRY WITH YOU

- State issued identification card
- Health Insurance Card(s)—private and Medicaid/Medicare
- Updated portable medical summary/emergency information sheet (1 page)
- Signature stamp (if you have trouble writing your name)

Before Your Appointment—Medical Information and Health Insurance

MEDICAL INFORMATION FOLDER/BAG—TAKE TO THE DOCTOR

Sometimes it's easier to have things organized ahead of time. Some folks have a plastic folder or canvas bag already packed with items they will need for their appointments.

- Copies of items listed in "Basic ID"
- Any new test results you have received, list of questions
- Money for co-pay, parking, taxi and phone calls

PREPARE A HEALTH FILE—KEEP AT HOME IN A SAFE PLACE

While your doctor and hospital will create and keep a file on you that has your medical information, immunization records and hospitalizations, what happens when YOU need information from these files when the office is closed or when you are out of state? It's a good idea for YOU to have a duplicate file at home. Here's what you need to file in a safe place. If you do not have these items ask your doctor for help in getting copies.

FILE 1—PERSONAL HEALTH INFORMATION

- Copy of your health insurance card(s) front and back
- Copy of state issued identification card (drivers' license or state ID card)
- Portable medical summary/emergency medical information sheet (1 page)

FILE 2—RECORDS AND TEST RESULTS

- Immunization records
- Copies of medical tests, blood work, height/weight chart and other test results

FILE 3—RECEIPTS

- Receipts from insurance company of bills they have paid or reimbursed you
- Receipts from out-of pocket medical expenses: co-pays for office visits, prescriptions, equipment and other related items (may be tax-deductible)

FILE 4—MEDICAL HISTORY & NOTES

- Copies of hospitalization discharge summaries
- Copies of discharge summaries from any specialists

FILE 5—NOTES FROM YOUR DOCTOR

- Copies of progress notes from your doctor
- Copies of letters that your doctor writes to other doctors about your health
- Ask your doctor to cc you on these letters

FILE 6—CORRESPONDENCE

- Copies of letters you have written: medical justification documentation, appeals to insurance companies, requests for services and other medically related correspondence
- Replies to your letters paper clipped or stapled to your letter

GETTING THE APPOINTMENT THAT WORKS FOR YOU	
PLAN TO MAKE AN APPOINTMENT	Before calling for an appointment, look at your schedule to see what dates/times work best for you—think about not missing school, work and arranging transportation. Write down dates/times that are good and ones that would not work due to something else being scheduled. Tell the scheduler your preferred dates/times.
CALL & GET THAT APPOINTMENT	When you talk to the person who schedules the appointment, tell him/her dates and times that work best for you and request an appointment time that you know you can be there.
MEDICAL TESTS	Sometimes during routine visits your doctor will want to order some tests. When you make this appointment ask if you will be having blood work or a urine specimen and, if so, should you eat or not before the visit. Some tests can't be done if you have eaten.
URGENT VISITS	While you may have routine appointments for "well visits," there may come a time when you are not feeling well and need to be seen the same day or as soon as possible. When you get connected to the person in charge of scheduling appointments you will need to identify that you have an urgent medical issue that requires you to be seen by the doctor today or as soon as possible. The office receptionist may ask you what the problem is—sometimes the doctor or nurse will call and talk to you and then decide if you need to come into the office or if a prescription can be called in to your pharmacy. ** If you have an emergency that is life threatening, go to the emergency room immediately. If you have time en route call your doctor's office and let them know you are going to the ER and why.
TRANSPORTATION	How are you getting to the doctor's office? Are you driving, or is someone else? Are you taking public transportation (cab, bus, subway)? Figure out how much time it will take to get to the office, then add 30 minutes to make sure you won't be late. Bring along money for parking or transportation fares. Remember it is better to arrive early than be late. (Some people call the office ahead of time to see if the doctor's appointments are running behind and then adjust when they will arrive.)

DAY OF YOUR APPOINTMENT	
BE ON TIME	Figure out how you will get to/from your doctor's visit, then allow some extra time (for example 30–60 minutes) for travel. Remember that your appointment time is when you need to actually be in the doctor's waiting area, already signed in—not the time that you arrive at the front door of the building or medical center. YOU ARE LATE—Always call ahead if you are unexpectedly running late (for example, due to transportation problems). DOCTOR IS LATE—It's okay to call the office and see if the appointments are running on time or if there has been a major delay due to an unexpected emergency. You can decide whether to reschedule or adjust your arrival time. Some people use this time to do homework, write letters or read.
HEALTH INFO	Bring your health insurance card, state-issued identification, updated portable medical summary, and signature stamp if you need one. See sample HRTW Portable Medical Summary—1 page (www.hrtw.org).
PAPERWORK	If this visit was a referral from your primary doctor, make sure to bring a copy of the referral (insurance companies require this).
MONEY/CHECK	Bring money or check to pay for transportation, phone calls, office co-pays that are not covered by your insurance.
PERSONAL CALENDAR	If you need to make another appointment, it is helpful to know the dates/times that are good for you. Dates can be written or logged in paper calendars, appointment books or PDAs.
MEDICAL UPDATE & QUESTIONS	Your updated portable medical summary will list any new medications or herbs, names of doctors you are seeing and other important medical information. Prepare a list of questions and concerns you would like to discuss with your doctor. It's easy to forget things once you are in the examining room. Write down, tape record or program your questions onto your communication device or ask someone to help you do this.
WHAT TO WEAR	Wear clothes and shoes that will make it as easy as possible for you to get undressed for your examination and dressed afterwards.

YOUR APPOINTMENT

- **ON YOUR OWN or NOT?** Decide ahead of time if your parent and/or guardian will come into the room with you, and let the office staff know this when you arrive for your appointment.
- **WHAT DO YOU NEED?** Speak up for the accommodations that you need, if they are not offered to you. For example, if you cannot get onto the examining table by yourself, there should be an adjustable height table in the room or staff to assist you. Your family members should not be expected to do this. If you cannot stand on the scale, there should be an alternate type of scale available. Not getting weighed is not an option and could possibly be bad for your health. If you are incontinent, be sure that appropriate bathroom facilities are available—lying on the floor is not an option! If you have requested an interpreter, always call a day or two beforehand to confirm that the interpreter will be present. Do not agree to let family members or friends interpret for you.
- **QUESTIONS NEEDING ANSWERS**—Let your doctor know that you have some questions to ask during your visit. Make sure your doctor communicates with you in a way that you can understand. Tell the doctor if he or she is using medical jargon or vocabulary that is hard for you to understand.
- **WHAT'S HAPPENING?** Keep asking questions—What are you doing to me? What is the name of that instrument? What test have you ordered and the reason for it? When will you have the results? Are there risks? What are my options? What are the side effects of the medicine you want me to take? When do I need to see you again and why? How can I reach you if I have questions or problems?

Web Sites Tips for Taking Charge of Your Health

INSTITUTE FOR COMMUNITY INCLUSION
www.communityinclusion.org
Offers transition manual for free download, includes section on health transitions, with fact sheets for families and YSCHN. "Taking Charge of Your Health Care" and "Communicating with Doctors and Other Health Care Providers."

KIDS AS SELF ADVOCATES (KASA)
www.fvkasa.org/health.asp
Written by and for teens and young adults with special health care needs, includes helpful articles such as "Communicating with Your Doctor," "Keeping a Health Diary," and "Transition to Adulthood." Lots of helpful resources, links, listserv and opportunities to get involved.

HEALTH CARING CARDS
www.savethepatient.org
Offers "Health Caring Cards" in five languages, available for free download. Designed to be printed out, filled in and folded down to pocket size. Includes rights of patients, getting ready for doctor visits, questions to ask at your appointment, questions about medicines for your doctor and pharmacist, and emergency information card.

VIRTUAL HOSPITAL
www.vh.org/adult/patient/familymedicine/prose
Information site for providers and patients offers "Communicating with Doctors: It's Their Job to Listen" as part of "Health Prose—One Minute Update for Your Health" feature. Many resources and links related to all aspects of health care.

Tip sheet developed by Patti Hackett, Glen Gallivan, KASA and Faye Manaster, 2003, revised 2007.
The HRTW National Resource Center is headquartered at the Maine State Title V CSHN Program and is funded through a cooperative agreement (U39MC06899-01-00) from the Integrated Services Branch, Division of Services for Children with Special Health Needs (DSCSHN) in the Federal Maternal and Child Health Bureau (MCHB), Health Resources and Services Administration (HRSA), Department of Health and Human Services (DHHS). Activities are coordinated through the Center for Self-Determination, Health and Policy at the Maine Support Network. The Center enjoys working partnerships with the Shriners Hospitals for Children and the KY Commission for CSHCN. HRSA/MCHB Project Officer: Elizabeth Mcguire, of the HRSA/MCHB, Rockville, MD. The opinions expressed herein do not necessarily reflect the policy or position nor imply official endorsement of the funding agency or working partnerships.

Appendix H
Elements of Effective Transition: Health & Life[a]

LIFE AREA	QUESTIONS TO ASK
Education **Community** **Recreation** **Employment**	• Has the illness necessitated any special accommodations? • Were certain times of day to perform a health care routine? • Did the illness affect attendance? • Did medication affect ability to concentrate or participate? (i.e., was the person more alert at certain times of the day, were frequent breaks required to take medications or rest?) • Was extra time necessary to complete tasks? • Was technology, such as computers, used in and out of the home helpful to complete tasks or communicate? • Was on-site assistance required such as a person to take notes?
Impact of Health Issues **Medical**	• Are any activities restricted? • Does the person require specialized medical care (for example, dialysis)? • Does the person require the coordinated care of many health care providers? • Is there a care routine, which must be done at a specific time of day? • Does the person have a care routine, which can only be done by a specially trained individual, such as a physical therapist, respiratory therapist or nurse? • Is there a medication schedule, which must be strictly adhered to? • Are required drugs difficult to find? • Is the care of a medical specialist required? How frequently? • Are there life-threatening allergies? (latex, nuts, prescription drugs, etc.)
Environmentally	• Do certain environmental factors such as heat, cold, molds, dust, odors, and humidity affect the student's health and well-being? • Does the person need to limit exposure to noise and distractions? • Does the person require a special living environment? • Are certain activities difficult? (i.e. walking long distances/climbing stairs)
Activities of Daily Living	• Is assistance getting out of bed required? • Is assistance with food preparation/eating needed? • Does the person require a special diet? • Does the person need assistance bathing or using the bathroom? • Is assistance with dressing or other personal care necessary? • Is assistance with mobility required?

[a] Adapted from an excerpt—resource paper written by the Adolescent Employment Readiness staff for the HEATH Resource Center, fall 1997.

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American Academy of Pediatrics

Developmental and Behavioral Pediatrics

Section on Developmental and Behavioral Pediatrics

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90000>
9 781581 102741