Acquired Aphasia in Children

Acquisition and Breakdown of Language in the Developing Brain

NATO ASI Series

Advanced Science Institutes Series

A Series presenting the results of activities sponsored by the NATO Science Committee, which aims at the dissemination of advanced scientific and technological knowledge, with a view to strengthening links between scientific communities.

The Series is published by an international board of publishers in conjunction with the NATO Scientific Affairs Division

A Life Sciences **B Physics**	Plenum Publishing Corporation London and New York
C Mathematical and Physical Sciences **D Behavioural and Social Sciences** **E Applied Sciences**	Kluwer Academic Publishers Dordrecht, Boston and London
F Computer and Systems Sciences **G Ecological Sciences** **H Cell Biology** **I Global Environmental Change**	Springer-Verlag Berlin, Heidelberg, New York, London, Paris and Tokyo

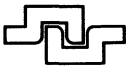
NATO-PCO-DATA BASE

The electronic index to the NATO ASI Series provides full bibliographical references (with keywords and/or abstracts) to more than 30000 contributions from international scientists published in all sections of the NATO ASI Series.
Access to the NATO-PCO-DATA BASE is possible in two ways:

– via online FILE 128 (NATO-PCO-DATA BASE) hosted by ESRIN, Via Galileo Galilei, I-00044 Frascati, Italy.

– via CD-ROM "NATO-PCO-DATA BASE" with user-friendly retrieval software in English, French and German (© WTV GmbH and DATAWARE Technologies Inc. 1989).

The CD-ROM can be ordered through any member of the Board of Publishers or through NATO-PCO, Overijse, Belgium.



Series D: Behavioural and Social Sciences - Vol. 60

Acquired Aphasia in Children

Acquisition and Breakdown of Language in the Developing Brain

edited by

Isabel Pavão Martins
Centro de Estudos Egas Moniz,
Lisboa, Portugal

Alexandre Castro-Caldas
Centro de Estudos Egas Moniz,
Lisboa, Portugal

Hugo R. van Dongen
Academic Hospital Rotterdam,
The Netherlands

and

Anne van Hout
Université Catholique de Louvain,
Belgium

Springer Science+Business Media, B.V.

Proceedings of the NATO Advanced Research Workshop on
Acquired Aphasia in Children:
Acquisition and Breakdown of Language in the Developing Brain
Sintra, Portugal
13–15 September 1990

Library of Congress Cataloging-in-Publication Data

Acquired aphasia in children : acquisition and breakdown of language in the developing brain / edited by Isabel Pavão Martins ... [et al.].
p. cm. -- (NATO ASI series. Series D, Behavioural and social sciences ; vol. 60)
"Published in cooperation with NATO Scientific Affairs Division."
Based on a workshop held in Sintra, Portugal, Sept. 13-15, 1990.
Includes index.
ISBN 978-94-010-5588-8 ISBN 978-94-011-3582-5 (eBook)
DOI 10.1007/978-94-011-3582-5
1. Aphasic children--Congresses. 2. Language acquisition--Congresses. I. Martins, Isabel Pavao. II. North Atlantic Treaty Organization. Scientific Affairs Division. III. Series: NATO ASI series. Series D, Behavioural and social sciences ; no. 60.
[DNLM: 1. Aphasia, Acquired--congresses. 2. Aphasia, Childhood--congresses. 3. Brain--physiopathology--congresses. 4. Language Development Disorders--congresses. WL 340.5 A1865]
RJ496.A6A29 1991
618.92'8552--dc20
DNLM/DLC
for Library of Congress 91-20796

ISBN 978-94-010-5588-8

Printed on acid-free paper

FOREWORD

One of the most fascinating problems in Behavioural Neurology is the question of the cerebral organization for language during childhood. Acquired aphasia in children, albeit rare, is a unique circumstance in which to study the relations between language and the brain during cerebral maturation. Its study further contributes to our understanding of the recovery processes and brain plasticity during childhood. But while there is a great amount of information and experimental work on brain–behaviour relationships in adult subjects, the literature about the effects of focal brain lesions in children is both exiguous and scattered throughout scientific journals and books.

We felt it was time to organize a meeting where scientists in this field could compare their experiences and discuss ideas coming from different areas of research. A workshop on Acquired Aphasia in Children was held in Sintra, Portugal, on September 13–15, 1990, and attended by 44 participants from 13 differents countries. The atmosphere was relaxed and informal and the group was kept small to achieve this effect. It was a very lively and pleasant meeting. Some consensus was indeed arrived at concerning methodological problems, definition of terms, and guidelines for future research.

The main contributions are collected in this book which, we hope, will serve the scientific community as a reference work on Childhood Aphasia.

I.P.M., A.C.C.

ACKNOWLEDGMENTS

First and foremost, I would like to thank Dr. Giovanni Venturi and the Scientific Affairs Committee of NATO, which he directs, for the backing and financial support received for this workshop on Acquired Aphasia in Children, held in Sintra, Portugal, during September 1990. The means awarded by NATO supported both the workshop itself and the expenses involved in the publication of this volume. Supplementary contributions to the meeting were made by the Portuguese INIC (Instituto Nacional de Investigação Cientifica) and JNICT (Junta Nacional de Investigação Científica e Tecnológica), to who we are also grateful. Thanks must also go to Miss Elisabete Mendes, who typed parts of this manuscript and gave secretarial support to the Workshop. Maria Clara Pires, Maria de Fátima Félix, and Rosalina Sá of the Library, Centro de Estudos Egas Moniz, were also very helpful.

Lastly, we would like to thank all participants for the high quality of their scientific contributions. The friendly atmosphere generated throughout the workshop, augurs well for future joint research.

I.P.M.

CONTRIBUTORS

Dorothy Aram
Department of Pediatrics,
Case Western Reserve University and
Rainbow Babies and Children's Hospital
Cleveland, Ohio 44106
U.S.A.

A. Castro Caldas
Laboratório de Estudos de Linguagem
Centro de Estudos Egas Moniz
Hospital de Santa Maria
1600 Lisboa
PORTUGAL

Michel Dugas
Service de Psycho Pathologie
de l'enfant et de l'adolescent
Hopital Robert Debré
48, Boulevard Sérurier
75935 Paris Cedex 19
FRANCE

Julie Eisel e
Department of Pediatrics,
Case Western Reserve University and
Rainbow Babies and Children's Hospital
Ceveland, Ohio 44106,
U.S.A.

José M. Ferro
Centro de Estudos Egas Moniz
Hospital de Santa Maria
1600 Lisboa
PORTUGAL

C. L. Gerard
Service de Psychopatologie de
de l'enfant et de l'adolescent
Hopital Robert Debré
48, Boulevard Sérurier
75019 Paris Cedex 19
FRANCE

Hallgrim Klove
University of Bergen
Dept. Biological and Medical Psychology
Division of Clinical Neuropsychology
Anstadveien 21
N 5009 Bergen
NORWAY

William Landau
Dept. of Neurology
Washington University
School of Medicine
660 South Euclid Avenue
St. Louis
Missouri 63110
MO, U.S.A.

Christa B. Loonen
Dept. of Neurology
Academic Hospital Rotterdam - Dijzigt
40 dr. Molewaterplein,
3015 CD Rotterdam
THE NETHERLANDS

Virginia Marchman
Dept. of Psychology C-009
University of California
San Diego
La Jolla, California 92093,
U.S.A.

Isabel P. Martins
Centro de Estudos Egas Moniz
Hospital de Santa Maria
1600 Lisboa
PORTUGAL

Ph. Paquier
Div. of Neurolinguistics
Universitair Ziekenhuis
Antwerpen
Wilrijkstraat 10
b-2520 Edegen
BELGIUM

Isabelle Rapin
Saul R. Korey Depatment of Neurology
Department of Pediatrics
Division of Child Psychiatry
Rose F. Kenneddy Center for Research in
Mental Retardation and Human Development
Albert Einstein College of Medicine
Bronx, New York 10461
U.S.A.

Daria Riva
Dept. Child Neurology
Istituto Neurologico C. Besta
Via Celoria 11
20133 Milano
ITALY

Paul Satz
Neuropsychiatric Institute and
Hospital Centre for Health Sciences
University of California, Los Angeles
760 Westwood Plaza
Los Angeles,
California 90024-1759
U.S.A.

H. R. Van Dongen
Dept. of Neurologie
Academic Hospital Rotterdam
Dijkzigt
40 dr. Molewaterplein,
3015 CD Rotterdam,
THE NETHERLANDS

Anne Van Hout
Service de Neurologie Pediatrique
Universite Catholique de Louvain
Cliniques Universitaires Saint-Luc
Avenue Hippocrate 10, UCL 10/1303
1200 Bruxelles
BELGIUM

E. Wijngaert
Royal Institute for Children with Communication
Disorders
Borggravevijverstraat 9,
3500 Hasselt,
BELGIUM

Bryan T. Woods
Department of Neurology, and Brain Imagings Centre
McLean Hospital,
115, Mill Street,
Belmont, MA 02178-9106
U.S.A.

TABLE OF CONTENTS

IV – HEMISPHERIC SPECIALIZATION

V – CLINICAL FEATURES OF ACQUIRED APHASIA IN CHILDREN

VI – PROGNOSIS FOR RECOVERY OF ACQUIRED APHASIA IN CHILDREN

VII – LATE SEQUELAE OF EARLY BRAIN LESIONS

VIII – LANDAU AND KLEFFNER SYNDROME

I-INTRODUCTION

INTRODUCTION

Acquired aphasia in children is fortunately rare. But if its rarity is a wisdom of nature, it also embarasses researchers on their will to understand the brain. For indeed each case is an "experiment de nature" and thus an unique source of information for those interested in the problems of brain/behavior and brain maturation/development relationships.

The interest on acquired aphasia in children started about a century ago with the work of Cotard (1868), Bernhardt (1885) and Freud (1897). In the following 50 years, and apart from scattered reports published in scientific journals describing either single cases or small series, little attention was paid to this matter. Early authors were particularly struck by the differences between child and adult cases, namely in what concerns their clinical presentation (stereotyped in children as consisting of transient nonfluent aphasia with good verbal comprehension) and their prognosis (deemed to be excellent). Such differences were attributed to the immature cerebral organization of the child's brain, and to the ability of its right hemisphere to take up language function after left hemisphere damage.

In the 1960's interest on this subject was revived, mainly through the work of Basser (1962), Alajouanine and Lhermitte (1965), Lenneberg (1967), and later Hécaen (1976), who published their own much larger series of children with acquired aphasia. For the first time children were assessed by standardized neuropsychological tests, (evaluating not only language but also non verbal functions, as well as intelligence), and the series were studied longitudinally. Long term consequences of aphasia on cognitive development and school achievement were now a focus of concern.

A major turning point, concerning the study of language functions' localization in the developing brain, was settled by the papers of Woods and Teuber (1978), and later by Carter et al (1982). These authors critically reviewed all previous reports (particularly in what concerns patient selection, etiology, handedness and unilaterality of lesion) and went on to demonstrate that the developmental hypothesis of hemipheric dominance was no longer tenable. This agreed well with evidence coming from other sources, such as neuroanatomy, neurophysiology, dichotic listening studies and studies of hemispherectomy.

In the last 10 years, new methods and techniques considerably enriched our understanding of childhood aphasia. Neuroimaging techniques partially solved the problem of determining lesions' localization, which in turn made clinico-pathological correlations possible.

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 3–12.

Neuropsychological, psychometric and neurolinguistic studies shed new light on cognitive processes in children. They also showed that early brain lesions were not as reversible as previously thought. Comparison of right versus left, and early versus late, hemispheric lesions also expanded our understanding of cerebral organization and its plasticity during childhood.

However, many issues remain controversial, poorly understood, or not formulated at all. Some of these issues will be discussed in the following chapters. Others will be merely formulated here.

1. The normal pattern of language acquisition. Linguistic and cognitive skills underlying such acquisition.

In order to analyse language and speech defects following a cerebral lesion, one must have a paradigm of normality. In the case of children this must portray normal language development at is different stages. Traditionally, language acquisition was represented on a linear time scale of major sequential achievements. But this was really only the tip of the iceberg. The cognitive and linguistic skills underlying the observable performance were unknown.

The paper by Marchman, in this volume, shows that it is difficult to define universal rules for language acquisition. There are wide variations in rate, strategy, and style, among normally developing children. Furthermore this acquisition is much more proctracted than previously thought. Children make use of syntactical forms that are different from the ones used by adults, although equally pragmatic. Such forms possibly express alternative cognitive/linguistic strategies that, in turn undergo progressive reorganizations and refinements during development.

This knowledge may prove essential for understanding some of the clinical features of acquired childhood aphasia and the functional processes of aphasia recovery. It will also help us understand differences between child and adult aphasia which, up to now have been abusively explained by a reductionist use of theories of cerebral organization.

2. Similarities and differences between child and adult acquired aphasia.

In articles published here, Martins and Ferro, Van Hout and Van Dongen and Paquier, describe in detail the clinical features of acquired aphasia, based on the personal observation of a large number of cases. It is apparent from these reports that aphasia in children is indeed very similar

to its counterpart in adults. All types of aphasia are described. Speech can be fluent, ranging from normal fluency to jargon. There are cases with auditory comprehension, or language repetition, defects. "Positive" signs are not uncommon, namely paraphasias, neologisms, perseverations, palilalia, "conduite d'approche" and anosognosia for speech defects.

There are, however, some features in children's aphasia that clearly stand out in contrast to those common in adults. Such is the case, for instance, for mutism and nonfluent types of speech, and for the lack of comprehension disorders found specially in the very young. Indeed, fluent aphasia is rare or absent in children under 3 years of age (if we exclude cases of Landau and Kleffner syndrome). This suggests some influence of age on clinical presentation. In order to be able to confirm such an influence of age one nevertheless needs to control several interveening factors and variables which could, by themselves, account for these findings.

The first difficulty concerns the analysis of speech in very young children. This is not an easy matter. A lot depends on their will to cooperate, something which can be difficult to secure from an acutely ill child. In addition, a properly controlled analysis requires specific tests designed for this age group, if meaninful quantitative data is to be gathered. Techniques of language and speech assessment in children, which have been refined in the last years, are fully described in the papers authored by Rapin and Alen, and Aram.

Second, one needs to take into account variables such as etiology and lesions' site and size. These do influence the clinical presentation. Moreover, these variables tend somehow to cluster, producing a bias in series which include mainly one type of pathology. Third, one must be careful about the selection of patients. As the papers by Woods and Ferro show in considerable detail, the inclusion of patients with bilateral lesions, or severe epilepsy, can further complicate the intrepertation of results.

Since acquired aphasia in children is a rare disease, major conclusions can better be drawn from a comparison of as large a sample of studies as possible. These studies should, ideally, control different variables and factors such as age of onset, site/size of lesion, etiology and time of examination, to list the most obvious ones. Only such detailed information will really enable future progress.

3. The neural basis of language organization during childhood.

Studies of acquired aphasia in children which include a clinico-anatomical correlation have so far shown that:
a) Language function is usually lateralized to the left hemisphere during childhood, and
b) The organization of the different language functions within the left hemisphere is identical in children and adults.

Indeed, both the most recently published series (Cranberg et al, 1987, Van Dongen et al, 1989, Van Hout, 1985) and the papers included here, showed that prerolandic and subcortical lesions are usually associated with nonfluent types of speech with normal auditory comprehension, while temporal lesions produce a fluent paraphasic speech with poor comprehension. This symptom pattern is exactly the one expected in adults with similarly located lesions.

On the other hand, it is difficult to understand, in terms of cerebral organisation, the symptoms that are peculiar to aphasia in this age group, such as the tendency for mutism and nonfluent speech. These symptoms can have several different explanations: a) They can merely result from particular patterns of lesions localization. Indeed, in the study of Martins and Ferro(in this volume), both mutism and nonfluent types of speech were found in association with frontal and subcortical lesions, which is the pattern one would expect to find in adults with such symptoms. Incidentally, this is also the explanation for the predominance of certain types of aphasia in young adults with stroke (Ferro and Crespo, 1988). ; b) Such symptoms could be due to distant metabolic effects secondary to the focal lesion. The techniques of regional metabolic assessment (SPECT, PET) have shown that, both in adults and children (Sahar et al,1990), dysfunctional areas are much larger than the areas of lesion; c) An alternative explanation is that the organization of some aspects of language within the left hemisphere, is different between children and adults. Namely, the neuronal networks subserving speech initation and fluency can be less localized in children; d) Finally, it can also happen that the pathological manifestations of a developing function are different from those found during the breakdown of that function after its full maturation. These alternative explanations can only be confirmed or rejected in future by a careful selection of patients and a more extensive imaging investigation.

In addition to the studies of acquired aphasia, other sorts of evidence would be welcome to complete our knowledge about language

representation in the brain. One of them is the inclusion of negatives cases (i.e. children with focal brain damage, whose lesions neither disrupt language functions acutely, nor impair the future acquisition of specific language skills). This sort of evidence could support arguments against theories defending a widespread/diffuse language representation in the developing brain.

But, all the above mentioned studies only help us understand cerebral functioning under pathologic conditions. To move foward it is necessary to supplement this information with studies of normal children. Woods, in his paper here, sheds some optimism in this new area of research by pointing out which methods and techniques will enable us to study brain functions in healthy subjects. These may help us, in future, not only to understand which neural structures subserve the normal acquisition of language, but also to know what is the structural basis underlying the different patterns of language acquisition.

4. The prognosis of acquired aphasia and the neural basis of aphasia recovery.

This issue can be divided in several questions.

4.1. The prognosis of aphasia acquired in childhood.

Three papers in this volume deal with this specific matter (the articles of Van Hout, Loonen and Van Dongen and Martins and Ferro). They revise previous studies and present their own series of aphasic children who were followed for variable periods of time.

All authors seem to agree that the prognosis of aphasia is better in children than in adults. One year after onset, 50% or more of all reported children had completely recoverd. Furthermore, all conclude that there are many factors which influence the prognosis. Such factors are the etiology of the lesion, its size, site and bilaterality, the nature of the aphasic symptoms, and the child's age.

On the other hand, if wee look at specific details, such as the influence of each of these factors in the prognosis, conclusions begin to diverge. Such results are not surprising if we realize that we are dealing with a rare entity. All series reported necessarily include only a small and heterogeneous number of cases and the series are not really comparable with each other. Just to state a few examples: a) the time of follow up is not always identical. Since these children may continue to recover for long

periods of time, this single fact could explain the different results; b) some series include children with progressive or fluctuating disorders, which compromise the prognosis; c) some of the etiologies included are known as capable of causing diffuse or bilateral damage, which may interfere with possible recovery mechanisms.

As has been stated about the clinical features of aphasia, only a large or a cooperative study, using very strict inclusion criteria, could possibly allow for a more homogeneous group of patients from which more firm conclusions could be drawn concerning the prognosis.

4.2. Does the ability to recover depend upon the child's age?

Since there is an overall difference in the prognosis between adults and children, one might logically expect to find a gradient of recovery related to age. This could be either a smooth linear relation or a stepwise, discontinuous process, punctuated by major functional or structural events such as language acquisition, corpus callosum myelination, or the acquisition of written language.

However, studies of acquired aphasia have not given so far,a definite answer to this question. This is possibly due to the enormous plethora of confusing variables. Case-control studies (including children matched for etiology and site/size of lesion and having age as a single variable) could well illustrate this point. But these studies are still lacking. A cooperative study could also help answer this question.

4.3. Does the ability to recover depend upon particular lesion sites?

The establishment of a connection between lesion site and the prognosis has only recently been made possible by new imaging techniques. This, in turn, is giving rise to new theoretical inquires and hopefully new research programs. From the studies presented in this volume, two major conclusions can be drawn:

a) Bilateral/severe lesions tend to harbour a worst outcome. This, thus raises the question of the participation of the right hemisphere in the recovery process;

b)The posterior language areas of the left hemisphere, namely Wernicke's area, need to be spared for recovery to take place . This, thus raises the question of the participation of these areas in the processes of intra hemispheric reorganization. One must also take into account that these areas (whatever their role in the processes of reorganization) can be essential for providing the verbal input to the reorganizing brain. Moreover, temporal lobe lesions may affect memory, undoubtely a fundamental

element for any sort of function acquisition. Unfortunatly, and apart from a few noteworthy exceptions (Aram and Ekelman,1988), the role of memory has frequently been neglected in studies of focal brain lesions in children.

4.4. What are the neural structures subserving recovery?

As I have just mentioned, two major hypothesis have been formulated. After a left hemispheric lesion, language may be transfered to the right hemisphere or may be taken over by intact neighbouring areas of the left hemisphere.These hypothesis are reviewed below by Satz, who presents corroborating evidence derived from studies other than those on acquired aphasia: studies of hemispherectomy, of the amital test in epileptics, dichotic listening studies, or the studies of aphasia following a lobectomy. All studies demonstrate that language can be transfered to the right hemisphere and this seems more likely to be carried out the earlier the lesion. However these studies are performed in subjects with poorly localized or ongoing disease (i.e. seizures). The effect of stactic, well localized, lesions on such transfer remains to be determined. Cerebral metabolic studies performed in children who recovered from aphasia can surely help us answer this question. Similarly, rare cases of sequential hemispheric lesions, as the case described by Castro-Caldas,also illustrate this point.

5. What are the late effects of early brain lesions?

Well beyond its interest in childhood aphasia, research about early cerebral lesions has been concerned with its late effects on cognitive and linguistic skills as well as school achievement. Evidence in this area comes from different sources, namely, 1) the study of children with congenital focal brain lesions; 2) the study of children with focal lesions acquired after birth and during the first year of life (before language acquisition); and 3)studies of children who recovered from acquired aphasia. Although these sorts of evidence are complementary of each other, they must be clearly distinguished. They represent different stages of cerebral maturation and the strategies involved in subsequent function acquisition can be different.

The papers by Riva et al and Eisele, in this volume, describe some of the cognitive, and linguistic deficits found after unilateral focal vascular lesions. Although these children are not aphasic, they show specific syntactic and semantic, or general cognitive deficits when compared to controls. Moreover these deficits clearly depend upon the side of brain lesion and upon the child's age. They point out the specific skills of each hemisphere

that cannot be overtaken by the intact areas at different stages of maturation.

The paper by Aram shows the contradictory evidence on scholastic achievement after early brain lesions. This contradiction stems from the heterogenous subject population studied. Her paper also stresses that it is necessary to know more about the linguistic and cognitive deficits underlying school failure in these children. These deficits have been little investigated apart from a few studies (Aram and Ekelman,1988). Moreover studies about the effects of age of lesion onset and lesion localization in school performance are also necessary. This is undoubtely an important field for future research.

6. The Landau and Kleffener syndrome.

This syndrome remains a misterious disorder of which only the clinical features are known. These are comprehensively reviewed by Dugas et al and by Van Dongen et al, based both on their personal series of cases and on those previously published.

This syndrome undoubtely affects restricted (focal) brain areas. Apart from aphasia, the neurological examination of these children is normal and intelligence is spared. The type of aphasia found here is quite peculiar, for it includes comprehension disorders (including auditory agnosia) and jargon even in very young children. Besides, the language disorder seems modality specific for some of these children can acquire written language. From what we know about localization in acquired childhood aphasia, we can infer that such aphasia type is most probably due to a temporal lobe lesion. A recent report of three cases studied with Pet Scan (Maquet et al,1990) corrobates such hypothesis.

Contrary to other types of aphasia in children, the prognosis here is variable but globally gloomy. It has been assumed from this lack of recovery that lesions must be bilateral. But poor recovery can just result from an ongoing pathological process.

Untill more is known about this disease, rehabilitation techniques are the only possible help to overcome the communication deficit. These are widely discussed by Gerard et al and by and Wijngaert in their papers below.

It is difficult to do justice to the richness of the contributions presented in this volume.By attempting to place them in the context, I have merely tried to touch upon some of the major issues posed.

REFERENCES

Alajouanine, TH. and Lhermitte, F. Acquired Aphasia in Children. Brain, 88, 653-662, 1965.

Aram,D.M. and Ekelman,B.L. Scholastic Aptitude and Achievement among Children with Unilateral Brain Lesions. Neuropsychologia, 26, 6 ,903-916,1988.

Basser,L.S. Hemiplegia of early onset and the faculty of speech with special reference to the effects of hemispherectomy. Brain, 85,427-460,1962.

Bernhardt, M., Urber die spastiche cerebral paralyse im Kindesalter, Nebst einem Excurse uber: "Aphasie bei Kindern". Archiv fur Pathologische Anatomie und Physiologie und fur Klinische Medicin 102, 26-80, 1885.

Carter, R.L., Hohenegger, M.K. and Satz, P. Aphasia and Speech Organization in children. Science, 218, 797-799, 1982.

Cotard, J. Étude sur l'atrophie partielle du cerveau. Paris: Thèse, 1868.

Cranberg, L. D., Filley, C. M., Hart, E. J. and Alexander, M. P. Acquired Aphasia in childhood: clinical and CT investigations. Neurology, 37, 1165-1172, 1987.

Ferro,J.M. and Crespo, M.. Young Adult Stroke: Neuropsychological Dysfunction and Recovery. Stroke,19, 982-986,1988.

Freud, S.. Infantile cerebral paralysis, Translated by L.A. Russin) Coral Gables: University of Miami Press (Original published, 1897), 1968.

Hécaen, H. Acquired Aphasia in Children and the Ontogenesis of Hemispheric Functional Specialization. Brain Lang. 3, 114-134, 1976.

Lenneberg, E. Biological Foundations of Language. New York, John Wiley, 1967.

Maquet, P. Hirsch, E., Dive, D., Salmon, E., Marescaux, C., and Franck, G. Cerebral Gluose Utilization During Sleep in Landau and Kleffner syndrome: A PET study. Epilepsia, 31 (6) 778–783, 1990.

Shahar, E., Gilday, D.L., Hwang, P.A., Cohen, E.K. and Lambert, R. Pediatric Cerebrovascular Disease. Alterations of Regional Cerebral Blood Flow Detected by TC 99m – HMPAO SPECT. Arch. Neurol., 47, 578–584, 1990.

Van Dongen, H. R., Loonen M. C. and Van Dongen K. J. Anatomical basis for acquired fluent aphasia in children. Ann. Neurol., 17, 306–309, 1985.

Van Hout, A., Evrard, Ph. and Lyon, G. On the positive semiology of Acquired Aphasia in Children. Develop. Med. and Child Neurol., 27, 231–241, 1985.

Woods, B. T. and Teuber, H. L. Changing patterns of Childhood Aphasia. Ann. Neurol. 3, 273–280, 1978.

II - NORMAL LANGUAGE DEVELOPMENT

THE ACQUISITION OF LANGUAGE IN NORMALLY DEVELOPING CHILDREN: SOME BASIC STRATEGIES AND APPROACHES

VIRGINIA A. MARCHMAN, Ph.D.
Center for Research in Language 0126
9500 Gilman Drive
University of California, San Diego
La Jolla, CA 92093-0126
USA

ABSTRACT. The field of language acquisition boasts considerable knowledge about the order and timing of linguistic achievements in the normally developing child. However, recent studies suggest that the course of acquisition is more *variable* and *protracted* than had once been thought. I overview two consequences of this shift in focus which challenge current viewpoints. First, even within normally developing populations, early lexical acquisition is marked by considerable diversity and variation. Recent studies outline normal extremes in *rate* of acquisition (e.g., late and early talkers), identifying the cognitive and linguistic skills that "hang together" or "break apart" at these ends of the normal continuum. These studies help to identify the *limits* on the form that the linguistic system can take in "normal" learning situations. Second, while young children can and do communicate quite effectively, linguistic skills *continue* to develop for many years. These protracted developments may reflect system-internal *reorganizations* in the discourse-based function of linguistic structures, e.g., the frequency of *use* of the English passive. In our view, language behavior is processing-based and dynamic, and language learning is continuous with developments in other cognitive and perceptual domains. Establishing the normal limits on individual variation and the functional basis for developmental change in linguistic performance are necessary for understanding the biological, cognitive and communicative foundations of language acquisition.

1. Introduction

Children are amazingly good at language learning. Indeed, many children begin to effectively communicate by the middle of their second year, and several basic linguistic structures are mastered with apparent ease by about four or five years of age. While detailed accounts have been offered regarding the transitions from babbling to words to sentences to stories, it has recently become more and more evident that the "normal" course of language acquisition may not be as normal as you think. In this paper, I briefly outline two ways in which standard views of acquisition have been challenged, presenting recent findings from projects conducted at the Center for the Study of the Neurological Basis of Language and the John D. & Catherine T. MacArthur Foundation Network for the Transition from Infancy to Early Childhood (San Diego). Both of these challenges

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 15–24.

derive from the fact that language usage and acquisition is *variable,* i.e., marked by considerable diversity and inconsistency across individuals and contexts.

The idea that language is variable and messy is not new. In fact, it was one of the primary assumptions put forth by Chomsky in the late 1950's. He notes that "the problem for the linguist, as well as the child learning language, is to determine from the data of performance the underlying systems of rules that have been mastered and that are put to use in actual performance" (1965, p. 3-4). As a consequence, it has proven to be less than straightforward to determine at what point children *should* acquire particular forms or structures, or even *whether or not* they have done so (see also Bates & MacWhinney, 1982; Snow, 1983; de Villiers, 1988). Over the history of the field, researchers have developed several very clever ways to minimize the mystery in the data collection process (e.g., structured elicited production, imitation, etc). However, while all are aware of the variability, there are disagreements among researchers with respect to how it should be managed, and above all, how it should be interpreted.

For some, including Chomsky, language variability is placed outside the domain of legitimate inquiry. Researchers working within this tradition are likely to seek experimental procedures that weed out those portions of linguistic behavior which are performance facts from those which truly reflect the linguistic abilities of an "ideal" speaker-hearer (e.g., Roeper, 1987). In general, these perspectives can be seen to adopt a content approach; that is, *what* do children know and *when* do they know it (cf. Bowerman, 1985). Here, the major goal of language acquisition research is to explicate the fact that children learn language, i.e., how could it be possible that children so effortlessly master such a complex linguistic system when the input is so messy and variable? (e.g., Borer & Wexler, 1987). Further, the emphasis is on establishing the relatively stable course of acquisition by identifying the *universal* biases for language learning that all speakers in all languages bring to the task.

In contrast, other perspectives do not make such a sharp division between linguistic abilities (i.e., competence) and the application of those abilities in real time (i.e., performance). These perspectives can even be seen to embrace the natural variability that is observed within and across speakers (e.g., Bates & MacWhinney, 1982, 1987). In this view, "language is created, maintained, used, and acquired in the service of communicative functions" (Bates & MacWhinney, 1982), and hence, the communicative context of language usage, as well as the diversity, creativity, and individual differences that are observed are crucial aspects of communicative and linguistic systems. From this viewpoint, acquisition research should explain how and why linguistic *behavior* changes across development: what is the r*ange* of possible forms that developing linguistic knowledge can take across individuals and how does *context* influence the ways in which knowledge is put to use in real time. In addition, this perspective assumes that children reach adult levels of communicative ability through the protracted integration of language specific, as well as general cognitive and communicative principles, into a system of working knowledge about how to effectively communicate with others.

The following sections outline two developmental consequences of the inherent variability of language acquisition and usage, and how they are managed within a functionalist perspective. The first concentrates on early milestones of acquisition, primarily early lexical development and the transition into word combinations in children ranging from 8 to 34 months. The second focuses on the changes in the frequency of passive usage in functionally defined discourse and event contexts in English speaking

children between 3 and 12 years of age. It should be noted that the emphasis on variation rather than universals does not imply the abandonment of the characterization of language as a biological system. Nor does it imply that language cannot or should not be described in formal terms. On the contrary, this approach seeks to shift the search for universals from content to *mechanism and process* (Bates & Marchman, 1988), in the attempt to provide a more feasible biological perspective for understanding language development (Bates, Thal, & Marchman, in press) and eventually, a formal characterization of language that is firmly grounded in real-time language processing. By explicitly acknowledging contextual variation and by examining the limits on the patterns that normal development can take, we hope to be in a better position to understand the limits on the plasticity that the linguistic system displays in typical and atypical learning conditions.

2. Within-language variation in lexical development

While the acquisition of language clearly must be determined by the biological endowments of the human species, recent evidence has shown that it is not the case that all children learn language in exactly the same way, according to a univeral course. There is considerable "normal" variation in the ordering and timing of the achievement of early language milestones across individuals and across languages. However, while some may disagree with the details, most would accept the following as an accurate description of the course of language acquisition (and concurrent non-linguistic developments) that is observed in the majority of normally developing English-speaking children (Bates, Bretherton, & Snyder, 1988; Bates, O'Connell, & Shore, 1987).

At about 6-9 months of age, children's vocalizations begin to shift from cooing (i.e., vowel-like sounds) to consonant-vowel sequences (i.e., canonical babbling). Systematic signs of word comprehension begin at around 9-10 months, when children start to respond to well-used words or phrases (e.g., their own name, "bottle", etc.). At this point, a child's intentional communication skills typically also includes gestural routines, e.g., bye-bye, patty-cake, etc. By 12-13 months of age, children begin to produce their first words, usually single nouns which name or request common objects. Children typically begin to produce recognitory gestures at this age as well (e.g., drinking from a cup). Over the next several months, comprehension and production vocabularies increase at a steady but relatively slow pace. Usually after about 50 words, there is a sudden burst in expressive vocabulary. Children now generally switch from naming and requesting objects with single nouns to also producing other types of lexical items, such as verbs (e.g., "go") or adjectives (e.g., "hot"). In addition, children now appear to be able to comment on the attributes of objects (e.g., "pretty") and talk about the relationships between objects (e.g, "daddy" when pointing at her father's briefcase). By this time, lexical items and gestural routines are becoming increasingly decontextualized (i.e., less bound to a particular object or situation). At this point, the first combinations typically appear, generally strings of content words that are "telegraphic" in character (e.g., "mommy sock"). In the beginning of the second year, function words and bound morphemes are added to the two- or three-word telegraphic phrases, as children are now beginning to fill in the constituent grammatical structure of their language.

It is important to emphasize that this "textbook" view of early language milestones is only applicable for the "textbook" English-speaking child. The majority of what is known about language acquisition is based on studies of English, yet recent crosslinguistic research (e.g., Slobin, 1985) has clearly shown that the language to be learned has a substantive impact on the nature of the course of acquisition. For example, children

learning Turkish (a case inflected language) are not likely to pass through a stage of telegraphic speech in which inflections and bound morphemes are omitted from their productions. In fact, Slobin (1982) reports that Turkish children typically include bound morphemes in their earliest utterances, and most Turkish children have mastered many of the basic case inflection contrasts by two years of age. In addition, children learning Italian, for example, are more likely to include verbs in their early single-word utterances (e.g., "mangia!"), not necessarily limiting their first words to object names (Bates, Caselli, & Casadio, 1990). Many other crosslinguistic counter-examples have been identified (see discussion in Bates & Marchman, 1988), and substantive progress is being made in this area.

However, even if one is in the position to evaluate only children learning English, the above textbook description must only be seen as outlining the timing and sequence of events *on the average*. These ages and milestones might be quite accurate for some children, however, others might follow a considerably different course. It is well-known, for example, that not all normally developing children produce clearly articulated single-word utterances that refer to common objects (e.g., "referential style"). Some children, in contrast, tend to produce long strings of babble that have sentence-like intonation contours. The vocabularies of these so-called "expressive style" children are typically heterogenous, comprised of both nouns and multiword phrases, as well as formulae (e.g., Nelson, 1973). These individual differences have been shown to be consistent across developmental periods, displaying themselves at every level of linguistic description, including phonology, lexical acquisition and grammar. Several hypotheses about the source of these differences have been postulated, including the child's preference for analyzed vs. unanalyzed units (e.g., "analytical" vs. "holistic"), or the tendency to parse the speech stream into small "word-sized" vs. larger "phrase-sized" units (see Bates, et al., 1988 for review).

For the researcher and clinician, it is clearly important to be aware of these sources of variation that can influence the normal course of acquisition. Without a substantive understanding of the origins of variation it would be impossible to identify acceptable ranges around the group average. That is, what degree and type of variation can be interpreted as a consequence of the natural differences between individuals versus a sign of language delay or precocity?

One first step in answering this question is to gather data from large numbers of children, across a range of social and economic groups. With this goal in mind, the MacArthur Norming project has devoted considerable energy to developing a parental report instrument on the achievement of early language milestones, primarily lexical development. To date, parental report data on vocabulary acquisition in infants (8-16 months) and toddlers (18-28 months) has been collected from approximately 1500 English-speaking children in several locations in the United States (San Diego, Seattle, Boston) sampling across several socioeconomic levels. These instruments consist of checklists of words commonly found in children's early vocabularies, as well as questions about children's progress in other communicative and non-verbal skills that have been shown to correlate with language milestones, e.g., turntaking, pointing, etc. For infants, parents are asked to indicate on the checklist which words their child "understands," and which words their child "understands and says." For toddlers, parents are simply asked to indicate which words their child "says." The parental report format in general, as well as these particular instruments, have been validated in several studies (Bates et al., 1988; Reznick & Goldsmith, 1989; Dale, Bates, Reznick, & Morisset, 1989). While parents sometimes tend to overestimate their child's achievements, these instruments have nevertheless proven to be excellent tools for assessing *relative* linguistic levels, correlating highly with laboratory production and comprehension measures which often

tend to underestimate children's abilities (e.g., Bates et al., 1988).

Results from these norming studies indicate that there is indeed considerable variation in the number of words that children at the same age are reported to comprehend and produce. For example, at 12 months of age, children typically comprehend about 71 words on average, however, that figure can range anywhere from 7 to 242. For production, a median-level 12 month old is reported to produce about 6 words, however, some children at this age haven't yet begun to produce words while others are reported to produce 50 or so. By 16 months of age, the variation in reported production vocabularies is considerable, ranging from 2 to 347 words with a median of 44. In the toddler group, variation in reported production vocabulary is still great. The average 20-month-old produces approximately 169 words, however, his/her agemates are reported to produce anywhere from 3 to 544 words! As we shall see shortly, however, the range can go even higher in special cases.

Interestingly, within-child comparisons reveal that comprehension and production levels sometimes go together (i.e., some children are either high or low in both production and comprehension), but sometimes they do not (i.e., some children are in the low range in production while either being high or low in comprehension). Again, it must be noted that these figures are only applicable for children learning English. Similar inventories have been constructed (and are currently in the process of being normed) for monolingual Spanish-speaking populations in San Diego and Mexico (Jackson-Maldonado, Marchman, Thal, & Bates, 1990) and Italian children in Rome (Bates et al., 1990). Preliminary crosslinguistic comparisons suggest, however, that within-age group ranges are similar for these language groups as well.

Recently, Donna Thal and her colleagues have followed two groups of English-speaking children who fall at the *extremes* of these normal ranges in vocabulary production (Thal and Bates, 1988a&b, 1991; Thal, Tobias & Morrison, in press). In these longitudinal studies, Early Talkers are defined as 11 to 20-month old children in the top 10th percentile in expressive vocabulary. Late Talkers are defined as 18 to 24-month old children in the bottom 10th percentile of expressive vocabulary. In order to understand the basis of these extremes, Thal and her colleagues have devoted considerable energy to exploring the non-linguistic correlates of early lexical development in these groups of children. In particular, they concentrate on identifying those linguistic and non-linguistic skills that display *associations* (i.e., that "hang together") vs. those abilities that reveal *dissociations* (i.e., "come apart"). In other words, what groups or clusters of abilities can be seen to lag behind or move ahead of other skills and abilities at these extremes in the normal population?

So far, these studies have revealed several interesting trends, however, I will mention only three briefly here. First, a cross-domain association has been identified between vocabulary production and the ability to imitate recognitory gestures. That is, children at both extremes are more likely to look like children who are at the same language level (even though they are different ages) when asked to produce single gestures modeled with toys or placeholder objects (e.g., sniff a flower/block, drink from a cup/block). Late talkers are delayed in recognitory gesture production, and early talkers are advanced. Thus, the relationship between symbolic gesture and vocabulary growth that has been identified in several studies (e.g., Bates, et al., 1988) has been also documented in children at the extremes of the normal range.

Second, for both late and early talkers (as well as in the middle ranges of the distribution), some children are equally precocious or delayed in both comprehension and production; however, other children display a dissociation between comprehension and production level (i.e., delayed or precocious in lexical production while comprehension is at age-level). Interestingly, Thal & Bates (1988a) found that late talkers, as a group,

display a significantly greater dissociation between comprehension and production than their language-matched controls. Thus, the match between late talkers and controls for reported production vocabulary did not hold for comprehension. Further, a one-year follow-up on these children (Thal, et al., in press) revealed that those children who continued to be delayed in lexical production (based on Toddler norms) were those children who were delayed in *both* comprehension and production at the first testing. Thus, the late talkers who displayed high comprehension abilities were the children who demonstrated "catch-up" in lexical development one year later.

Finally, this work reveals that just because children achieve lexical milestones at the same rate, albeit faster or slower than their agemates, individual differences might also exist. The best example is one reported in Thal and Bates (1988b) in which two early talkers produce very different types of utterances. M (21 months) has a reported production vocabulary of 627 words; S (17 months) is reported to produce 596 words. An examination of their productive speech in the laboratory quickly reveals that their utterances are quite different in average length (MLUs of 1.19 vs. 2.39, respectively). M is likely to produce single words (e.g., "hungry," "cute" or "falling"); whereas, S is more likely to produce phrase-long expressions that might derive from idioms (e.g., "see Becky in the morning" or "you little monkey"). While both children produce closed-class morphemes, verbs, and other non-noun content words indicative of their precocity, they nevertheless differ in the general character of their utterances -- one, focusing on small units of primarily single words, and the other, concentrating on larger sentence-level units. So vocabulary size and utterance length can be seen to "break apart," even though they are highly correlated in most children (approx. r=+.80). Does this type of individual difference represent a dissociation between semantic and grammatical development? Not necessarily, as both children are using grammatical morphology in a productive way (e.g., "talks" vs. "talking" in M's single word utterances). These two children serve as yet another reminder that there is variation even *within* the extremes, and that it is crucial to continue to examine the nature of cognitive and linguistic developements at these extremes.

In summary, these and other studies explicitly examine the variability across individuals in the timing and style of the achievement of linguistic milestones. Such work reinforces the idea that all children do not solve the problem of language acquisition in exactly the same way. However, with the help of tools such as the MacArthur parental report instruments, in conjunction with fine-grained longitudinal analyses about which aspects of development tend to hang together or come apart, we can begin to meet the challenge of understanding what "normal" development, and hence normal variation, really is.

3. Contextual variation in the frequency of passive usage

Variation in linguistic performance can also result from the fact that languages typically provide a range of means to solve the same communicative problem. While this feature allows the productive and creative use of language, it nevertheless can make the accurate assessment of linguistic production abilities more challenging than might be hoped. To take a simple example, in a picture description task, the range of appropriate responses to the prompt "tell me about the _____" span from a single word or simple phrase to many paragraphs of detailed description. In the adult, these kinds of variations would be attributed to mood, level of literacy, or cultural background, to name only a few. These factors can also influence a child's output, however, for interesting *other* reasons, children may provide perfectly legitimate solutions to communicative problems that differ quite

dramatically and consistently from what would be produced by an older child or adult. Unfortunately, then, it cannot necessarily be concluded that a child who does *not* produce or comprehend a particular form or structure in a given experimental context or observational period necessarily *lacks* the linguistic ability that underlies the production of that structure.

The converse is also true. A child who correctly and appropriately produces a particular linguistic form should not necessarily be held accountable for the entire range of linguistic knowledge that is said to govern its usage in the mature speaker. For example, take the two-and-a-half year old who correctly says "daddy ate" when describing her father's activities around the dinner table. However, a year later, this same child, who presumably is more linguistically and cognitively advanced, is much more likely to produce the incorrect utterance "daddy eated." By the time that this child is 5 or 6, she has returned to using the appropriate form "ate." Most interpretations of this U-shaped learning phenomenon suggest that the mechanisms which underlie the "correct" output at two years of age are in several respects qualitatively different from those that underlie the production of the very same form at the age of 6 (Plunkett & Marchman, in press). With development and experience, then, the child's linguistic system undergoes substantive *reorganization and refinement* beyond the point when they may appear to be linguistically sophisticated.

Another classic example of protracted linguistic change involves the passive construction in English (e.g., the girl got kissed). In English, passives are used to signal that the patient (i.e., what is acted upon) is the focus or topic of the discourse. The agent (or actor) is sometimes, but not obligatorily, mentioned at the end of the clause in a by-phrase (e.g., the tiger was licked *by the bear*). According to most reports, normally developing children productively use passives in naturalistic speech by the time they reach 3 or 4 years of age (e.g., Bowerman, 1982). Further, when passives are used by children, they generally occur in the appropriate discourse contexts. However, there is considerable evidence to suggest that the task of passive acquisition is far from complete by this time. Children are much less likely to use passives than adults (Horgan, 1977) and they are much more likely to produce truncated forms than passives which include a by-phrase (e.g., Beilin, 1975). In addition, they are much less likely than adults to produce a passive when the patient of the action is inanimate (e.g., the flower was given to the lady) or when the sentence includes a non-reversable action (e.g., the rocks got crawled on). These studies suggest that consistent and identifiable developmental changes in the *frequency* of passive usage occur across event and discourse contexts. Thus, it is the *performance facts* of passive usage which undergo change across development, beyond the point when children can produce their first passive.

In a recent study reported more fully in Marchman, Bates, Burkardt, & Good (in press), we explored several functional contraints on passive usage, in the attempt to outline the basis of the developmental changes in the frequency of passive usage that are observed in normally developing English-speaking children. In order to elicit passive usage in a relatively naturalistic context, we used a video which contained 24 scenes depicting animals and people performing simple single-action activities (e.g., a tiger licks a bear), and complex two-action activities (e.g., a camel hits a bear and then the bear hits the horse). Some of the scenes contained only animate characters, depicting reversible transitive actions. Other scenes contained inanimate objects and/or non-reversible actions (e.g., a snake crawls on a rock). After the child watched each scene, the video was paused and the experimenter altered the focus of the discourse by asking the child to "tell me about the _____." The procedure was run in two conditions: one in which each character was probed, and a second in which only the patient in the scene was probed. It should be noted that while each scene depicts clearly identifiable action(s), the child was free to

describe the scenes in any way that s/he wished. 108 children ranging in age from 3 to 11 participated in the study, as well as a sample of 24 undergraduates at UCSD.

First, this procedure was indeed successful at eliciting passives in even three-year-old children. For example, 67% of the three-year-olds produced at least one passive in response to a probe about the patient. Thus, the results confirm past findings that children can produce passives fairly early in development. However, the frequency of passive usage changed considerably across the age period tested here. For example, three year olds produced a passive only about 17% of the time. In older children and adults, in contrast, passives were used quite frequently, comprising approximately 62% and 87% of the responses in the eleven-year-old and adult groups, respectively. In addition, passives were much less likely to occur when the scene contained inanimate characters or non-reversible actions, hovering around 20% of the time for the children, increasing only slightly for adults (41%).

These data confirm that passives do not occur equally often in all discourse and event contexts. Further, the general tendency to produce a passive does increase in frequency across development. In this study, two additional questions were addressed. First, when these children are not using a passive, what are they doing *instead*? The results indicate that in place of the passive, children typically produce an alternative, yet appropriate, means to describe the scenes from the point of view of the patient. For example, instead of "the tiger got licked," a young child might instead choose "the tiger is sitting there while the bear licked him" or "it was the tiger that the bear licked." Interestingly, these alternatives fulfill the discourse requirements of the task (i.e., they structure the response around the patient), and therefore, must be viewed as appropriate responses. Further, these alternatives can be considered to be quite sophisticated syntactically, sometimes involving two-clause or relative-clause constructions. Thus, young children appear to *know* how to produce the passive, however, they do not necessarily *like* to do so.

Second, when children do use a passive, are they identical in form to adults' passives? These data confirmed previous findings that children were less likely than older children and adults to mention the agent of the action in a by-phrase. In the youngest three age groups, a clear majority of the passives produced (76%) did not include a by-phrase (i.e., truncated passives). After age 7, the use of the by-phrase increased in frequency but it was only the adults who included it consistently (e.g., only 65% in the 11-year-olds). The by-phrase is an optional component of the English passive, but it nevertheless serves a substantive discourse function, i.e., to inform the listener of the identity of the agent of the action. Do children avoid by-phrases because they are not sensitive to its function to identify who did the action? Interestingly, these data suggest that this is not the case. Children of all ages used a by-phrase more frequently in this scenes in which it might be useful to identify the agent. That is, by-phrases were used more often when describing the complex scenes (in which two remaining characters could be the agent), than when describing the simple scenes (in which the agent is unambiguously the non-patient).

In general, then, these findings reinforce the notion that the acquisition of syntactic abilities, such as the passive, should be viewed as a protracted event. While children can communicate quite effectively early in development, their linguistic skills continue to develop and refine throughout childhood and perhaps even into adolescence in subtle, but nevertheless substantive, ways. Thus, adult-like performance can be seen to incorporate more than the ability to produce a given syntactic structure. From a functionalist perspective, it is important to look beyond the acquisition of grammatical structures *per se* and account for the development of linguistic behavior with respect to the cognitive, communicative and processing factors which determine how and when linguistic knowledge is accessed during language use.

4. Conclusion

The variability that is observed across individuals and across contexts presents challenges to an understanding of the acquisition of language in normally developing children. However, in our view, considerable progress has been made in explicitly acknowledging and systematically outlining the sources of that variation, as well as understanding its limits in the normal population. By developing new techniques, and by outlining the functional basis for linguistic change across development, researchers will be in a better position to define the conceptual and communicative foundations of normal acquisition.

References

Bates, E., Bretherton, I., & Snyder, L. (1988). From first words to grammar: Individual differences and dissociable mechanisms. New York: Cambridge Univ. Press.

Bates, E., Caselli, M.C., & Casadio, P. (1990). 'A crosslinguistic study of early lexical development using parental report.' In C. Rovee-Collier (Ed.), Abstracts of the Seventh International Conference on Infant Studies: Special Issue of Infant Behavior and Development, 13, pg. 258.

Bates, E., & MacWhinney, B. (1982). 'Functionalist approaches to grammar.' In E. Wanner & L. Gleitman (Eds.), Language Acquisition: The State of the Art. New York: Cambridge Univ. Press.

Bates, E., & MacWhinney , B. (1987). 'Functionalism and the competition model.' In B. MacWhinney (Ed.), Mechanisms of language acquisition. Hillsdale, N.J.: Erlbaum.

Bates, E. & Marchman, V. (1988). 'What is and is not universal in language acquisition.' In F. Plum (Ed.) Language, Communication and the Brain. New York: Raven Press.

Bates, E., Thal, D., & Marchman, V. (in press). 'Symbols and syntax: A Darwinian approach to language development.' In N. Krasnegor, D. Rumbaugh, R. Schiefelbusch, and M. Studdert-Kennedy (Eds.), The Biological Foundations of Language Development. Oxford: Oxford University

Bates, E., O'Connell, B., & Shore, C. (1987). 'Language and communication in Infancy.' In J. Osofsky (Ed.), Handbook of Infant Competence, 2nd Edition. New York: Wiley.

Beilin, H. (1975). 'Experiments on the passive.' In H. Beilin (Ed). Studies of the cognitive basis of language development. New York: Academic Press.

Borer, H. & Wexler, K. (1987). 'The maturation of syntax.' In T. Roeper & E. Williams (Eds.), Parameter Setting. Dordrecht, Holland: Reidel.

Bowerman, M. (1982). 'Reorganizational processes in lexical and syntactic development.' In E. Wanner & L. Gleitman (Eds.), Language acquisition: The State of the Art. Cambridge: Cambridge University Press.

Bowerman, M. (1985). 'What shapes children's grammars?' In D.I. Slobin (Ed.)., The crosslinguistic study of language acquisition: Volume II: Theoretical Issues. Hillsdale, N.J.: Erlbaum.

Chomsky, N. (1965). Aspects of a Theory of Syntax. Cambridge, MA: MIT Press.

Dale, P., Bates, E., Reznick, S., & Morisset, C. (1989). 'The validity of a parental report instrument of child language at 20 months'. Journal of Child Language, 16, 239-250.

de Villiers, J. (1988). 'Faith, Doubt, and Meaning.' In F. Kessel (Ed.), The development of language and language researchers: Essays in honor of Roger Brown. Hillsdale, N.J.: Erlbaum.

Horgan, D. (1977). 'The development of the full passive.' Journal of Child Language, 5, 65-80.

Jackson-Maldonado, D., Marchman, V., Thal, D., & Bates, E. (1990). 'Adaptation of parental report language inventories for Spanish-speaking infants and toddlers.' Poster presented at the International Conference on Infant Studies, Montreal 1990.

Marchman, V., Bates, E., Burkardt, A., & Good, A. (in press). 'Functional constraints on the acquisition of the passive: Toward a model of the competence to perform. ' First Language.

Nelson, K. (1973). 'Structure and strategy in learning to talk.' Monographs of the Society of Research in Child Development, 38, Serial No. 149.

Plunkett, K. & Marchman, V. (in press). U-shaped learning and frequency effects in a multi-layered perceptron: Implications for language acquisition. Cognition.

Reznick, S. & Goldsmith, S. (1989). Assessing early language: A multiple form word production checklist. Journal of Child Language, 16, 91-100.

Roeper, T. (1987). 'The acquisition of implicit arguments and the distinction between theory, process and mechanism.' In B. MacWhinney (Ed.) Mechanisms of Language Acquisition. Hillsdale, N.J.: Erlbaum.

Slobin, D. I. (1982). 'Universal and particular in the acquisition of language.' In E. Wanner & L. Gleitman (Eds.), Language Acquisition: The State of the Art. Cambridge: Cambridge Univ. Press.

Slobin, D. I. (1985). The crosslinguistic study of language acquisition. Hillsdale, N.J.: Erlbaum.

Snow, C. (1983). Saying it again: The role of expanded and deferred imitations in language acquisition. In K.E. Nelson (Ed.) Children's Language, Volume 4. Hillsdale, N.J.: Erlbaum.

Thal, D. & Bates, E. (1988a). 'Language and gesture in late talkers.' Journal of Speech and Hearing Research, 31, 115-123.

Thal, D. & Bates, E. (1988b). 'Relationships between language and cognition: Evidence from linguistically precocious children. ' Paper presented at the Annual Convention of the Americal Speech-Language-Hearing Association, November, Boston, MA.

Thal, D., & Bates, E. (1991). 'Continuity and variation in early language development.' In J. Colombo & J. Kagan (Eds.) Individual differences in infancy: Reliability, stability and prediction. Hillsdale, N.J.: Erlbaum.

Thal, D., Tobias, S., & Morrison, D. (in press). 'Language and gesture in late talkers: A one year follow-up. ' Journal of Speech and Hearing Research.

III - METHODOLOGICAL PROBLEMS IN STUDIES OF ACQUIRED APHASIA IN CHILDREN

PATIENT SELECTION IN STUDIES OF APHASIA ACQUIRED IN CHILDHOOD

Bryan T. Woods, M.D.
McLean Hospital, Department of
Neurology and Brain Imaging Center
115 Mill Street
Belmont, MA 02178, U.S.A.

ABSTRACT. The history of the study of acquired aphasia of childhood is filled with examples of erroneous conclusions that could be traced to an imperfect understanding of the underlying pathological process that caused the aphasia. Ideally, one wishes to be certain about both the spatial and temporal extent of the pathological process. Strokes, with their restriction to one major vessel territory, sudden onset, and lack of progression best fit these criteria. Other lesions can also be utilized effectively, but the greatest single confounding clinical variable when one wishes to study effects of discrete lesions is severe or prolonged generalized seizures.

1. INTRODUCTION

Acquired childhood aphasia has always been an uncommon medical problem, but its study assumes much greater importance than the numbers alone might warrant because of its potential for giving otherwise unavailable insights into both the development of normal higher-order brain functions and into the far more common congenital disorders of that development. Somewhat paradoxically a history of acquired aphasia, defined simply as a loss or reduction in previously achieved levels of language functions, is not a necessary condition for patient inclusion in such studies. What is a necessary condition, however, is the occurrence of a pathological process involving the patient's cerebrum, and it is my belief that progress in understanding childhood aphasia has resulted in large part from an increasingly precise specification of the pathological processes that not only produce it but fail to produce it.

2. EXAMPLES

The first example of this proposition can be taken from the beginnings of modern aphasiology; the observation that patients with infantile hemiplegia due to left hemisphere lesions involving the

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 27–34.

language area are not aphasic (Cotard, 1868). The second example is the increasing awareness about 15 years ago by ourselves and others that lesions truly localized to the right hemisphere rarely produce even short-term aphasia in right-handed children. A review of the older literature led us to speculate that the earlier observations had been confounded by looking at patients with pathological processes that appeared to be lateralized but actually affected functions bilaterally (Woods and Teuber, 1978).

The final example of the importance of precise pathological localization is a recent patient in whom the presence of the pathological process was only initially revealed by magnetic resonance imaging (MRI). The patient was a 29-year-old, left-handed man admitted to a psychiatric facility because of impulsive and inappropriate behavior, including making obscene remarks to strangers in public places. Initially his only reported neurological history was of a single grand mal seizure at age 21. A Sleep/Awake EEG was normal but an MRI scan revealed an old infarction in the territory of the left posterior cerebral artery (Fig. 1). Subsequent inquiry from surviving family members (unfortunately limited to younger siblings) elicited the information that he had had a sudden neurological "problem" when very young that had left him with a "lazy" eye. The best estimate was that this occurred at about age two. The patient had performed adequately in elementary school and had gone on to finish high school and complete several semesters of college before dropping out.

Neurological examination confirmed the presence of a previously undocumented right upper quadrant visual field defect, but good reading skills and intact color naming (9/9). Figure 1 shows axial plane MRI images showing the extent of the lesion's involvement of the left lingual and parahippocampal gyri, calcarine cortex, and the forceps major outflow from the splenium of the corpus callosum. This is the classic lesion location to produce the adult alexia without agraphia syndrome. The almost complete absence of surrounding gliosis on other appropriate scan indicated the early-life origin of the injury.

Anatomically more extensive abnormalities were elicited by the auditory and visual evoked response portion of brain electrical activity mapping (BEAM). The abnormalities were centered in the more lateral portion of the posterior left temporal lobe.

Table 1 shows a summary of the results of neuropsychological testing. The first striking finding is the 38-point superiority of verbal IQ score over performance IQ score. The next point of note is the low memory quotient, due primarily to problems with memory passages, associate language, and visual reproductions. Other test results show a mixture of visual and verbal deficits intermixed with intact performance on tasks such as face recognition.

Although single case interpretation must be tentative the clinical and neuropsychological findings in this patient are consistent with a shift of both handedness and control of language function to the intact right hemisphere, with normal reading and color-naming but incomplete compensation for left medial temporal functions, primarily

verbal memory, and considerable impairment of a number of complex visuospatial functions. (The alternate hypothesis that he was always right-brain dominant is thought less likely because of the absence of family history of left-handedness and because of the presence of a rather severe impairment of verbal memory.)

Pathologically, the anatomy of the lesion is restricted to a portion of a single major vessel distribution, but the electrophysiological findings indicate more extensive lateral and anterior dysfunction that includes most of the left temporal lobe. This observation that the functional effects of a brain insult extend beyond its apparent anatomical borders is becoming commonplace as techniques that image metabolic activity become more widely available.

3. SELECTION PRINCIPLES

Given these examples and the concerns they embody, what then should be the basic principle of patient selection for studies of childhood aphasia? It would seem that above all else one should be able to define as precisely as possible both the temporal and spatial extent of each patient's pathology. Patients with a single stroke involving the territory of one major vessel and no subsequent seizure disorder come closest to the ideal. The MRI scan of a patient from our initial study (Woods and Teuber, 1973) is shown in Figure 2. The scan shows a large defect in the territory of the posterior division of the left middle cerebral artery, probably acquired at seven months gestational age. Even such patients, as our third example shows, may have functional deficits that extend beyond the discernible anatomical borders of the lesion.

Another problem in defining the nature and extent of the pathology is that some early lesions may leave very little focal residual change. Figure 3 shows the MRI of another patient from the initial study with right hemiparesis with sudden onset at 14 months of age; the whole left hemisphere is quite small (Dyke-Davidoff-Masson syndrome) and the left ventricle large but, except for a small linear area in the thalamus, the lesion locus is not seen. This is because characteristic gliotic changes present after later lesions may simply not be present after early lesions, and if the lesion doesn't leave a "hole" (Figure 2), but rather results in tissue shrinkage, then the only residual may be decreased brain volume and increased ventricular volume; this may be so even if the lesion is vascular (Taveras and Wood, 1976).

Tumors are still more problematical pathologically because of imprecision in both the temporal and spatial dimensions; the former because of uncertainties as to time of onset and rate of prior progression, and the latter because of edema. Similar time course uncertainty is also present with arteriovenous malformations (AVMs). To some extent, these problems are minimized if the tumor is a benign one that is surgically excised, or if the AVM hemorrhages and is then surgically removed. In these situations one can conclude that the significant lesion occurred "no later than" the time of surgery. If the lesion is continuing to progress all interpretations are confounded.

Infections also pose major difficulties for localization. With the exception of single abcesses or herpes simplex encephalitis, which is often unilateral, and has a characteristic CT/MRI appearance, one ordinarily has to assume bilateral involvement. Similar considerations apply to traumatic injury. Our original series included one child with a gunshot wound with entrance and exit both on one side of the head, but for the most part trauma in children involves blunt objects, and it is extremely difficult to exclude bilateral injury, even with brain imaging.

A major difficulty can arise from secondary epilepsy. Both recurrent generalized seizures themselves, and the medication used to treat them, may have diffuse effects on the developing nervous system that can confound the interpretation of cases with otherwise focal lesions, both because the lesion becomes ill-defined spatially, and because even if the seizures are focal, they may result in a progressive lesion. In our study all of the children with FSIQ scores below 70 (n = 11) also had a history of severe or prolonged seizures (Woods, 1980). Nevertheless, it would seem that one need not take an all-or-none position, and that a history of a few seizures after a lesion, and even ongoing anticonvulsant treatment that is effective at non-toxic levels, need not disqualify a child from inclusion in a series.

The question remains, however, whether a truly unilateral brain lesion during early childhood that is not complicated by significant seizures is ever sufficient to lower the full scale IQ score below the borderline retardation range. If not, then it could be argued that an FSIQ below 70 should be an exclusion criterion if lesions are to be interpreted as being strictly unilateral. In older children where there is persistent aphasia, or where the PIQ is very low and "drags down" the total score, this inference of bilaterality based on FSIQ below 70 would not necessarily hold.

One final relevant point is the distinction between inclusion and exclusion criteria. Evidence of significant lesion bilaterality and continued lesion progression are widely used specific exclusionary criteria, but what about specific inclusionary criteria; is it necessary that the lesion be visible on CT/MRI scan? My view is that this is too restrictive; valid evidence of a unilateral lesion could come from clinical neurological examination, from clinical neurophysiological procedures including electroencephalography, evoked responses and brain mapping; or from brain imaging. What I do think would be unacceptable is using the dependent variables to define the lesion; for example, use of a low PIQ score or a low left ear dichotic score as validating criteria for right hemisphere lesions.

4. CONCLUSION

In general, appropriate patient selection for studies of childhood aphasia will be assured if four requirements are met: First, a clear purpose for the study, which means a well-phrased question; second, an awareness of relevant neuropathological considerations; third, a

familiarity with what has already been observed by others (even if one rejects their conclusions); and finally, an adherence to the generally accepted principles of sound scientific methodology in the design of the study.

5. REFERENCES

Cotard, J. (1868) 'Étude sur l'atrophie partielle du cerveau', Thèse de Paris.

Taveras, J.M., Wood, E.H. (1976) Diagnostic Neuroradiology, Second Edition, Vol. 1. The Williams and Wilkins Co., Baltimore, p.375.

Woods, B.T. (1980) 'The restricted effects of right hemisphere lesions after age one: Wechsler test data', Neuropsychologia 18, 65-70.

Woods, B.T., Teuber, H-L. (1973) 'Early onset of complementary specialization of cerebral hemispheres in man', Trans. Ann. Neurol. Assoc. 98, 59-63.

Woods, B.T., Teuber, H-L. (1978) 'Changing patterns of childhood aphasia', Ann. Neurol. 3, 273-280.

6. FIGURES AND TABLES

6.1 Figures

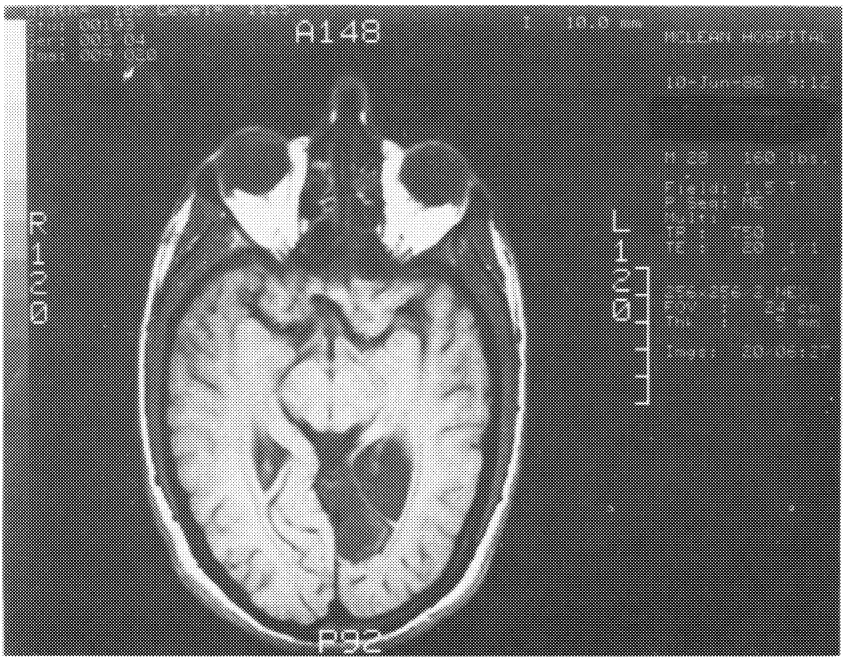


Figure 1: Axial MRI section showing area of infarction in the territory of the left posterior cerebral artery.

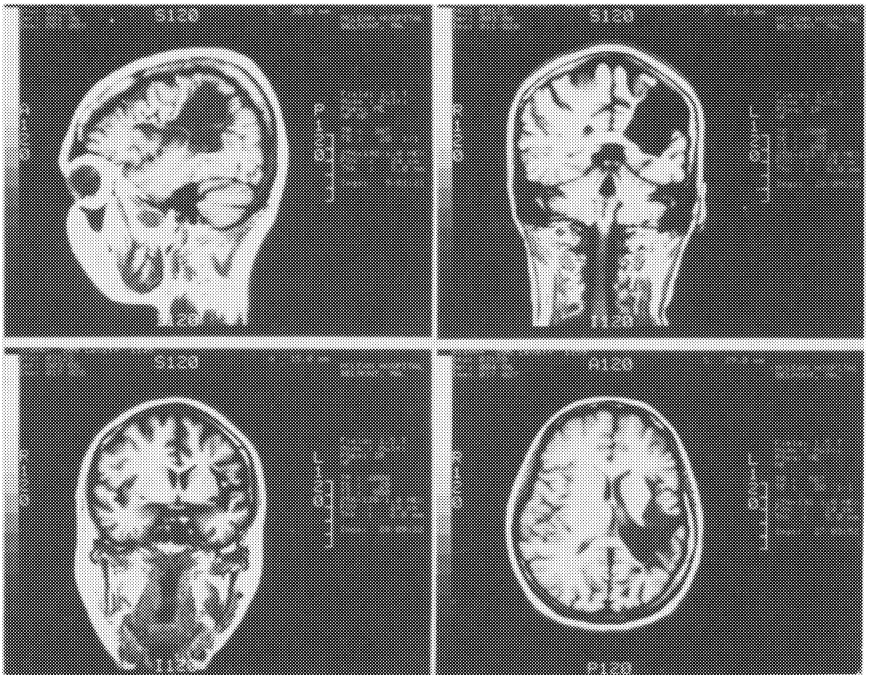


Figure 2: Sagittal (upper left), axial (lower left), posterior (upper right), and anterior (lower left) coronal views of a lesion involving the vascular territory of the left middle cerebral artery.

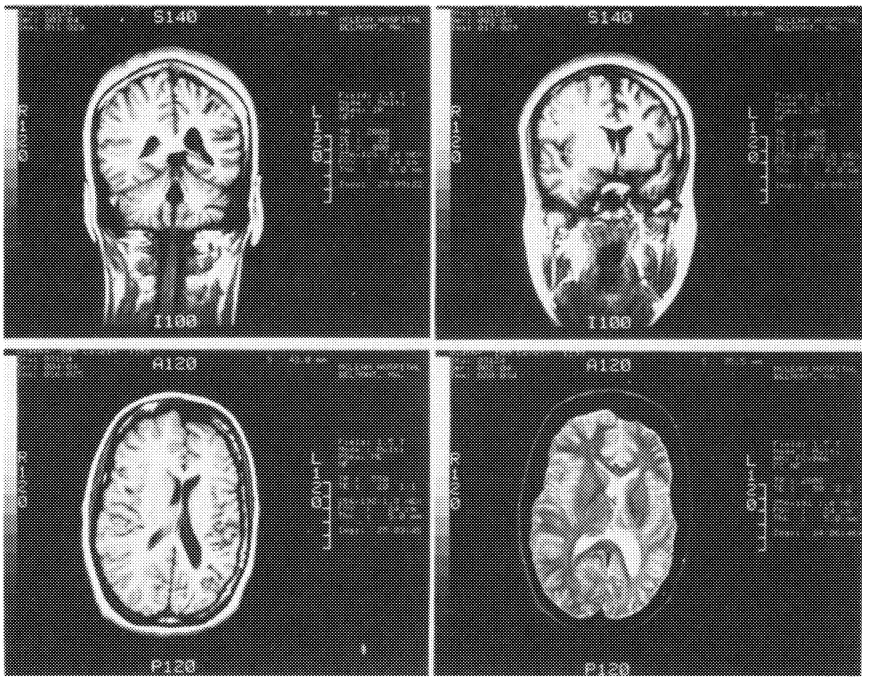


Figure 3: Coronal (upper) and axial (lower) views showing thicker skull, larger ventricle, and smaller hemisphere on the left. The T-2 weighted scan on the lower right shows a bright diagonal linear streak in the left thalamus, medial and parallel to the posterior limb of the internal capsule. That is the only focal lesion seen in the scan.

6.2 Tables

TABLE 1. Summary of results of neuropsychological testing.

WAIS-R					
VIQ	103	Inf.	13	Pict. Comp.	5
PIQ	65	Dig. Sp.	9	Pict. Arr.	4
FSIQ	84	Voc.	13	Bl. Des.	4
		Arith.	12	Obj. Assem.	3
		Comp.	9	Dig. Sym.	4
		Sim.	10		
MQ 75					
Inf.	6/6	Vis. Rep.			
Orient.	4/5	Immed.	4/14		
Ment. Con.	9/9	Delay	1/14		
Mem. Pass.					
Immed.	10/24	Assoc. Lang.			
Delay	5.5/24	Immed.	3.5/21		
		Delay	3/21		
FAS 17 (2nd %ile)					
Bost. Naming	13/15				
WRAT-R					
Reading	88th %ile				
Spelling	91st %ile				
Gray Oral Reading	Grade 10.8				
Visuospatial Tests					
Hooper 13/30					
Rey-Osterrieth					
Copy	28 (below 25th %ile)				
Immed. recall	0				
Delayed recall	.5				
Facial recog.	49/54				

ANALYSIS OF LESION LOCALIZATION AND SIZE

José M. Ferro
Faculdade de Medicina de Lisboa
Hospital de Santa Maria, S. Neurologia
1600 LISBOA, PORTUGAL

ABSTRACT. Methodological issues on the study of the brain / behaviour relationships are reviewed. Present studies intented to evaluate the structural correlates of childhood aphasia should describe the selection of the study sample, aetiology, time interval from onset to neuropsychological examination and imaging studies and the presence of previous and concomitant disorders. CT and MRI make possible a detailed anatomical analysis of lesion site and size. The extent of damage to each relevant anatomical structure should be quantitated.

Correlation of neuropsychological and imaging data can be made through visual methods - overlaping individual lesions - or through chi square, t tests and multiple regression analysis. Whenever available, functional studies, such a SPECT or PET should also be performed, specially in subcortical lesions.

The study of the structural correlates of aphasia has been, since Broca (1861), one of the major aims of these who study brain / behaviour relationships. In the following pages we will adress some of the methodological problems of this research. Appropriate selection of subjects, careful analysis of lesion and adequate statistical methods are necessary to relate language disturbance to damage to localized brain sites.

Aetiology, time from onset and referral bias should be considered when describing the study population. Bias should be avoided when selecting comparable controls. For the study of childhood aphasia cases

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 35–44.

and controls should be matched not only for sex and age, but also for school grade, social class and, if possible, IQ, (or performance IQ).

Aetiology interacts with lesion location. This may be specially relevant if a center is working with a particular selected population. Non penetrating head trauma tends to affect mainly the basofrontal and the temporal lobes. Herpes encephalitis causes damage to both temporal lobes. With ischemic stroke, which is the best aetiology to study acquired aphasia, the chances of getting lesions on different parts of the brain depends mainly on the physiopathology of stroke. Embolism produces cortical or large subcortical infarcts, while those having an hemodynamic pathogenesis are localized in terminal or border zones (Zeumer and Ringelstein, 1987). In children, the majority of strokes are secondary to intracranial occlusive disease are localized in the basal ganglia (Zimmerman et al, 1987), while those related to congenital heart disease or cardiac surgery are either cortical or of the hemodynamic type (Furlan and Jones, 1987).

What methods can we use to detecte and localize brain lesions ? They can be divided in morphological and functional. Morphological methods include CT scan and MRI. MRI does not expose subjects to radiation and it is clearly superior to CT scan, specially for posterior fossa imaging, grey-whitte matter differentiation, visualization of gyri details, detection of small deep lesion. MRI provides not only conventional axial slices, but also coronal and sagital cuts. Functional methods include electrophysiological procedures, such as EEG and evoked potentials, and perfusion / metabolic studies such as measurements of regional cerebral blood flow that can be imaged in axial tomographic slices, SPECT and PET. The main problem with electrophysiological studies is that they cannot differentiate between the activities of cortical and subcortical structures, while this is possible using with methabolic / perfusion methods. PET is much more expensive and less available than SPECT. With those functional methods patients can be studied at rest and while performing some simple tasks (Rousseau et al, 1989). These activation procedures may be at great interest to study the mechanisms of recovery.

Several parameters should be considered and described when reporting the results of localization studies namely: time from onset, lesion location and size, distant effects of localized lesions and concomitant disorders.

Despite data from several adult series (cf: Kertesz, Harlock and Coates, 1979; Knopman et al, 1983; Ferro and Crespo, 1988) has clearly demonstrated that neuropsychological defects improve over time and, consequently, that the structural correlates of these disturbances are

different at one, three, six or more months after onset, some authors continue to lump data from subjects seen at different time from onset. Some consider appropriate for research only those subjects seen some months after the insult (Alexander, Naeser and Palombo, 1990), negleting the fact that by using this type of selection, one may loose mildly affected individuals, who recover almost completely in a few weeks / months.

Imaging has also an optimal "window" to be obtained. In ischemic stroke the best time to perform a CT scan is between the 3rd and the 6th week post onset (Savoiardo, 1986). Early scans have a poor definition of lesion boundaries, while in very chronic scans, there is a problem of scarring and rectraction, which is particulary relevant for lesions close to the fissures, sulci or ventricles.

There are several methods and atlas which can help researchers in establishing detailed lesion location. The Matsui and Hirano' atlas (1978) is specialy appropriate because it presents diagrams for different orbito-meatal angles. The Damásios' atlas (1982) despites Broadman's areas in axial CT slides, while Naeser's diagrams (1981) indicates the location of the areas relevant to language prodution and comprehension in different CT cuts. In more recent studies the Boston group (Alexander, Naeser and Paloumbo, 1987, 1990) further detailed their previous anatomical analysis, trying to identify more than 20 regions of interest. This level of detail, although increasing our knowledge of the structural correlates of language, has several drawbacks. First it decreases inter-judge reliability in CT evaluation. Second, in some regions, even with high resolution scans, differentiation of contigous areas is very difficult or hazardous. Three areas are specially treacherous: the rolandic operculum, the temporal pole, the periventricular zone and the mesial occipital lobe. In the rolandic operculum distinction between Broca's areas and the narrow strips of areas 6 and 4 is rather difficult. In chronic scans, gliotic periventricular lesions produce retraction of the ventricular wall and distortion of the usual anatomy of the periventricular tracts and nuclei, which became difficult to locate and size. Because of its location, and of the shape of the cerebellar tent, evaluation of mesial occipital lesions demands careful attention to the CT angle. Otherwise, cerebellar and occipital lesions will be confounded and mesial occipital damage incorrectly located.

To obtain an estimate of lesion size one can calculate the area of the lesion in each CT cut, by multiplying the largest and smallest axis of the lesion, multiply these areas for the thickness of the CT slices, and adding these partial volumes. More precise measurements require either a cooperating neuroradiologist who traces the lesion on the CT screen, the

volume beeing calculated by the computer, or the use of a digital table and a cursor to follow the lesion-contour and appropriate software to calculate the volume (Kertesz, Harlock and Coates, 1979). The measurements can be made for CT hard copies, taking in consideration the minimification factor (the ratio between actual head size and its dimension on CT copies). Alternatively, if we want also obtain composite owverlaps of the individual lesions the actual images have to be projected with the help of a photographic enlarger on standard diagrams or templates.

In most studies the extent of damage to relevant areas is more important than the total lesion size. A five (Selnes et al, 1985) or four (Ferro, Kertesz and Black, 1987) point-scale (eg: 0 - no damage; 1 - < 50 %; 2 - > 50 %; 3 - total damage) is preferable to more detailed (eg: 7 or 8 points) (Naeser et al, 1987, 1990) scales, which are less reliable.

In ischemic stroke there is a generally overlooked interaction between lesion site and size. Because lesions are confined to vascular boundaries, areas located in the core of a vascular territory are more likely to be involved by small to medium size lesions while those localized at the periphery are involved mostly by large lesions. For example the majority of middle cerebral artery (MCA) infarcts involve the rolandic operculum and the insular region, while the caudate nuclei or the superior parietal lobe are damaged either by uncommon small infarcts, or extensive damage in the MCA territory (Zulch and Hossman, 1988).

PET (Metter et al, 1988), SPECT (Perani et al, 1987) and other dynamic imaging methods demonstrated that the effects of a brain lesion are more widespread than was is shown by CT or MRI. Distant effect of a localized lesion (diaschisis) (Feeney and Baron, 1986) include cortical hypometabolism after subcortical lesions and vice-versa, contralateral cerebellar and hemispheric (mirror) hypofunction. These methods have been rarely used in cases of childhood aphasia.

A detailed anatomical analysis based only on "static" imaging methods - CT, MRI - gives imcomplete information about the non-functional area which is better show as a zone of hypometabolism / hypoperfusion by PET / SPECT.

These distant effects probably do not have the same significance in very early lesions and in the chronic stage. In many acute cases, in the first hours and days, ipsilateral hypoperfusion may represent incomplete ischemia, in zones that are going to remain viable; sometime later it can be related to disconnection, interruption of fibers linking cortical and subcortical stuctures, while many late hypoperfusion defects can be

secondary to neuronal loss due to transynaptical and axonal degeneration (Feeney and Baron, 1986).

The influence of concomitant disorders should also be thought of. Both pre and perinatal insults and post-natal disorders must be checked. Epilepsy and congenital heart disease represent special problems. Populations afflicted by these disorder are not interely comparable to others with similar lesions but different aetiologies. Anoxia due to frequent generalized seizures, the effect of anticonvulsivants, brain lesions such as neuronal migration defects or gyri malformations that can be the cause of epilepsy but are too subtle to be detected by CT, but and the kindlig effect in the focus area, all case give rise to brain functional reorganization and modify the standard lesion / dysfunction relationship (Trimble and Reynolds, 1987).

Congenital heart disease is a potential source of cases of acquired aphasia (Aram, 1988). Stroke in these patients occurs in relation to several different mechanisms: embolism from the heart, paradoxical embolism in right-to-left shunts, high blood viscosity due to the elevated hematocrit. Complication of heart surgery, such as brain embolism, with large or micro-embolism, hypoperfusion related to prolonged hypotension and hypothermia can affect the brain locally or difusely. The effects of long standing low oxigen hemoglobin saturation and high hematocrit are not known (Furlan and Jones, 1987). Furthermore cardiac abnormalities can be associated with CNS malformation, that are usually not shown in CT or MRI.

When we have a series of patients to study the correlation between lesion site and neurobehavioural performance we must overlap the individual lesions to obtain the area of maximal overlap. Because scans are usually performed in different machines with various orbito-meatel angles, lesions should first be projected, though photographical enlargers or other devices, in standard (Kertesz, Harlock and Coates, 1979) templates. This overlap method, although commonly used, has a underecognized problem. When we work with ischemic vascular lesions, the more cases we overlap the greater the chance of getting an area of maximal overlap that is determined by "vascular" anatomy and not functional reasons. ie, of getting an area of maximal overlap that corresponds to the "core" of the MCA territory (Kertesz, 1983). Lesions can also be projected in diagrams of the lateral view of the brain, using the method described by Mazzochi and Vignolo (1978).

If we want to identify areas that are important for one function or to contrast zones that are crucial for two different functions, we can use the some called "subtraction" method (Blunk et al, 1981). First overlaps of

the individual lesions from the subjects of the two groups we are comparing (impaired vs non impaired in a particular function; impaired in one task vs impaired in others tests) are obtainen; then these overlaps are compared, to distinguish between a common area important for both functions and "subgroup" areas which are related to innability to perform a particular task (Ferro et al, 1983). The Aachen group developed software to perform these overlaps and (Poeck et al, 1984) substractions in a computer. This visual or "analogic" way of analysing CT or MRI should be complemented by a analytic "digital" procedure. For that we should have a check-list of anatomical sites we are interested in and look for its involvement in individual lesions.

The numerical analysis of imaging / behavioural data is difficult. One method consists of comparing,
using x^2 statistics, the frequency of damage to the area of interest in impaired and non impaired subjects. A more sensitive method consists of contrasting, with t tests, the scores in the test we are studying, of subjects having the area of interest destroyed or intact (Gordon and Rosencrantz, 1982). Repeating this procedure for different areas, we can obtain the matrix of areas that are critical to the performance of that particular function. To avoid a type I error, due to multiple comparisons, one should either use the Bonferroni correction, choose the 0.01 instead of the 0.05 limit for statistical significance or collect a large sample.

These univariate methods do not take in consideration the interaction between lesion site and size and the fact that some areas tend to be damaged simultaneously, because they belong to the same vascular territory, nor can appreciate the effect of combined lesions in more than one area. Multiple regression analysis, using a dummy coding forthe presence / absence of lesions in each area of interest is the procedure of choice (cf: Kerliger and Pedhazur, 1973). However regression analysis results are sometimes difficult to interpret and can provide confusing results for functions that may have a bihemispheric representation. Furthermore regression analysis needs a very large sample, specially if there are many anatomical sites to include in the analysis.

I shall conclude with a couple of suggestions for studies intented to evaluate the stuctural correlates of childhood aphasia:

1. Description of the study sample on what concerns
   - A. Population from which cases (and controls) were selected.
   - B. Aetiology
   - C. Timing of neuropsychological and imaging studies
   - D. Previous and concomitant disorders

2. Detailed anatomical analysis of lesion site and size with CT or MRI

A. For each CT/MRI slice, a list of anatomical structures should be screened and the extent of damage quantitated

B. Correlation of neuropsychological and imaging data through visual (overlap) and statistical analysis.

3. Whenever available, functional studies (SPECT) should be performed, specially in subcortical lesions.

REFERENCES

Alexander MP, Naeser MA, Palumbo CL. Broca's area aphasias: aphasia after lesions including the frontal operculum. Neurology 40: 353-362, 1990.

Alexander MP, Naeser MA, Palumbo CL. Correlations of subcortical CT lesion sites and aphasia profiles. Brain 110: 961-991, 1987.

Aram DM. Language sequelae of unilateral brain lesions in children. In Plum F (ed). Language, Communication and the Brain, Raven Press, New York, p. 171-197, 1988.

Blunk R, De Bleser R, Willmes K, Zeumer H. A refined method to relate morphological and functional aspects of aphasia. Eur Neurol 20: 69-79, 1981.

Broca P. Remarques sur le siège de la faculté du langage articulé. Bull Anthropol 6: 377-393, 1865.

Damásio H. A computerized tomography guide to the identification of cerebral structures. Arch Neurol 40: 138-142, 1982.

Feeney DM, Baron JC. Diaschisis. Stroke 17: 817-830, 1986.

Ferro JM, Crespo M. Young adult stroke: neuropsychological dysfunction and recovery. Stroke 19: 982-986, 1988.

Ferro JM, Kertesz A, Black SE. Subcortical neglect: quantitation, anatomy and recovery 37: 1487–1492, 1987.

Ferro JM, Martins IP, Mariano G, Castro Caldas A. CT scan correlates of gesture recognition. J Neurol Neurosurg Psychiat 46: 943–952, 1983.

Furlan AJ, Jones SC. Central nervous system complications related to open heart surgery. In Furlan AJ (ed). The heart and Stroke. Springer–Verlag, Berlin, p. 287–304, 1987.

Golden GS. Stroke syndromes in children. Neurol Clin 3: 59–75, 1985.

Gordon WP, Rosencrantz PL. Statistical methods of analysing CT scan data based upon neuroradiological localization. 10th Annual INS Meeting Pittsburg, USA, 1982.

Kerlinger FN, Pedhazur EJ. Multiple Repression in Behavioral Research, New York, Holt, Rinehart and Winston, 1973.

Kertesz A. Issues in Localization. In Kertesz A (ed). Localization in Neuropsychology. Academic Press, New York, 1983.

Kertesz A, Harlock W, Coates R. Computed tomographic localization, lesion size and prognosis in aphasia and non verbal impairment. Brain and Language 8: 34–50, 1979.

Knopman DS, Selnes OA, Niccum N, Rubens AB, Yock D, Larson D. A longitudinal study of speech fluency in aphasia: CT correlates of recovery and persistant nonfluency. Neurology 33: 1170–1178, 1983.

Matsui T, Hirano A. An Atlas of the Human Brain for Computorized Tomography. New York, Igaku–Shoin, 1978.

Mazzochi F, Vignolo LA. Computer assisted tomography in Neuropsychological research: a simple procedure for lesion mapping. Cortex 14: 136–144, 1978.

Metter EJ, Riege WH, Hanson WR, Jackson CA, Kempler D, Lancker D. Subcortical studies in aphasia. An analysis based on (F-18) fluorodeoxyglucose, positron emission tomography and computed tomography. Archives of Neurology 45: 1229-1234, 1988.

Naeser MA, Gaddie A, Palumbo CL, Stiassny-Eder D. Late recovery of auditory comprehension in global aphasia. Archives of Neurology 47: 425-432, 1990.

Naeser MA, Hayward RW. Lesion localization in aphasia with cranial computed tomography and the Boston Diagnostic Aphasia Exam. Neurology 28: 545-551. 1978.

Naeser M, Helan-Estabrooks N, Haas G, Averbach S, Srinirasan M. Relationship between lesion extent in "Wernicke's area" on computed tomography scan and predicting recovery of comprehension in Wernicke's aphasia. Archives of Neurology, 44: 73-82, 1987.

Perani D, Vallar G, Cappa S, Messa C, Fazio F. Aphasia and neglect after subcortical stroke: a clinical / cerebral perfusion correlation study. Brain 110: 1211-1229, 1987.

Poeck K, De Bleser R, Von Keyserling DG. Computed Tomography localization of standard aphasia syndromes. In Rose FC (ed). Advances in Neurology volume 42: Progress in Aphasiology. Raven Press, New York, p. 71-89, 1984.

Rousseaux M, Steinling M, Kassiotis P, Lesoin F. Débit sanguin cérébral et infarctus thalamiques. Revue Neurologique 145: 140-147, 1989.

Savoiardo M. CT Scanning. In Barnett HJM, Mohr JP, Stein BM, Yatsu FM (eds). Stroke, Volume 1, Churchill Livingstone, New York, p. 189-219, 1986.

Selnes OA, Knopman DS, Niccum N, Rubens AB. The critical role of Wernicke's area in sentence repetition. Ann Neurol 17: 549-557, 1985.

Trimble MR, Reynolds EH (eds). Epilepsy, Behaviour and Cognitive Function. John Wiley and Sons, Chichester, 1988.

Zeumer H, Ringelstein EB. Computed tomography patterns of brain infarctions as a pathogenetic key. In Poeck K, Ringelstein EB, Hacke W (eds). New Trends in Diagnosis and Management of Stroke. Springer-Verlag, Berlin, p. 75-85, 1987.

Zimmerman RA, Bilaniuk LT, Packer RJ, Goldberg HI, Grossman RI. Computed tomographic correlates in acute basal ganglionic infarction of childhood. Neuroradiology 24: 241-248, 1983.

Zulch K-J, Hossman V. Patterns of cerebral infarctions. In Vinken PJ, Bruyn GW, Klawans HL (eds). Handbook of Clinical Neurology, Volume 53, Elsevier, Amsterdam, p. 175-198, 1988.

CLINICAL TECHNIQUES TO ASSESS LANGUAGE IN YOUNG CHILDREN

I. RAPIN AND D.A. ALLEN
Saul R. Korey Department of Neurology, Department of Pediatrics, Division of Child Psychiatry, and Rose F. Kennedy Center for Research in Mental Retardation and Human Development
Albert Einstein College of Medicine
1300 Morris Park Avenue
Bronx NY 10461, USA

ABSTRACT. Clinical evaluation of language in a young child may be difficult because of shyness, negativism, or hypospontaneity of speech. Assessment of spontaneous language during play often overcomes these obstacles and provides information about conversational skills, supplementing formal quantitative tests of language. A language checklist for scoring the various aspects of language live or from recordings lends itself to efficient and serial assessment and ensures systematic data collection. The requirements for reliable scoring are to limit the list to unambiguous readily observable items and to use either dichotomous scores or allow no more than three levels of severity with a continuous scale. Such a checklist is ideal for following recovery from acquired aphasia. It may enable investigators to pool data and, possibly, to carry out cross-linguistic studies of children with acquired aphasias and developmental dysphasias.

1. Introduction

In the practice of physicians who deal with young children, a frequent complaint is that a child has failed to acquire language at the appropriate age or a child who had begun to speak has lost whatever language s/he had acquired. Assessing the language status of a young child may be difficult for professionals who are not specialists in early language development. The problem is further complicated by the fact that many young children are reluctant to speak to strangers, particularly in the physician's office or hospital setting, environments they perceive as hostile. It is extremely important, therefore, to use the parents as primary informants in gathering information needed to determine whether the child has a developmental dysphasia, an acquired aphasia, autism, or generalized cognitive deficits, or whether the child is simply negativistic, shy, or fearful. The parents' answers to the physician's questions will guide the course of the evaluation, but the questions themselves must be based on an understanding of the normal developmental processes in the acquisition of language.

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 45–53.

It is crucial to be cognizant of the variability among normal children in the milestones of early language acquisition if one is to evaluate the language of a preschoool child with a developmental language disorder or a focal brain lesion and an acquired aphasia. These milestones have been the subject of intensive study. There exist screening instruments suitable for use in well-child clinics to enable non-specialists to decide which child's language is sufficiently defective to warrant referral for formal assessment (Hedrick et al., 1984, Capute et al., 1986, Coplan, 1989). Standardized word lists that parents can use to record the vocabulary and comprehension of their child systematically are an effective means for collecting reliable data on the growth of a very young child's language abilities (Bates et al., unpublished).

Quantitative tests administered by speech and language pathologists and psycholinguists provide rich data on the acquisition of vocabulary and syntax (Aram, this volume) but not on the child's language use in a conversational setting or on his/her ability to formulate a complex verbal message coherently. Also, quantitative tests do not lend themselves to repeated day-to-day follow-up of a child recovering from acquired aphasia whose skills may be changing very rapidly. Yet purely informal bedside assessment has clearly proved insufficient to evaluate these changes because of lack of systematic attention to the many aspects of language. What we would like to propose is that investigators and clinicians use a checklist to record these day-to-day changes in an efficient yet standardized manner so as to collect data that will lend themselves to later analysis.

The best approach is to obtain a spontaneous language sample by engaging the child in conversation in a naturalistic setting during play. One is often surprised to hear a previously mute child speak when characters from favorite TV shows are used as props. It is critically important to use an unobtrusive audiotape recorder to record what the child actually said in response to the examiner's attempts to engage him/her in conversation. The tapes can be listened to later and scored along the dimensions to be discussed below. Such recordings provide objective evidence for development or recovery of language over time which might be missed or overestimated if only the examiner's imprecise memory of what transpired is available.

2. Dimensions of Language Scored on Checklist

The dimensions to pay attention to are listed in Table 1. They include (A) comprehension; (B) amount, fluency, and rate of the child's speech; (C) intelligibility and grammaticality; (D) range of vocabulary and meaning, word retrieval; (E) repetition of words and sentences; (F) formulation of spontaneous language; (G) language use and communicative skill; (H) abnormal features such as echolalia, perseveration, paraphasias, jargon.

2.1 A. ASSESSMENT OF COMPREHENSION

In young normal children language production is critically dependent on comprehension. This is not the case in children with the semantic-pragmatic form of developmental language disorder (Rapin and Allen, 1983, Allen, 1989), and in those with acquired transcortical sensory aphasia (Van Hout et al., 1985) in whom spontaneous production of well-formed overlearned sentences is far superior to comprehension. On the other hand,

almost all children deafened by meningitis below the age of 5 years stop speaking within days and savings in terms of relearning language are often quite limited. Rapid loss of verbal expression is also typical of children with the Landau-Kleffner syndrome (acquired epileptic aphasia) (Landau and Kleffner, 1957). It is equally typical of young autistic children whose parents report regression of language, sociability, play, and cognitive skills in the toddler or early preschool years in the absence of known epileptic activity or encephalopathic event (Rapin, in press). It is logical, therefore, to start one's investigation of a child's language by assessing comprehension, even though comprehension is the most difficult aspect of language to evaluate reliably.

There is no means for assessing comprehension that does not require a motor or verbal response to a verbal input. Since lack of response may have many causes besides failure to comprehend, assessment of comprehension is always, of necessity, inferential. The usual approach is to see whether a cooperative child will carry out verbal commands without the need for visual cues or will point to body parts, objects, or pictures the examiner names.

In order to evaluate comprehension in preschoolers, the examiner must know something of the manner in which comprehension of questions develops (Brown, 1973). Early in the second year normal children learn to shake their head "No" when asked questions requiring a yes/no response such as, "Do you want ice cream?", "Are you hungry?", and so on. Typically several months will elapse before the child can nod "Yes" and even later can respond to either/or questions such as "Do you want milk or juice?".

Soon to appear are one word answers to requests for labels in response to "Who's that?", "What's that?", "Where's your nose?" questions. Answers to where questions and what

TABLE 1. Clinical Checklist for Assessing Child Language*

A.	Comprehension
1.	Comprehension of simple commands and questions
2.	Comprehension of abstract language and complex questions
B.	Amount, fluency, rate of speech
3a.	Amount of speech: Less than expected
3b.	" " " : More than expected
4.	Fluency of speech
5a.	Rate of speech: Decreased
5b.	" " " : Increased
C.	Intelligibility, grammaticality
6.	Intelligibility of speech
7.	Speech articulation

TABLE 1. (Continued)

8. Grammar: Word order, presence of "little words" (articles, prepositions, conjunctions, auxiliaries, etc.)
9. Grammatical markers, e.g., for tense, plural, possessive, gender, etc.

D. Range of vocabulary, word retrieval, meaningfulness

10. Range of vocabulary
11. Word retrieval (naming)
12. Word recognition (pointing)
13. Meaningfulness of the communication

E. Repetition

14. Repetition of single words or digits
15. Repetition of sentences

F. Formulation

16. Formulation of a coherent conversation or narrative

G. Language use and communicative skill

17. Communicative use of speech
18. Communicative use of gestures and facial expression
19. Affective prosody: Decreased and/or inappropriate
20. Intonation

H. Abnormal features

21. Echolalia
22. Perseveration
23. Verbal paraphasias
24. Phonemic paraphasias
25. Neologisms
26. Jargon

* Data are most reliable if scored in a yes/no, present/absent, normal/abnormal format. If quantitative observations are desired, we suggest limiting oneself to only 3 levels: (1) Normal; (2) Somewhat abnormal; (3) Very abnormal or cannot do. If desired, such a scale can be collapsed later to a dichotomous normal/abnormal score. All items are referred to what the clinician would consider normal for age.

TABLE 1. (Concluded)

For each item describing language production, there must be an opportunity to score "Nonverbal" for children who do not speak. Also, there needs to be an opportunity to score "No data" for all items since there may be no opportunity to make certain observations on a given day, certain items may be overlooked, etc. "No data" is a score that is clearly distinct from "Absent" or "No" which are valid observations. The distinction between "Cannot do" and "Won't do" is often tenuous. If desired, "Won't do" may be scored as such or it can be scored as "No data". The form should be designed so that there is space to describe salient items and make comments on the child's alertness, cooperation, etc.

questions when the person or object refered to are immediately present are answered earlier than when they are not. More subtle comprehension deficits are evaluated by asking complex information questions (when, why, and how). These questions are answered later since correct answers require an understanding of temporal sequence and causation. Failure to respond to these questions may reflect either cognitive incompetence or inadequate comprehension. It is important, therefore, to observe non-language problem-solving abilities and to determine whether these abilities are superior to verbal comprehension.

The situation in which one is most likely to miss inadequate comprehension is in verbal children who respond to one or another word in a question but miss its full intent. It is very important to pay close attention so as not to miss answers that are only approximately correct. It is in such cases that the use of open-ended higher order questions is most useful.

2.2 B. AMOUNT, RATE, FLUENCY OF SPEECH

The first question to answer is whether the child talks, and if s/he does, how much and how fluently. Of course one must guard against putting the child in a situation where single words or head nods will suffice to fill the requirements of the conversation. That is, asking a child questions that require only yes/no answers or labels is less informative than asking the child about a favorite story or TV show and to use such responses as "Uhuh", "Tell me more", or "I see" to encourage the child to continue speaking.

Lack of fluency, that is, intrusive pauses and repetition of syllables or of words, may reflect either difficulty with the formulation of what the child wants to say, with the retrieval of words, or with the programming and production of speech. Stuttering is often characterized by pauses not only between words but also within words and by effortful production of speech, sometimes accompanied by spastic contraction of the face, neck, and mouth muscles, with poor control of the breath, and also by repetition of syllables not only at the start but within words. Pseudostuttering due to anomia or word retrieval difficulty is more likely to be characterized by pauses and repetition of whole words or phrases.

There are occasional children with language disorders who speak too much (and often

too fast) rather than not enough. These chatter-boxes are likely to be repetitious and often use overlearned scripts to fill in when they are aware that they should be saying something and have difficulty generating novel utterances. Many of these verbose children have less than perfect comprehension and very defective interpersonal conversational skills.

2.3 C. INTELLIGIBILITY AND GRAMMATICALITY

The next aspect of speech to which to pay attention is intelligibility. The question to ask is not only whether the examiner understands what the child is trying to say but whether the parents understand. Is the child intelligible only in context, or out of context as well? Immaturities of speech production may interfere significantly with intelligibility yet not be the sign of either dysphasia or aphasia. Detailed assessment of phonologic production is beyond the reach of non-specialists in early language assessment, but non-specialists can at least ask the parents whether they have trouble understanding their child and note how much they themselves understand. Availability of an audio tape is a necessity if one wishes to make any statement beyond these very rough ones. Deciding whether errors are systematic and defining their origin may require a phonetic transcription, a labor-intensive endeavor that is most useful to speech pathologists planning remediation.

Next, one needs to pay attention to the length, complexity, and grammatic correctness of the child's utterances. Again, non-specialists can only expect to be sensitive to major dimensions of syntax. Does the child speak using mostly nouns and verbs, in a telegraphic style missing the "little words" like articles and prepositions? Does the child mark the plural and the past and future tenses? Are sentences produced with words in the order demanded by the child's native language? Defective syntax is often associated with defective phonology, so that diagnosis of the type of aphasia or dysphasia does not ride solely on the characteristics of the child's grammatic productions. Much information is available on the acquisition of syntax in very young children so that in preschoolers one must take these normal milestones into account when making a judgement about the grammaticality of the child's utterances (Bates et al., 1988). Once again a recorded speech sample provides the opportunity to verify one's clinical impression of the child's competence and to follow it over time.

2.4 D. RANGE OF VOCABULARY, WORD RETRIEVAL, MEANINGFULNESS

If the emergence of syntactic competence is mainly a task for the third year, growth of vocabulary starts in the second year, with a spurt preceding the emergence of the first two-word utterances. By two years of age normal children understand anywhere from a few dozen to several hundred words and produce a slightly lesser number (Bates et al., 1988). By three years, their vocabularies have enlarged to the point where keeping track of how many words they know becomes impossible.

In addition to the number of different words produced in spontaneous language, assessment of the richness of a child's vocabulary may take the form of pointing to pictures or naming them. These two types of tasks enable one to evaluate the dissociation between knowledge, i.e., word recognition, and production, i.e., word retrieval. Children (and adults) recognize many more words than they can retrieve in any given context. Having children spontaneously list as many animals, or articles of clothing, or furniture as they can provides an easy way to evaluate word retrieval and the richness and semantic organization

of the child's vocabulary. Words within a semantic (meaning) category tend to be linked in the lexicon (repository of word meanings) so this type of task provides a clinical way to access this dimension of language.

Word finding difficulties are ubiquitous in all forms of acquired aphasia and developmental dysphasia. Anomia is particularly salient in the nonfluent aphasias but can also be marked in conjunction with receptive deficits. In dysphasic children, anomia, together with difficulty formulating verbal expression, is a prominent feature of one of the clinical syndromes proposed by Allen (1989) and Rapin and Allen (1983).

Besides evaluating the richness of a child's vocabulary, it is also important to determine whether s/he uses this vocabulary to convey meaningful verbal messages. One needs to be attuned to sentences that do not make sense even in a child who uses sophisticated words.

2.5 E. REPETITION OF WORDS AND SENTENCES

Typically, repetition is not investigated when collecting a spontaneous language sample. Inability to repeat in the face of adequate comprehension is a feature of so-called conduction aphasia in adults and denotes a deficit in short-term verbal memory. Repetition failure is typically more marked for strings of single meaningless items than for meaningful grammatical sentences. Inability to repeat what has just been said (immediate recall) is also sensitive to attentional deficits. Inadequate repetition of what could be repeated immediately after an interval occupied by distracting activities denotes defective short term verbal memory. There are many formal tests of verbal memory that provide information about its various components.

2.6 F. FORMULATION

The ability to retell a story or to explain how to play a game is a good way to evaluate the child's ability to formulate expressive language coherently. This type of assessment is inappropriate for children with a very limited output but is very effective for detecting subtle formulation deficits in fully verbal children.

2.7 G. LANGUAGE USE AND COMMUNICATIVE SKILL

Pragmatics, that is the ability to use language communicatively to ask, tell, show, answer, initiate and sustain a conversation which stays on topic, is usually preserved in expressive aphasia. Pragmatics is always deficient in autistic children. It is often impaired in children acquired fluent aphasias and in non-autistic hyperverbal children with the semantic-pragmatic variant of developmental dysphasia (Rapin and Allen, 1983, Allen, 1988). Pragmatics is also impaired in some children with acquired epileptic aphasia who are at high risk for developing frankly autistic features (Rapin, in press). Intonation and pitch (prosody) are often abnormal in verbal children with autistic features: their voice may be high pitched and sing-song, and declarative sentences may sound like questions. The rhythm of their speech may be choppy or overly monotonous, too rapid or too slow, and they may fail to communicate the affective intent of their communication.

In normal speakers, oral language often violates the rules of syntax but uses prosody and gestures to repair these breaches. Tone of voice, mutual gaze, facial expression, and

gestures are crucial to enrich and clarify the intent of verbal utterances. In the absence of these pragmatic supports, miscommunication would be even more frequent than it is. Since communicative intent is conveyed by visual as well as acoustic cues, research on pragmatics requires video recording the conversational partners rather than just audiotaping them.

2.8 H. ABNORMAL FEATURES

There are abnormal features whose occurrence must be noted because of their diagnostic importance in the acquired apahasias. These include paraphasic errors, both literal or phonemic, where one speech sound is substituted for another, and verbal or semantic paraphasias in which an entire word is substituted for another. The substituted word is related in meaning to the target word or may be a more general superordinate or filler word such as "animal" or "thing". Paraphasias were said not to occur in childhood aphasia, in contrast to adult aphasia, but the work of Van Hout et al. (1985) and others has definitively demonstrated that this is untrue. Perseveration, the repetition of words or sentences, and jargon, the production of meaningless speech-like utterances, are other abnormal features of the language of children with either developmental dysphasia or an acquired aphasia.

3. Utility of a language checklist

The clinician interested in determining the type of a child's developmental dysphasia or acquired aphasia and in following his/her progress or recovery over time needs to collect systematic data efficiently at the bedside or in the office. Ideally, s/he should also record the child's language. With the help of several of our colleagues who are participating with us in a project concerned with the differential diagnosis and subtyping of preschool children with dysphasia, autism, and mental deficiency, we have developed an as yet unpublished checklist for rapid clinical scoring of children's language skills. With minimal changes, the checklist is also suitable for following children with acquired aphasia. It can be used live at the bedside or in the office, as well as for the scoring of recorded spontaneous language samples. The checklist is not meant to substitute for formal tests of language but, used systematically, it will enrich formal test data and ensure that adequate and comparable data are collected over time in a given child as well as across different children.

This "low-tech" clinical instrument has the potential for enabling multiple users to compare observations. Since acquired childhood aphasia is relatively rare, there is a real need for investigators to pool their observations. Unfortunately, most published case reports of acquired aphasia in children are incomplete because investigators concentrated on particular aspects of language and did not record systematic observations. Checklists are a convenient and economical way to ensure a minimum of uniformity in clinical data collection. Whether such a modest instrument can be used by speakers of different languages to carry out cross-linguistic studies of acquired aphasia in children remains an exciting challenge for the future.

4. Acknowledgement

Preparation of this chapter was supported in part by Program Project Grant NS 20489 from the National Institute of Neurologic Diseases and Stroke, United States Public Health Service. D. M. Aram and J. Flax collaborated in the development of the checklist to assess the spontaneous language of dysphasic children.

5. References

Allen, D.A. (1989) "Developmental language disorders in preschool children: Clinical subtypes and syndromes", School Psychology Review 18, 442-451.

Bates, E., Bretherton, I., and Snyder, L. (1988) From First Words to Grammar: Individual Differences and Dissociable Mechanisms, Cambridge University Press, New York.

Bates, E., Thal, D., Fenson, L., Dale, P., and Reilly, J. (Unpublished) McArthur Communication Development Inventory: Infant and Toddler Versions. (Available from L. Fenson, Developmental Psychology Laboratory, San Diego State University, San Diego CA 92182, USA).

Brown, R. (1973) A First Language, Harvard University Press, Cambridge MA.

Capute, A., Palmer, F., Shapiro, B., Wachtel, R., Schmidt, S.,and Ross, A. (1986) "Clinical linguistic and auditory milestone scale: Prediction of cognition in infancy". Developmental Medicine and Child Neurology 28, 762-771.

Coplan, J. (1989) ELM Scale: The Early Language Milestone Scale (Revised), Pro-Ed, Austin TX.

Hedrick, D.L., Prather, E.M., and Tobin, A.E. (1984) Sequenced Inventory of Communication Development, University of Washington Press, Seattle.

Landau, W. and Kleffner, F.R. (1957) "Syndrome of acquired aphasia with convulsive disorder in children", Neurology 7, 523-530.

Rapin, I. (in press) "Autistic children: Diagnosis and clinical features" Pediatrics.

Rapin, I. and Allen D.A. (1983) "Developmental language disorders: Nosologic considerations" in U. Kirk (ed.), Neuropsychology of Language, Reading, and Spelling, Academic Press, New York, pp. 155-184.

Van Hout, A., Evrard, P., and Lyon, G. (1965) "On the positive semeiology of acquired aphasia in children", Developmental Medicine and Child Neurology 27, 231-241.

TEST BATTERY FOR LANGUAGE AND SPEECH ASSESSMENT

DOROTHY M. ARAM
Department of Pediatrics
Case Western Reserve University
2101 Adelbert Road
Cleveland, Ohio 44106

ABSTRACT. Psychometric tools available for assessing children's language are reviewed from the perspective of the aspect of language comprehension or production addressed including: phonology; syntax and morphology; the lexicon and semantics; and pragmatics. A framework for speech and language assessment is outlined and assessment batteries appropriate for different ages are suggested. English-based tools will be used to exemplify aspects targeted for assessment.

INTRODUCTION

When I was originally asked to present a paper on speech and language test batteries, I initially suggested that I might speak on a different topic which I thought might be of more interest to this group. Dr. Martins' response to me commented upon the "need for a good characterization of the language defects in children, since many studies suffer from a limited description of them." I therefore agreed to this topic; however, again when I began organizing my thoughts for this paper the same reservations which I initially had in addressing this topic arose. These reservations centered around three problems. First, reviewing the list of participants, I realize that the majority are not linguists, speech-language pathologists or others primarily concerned with speech and language, and I was concerned about the level of detail and potential irrelevance of much of what I could say to many in this group. Second, descriptions and study of speech and language among children with acquired brain lesions may serve multiple purposes, and no one approach will serve all of these purposes. And third, as I have worked only with an English-speaking population, I am knowledgeable principally about speech and language batteries in English, and am aware of the limited comparable tests with normative data in other languages. All of these hesitancies left me wondering what I could say in a half-hour to a group of investigators representing multiple disciplines, purposes and languages. What I decided to do, and will present in the next several minutes, is to: first, briefly comment upon three levels of

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 55–69.

description, depending on the investigators purposes; second, present a framework for summarizing aspects of language which can be addressed at various levels of detail and is not bound to a specific language; and finally, illustrate each aspect of this framework through summarizing some of the specific tests that have been used in studies of children with acquired lateralized lesions and suggesting alternative tests which are available.

DIFFERENT PURPOSES - DIFFERENT LEVELS OF DESCRIPTION

In studies of children with acquired brain lesions in which these children's speech and language have been described, at least three different levels of description/study can be discerned: clinical rating, psychometric assessment and neurolinguistic study. In part, these levels of description represent different investigators' backgrounds and purposes, and in part, I believe they reflect the evolution of an area of study, still in relative infancy in comparison to, for example, adult aphasiology. In no way do I want to suggest that any one level of description is more "correct" than the others; each serves different purposes, and each has its trade-offs. These three levels of description can be ranked in terms of: the time required to accomplish each; the amount of detailed data preserved through each; and the degree of specialized training required to perform each.

Clinical Ratings

Clinical ratings take the least time, preserve the least amount of information and require the least amount of specialized training to administer. These characteristics clearly are beneficial for some purposes but detrimental for others. Benefits include: the ease of collecting such information through rating scales in clinical settings on a large number of children; the user-friendly nature of rating scales making their use and adaptation applicable to a range of clinical settings for a variety of investigators with differing backgrounds; and the real world practicality of summary judgments regarding the presence or absence of a deficit in a given ability area. Experienced clinicians may well be able to arrive at global judgments of a child's functioning in designated areas that are as, or more valid than an array of psychometrically sound tests. Limitations, however, include the variable reference points for judgments largely dependent on the observers' background, interest and experience. The information lost is perhaps the most serious downside of clinical ratings, at least from the perspective of another investigator trying to interpret, replicate or extend the reported findings.

Nonetheless, most of the early reports of children's speech and language following lateralized brain injury have been based on clinical judgments presumably kept in some sort of a rating format, although typically the actual format in which the clinical judgments were recorded is not reported. The reports of Guttmann (1942), Basser

(1962), Alajouanine and Lhermitte (1965), Annett (1973) and Hecaen (1976, 1983) all have been based on clinical ratings of speech and language abilities, and have formed the basis of our knowledge about speech and language following acquired childhood lesions. Clearly if all investigators would record clinical judgments relative to an agreed upon set of speech and language characteristics, on a composite basis we would have a much more extensive data base for addressing some fundamental questions in this area, such as the incidence of various kinds of speech and language disorders following lesions of known laterality, site and age of onset. As Dr. Rapin has addressed clinical ratings in detail, these will not be discussed further here, other than to agree with her position that clinical rating scales have an important function in the initial description of speech and language abilities among children following lateralized brain injury.

Psychometric Assessment

Since Woods and Carey's (1979) seminal paper, we have been begun to see reports of psychometric assessment of the speech and language of children with lateralized brain lesions. Prior to this time, numerous investigators reported results of intelligence tests and used verbal intelligence scores to quantify verbal abilities. As far as I can determine, however, Woods and Carey (1979) were the first to report results of a test battery specifically addressing speech and language abilities apart for intelligence. Since that time a number of us have followed suit including Riva and Cazzaniga (1986), van Dongen and colleagues (1977, 1985, 1987), Vargha Khadem, O'Gorman and Watters (1985), Kiessling, Denckla and Carlton (1983) and some of our studies (Aram and Ekelman, 1987; Aram, Ekelman, Rose and Whitaker, 1985).

Psychometric test batteries are intermediate between clinical ratings and neurolinguistic studies in terms of the amount of time required for administration and analysis, amount of detail recorded and the amount of specialized training required for their use. Psychometric test batteries have been used predominately when the question addressed is whether or not children with lateralized lesions have deficits in a given area relative to their peers; the question basic to much of the work concerned with the degree of early brain specialization versus plasticity for language functions. This question requires comparison to a normative sample or alternatively to a control group matched on the basis of specified criteria.

The benefits of using psychometric batteries are several. First, standardized batteries impose a systematic examination on a given area, forcing an investigator to base judgments on a constant set of variables, with a constant set of stimuli and scored in a uniform manner. Standardized batteries afford constant coverage, consistency and replicability. As mentioned, probably their greatest value is permitting comparison to "normal" children, a necessary comparison for both theoretical (e.g. degree of recovery of brain functions possible when a lesion is incurred at a given site and at a given age) and practical reasons (e.g. how well can we predict this child will

function if returned to a normal classroom). The chief drawback of psychometric tests is that they typically focus upon right or wrong answers, and do not address process or strategy through which an answer is determined. The question addressed is "Can a child perform?" not "How does a child perform?" Thus a psychometric battery approach to description is limited in the questions it can address and the degree to which sophisticated neurolinguistic issues can be pursued.

Neurolinguistic Study

I am using the term, neurolinguistic study, to refer to the indepth study of language abilities, typically involving an array of experimental tasks designed by the investigator to address hypotheses relevant to language performance. Of the three levels of description, this one generally requires the greatest time to administer and/or analyze, the most specialized training in language, and offers the greatest detail of speech and language functions. The chief advantages of such study are the level of detail provided in the language description and the contributions in understanding the processes and strategies through which the observed level of performance is achieved. Hypothesis-driven neurolinguistic study undoubtedly advances our understanding of theoretical issues related to brain and language relationships more than either clinical ratings or psychometric approaches to description of speech and language. Further, the level of detail related to both language performance and process also has the potential of providing an informed basis for developing intervention programs. For some clinical purposes, however, this level of detail is not needed and may be an "overkill", lacking real-world practicality. The frequent lack of normative data for many of the experimental measures used also is a drawback for some clinical purposes.

Although neurolinguistic description has a long tradition in adult aphasiology, beyond a few case studies (e.g. Dennis, 1980), thus far there have been very few such studies with children with acquired lateralized brain lesions (with the notable exception of the work of Maureen Dennis and her colleagues with hemispherectomized children). Perhaps the sparsity of such studies evidences the relative level of maturity of investigations in this area. With the recent work of investigators such as Van Hout, Evrard and Lyon (1985), Marchman, Bates and colleagues (1989) and a couple of our own studies (Aram, Ekelman and Whitaker, 1986; 1987), we are beginning to see more neurolinguistic analyses of children with brain lesions. We are fortunate to have included in this symposium additional papers representing this level of language description, for example the paper presented by Julie Eisele.

A FRAMEWORK FOR SPEECH AND LANGUAGE ASSESSMENT

Turning from these three levels of speech and language description, I have found it useful to view assessment of language from the following

framework. Given time constraints I will not be addressing speech assessment, although clearly this too is in need of description. When approaching assessment of any child's language performance, be they children with developmental or acquired speech and language problems, I consider a comprehensive assessment to include observations or measurement of the following:

TABLE 1. Framework for language assessment

	Comprehension	Production
Pragmatic		
Lexicon/semantics		
Syntax/morphology		
Phonology		

First, I consider it important to make a distinction between comprehension and production of a specific aspect of language; however, surprisingly few studies of children with lateralized brain lesions have addressed language comprehension. Although production often assumes comprehension, this is not always the case, for example in early language acquisition where children may use a word with very limited comprehension of its meaning, or in some forms of language pathology, for example sensory transcortical aphasia or the semantic-pragmatic subtype of children with developmental language disorders described by Drs. Rapin and Allen. Production can be further subdivided between self-generated language or repetition, although again contrasts between self-generated verses repeated language are rarely drawn in the studies of children with acquired brain lesions.

Although the true components of language are a matter of debate, I believe most could agree to at least four aspects: a functional aspect, pragmatics; a meaning component consisting of the lexicon and the meaningful relationship between words, lexicon/semantics; a grammatical structure component, including word order and inflectional endings, syntax/morphology; and a sound component, including sound units and rules for ordering those sounds, phonology. Presumably the participants at this symposium are knowledgeable about these components of language; therefore these will not be defined further, except as exemplified through specific test tools. In addition to language, individual investigators may wish to include aspects of speech, such as articulation, fluency and voice, although because of time constraints these aspects of speech will not be discussed here.

This schema, or framework, then is suggested as a guide for observations and selection of tests. As clinicians, I believe we should attempt to cbtain observations, at least at the level of a rating scale, to judge at a minimum the gross adequacy of each of these components of speech and language. Observations of spontaneous language will permit some degree of judgement for all levels of

language production and motor speech articulation. However, comprehension of each component needs to be inferred from either the child's productive language or from secondary motor responses such as complying with commands, pointing to pictures, or in some other manner responding appropriately. Although a global estimate of comprehension can be ascertained through observation and rating scale formats, assessing comprehension is often difficult without the aid of testing instruments which control stimuli presentation. As a clinical speech and language pathologist, therefore, I supplement observations with standarized tests, some of which will be reviewed shortly. On a clinical basis, I believe we have a responsibility to attempt to screen comprehensively all of these aspects of language. As investigators, however, we may choose to focus on selected components, for example syntactic comprehension, achieved through psychometric assessment and/or neurolinguistic study.

TEST BATTERY EXEMPLARS

It is impossible and probably not useful to attempt to review all the available tests one could use to assess each of these components of speech and language for several reasons. The purposes investigators have and the level of detail desired vary widely. Also a number of practical concerns, beyond the content, dictate which specific test is most appropriate, central considerations being the age of the children tested, and the time one can devote to assessing a specific area. And finally the ever growing number of new tests available, (at least in English) precludes a comprehensive review. For example in our Child Language Disorders book (Aram and Nation, 1982) we devoted 28 pages to cataloging tests available for assessing speech and language in children; undoubtedly the list has expanded greatly since that time.

Before focusing on specific language components, I would like to say a few words about three types of language tests that have been used with children with acquired brain lesions: verbal intelligence measures; adult aphasia batteries; and composite batteries. Reports of Verbal IQ's or scores on verbal subtests are probably the most frequently reported measures of language among children with brain injury. With very few exceptions the verbal subtests of intelligence tests only measure the informational or semantic aspect of language and do not differentiate between comprehension and production, despite subtests labelled "Comprehension". Although some intelligence tests include Memory for Sentences subtests, which could be viewed as tests of syntactic repetition, the syntactic principle motivating these tests is typically obscure, as length seems to be the dominant feature under test. Thus verbal IQ measures address only very limited aspects of language - no phonology, little syntax and no pragmatics. A few investigators have reported attempts to use aphasia batteries developed for adults with their brain lesioned children, for example Dennis (1980) reported using the Spreen Benton Aphasia Battery with her 9 year-old, left-lesioned girl, and van Dongen and Visch-Brink (1985) reported using the Boston Diagnostic Aphasia Battery with their aphasic children. The limitations of these adult aphasia batteries

with children are two-fold: first, except for the Boston Naming Test and portions of the Benton, I am unaware of normative data for children, against which to judge the adequacy of their performance; second is the content which in several instances is not appropriate for children. Finally, several have used composite language batteries developed for children. In several instances these composite batteries do provide relatively good coverage of language comprehension and production, and provide quite appropriate measures particularly when subtests addressing various components of comprehension and production can be broken out and scored independently (for example, the Test of Language Development [TOLD, Newcomer and Hammill, 1982]). Other composite language batteries developed for use with children provide only a very limited number of items assessing any one aspect of language or permit only a single composite language score. Such batteries may fail to be sensitive to subtle, or circumspect language deficits and do not permit comparison of ability level among components of language. We will now turn to exemplars of each language component.

Pragmatics

Other than the case description of a nine-year old girl with a left temporo-parietal infarct reported by Dennis (1980) in which communicative intent was discussed, there have been no studies, of which I am aware, of pragmatic abilities among children with lateralized brain lesions. A number of studies of pragmatic abilities in adults following acquired brain injury, however, have been reported. Several systems for categorizing communicative intent or for analyzing other aspects of pragmatics in children have been reported in the research literature. Recently a couple of these have been standardized, providing normative data. The Test of Pragmatic Skills - Revised (Shulman, 1986) and the Pragmatics Screening Test (Prinz and Weiner, 1989) have been developed for preschoolers using naturalistic, play oriented tasks. The Let's Talk Inventory for Children (Bray and Wiig, 1987) is for use with the older child or adolescent and consists of an extensive protocol requiring a least an hour to administer.

Semantics

Semantics has been addressed predominantly through administration of various vocabulary measures and through subtests of verbal intelligence. For this aspect of language investigators working with children with brain lesions have differentiated between comprehension and production to some degree. Several groups have reported performance on the Peabody Picture Vocabulary Test (PPVT, Dunn, 1965) (Aram, Ekelman, Rose and Whitaker, 1985; Cooper and Flowers, 1987; Kiessling et al., 1983; Levine, Huttenlocher, Banich and Duda, 1987; and Riva and Cazzaniga, 1986), or comparable translations, which have served as measures of vocabulary comprehension. Similarly several naming tasks, such as the Oldfield-Wingfield Picture Naming Test, the

Boston Naming Test and the Expressive One-Word Picture Vocabulary Test (EOWPVT, Gardner, 1979) have provided measures of expressive vocabulary. While valuable within a more comprehensive battery of language measures, these naming tasks are limited to relative concrete, pictureable lexical items, predominantly noun forms. Several investigators also have reported performance on the vocabulary subtest of various intelligence tests. Although vocabulary subtests typically do not differentiate between comprehension and production vocabularies, they do probe lexical knowledge at a more abstract level, for example on the WISC-R requiring definitions for words such as join, brave, compel, and obliterate. Except for case studies, few investigators in this area have gone beyond semantic comprehension of single words. Cooper and Flowers (1987) are among the few who have, assessing comprehension of contextual language through the Processing Spoken Paragraphs subtest of the Clinical Evaluation of Language Functions test (CELF; Semel and Wiig, 1980). A few investigators have used experimental measures to describe other aspects of semantics. For example Woods and Carey (1979) included a Relations Task in which their subjects were asked to determine on semantic or syntactic grounds, the truth value of sentences such as: "My father's sister is my aunt." We (Aram, Ekelman and Whitaker, 1987) used the Rapid Automatized Naming Test (Denckla and Rudel, 1976) and Word Finding Test (Wiegel-Crump and Dennis, 1984) to assess latency and the effect of type of cue on lexical retrieval. Thus beyond vocabulary measures, little investigation of semantic abilities of brain lesionsed children has been systematically pursued.

Suggestions for tests of semantic abilties to consider in a comprehensive language battery are as follows. First for very young children (i.e. infancy and toddlers), Bates and her collegues have developed a very useful tool based on parental report, The MacArthur Communicative Development Inventory (1989); two forms for successive developmental levels are available. The inventory for infants differentiates between comprehension and production of words in a range of semantic categories and also surveys use of actions and gestures. The toddler inventory tracks only the productive use of words, but also includes a section on early sentences and grammar, assessing predominantly morphological forms and early sentence forms. These inventories should be readily translatable, and normative work in languages other than English may be underway. Beyond the early toddler stage, a standardized test of vocabulary comprehension and one of productive vocabulary probably should be included, such as the Peabody Picture Vocabulary Test (Dunn, 1965) for the former and the Expressive One-Word Picture Vocabuary Test (Gardner, 1979) for the latter, although there are numerous alternatives available in several languages. Because subgroups of children with language problems may understand concrete words relatively well and label pictures readily, but fail to see relationships between words, including semantic assessment tools which go beyond vocabulary assessment is important. Of the standardized, commercially available alternatives, I suggest two. The Clinical Evaluation of Language Fundamentals - Revised (CELF-R, Semel, Wiig and Secord, 1987) includes three subtest

assessing semantic comprehension: the Word Classes subtest asks children to identify which 2 of 4 words go together; the Semantic Relationships subtest assesses comparative, spatial, passive and temporal relationships; and the Listening to Paragraphs subtest assesses comprehension of content in extended contexts. The second suggestion, The Word Test: A Test of Expressive Vocabulary and Semantics (Jorgensen, Barrett, Huisingh and Zachman, 1981), includes subtests assessing word associations, (identify which word does not belong and why: empty, story, sunny, windy), synonyms, semantic absurdities (tell me what doesn't make any sense about this: He sprinkled a glass of milk for me.), antonyms, definitions, and multiple definitions. Numerous other tests are available, however, in both standardized form and in the research literature (refer to Aram and Nation, 1982; Lahey, 1988, for a listing).

Svntax/Morphology

A variety of tests have been used sporadically to assess comprehension and/or production of syntax and morphology among children with lateralized brain lesions; yet given the centrality of syntactic comprehension, it is surprising that so little assessment of this area has been undertaken with children with brain lesions. Probably the most commonly used measure of comprehension of conected language used with both children and adults has been the Token Test (Disimoni, 1978; McNeil and Prescott, 1978) which, as I am sure all are aware, comes in an array of forms in an array of languages. Varying forms have been used with children with lesions including reports by van Dongen and Loonen, 1977; Vargha-Khadem et al., 1985; Riva and Cazzaniga, 1986; and Aram and Ekelman, 1987. The advantanges and disadvantages of the Token Test have been discussed extensively. Primary among the advantages are the limited and controlled vocabulary, the independent assessement of the effect of length versus sentence complexity, and the extensive literature against which to compare results. Limitations include: the concrete nature of the commands; and depending on the form used, the limited range of syntactic forms or limited number of exemplars assessed. Other experimental measures of syntactic comprehension reported with brain lesioned children include Kiessling et al.'s (1983) use of a syntactic awareness test composed of 64 sentences in which the child is asked to identify whether each sentence sounds funny (syntactically incorrect) or "O.K." (syntactically correct); and Woods and Carey's (1979) inclusion of two experimental tests addressing syntactic comprehension, a that-clause task (comprised of 20 sentences each containing a relative clause beginning with that, 12 of which were anomalous because of subject-object reversal either within or across relative clause boundaries, e.g., "The boat that built the boy went in for lunch."), and an ask-tell distinction task. Dennis and Kohn (1975) have also developed an Active-Passive Task which they have used with children with hemispherectomies. Later in this symposium we will learn of a further experimental study of comprehension of complex language structure in brain lesioned children presented by Julie Eisele. All of these tasks

have addressed relatively circumsribed aspects of syntactic comprehension and few have extensive normative data, beyond the Token Test.

It may be difficult to arrive at a common assessment battery for syntax given the syntactic demands of varying languages and the few standardized tools available in most languages. The Token Test would appear to be an appropriate inclusion in an assessment battery, provided its limitations in terms of structures assessed and exemplars provided are kept in mind. Forms of the Token Test are available which provide norms for children (DiSimoni, 1978) and which permit analysis of subtle response variations, (McNeil and Prescott, 1978). Other standardized tests of syntactic comprehension available in English include: the Northwestern Syntax Screening Test (Lee, 1971); the TOLD Grammatic Understanding Subtest (Newcomer and Hammill, 1982); and the Linguistic Concepts, Sentence Structure and Oral Directions subtests of the CELF-R (Semel, Wiig and Secord, 1987).

Regarding productive syntax in children with lateralized lesions, here too surprisingly little systematic survey of abilities has been reported beyond case reports. Again, the few reports available have relied on either subtests from intelligence tests, for example use of the Binet Sentences (Kiessling et al., 1983), or use of various experimental measures including a repetition test used by van Dongen and Loonen (1977), a sentence completion task reported by Woods and Carey (1979), and a speech shadowing task reported by Woods (1987), in which patients with unilateral lesions were asked to report syntactically correct sentences or random word order sentences presented at differing rates. As with syntactic comprehension, each of these tasks assesses only a limited range of a child's productive syntax. We (Aram, Ekelman and Whitaker, 1986) analyzed the spontaneous spoken language of children with unilateral brain lesions, reporting use of both inflectional morphology and sentence complexity. While the degree of detail and coverage provided through spontaneous language samples is a strength for some research questions, the time and training required for analysis renders such analyses inappropriate for many clinical settings. Fortunately several computer based programs are now available to assist syntactic analysis of spontaneous productive language, for example, Systematic Analysis of Language Transcriptions, Lingquest, and CHILDES, yet the data produced may be too massive for many purposes.

Several normed alternatives are available to screen productive syntax, although each only taps a small portion of the possible structures. As previously mentioned, for toddlers the MacArthur Communicative Development Inventory (1989) is a well-standardized inventory based on parent report. Suggested alternatives for older children include: the Northwestern Syntax Screening Test (Lee, 1971); the TOLD Grammatic Completion subtest, (Newcomer and Hammill, 1982); and three subtests from the CELF-R (Semel, Wiig and Secord, 1987), the Formulated Sentences subtest, (requiring spontaneous generation of a sentence comprising a stimulus word), the Recalling Sentences subtests, (a sentence repetition task), and the Sentence Assembly subtest (ordering sentencs given written sentence fragments).

Phonology

Finally, the phonological aspect of language has received the least descriptive attention. Beyond a few case reports of phonemic paraphasias, particularly acutely following acquired brain lesions in children (Van Hout, Evrard and Lyon, 1985; Visch-Brink and Sandt-Koenderman, 1984), few studies have addressed the speech characteristics of children with lateralized brain injury. Although many investigators have commented upon the incidence of speech or articulation disorders among children with lateralized brain injury, rarely is the form of screening or any specific assessment tool cited. Other than case reports, I was unable to locate any study except our own work which provided actual test data on any standardized measure for either phonological compehension or production. In a study of reading, we (Aram, Gillespie and Yamashita, in press) assessed phonetic analysis and segmentaion stills in children with unilateral lesions using the phonetic analysis subtest of the Stanford Diagnostic Reading Test (Karlsen, Madden, Gardner and Karlsen, 1976) and the Sound Analysis and the Sound Blending subtests from the Goldman-Fristoe-Woodcock Test of Auditory Discrimination (1972). In a study just completed, we (Shriberg and Aram, under review) addressed the spoken phonology of children with unilateral brain lesions based on responses to the Photo Articulation Test (Pendergast, Dickey, Selman and Soder, 1969) (both spontaneous naming and imitation), repetition of multisyllabic words and analysis of a spontaneously produced, connected speech sample - all transcribed phonetically and aided by a computer assisted analysis program developed by Larry Shriberg (PEPPER, 1986). Clearly this level of analysis is not indicated except for interesting case studies or particular experimental studies; yet some screening of phonological skills should be included in a comprehensive language battery. Numerous normed tests are available purporting to assess some aspect of phonological skills, often included as parts of reading batteries or auditory processing batteries (see Lahey, 1988, or Aram and Nation, 1982, for a summary of some of these tools). Of the multitude of tests available in English, I would suggest either subtests from the Goldman-Fristoe-Woodcock Test of Auditory Discrimination (1972) or the speech discrimination subtest included on several of the composite language batteries developed for children such as the Test of Language Development (TOLD, Newcomer and Hammill, 1982). For articulated phonology, again numerous tests are available and everyone seems to have their favorite. The Photo Articulation Test (Pendergast et al., 1969), the Templin-Darley Tests of Articulation (Templin and Darley, 1969) and the Kahn-Lewis Phonological Analysis (Kahn and Lewis, 1986) for use with the Goldman-Fristoe Test of Articulation (Goldman and Fristoe, 1986) are three frequently used measures.

SUMMARY

In summary, I would like to reiterate three central points. First, I have suggested that description of children's speech and language may

proceed at varying levels, outlining three: clinical rating, psychometric assessment and neurolinguistic study. These levels of description differ in terms of the time required to accomplish each, the amount of data preserved through each and the degree of specialized training required to perform each. Secondly, a uniform framework has been outlined, suggested for investigators in this area as a guide for observations and selection of tests to assess various aspects of language comprehension and production. Third, since this paper was supposed to address standarized batteries for language assessment, suggestions have been made for a composite battery for English-speaking children or from which adaptations might be made for other languages. Finally, whatever level of description used or specific tests selected or developed, the overwhelming conclusion that emerges from review of work in this area is that as an area of study, we have only begun to describe and understand factors related to the language of brain-lesioned children. Hopefully this symposium and the papers presented here will stimulate further work in this potentially rich area of study.

ACKNOWLEDGEMENT

Preparation of this manuscript was supported by a National Institute of Neurological Disorders and Stroke Grant NS 17366 to Dr. Aram.

REFERENCES

Alajouanine, T.H., & Lhermitte, F. (1965). Acquired aphasia in children. Brain, 88(4), 653-662.

Annett, M. (1973). Laterality of childhood hemiplegia and the growth of speech and intelligence. Cortex, 9, 4-33.

Aram, D.M., & Ekelman, B.L. (1987). Unilateral brain lesions in childhood: Performance on the Revised Token Test. Brain and Language, 32, 137-158.

Aram, D.M., Ekelman, B.L., Rose, D.F., & Whitaker, H.A. (1985). Verbal and cognitive sequelae following unilateral lesions acquired in early childhood. Journal of Clinical and Experimental Neuropsychology, 7, 55-78.

Aram, D.M., Ekelman, B.L., & Whitaker, H.A. (1986). Spoken syntax in children with acquired unilateral hemisphere lesions. Brain and Language, 27, 75-100.

Aram, D.M., Ekelman, B.L., & Whitaker, H.A. (1987). Lexical retrieval in left and right brain lesioned children. Brain and Language, 31, 61-87.

Aram, D.M., Gillespie, L.L., & Yamashita, T.S. (Under review). Reading among children with left and right brain lesions. Developmental Neuropsychology.

Aram, D.M., & Nation, J.E. (1982). Child Language Disorders. St. Louis: C.V. Mosby Co.

Basser, L.S. (1962). Hemiplegia of early onset and the faculty of speech with special reference to the effects of hemispherectomy. Brain, 85, 427-460.

Bates, E., Beeghley, M., Bretherton, I., McNew, S., Oakes, L., O'Connell, B., Reznick, S., Shore, C., Snyder, L., Volterra, V., & Whitesell, L. (1985). Language and gesture inventory. Unpublished test materials, University of California, San Diego.

Bray, C., & Wiig, E. (1987). Let's Talk Inventory for Children. San Antonio, Texas: Psychological Corp.

Cooper, J.A., & Flowers, C.R. (1987). Children with a history of acquired aphasia: Residual language and academic impairments. Journal of Speech and Hearing Disorders, 52, 251-262.

Denckla, M.B., & Rudel, R.G. (1976). Rapid "automatized" naming (R.A.N.): Dyslexia differentiated from other learning disabilities. Neuropsychologia, 14, 471-479.

Dennis, M. (1980). Strokes in childhood I: Communicative intent, expression, and comprehension after left hemisphere arteriopathy in a right-handed nine-year old. In R.W. Rieber (Ed.), Language development and aphasia in children (pp. 45-67). New York: Academic Press.

Dennis, M., & Kohn, B. (1975). Comprehension of syntax in infantile hemiplegics after cerebral hemidecortication: Left hemisphere superiority. Brain and Language, 2, 472-482.

DiSimoni, F. (1978). The Token Test for Children. Boston: Teaching Resources Corp.

Dunn, L.M. (1965). Peabody Picture Vocabulary Test. Circle Pines, Minnesota: American Guidance Service.

Gardner, M.F. (1979). Expressive One-Word Picture Vocabulary Test. Novato, California: Academic Therapy Publications.

Goldman, R., & Fristoe, M. (1986). The Goldman-Fristoe Test of Articulation. Circle Pines, Minnesota: American Guidance Service.

Goldman, R., Fristoe, M., & Woodcock, R. (1972). The Goldman-Fristoe-Woodcock Test of Auditory Discrimination. Circle Pines, Minnesota: American Guidance Service.

Guttmann, E. (1942). Aphasia in children. Brain, 65, 205-219.

Hecaen, H. (1976). Acquired aphasia in children and the ontogenesis of hemispheric functional specialization. Brain and Language, 3, 114-134.

Hecaen, H. (1983). Acquired aphasia in children: Revisited. Neuropsychologia, 21(6), 581-587.

Jorgensen, C., Barrett, M., Huisingh, R., & Zachman, L. (1981). The Word Test: A Test of Expressive Vocabulary and Semantics. Moline, Illinois: LinguiSystems.

Kahn, L., & Lewis, N. (1986). Kahn-Lewis Phonological Analysis: for Use with the Goldman-Fristoe Test of Articulation. Circle Pines, Minnesota: American Guidance Service.

Karlsen, B., Madden, R., Gardner, E.G., & Karlsen, B. (1976). Stanford Diagnostic Reading Test. New York: Harcourt, Brace, Jovanovich.

Kiessling, L., Denckla, M., & Carlton, M. (1983). Evidence for differential hemispheric function in children with hemiplegic cerebral palsy. Developmental Medicine and Child Neurology, 25, 724-734.

Lahey, M. (1988). Language Disorders and Language Development. New York: MacMillan Publishing Co.

Lee, L. (1971). Northwestern Syntax Screening Test. Evanston, Illinois: Northwestern University Press.

Levine, S.C., Huttenlocher, P., Banich, M.T., & Duda, E. (1987). Factors affecting cognitive functioning of hemiplegic children. Developmental Medicine and Child Neurology, 29, 27-35.

MacArthur Communicative Development Inventory. (1989). San Diego, California: Developmental Psychology Lab, San Diego State University.

Marchman, V., Miller, R., & Bates, E. (1989). Prespeech and babble in children with focal brain injury. Paper presented at the Society for Research in Child Development Bicentennial Conference, Kansas City, Missouri.

McNeil, M.R., & Prescott, T.E. (1978). Revised Token Test. Austin, Texas: Pro-Ed, Inc.

Miller, J., & Chapman, R. (1982-87). SALT: Systematic Analysis of Language Transcripts. A computer program designed to analyze free speech samples - Apple II Version. Madison, Wisconson: University of Wisconsin, Language Analysis Laboratory, Waisman Center.

Newcomer, P., & Hammill, D. (1982). Test of Language Development - Primary Edition. San Antonio, Texas: Psychological Corporation.

Pendergast, K., Dickey, S.E., Selman, J.W., & Soder, A. (1969). Photo Articulation Test. Danville, Illinois: The Interstate Printers & Publishers Inc.

Prinz, P., & Weiner, F. (1989). Pragmatics Screening Test (PST). San Antonio, Texas: Psychological Corporation.

Riva, D., & Cazzaniga, L. 1986. Late effects of unilateral brain lesions sustained before and after age one. Neuropsychologia, 24(3):423-428.

Semel, E., & Wiig, E. (1980). Clinical Evaluation of Language Functions. San Antonio, Texas: Psychological Corp.

Semel, E., Wiig, E., & Secord, W. (1987). Clinical Evaluation of Language Fundamentals - Revised. San Antonio, Texas: Psychological Corp.

Shriberg, L.D. (1986). PEPPER: Programs to Examine Phonetic and Phonologic Evaluation Records. Madison, Wisconsin: University of Wisconsin-Madison Software Development and Distribution Center.

Shriberg, L.D., & Aram, D.M. (Under review). Speech characteristics of brain lesioned children.

Shulman, B. (1986). Test of Pragmatic Skills - Revised. Tucson, Arizona: Communication Skill Builders.

Templin, M.C., & Darley, F.L. (1969). Templin-Darley Tests of Articulation (2nd ed). Iowa City, Iowa: Bureau of Educational Research and Service, University of Iowa.

Thal, D., Marchman, V., & Bates, E. (1989). Early communication and language. Paper presented at the Society for Research in Child Development Bicentennial Conference. Kansas City, Missouri.

van Dongen, H.R., & Loonen, M.C.B. (1977). Factors related to prognosis of acquired aphasia in children. Cortex, 13, 131-136.

van Dongen, H.R., & Visch-Brink, E.G. (1988). Naming in aphasic children: Analysis of paraphasic errors. Neuropsychologia, 26, 629-632.

van Dongen, H.R., Loonen, M.C.B., and van Dongen, K.J. (1985). Anatomical basis for acquired fluent aphasia in children. Annals of Neurology, 17, 306-309.

Van Hout, A., Evrard, P., & Lyon, G. (1985). On the positive semiology of acquired aphasia in children. Developmental Medicine and Child Neurology, 27, 231-241.

Vargha-Khadem, F., O'Gorman, A.M., & Watters, G.V. (1985). Aphasia and handedness in relation to hemispheric side, age at injury and severity of cerebral lesion during childhood. Brain, 108, 677-696.

Visch-Brink, E.G., & Sandt-Koenderman, M. (1984). The occurrence of paraphasias in the spontaneous speech of children with an acquired aphasia. Brain and Language, 23, 258-271.

Wiegel-Crump, C.A., & Dennis, M. (1984). The Word Finding Test. Toronto: The Hospital for Sick Children (unpublished test).

Woods, B.T. (1987). Impaired speech shadowing after early lesions of either hemisphere. Neuropsychologia, 26, 519-525.

Woods, B.T., & Carey, S. (1979). Language deficits after apparent clinical recovery from childhood aphasia. Annals of Neurology, 6(5), 405-409.

IV - HEMISPHERIC SPECIALIZATION

THE ONTOGENESIS OF HEMISPHERIC SPECIALIZATION: INSIGHTS FROM ACQUIRED APHASIA OF CHILDHOOD

Bryan T. Woods, M.D.
McLean Hospital, Department of Neurology
and Brain Imaging Center
115 Mill Street
Belmont, MA 02178, U.S.A.

ABSTRACT. The current state of knowledge about the developmental course of hemispheric specialization in man rests in large part on concepts derived from the study of acquired aphasia of childhood. Basic questions center about the concepts of inter- and intra-hemispheric functional equipotentiality, progressive lateralization/localization of function, and regional functional commitment. It is suggested that the two hemispheres begin with a high degree of equipotentiality that is gradually lost beginning with late prenatal development, albeit with an asymmetric time-course. It is further suggested that progressive loss of regional potential for one function comes with progressive actualization of another rather than through progressive localization of initially widespread functional activity. Finally, it is suggested that newer techniques for imaging brain metabolic activity will permit experimental testing of these assumptions.

1. INTRODUCTION

Historians of science tell us that progress in any area is rarely smooth and steady, but rather marked by an irregular pattern of sudden accelerations interspersed with long periods of slow and at times barely discernible advances. The study of acquired aphasia in children certainly exemplifies such an irregular rate of progression. After a long era of near stasis in the clinical neurological understanding of the problem during the first half of this century, fresh and apparently contradictory insights began to emerge from the newer methods of psycholinguistics, neuropsychology, and clinical neurophysiology. Beginning about 20 years ago the conflict between theories based on clinical observations and those based on these other methods began to be resolved as acquisition of newer clinical data and critical reappraisal of the older data led to changes in the clinical theories that brought them much closer to the experimentally-based views.

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 73–81.

At this point it would appear that there is considerable congruence of opinion in the field as to what we know and we don't know about the normal developmental course of language localization and the modification of this course by early brain injuries. Based on this perceived congruence it is possible to sketch the outlines of a model that I believe to be reasonably consistent with the currently available data. Of course those of you who are familiar with the history of this field are aware that the landscape is littered with the wreckage of previous models that were also thought to be "consistent with the currently available data." Why then try again? I would suggest to you that the reason to try again is that the earlier models shared one supremely valuable attribute; they made clear predictions that could be falsified by further observations and newer methodologies. It is just because I believe we are on the verge of having available to this field such a powerful critical new methodology that I think it worthwhile to try to provide some testable and falsifiable hypothesis about the ontogeny of language lateralization.

I would like first to discuss some key concepts in the field, and define them as they are applicable to the hypothesis. Second, I shall sketch the model itself. Finally, I shall turn to the potential offered by new methodology.

2. CONCEPTS

2.1. Equipotentiality

"Competence on demand"--the ability of one brain region to substitute for another damaged or nonfunctional region and to carry out fully a function that would in the normal order of development have been localized to the damaged region.

2.1.1. Operationally, albeit not semantically, equipotentiality can be usefully thought of as a property with gradations ranging from zero to unity; this permits us in principle to say things like "at 40 weeks gestational age the mean language equipotentiality quotient of the right hemisphere is .9."

2.1.2. Equipotentiality can only be observed under pathological conditions--the region normally subserving a function must be unable to do so.

2.1.3. We can ask about the functional equipotentiality of any brain area, whether homologous or not, and whether ipsilateral or contralateral.

2.2. Progressive Lateralization

As originally used, this term referred to the developmental progression of language function localization from both hemispheres

to only the left hemisphere. It seems reasonable, however, to think of progressive lateralization as a special case of progressive localization (cf Satz et al. 1990), by which is meant a gradual constriction during normal development of the brain area involved in sustaining a function at its full operational level.

2.2.1. Progressive localization is not related to pathology, and, contrary to equipotentiality, it should in principle be observable in normal individuals over the course of development. Even though it was originally postulated as the mechanism underlying early equipotentiality for language it is logically independent of it.

2.2.2. As a concept, progressive localization should be kept distinct from the process of progressive automatization that takes place during learning. Progressive localization is related to developmental changes in the anatomical localization of the neural mechanism underlying an unchanging function, whereas progressive automatization implies a functional change and can presumably occur at any age.

2.3. Progressive Substitution

This term refers to the functional replacement of one brain region by another over the course of normal development. This mechanism, when applied to different species, is a mainstay of evolutionary biology, and is embodied in terms such as progressive encephalization. In recent years its applicability to individual development has been elucidated by the work of Goldman-Rakic and colleagues studying "frontal lobe" functions in monkeys (Goldman-Rakic et al. 1983). Progressive substitution differs from progressive localization in that in the latter case the ultimate area of localization is from the outset part of a broader area subserving the function, while in the case of progressive substitution the final area of localization plays no substantive role in the initial exercise of the function.

3.1.1. The concept of progressive substitution is intriguing, but the supporting evidence, derived from the apparent functional equivalence of earlier and later test performance, may be due to limitations of the testing procedure; if the testing cannot distinguish between the use of different strategies to accomplish the same task, then one may erroneously conclude that the different brain areas are performing truly identical functions at different points in development (Kinsbourne and Hiscock, 1983).

2.4. Functional Commitment

The idea that a brain region has the potential to substitute for another after a pathological insult to the latter only so long as it was not committed to its own proper function has a very long history (Kinsbourne and Hiscock, 1983). However, recent quantitative studies of numbers of neurons, dendrites, and synapses in discrete brain

regions at different stages of development indicate that while neuron number and dendritic length first increase and then level off, synaptic number decreases sharply at about the time in development that the region being studied becomes fully functional (Huttenlocher, 1990). This observation leads to the suggestion that synaptic pruning may parallel regional commitment. According to this notion the presence of an excess of synapses confers the potential for a variety of alternate functional organizations of the same area, but actualization of one pattern of organization would lead to a dropout of unutilized and superfluous synapses and loss of potential for alternate functional patterns.

3. THE MODEL

Given these basic conceptual building-blocks, what is the most plausible working model of the developmental course of localization and lateralization of language and other higher functions under not only normal but also pathological conditions? Table 1 shows a schematic outline for language and for a generic right-lateralized visuospatial function that is postulated to develop during the sixth post-gestational year. The table indicates the state of affairs at different time intervals for each hemisphere with regard to both actual functional activity and potential for taking over function after lateralized brain injury.

Several points should be noted. First, I have postulated true equipotentiality in the early stages of embryonic and fetal development, even though empirical data to support this directly are largely lacking; the assumption is based on both logical considerations arising form the dissociability of handedness and speech lateralization and on a backward extrapolation from empirical data for later developmental stages. The logical argument is as follows: Fully adequate speech can develop not only in the right hemisphere of a large minority of left-handers but in that of a small number of right-handers. This pattern of atypical lateralization is not known to be on a genetic basis, especially for the right-handers, and so presumably arises from some unusual stimulus early in development; a stimulus that shifts language to an at that time equipotential right hemisphere.

Second, the loss of equipotentiality is shown as gradual rather than all-or-none. The original study by Woods and Carey (1980) could not conclusively demonstrate specific language impairments in children with left hemisphere lesions acquired before age one, but the work of Dennis and Kohn (1975) and Vargha-Kadem et al. (1985) suggest that this younger group does not escape at least mild language impairment, implying that the loss of right-sided equipotentiality for language occurs gradually, rather than at some threshold (cf Lenneberg 1967, Krashen 1973; for still another view, cf Aram, 1988).

The third point to note is that the hemispheric loss of equipotentiality is asymmetric and determined by the differential

Table 1: Functional activity and functional potentiality by hemisphere for language and a prototypical visuospatial function at different developmental ages.

AGE	- - - - - LANGUAGE - - - - -		- - VISUOSPATIAL FUNCTION - -	
	LH	RH	LH	RH
Early Prenatal	No actual function.	Full potential.	Full potential.	No actual function.
Late Prenatal-Perinatal	Early beginnings of activity.	No actual function. Slight loss of potential.	Slight loss of potential.	No actual Function
Age 1-5	Essentially full development of spoken language.	Slow regression to mild loss of potential.	Regression to moderate loss of potential.	Early beginnings of activity.
Age 5-8	Development of ancillary functions; reading, writing.	Further slow regression to moderate loss of potential.	Marked loss of potential.	Essential full development of function
Age 8+	Adult-level function.	Regression to adult-level residual potential.	Adult-level residual potential.	Adult-level function

rate at which the relevant regions of the hemispheres are committed to their respective functions. This conclusion is supported in large part by Wechsler test data which indicate that right hemisphere lesions already have a restricted effect on verbal IQ (VIQ) scores (i.e., left hemisphere function) at an age (of injury) when left hemisphere lesions still lower performance IQ (PIQ) scores (i.e., right hemisphere function) as well as VIQ. The degree of this asymmetry remains less certain. Our results (Woods, 1980) suggest that prior to age one, both hemispheres have similar degrees of equipotentiality for contralateral functions, but another study using Wechsler data (Riva and Cazzaniga, 1986) suggests a still earlier loss of left hemisphere potential for taking over the functions tested by PIQ.

There remain several critical questions. First, what is necessary to bring about a shift in lateralization of a function? It is tempting to postulate that part of the mechanism of lateralization of functions to regions which have approximately equipotential contralateral homologies involves some kind of reciprocal inhibitory mechanism; the first area to be active inhibits its contralateral homologue. It follows that only a lesion severe enough and long-lasting enough to turn off inhibition of the contralateral area and allow it to begin functioning will lead to a shift in lateralization; a lesion that spares contralateral inhibition will not trigger a shift. It follows as a corollary that equipotentiality also includes this contralateral inhibition capacity, and that once the shift takes place the newly active area will in turn inhibit the damaged area and prevent it from becoming active again, even if some recovery takes place. Moreover, the gradual loss of equipotentiality will pari passu raise the threshold of damage which will result in a permanent shift of function to the contralateral side, because that side is gradually less and less able to inhibit even a damaged region of origin.

One further question that is currently being actively investigated (Hecaen, 1983; Satz et al. 1990) is the extent to which equipotentiality and progressive localization may be relevant to the ipsilateral hemisphere. Just as the clinical basis for the progressive lateralization hypothesis was the apparent excess of crossed aphasias in children, the basis for a progressive localization hypothesis is the putatively uniform motoric aphasia seen in these young patients. And just as the paucity of crossed childhood aphasia cases in modern series undercut the concept of progressive lateralization, recent reports of anterior subcortical aphasia (Aram et al. 1983), conduction (Martins and Ferro, 1987), and even jargon aphasia (Van Haut and Lyons, 1985; Visch-Brink and Van de Sandt-Koenderman, 1984) in children should be cause for grave reservations about the claim that language is differently organized in the immature brain.

As regards equipotentiality of brain tissue adjacent to lesions of active speech areas, it should first of all be said that small lesions of speech areas in adults can be followed by apparent recovery. The relevant question should then be whether or not, given

lesions of equal size and location, the size of the surrounding region that can support an ipsilateral return of language function is inversely related to age at time of lesion; i.e., is larger in the young child. Although one would suspect that the answer to this is yes, it may not prove easy to confirm it.

4. FUTURE INVESTIGATION

I now turn to the ultimate question for this or any model, "Can it be tested and disproven?" I believe that the answer to this is yes. The basis for my optimism is as follows: In the last ten years there has begun to emerge data on changes in regional cerebral energy metabolism induced by changes in cerebral function. In the last few years investigators have reported data on brain activation during linguistic tasks by normal volunteers acquired with Positron Emission Tomography (PET) scanning, using the tracer isotope oxygen-15 (Petersen et al. 1990). The technique of PET scanning has also been applied to children, but because of appropriate ethical concerns, can only be used in certain clinically ill patients (Chugani and Phelps, 1986). There is, however, currently becoming available another technology that is non-invasive and carries no risk of radiation. Magnetic resonance spectroscopy (MRS) actually was developed long before magnetic resonance imaging (MRI), but its application in vivo has been slowed by a variety of technical difficulties. Within the last two years, however, it has progressed dramatically in two respects: first, in the ability to provide quantitative data about short-term changes in localized brain energy metabolism (i.e., turnover rates) in response to stimuli, and second, in the ability to acquire brain metabolic information simultaneously over whole brain slices with a resolution of about one cubic centimeter (so-called chemical shift imaging).

Two laboratories (Prichard et al. 1989; Sappey-Marinier et al. 1990) have already shown measurable increases in lactate levels in normal human primary visual cortex in responses to flashes of light (photic stimulation), and the basic tools and methods are at hand that will permit temporally and spatially resolved imaging of localized human cerebral functions both at rest and when engaged in specific tasks. Moreover, since the process is non-invasive and involves no exposure to ionizing radiation, its application to normal adults and children does not pose ethical problems.

It is this technology that should allow testing and perhaps falsification of the model I have sketched. By following normal children during development it should at last be possible to determine directly whether or not language functions undergo progressive lateralization. By studying individuals with early brain lesions it should be possible to determine how the functional organization of the damaged nervous system differs from the normal. Similar intriguing questions about male-female or right-hander--left-hander language organization may also begin to be answered.

One final observation must be made about this new technology. It will provide an enormously powerful tool for studying the organization of higher-order brain functions, but its productive use will require a thorough understanding of neuropsychology, cognitive psychology, behavioral neurology, and neuropsychiatry. As a general rule, the people who have up to now taken the lead in implementing the technology have relatively little experience in the design of experiments that address some of the vexing but interesting questions about human behavior and higher-order brain function. If the PET example teaches us anything, it is that only the research groups that have established truly effective collaborations with behavioral scientists are likely to generate much in the way of clinically or theoretically interesting results in the area of brain-behavior relationships.

5. REFERENCES

Aram, D.M., Rose, D.F., Rekate, H.L., Whitaker, H.A. (1983) 'Acquired capsular/striatal aphasia in childhood', Arch. Neurol. 40, 614-617.

Aram, D.M. (1988) 'Language sequelae of unilateral brain lesions in children', in F. Plum (ed.), Language, Communication, and the Brain, Raven Press, New York.

Chugani, H.T. and Phelps, M.E. (1986) 'Maturational changes in cerebral function in infants determined by ^{18}FDG positron emission tomography', Science 231, 840-843.

Dennis, M. and Kohn, B. (1975) 'Comprehension of syntax in infantile hemiplegics after cerebral hemidecoritcation: Left hemisphere superiority,' Brain and Language 2, 472-482.

Goldman-Rakic, P.S., Isseroff, A., Schwartz, M.L., Bugbee, N.M. (1983) 'The neurobiology of cognitive development', in P. Mussen, M. Haith, J. Campos (eds.), Handbook of Child Psychology, (4th Ed), J. Wiley & Sons, New York.

Hecaen, H. (1983) 'Acquired aphasia in children revisited', Neuropsychologia 21, 581-587.

Huttenlocher, P.R. (1990) 'Morphometric study of human cerebral cortex development', Neuropsychologia 28, 517-527.

Kinsbourne, M. and Hiscock, M. (1983) 'The normal and deviant development of functional lateralization of the brain', in P. Mussen, M. Haith, J. Campos (eds), Handbook of Child Psychology, (4th Ed), J. Wiley & Sons, New York.

Krashen, S. (1973) 'Lateralization, language learning, and the critical period: some new evidence', Language Learning 23, 63-74.

Lenneberg, E.H. (1967) Biological Foundations of Language. J. Wiley & Sons, New York.

Martins, I.P. and Ferro, J.M. (1987) 'Acquired conduction aphasia in a child', Devel. Med. Child. Neurob. 29, 529-540.

Petersen, S.E., Fox, P.T., Snyder, A.Z., Raichle, M.E. (1990) 'Activation of extrastriate and frontal cortical areas by visual words and word-like stimuli', Science 249, 1041-1044.

Prichard, J., Rothman, D., Novotny, E., Petroff, O., Avison, M., Howseman, A., Hanstock, C., Shulman, R. (1989) 'Photic stimulation raises lactate in human visual cortex', Soc. Mag. Res. Med. Abstracts; Works in Progress, p. 1071.

Riva, D. and Cazzaniga, L. (1986) 'Late effects of unilateral brain lesions sustained before and after age one', Neuropsychologia 24, 423-428.

Sappey-Marinier, D., Calabresa, G., Hugg, J., Deicken, R., Fein, G., Weiner, M. (1990) 'Increased lactate in human visual cortex during photic stiumlation', Soc. Mag. Res. Med. Abstracts, Vol. 1, p. 106.

Satz, P., Strauss, E., Whitaker, H. (1990) 'The ontogeny of hemispheric specialization: Some old hypotheses revisited', Brain and Language 38, 596-614.

Van Hout, A. and Lyon, G. (1985) 'Wernicke's aphasia in a ten-year-old boy', Brain and Language 29, 268-285.

Vargha-Khadem, F., O'Gorman, A.M., Watters, G.V. (1985) 'Aphasia and handedness in relation to hemispheric side, age of injury, and severity of cerebral lesion during childhood', Brain 108, 677-696.

Visch-Brink, E.G. and Van de Sandt-Koenderman, M. (1984) 'The occurrence of paraphasias in the spontaneous speech of children with an acquired aphasia', Brain and Language 23, 258-271.

Woods, B.T. (1980) 'The restricted effects of right-hemisphere lesions after age one; Wechsler test data', Neuropsychologia 18, 65-70.

Woods, B.T. and Carey, S. (1979) 'Language deficits after apparent clinical recovery from childhood aphasia', Ann. Neurol. 6, 405-409.

Woods, B.T. and Teuber, H-L. (1978) 'Changing patterns of childhood aphasia', Ann. Neurol. 3, 273-280.

CROSSED APHASIA AS A MODEL OF ATYPICAL SPECIALIZATION

Alexandre Castro-Caldas MD
Centro de Estudos Egas Moniz
Hospital de Santa Maria
1600 Lisboa
PORTUGAL

ABSTRACT. Crossed aphasia in dextrals is a rare clinical entity . Due to its rarity it is difficulty to understand all the problems involved in these syndromes. The explanations for atypical hemispheric specialization and some of the peculiarities of the clinical profile of crossed aphasia are reviewed and discussed.

INTRODUCTION

Crossed aphasia in dextrals has been consistently found in large series of brain lesioned patients for the past 100 years. It can thus be considered the result of a normal variant of brain specialization occuring in a small percentage of the population, ranging from 1% to 18% according to different series.

Recent studies provided information that allowed to change some of the ancient concepts about these syndromes. It is acceptable today to say that in most cases this atypical organization of brain function is genetically determined, like left handedness.

The study of the hereditary rules for this deviant arrangement is understandably difficult. Cerebral dominance for language is not easely assessed. To our knowledge, the few laboratory tests, like dichotic listening and tachistoscopy, which make the suspicion of abnormal specialization

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 83–93.

possible, had never been performed in the families of these patients. Invasive tests, like intra carotid injections of sodium amytal can not, of course, be performed in normal subjects. Finally, aphasia in relatives occurs very rarely and no familial cases of crossed aphasia has ever been reported.

The idea of this atypical specialization being genetically determined is only possible after ruling out other hypothesis that will be discussed in the following pages.

Before discussing some of the more relevant issues concerning crossed aphasia it is, however, important to criticise two aspects that were commonly neglected in older studies.

1) The systematic use of new techniques for studying brain lesions contributed significantly to change some of the old concepts. The most striking aspect was the clinical reported absence of fluent cases of crossed aphasia. This was due, most probably, to the absence of other localizing clinical signs of brain dysfunction besides fluent aphasia. Wernicke's aphasia, for instance, was never thought to be due to right hemisphere lesion before CT scan. Being a rarity only by chance would a case of crossed Wernicke's aphasia come to pathological study and this, to my knowledge, has never happened in the past. We know now that such syndromes exist (Henderson 1983) and probably as frequently as in aphasia due to left hemisphere lesions.

2) the other topic that has to be criticised in the former literature is the inclusion of traumactic cases in the series. Mirror foci of brain contusion are frequent and sometimes they are neglected if the major lesion is vary large. We have recently reported a case of post-traumactic fluent aphasia in which the CT scan revealed a right frontal heamorrage (Castro-Caldas et al 1986). However the electroencephalography also revealed a left occipito-temporal focus. This was in accordance with the report by Heilman et al (1971) on aphasia following closed head trauma. The authors found that fluent aphasia was common when the impact was right orbito-frontal. Cases like this explain the larger incidence of crossed aphasia in post-traumactic populations. Mohr et al (1980), for example, reported that 18% of the 244 aphasias due to penetrating head injuries had lesions on the right side of the brain. This is very high if compared, for instance, with our own stroke series of more than 1,000 patients in which the incidence is less than 1%.

Taking these two aspects in consideration, which are very important on what concerns the reliability of the data we have, we may procede to the following aspects raising several questions.

A. Why is language represented in the right hemisphere?

The following case studied in our Laboratory is a good example for discussing this point.

A 56-year-old woman suffered a stroke of the left hemisphere when she was 2 years old. The symptoms were right hemiparesis and language disturbance. Although before this incident, the relatives had never noticed a preferencial use of the left hand, the patient became left handed after this event. No familial left handeness was reported. According to relatives her language was disturbed until the age of 7 or 8. Due to a low social background the patient never went to school (the same happened to her brother and sister).

At the age of 56 she had a right temporal stroke and became aphasic. The characteristics of the speech disturbance were in accordance with the temporal lesion i. e. - fluent speech and poor comprehension (Wernicke's aphasia) (Guerreiro and Castro-Caldas, 1989). The CT scan revealed an old subcortical infarct lateral to the head of the left caudate nucleus and a recent one on the right temporo-parietal areas.

In this case language was in the right hemisphere most probably due to an early left hemisphere lesion. Therefore we can consider that, like in handedness, we have cases of pathological crossed aphasia. This lady is also an example of pathological left handness (Satz et al, 1985).

It is necessary to be careful in the investigation of early language difficulties and learning disabilities in crossed aphasics to prevent the misinterpretation of such cases. In the great majority of the cases reported in the current literature such antecedents are investigated but not always reported in detail.

Going back to our previous discussion on the genetically determined right hemisphere dominance for language, we can thus consider that there is a group of cases in which language functions were shifted to the right as a result of a left hemisphere lesion, and a group of patients in which this possiblity is ruled out. As it has also been pointed out by Alexander et al (1989) crossed aphasia can be considered anomalous or a mirror image of left hemisphere aphasia.

In cases of pathological crossed aphasia we do not know exactly how language shifts and which areas of right hemisphere are called to contribute to this novel task. In our case, reported above, the right temporal lobe was apparently involved in the process of auditory comprehension and not in fluency as it is usual in the left hemisphere. It is, however, possible to admit that in certain cases this is not the rule. Furthermore the shifting process may depend on the age of the patient and or the areas and functions disturbed by the first lesion.

Including cases of pathological crossed aphasia in studies of series of patients may constitute an important bias. This has to be taken in to

account while discussing, for instance, the recovery profiles and the localization of lesions.

B. What is the relationship between language lateralization and handedness?

It is well established that handedness is not a single variable and that between strong right handers and strong left handers it is possible to find sub groups. It is also well accepted that aphasia following right hemiphere lesions is more frequent in left handers than in right handers.

Although there seems to be a certain tendency for both language and control of handednees being represented in the same side of the brain they are sometimes dissociated.

In many cases of crossed aphasias in dextrals a family history of left handedness is reported, suggesting a genetic predisposition for some kind of atypical brain representation of functions.

Limb apraxia following a brain lesion has also a certain correlation with handedness and its incidence in crossed aphasia deserves some comment. Limb apraxia is present in 40% of the right handed aphasic patients in the first three months following left hemisphere stroke (Ferro 1983). Although there was the suggestion that it correlated with the severity of aphasia (Kertesz and Hooper, 1982) some cases have been reported of limb apraxia without aphasia in left handers with right hemisphere lesions (Poeck and Kerschensteiner, 1971; Heilman et al, 1973; Valenstein and Heilman, 1979). This may illustrate a relationship between handedness and limb apraxia. The evidence of crossed aphasia in dextrals is that limb apraxia is a rarety, as first suggested by Brown and Wilson (1973).

In our recent review of this topic (Castro-Caldas et al 1987) limb apraxia was present in only 9 cases of the 66 cases reviewed. In three of these cases familial left handedness was mentioned (Barroche et al 1979; Assal et al 1981, Henderson 1983). This also illustrates that higher motor control of the hand correlates better with handedness than with language.

Higher motor control of the hand and language seem therefore to be relatively separate variables that may fall independently either on the right side of the brain or on the left, according to genetic determination.

This is an interesting arrangement of functions, which contributes to the unexpected fluent writing with the right hand by non-fluent dextral crossed aphasics (Assal, 1982). Although disturbed in its linguistic content the written language is fluent and the drawing of individual letters and small words is generally correct in these patients. Studying the ability to write by non-fluent left hemisphere aphasics is most of the times impossible due to

the accompanying motor deficits on the right limbs, either paretic or apraxic.

C. Is there a continuum in brain specialization with age or a stable pattern since childhood?

Brown and Jaffé (1975) and Brown and Hecaen (1976) suggested that age could be an important variable in brain lateralization. However, concepts like crossed aphasia being more frequent in children and crossed aphasics being younger than other cases of aphasia, are no longer acceptable. In children crossed aphasia seems to be as rare as in adults. In her series of 31 aphasic children Martins has only one case of crossed aphasia, (Martins et al 1987). On the other hand, reviewing the published cases of stroke crossed aphasia and our own cases, we were able to demonstrate that not only was the age of patients similar to the age of left hemiphere stroke aphasics but also that the age difference between Broca's aphasia and fluent aphasia was present (Castro-Caldas and Confraria 1984; Obler et al, 1978; Castro-Caldas et al 1979). These findings suggested a similar pattern of distribution of the lesions in the antero-posterior axis, which is known to be influenced by age in stroke (Ferro et al, 1988).

The hipothesis of a continuum of hemispherical specialization does not seem to be supported by this evidence. Many studies proved that plasticity to adapt to a brain lesion decreases with age but this does not mean that certain areas are not predisposed to operate specific tasks since very early brain development.

D. Does the localization of lesions responsible for crossed aphasia mirror the localization of lesions responsible for left-hemisphere aphasia?

Lesion localization and its effect on functions can be accepted as an hint for functional arrangement of brain. Hence, the question of lesion localization in crossed aphasia is a relevant one.

Some authors have suggested that the localization of the lesions responsible for the different types of crossed aphasia did not mirror the distribution of lesions of left-hemisphere lesion. Habib et al (1983), for instance, considered that the lesions had a tendency to be deeper in the white matter. We have recently reviewed this subject. For each case of stroke crossed aphasia on which we had enough information concerning the clinical diagnosis and the localization of the lesion in the CT scan we selected, sequentially, in our files of left hemisphere aphasias, a control subject. These controls were matched for type and severity of aphasia, age, sex and educational level. The lesions were drawn on standard templates in a blind fashion and than overlapped in each pair of subjects (the drawing

correspondent to the crossed case was, of course, reversed). Comparison on what concerns the size of the lesions, the localization in the antero-posterior axis and the involvement of subcortical structures was judged by three of us independently. The results showed a high consistency among the three judgments. It was also concluded that it was impossible to find a difference between crossed and non-crossed aphasia on the three variables that were studied.

We can conclude by saying that in the great majority of the cases the brain lesion in crossed aphasia, shown in the CT scan, mirrors the lesion responsible for left hemisphere aphasia.

E. Are the anatomical brain asymmetries different in crossed aphasia?

It is known, after the preliminary work of Geschwind and Levitsky (1968), that the two cerebral hemipheres are anatomically different. A correlation between a "normal" and a "reversed" pattern of asymmetry failed to show significant differences between right-handers and non-right-handers (Chui and Damásio, 1980). These authors concluded that "the noted asymmetries are more likely to be direct correlates of cerebral language dominance than handedness". The correlation of a "reversed" pattern with right hemisphere representation of language seemed thus to be the most probable one according to the original hypothesis for this phenomenon. However, Henderson et al (1984) failed to demonstrate this correlation by studying the CT Scans of crossed aphasics. On the other hand, studying the same population we found a correlation with the presence or absence of left side neglect (Castro-Caldas et al 1985). Neglect, which is a major sign of right hemisphere dysfunction is commonly found in crossed aphasia (see below). In cases where neglect was present the CT scan showed the normal pattern of assymmetry. In those cases without neglect the pattern was reversed suggesting that the asymmetries that were found in the CT Scan are probably more related with non-verbal functions than with verbal ones.

F. Does the information content of the brain influence the inter-hemispheric organization?

Many of the cases reported in the literature of crossed aphasia occured in multi-lingual subjects, and there was a tendency to consider that in these cases the right hemisphere played a stronger role in language processing. Therefore right hemisphere lesions would have a higher chance of producing aphasia in poliglote subjects. On the other hand, illiteracy has also been claimed to be an important factor to influence the balance of competency between the two hemispheres on what concerns language.

Our opinion is that both these claims are no longer supportable. In the first case it is important to say that most of the cases reported in the last five years are not bilingual. In the past there was probably a tendency to report more complex cases, which biased the information as a group. In the second case, we have a great experience with aphasia in illiterate patients and as far as we can say the incidence is similar both in illiterate and literate subjects.

G. Which are the more common non-verbal disturbances in crossed aphasia?

The analysis of non-verbal accompanying disturbances in crossed aphasia allows us to understand what is represented in the right side of the brain in these subjects besides language. We reviewed 66 cases of the literature on what concerns limb apraxia, bucco-facial apraxia, left side neglect and constructional apraxia and the results are summarized in the table (Castro-Caldas et al 1987).

Non-verbal disturbances in 66 crossed aphasics

	Limb apraxia		Bucco-facial apraxia		Left-sided neglect		Construc-tional apraxia	
P	9	17%	14	52%	36	82%	29	76%
		80%		41%		67%		58%
A	44	83%	13	48%	8	18%	9	24%
U	13	20%	39	59%	22	33%	28	42%

P - Present A - Absent U - Unknown

As we mentioned above limb apraxia is rare. On the other hand, bucco-facial apraxia was present in half of the patients, which is a rule for left hemiphere aphasia (Ferro 1983). Left sided neglect was present in the great majority of the cases. This is the most consistent finding in crossed aphasia.

Neglect can be associated with lesions in either hemisphere but it is characteristically more severe and more persistent when the lesion is in the right hemisphere (Heilman 1979). Left-sided neglect occurs in 33-46% of the cases with right hemiphere lesions (Hier, Mondlock and Caplan

1983). From among all cases with crossed aphasia included in this study, important signs of left-side neglect were reported in 82% of the cases. The findings suggest that, as far as the representation of functions responsible for visuo-spatial activities is concerned, the right hemisphere of these crossed aphasic patients is similar to right hemisphere of most people whose language functions are located in the left hemisphere. The high incidence of neglect in crossed aphasia may suggest that these functions are even more lateralized in these subjects than in right-handed aphasics with left hemisphere lesions.

Constructional apraxia was present in 76% of crossed aphasics. Ferro and Castro-Caldas (1979) found constructional apraxia in 45% of 236 right-handed stroke aphasics with left hemisphere lesions. However, constructional apraxia is also frequent after right hemisphere lesions, often in association with left-sided neglect. The apparently higher incidence of constructional apraxia and neglect in crossed aphasia may reflect what Humphrey and Zangwill (1952) called "dual", i.e. major and minor hemispherical symptomatology following a lesion of the right hemisphere alone.

Our final conclusion is that the representation of language in the right hemisphere may be, in rare cases, the result of a recovery process of an early left hemisphere lesion. However, the majority of the cases represent a stable variant of language representation. This right hemisphere representation mirrors the left side language processing on what concerns the relation with development, the influence of learning, the envolved brain structures and the signs of language disturbance following the lesion. On the other hand, typical signs of right hemisphere disfunction are frequent, suggesting that these functions are not crossed.

Cases like these are probably more prone to have other variants on brain representation of functions. There are probably functions, or clusters of functions, which can be genetically determined to be represented on the left or on the right side of the brain and several combinations are most probably possible.

REFERENCES

Alexander, M.P., Fischette, M.R. and Fischer, R.S.: Crossed aphasics can be mirror image or anomalous. Brain 112: 953-973, 1989.

Assal, G.: Étude neuropsychologique d'une aphasie croisé avec jargonographie. Revue Neurologique, 138: 507-517, 1982.

Assal, G. Perentes, E. and Dervaz, J. P.: Crossed aphasia in a right handed patient: post-mortem findings. Archives of Neurology, 38: 455-458, 1981.

Barroche, G., Presles, O., Ramel, P. Weber, M. and Arnould, G.: Revue d'Oto-neuroophtalmologie, 51: 251-262, 1979.

Brown, J.W., and Jaffé, J.: Hypothesis on cerebral dominance. Neuropsychologia, 13: 107-110: 1975.

Brown, J.W., and Hécaen, H.: Lateralization and language Representation. Neurology, 26: 183-189, 1976.

Brown, J.W., and Wilson, F.R.: Crossed aphasia in a dextral. A case report. Neurology, 23: 907-911, 1973.

Castro-Caldas, A., Ferro, J.M. and Grosso, J.T.: Age, sex and type of aphasia in stroke patients. Paper presented at International Neuropsychological Society (European Conference), 1979.

Castro-Caldas, A., and Confraria, A.: Age and type of Crossed Aphasia in Dextrals Due to Stroke. Brain and Language, 23: 126-133, 1984.

Castro-Caldas, A., Confraria, A., and Mariano, M.G.: CT criteria of hemisphere asymmetry fail to predict language laterality. Neurology, 35: 935, 1985.

Castro-Caldas, A., Confraria, A., Paiva, T. and Trindade, A.: Contrecoup Injury in the Misdiagnosis of Crossed Aphasia. Journal of Clinical and Experimental Neuropsychology, 8: 697-701, 1986.

Castro-Caldas, A., Confraria, A., Poppe, P.: Non-verbal disturbances in crossed aphasia. Aphasiology, 1: 403-413, 1987.

Chui, H.C. and Damásio, A.R.: Human cerebral asymmetries evaluated by computed tomography. Journal of Neurology, Neurosurgery and Psychiatry, 43: 873-878, 1980.

Ferro, J.M.: Localization, lesion size and cerebral asymmetry in ideomotor apraxia. Paper presented at International Neuropsychological Society (European Conference), 1983.

Ferro, J.M., and Castro-Caldas, A.: Recovery from constructional apraxia in aphasia. Paper presented at International Neuropsychological Society (European Conference), 1979.

Ferro, J.M., and Crespo, M.: Young adult Stroke Neuropsychological Dysfunction and Recovery. Stroke, 19: 982-986, 1988.

Geschwind, N. and Levitsky, W.: Human brain, left right asymmetries in temporal speech region. Science, 161: 186-187, 1968.

Guerreiro, M., and Castro-Caldas, A.: Aphasia following right hemisphere lesion in a woman with left hemisphere injury in childhood. Paper presented at International Neuropsychological Society (European Conference), 1989.

Habib, M., Joanette, Y., Ali-Cherif, A. and Poncet, M.: Crossed Aphasia in dextrals: a case report with special reference to site of lesion. Neuropsychologia, 21: 413-418, 1983.

Heilman, K.M.: Neglect and related disorders. In K.M., Heilman and E. Valenstein (ed). Clinical Neuropsychology. Oxford University Press, 1979.

Heilman, K.M., Safran, A. and Geschwind, N.: Closed head trauma and aphasia. Journal of Neurology, Neurosurgery and Psychiatry, 34: 265-269, 1971.

Heilman, K.M., Coyle, J.M., Gonyea, E.F., and Geschwind, N.: Apraxia and agraphia in a left-hander. Brain,96: 21-28, 1973.

Henderson, V.W.: Speech fluency in crossed aphasia. Brain, 106: 837-857, 1983.

Henderson, V.W., Naeser, M.A., Weiner, J.M., Pieniads, J.M. and Chui, H.C.: CT criteria of hemisphere asymmetry fail to predict language laterality. Neurology, 34: 1086-1089, 1984.

Hier, D.B., Mondlock, J. and Caplan, L.R.: Behavioral abnormalities after right hemisphere stroke. Neurology, 33: 337-344, 1983.

Humphrey, M.E. and Zangwill, O.L.: Effects of a right-sided occipito-parietal brain injury in a left-handed man. Brain, 75: 312-324, 1952.

Kertesz, A. and Hooper, P.: Praxis and Language: the extent and variety of apraxia in aphasia. Neuropsychologia, 20: 275-286, 1982.

Martins, I.P., Ferro, J.M., and Trindade, A.: Acquired Aphasia in a Child. Development Medicine and Child Neurology, 29: 96-109, 1987.

Mohr, J.P., Weise, G.H., Caveness, H.F., Dillon, J.D., Kistler, J.P., Meirowsky, A.M., and Rish, B.L. Language and motor disorders after penetrating head injury in Vietnam. Neurology, 30: 1273-1279, 1980.

Obler, L., Albbert, M., Goodglass, H. and Benson, D.F.: Aphasia type and aging. Brain and Language, 6: 318-322, 1978.

Poeck, K. and Kerschensteiner, M.: Ideomotor apraxia following right-sided cerebral lesion in a left-handed subjet. Neuropsychologia, 9: 359-361, 1971.

Satz, P., Orsini, D.L., Saslow, E., and Henry, R.: The Pathological Left-Handedness Syndrome. Brain and Cognition, 4: 27-46, 1985.

Valenstein, E. and Heilman, K.M.: Apraxic agraphia with neglect-induced paragraphia. Archives of Neurology, 36: 506-508, 1979.

SYMPTOM PATTERN AND RECOVERY OUTCOME IN CHILDHOOD APHASIA: A METHODOLOGICAL AND THEORETICAL CRITIQUE

Paul Satz, Ph.D.
Department of Psychiatry
University of California
760 Westwood Plaza
Los Angeles, CA 90024

ABSTRACT. Two of the most distinguishing features attributed to the childhood aphasias are the symptom pattern and recovery course (Hecean, 1976; Satz & Bullard-Bates, 1981). With respect to symptom pattern, childhood aphasia, in contrast to adult aphasia, has long been characterized by its nonfluency. This position dates from the early reports of Bernhardt (1885) and Freud (1897/1968) who both noted the poverty of spontaneous speech and the telegraphic expression following left unilateral brain injury. They described the aphasia as primarily motor in type, with initial mutism followed by reduced initiative for speech, hesitations, dysarthria, and impoverishment of lexical stock. These early reports also noted the absence or rarity of logorrhea, paraphasia, and auditory comprehension defects, which suggested a dissociation between output (expression) and input (comprehension). This position has largely prevailed throughout the twentieth century (Assal & Campiche, 1973; Basser, 1962; Benson, 1972; Branco-Lefere, 1950; Byers & McLean, 1962; Geschwind, 1973; Guttmann, 1942; Poetzl, 1926).

INTRODUCTION

While most studies have essentially confirmed the predominant nonfluent pattern in aphasic children, particularly the absence or rarity of logorrhea and paraphasia, some investigators have continued to challenge this view by noting the presence of auditory comprehension and related fluency defects in a subset of aphasic children (Alajouanine and Lhermitte, 1965, Collignon, et al., 1968; Hecaen, 1976; Dennis, 1980; Woods & Teuber, 1978; Van Dongen & Loonen, 1979; Loonen and Van Dongen, 1989; Van Hout, 1989; Martins, 1990; Paquier, 1990). However, when one computes the frequency (prevalence) of such cases (i.e., fluent), it appears that the predominant symptom pattern in childhood aphasia is still largely non-fluent, even in studies where more rigorous linguistic assessments have been made. For example, in a recent study Loonen and Van Dongen (1989) observed only 4/28 cases of fluent aphasia, two of which recovered completely. Similarly, Paquier (1990) reported 7 cases of fluent aphasia in their series, but neglected to note that this subset was only part of a larger cohort of 48 childhood aphasia cases, the vast majority of whom were nonfluent (7/48). Finally, several studies reporting fluent cases have largely been single case reports (e.g., Dennis, 1980; Woods and Teuber, 1978; Van Dongen and Loonen, 1979).

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 95–114.

Additional support for the low prevalence of fluent aphasia in childhood was reported recently for a large and well controlled series of stroke cases in young adults (ages 15-50) who were still well below the age risk for stroke in the general population (Ferro & Crespo, 1988). The authors correctly noted that young adults in this age range represent only a small fraction (approximately 4%) of strokes in North America. The authors investigated etiology, neuropsychological pattern, aphasia type and recovery retrospectively in 254 patients, 166 of whom were aphasic at insult. With respect to aphasia type, they found that Broca's aphasia was significantly more common (127/166=77%) while Wernicke's and transcortical aphasia were significantly less frequent in this younger age cohort (39/166=23%). They also found that fluent aphasias and aphasias with comprehension deficits were significantly less common than in the older overall stroke population (>50 years).

At the present time, it seems reasonable to conclude that the symptom picture in childhood aphasia is predominately non-fluent and, based on estimates from the preceding three recent large-scale series, the prevalence is probably in excess of 85%, at least in children. Given these estimates, one should attempt to ask a more basic question as to why the symptom picture in childhood aphasia is different than in adult aphasia. The symptom pattern is even more puzzling given the multitude of potential confounders that could affect the onset and type of aphasia in children (e.g., type of lesion, age of insult, handedness, sex, lesion severity, lesion localization, etc.). Before addressing this issue, we will look briefly at the second most distinguishing feature attributed to the childhood aphasias -- namely, recovery and prognosis.

It has long been reported that the symptoms are transient and rarely permanent. Recovery from childhood aphasia is much more dramatic in comparison to the adult aphasias. These observations date from the early reports by Clarus (1874), Bernhardt (1885) and Freud (1897) and have been accepted generally by most twentieth-century clinicians (Alajouanine and Lhermitte, 1965; Basser, 1962; Benson, 1972; Byers and McLean, 1962; Geschwind, 1974; Guttmann, 1942; Hecaen, 1976; Lenneberg, 1967; Woods and Teuber, 1978).

The first major twentieth-century paper was by Guttmann (1942). This study comprised 30 unilateral cases (trauma, abscess, thrombosis), only 10 of which were available at follow-up. Moderate to complete recovery in speech was observed for all but one of the cases (vascular). Basser (1962) reported follow-up data on 30 children who sustained an acute hemiplegia (left =15, right = 15) after the onset of speech. An initial non-fluent aphasia was observed in most of the cases. At follow-up (3 months-2 years), Basser (1962) reported significant recovery in all cases. Despite the robust recovery in speech, the

intellectual levels were in the retarded range for a majority of the sample, regardless of lesion side. Unfortunately, no additional information was available with respect to school placement or achievement.

A similar recovery course was reported in a subsequent paper by Lenneberg (1967). Age at insult ranged from 3 to 18 years. Etiology was mixed and included cases of trauma (N = 4), stroke (N = 3) and neoplasm (N = 1). All of the children were aphasic at the initial examination, but only two were aphasic at follow-up. These two were in adolescence when the lesion (one trauma, one neoplasm) occurred (ages 15 and 18 years, respectively). Recovery was complete for all children who were under age 11 years at the time of injury. Lenneberg (1967) provided no data on intellectual level or on school achievement.

Alajouanine and Lhermitte (1965) reported a 1-year follow-up of 32 children (ages 6-15 years), all of whom were aphasic at initial contact following injury to the left hemisphere (trauma, neoplasm, stroke). At follow-up 24 of the children had regained `a normal or nearly normal language' (p. 659). No agrammatism was observed in these children. In contrast the remaining eight children showed a more unfavorable course. Most of these children had large cerebral lesions. On the basis of these results, Alajouanine and Lhermitte concluded that recovery is an indisputable fact and one very particular to children. This conclusion, however, should be tempered by an additional finding concerning intellectual level and school achievement. The authors were the first to note that, despite a dramatic remission in aphasia, none of the children could follow a normal progress in school. Unfortunately, no data were available on reading status, although it was suggested that most of the academic difficulties were due to intellectual impairment.

Byers and McLean (1962) also observed persisting cognitive impairments, despite complete restitution of speech functions in their follow-up study of ten children with unilateral vascular lesions. Unfortunately, no information was provided on reading status and no quantitative assessment of cognitive status was employed.

Hecaen (1976) reported follow-up data on the language status of 17 children (ages 3-19 years) who became aphasic following unilateral left brain injury. Spontaneous recovery was noted in a majority of the cases. However, most of the patients were reported to show long-term difficulties in writing and naming, although no data were presented to support this claim.

Woods and Teuber (1978) reported follow-up data on 25 aphasic patients who ranged in age from 2 to 15 years at onset. The cases were mostly of vascular origin but included trauma cases as well. At follow-up four years later, 21 of the cases showed spontaneous

recovery of speech. In each of these cases, the lesion occurred before 8 years of age. The four cases who remained aphasic ranged in age from 8 to 13 years at the time of the lesion. No information was provided on school achievement.

In a later study of 27 children with predominant vascular injury to the left hemisphere, Woods and Carey (1978) reported nearly complete sparing of speech in those children whose lesion onset was before the first birthday (N = 11) and in approximately half of those children whose onset was later (N = 7). In those children who became aphasic post-insult, all (N = 7) recovered speech if their lesion was before age 8 years. With respect to linguistic functions, deficits were observed only in those children who were initially or later aphasic. No information was reported on academic skills.

Van Dongen and Loonen (1979) reported aphasia status three years after brain injury in 14 right-handed children who ranged in age from 4 to 14 years at onset. Nine of the cases were trauma. Spontaneous recovery was observed in half of the cases. Unfortunately, it is unclear whether recovery was more associated with time of lesion onset (i.e. early), side of lesion (unilateral vs. bilateral) or lesion type (trauma vs. vascular). Also, no information was provided on follow-up academic status.

Martins and Ferro (1990) examined a number of factors influencing recovery of acquired aphasia in 29 children with unilateral left brain injury after language acquisition. Quantitative measures of language status were used at baseline and at follow-up. Twenty-two children (76%) recovered completely from the aphasia. Better recovery was found for vascular and trauma cases than for encephalitis which is more likely to cause severe and diffuse brain injury. Interestingly, recovery was closely associated with age at insult with 100% aphasia recovery in children under seven. In fact all of the persistent cases of aphasia were observed in children whose lesions occurred after age 7 years. Again, no information was provided for academic status.

The importance of age of insult in predicting recovery from aphasia was documented carefully in a recent study by Vargha-Khadem, O'Gorman and Watters (1985). The authors measured aphasia and language status in 3 groups of children: 28 patients with left hemisphere injury, 25 with right hemisphere injury, and 15 normal control subjects. Both lesioned groups (vascular, tumor, cysts, trauma) were also divided by age of insult (Early = prenatal to 5 years and Late = 5-14 years). All children were assessed at least 2 years post insult. With respect to aphasia course, all children in the early left hemisphere group recovered completely and only 1 child in the older left hemisphere group remained aphasic. With respect to language course, subtle naming difficulties were observed in the early left-lesioned group. However, the impairments were most pronounced for the older left-

lesioned group, "...with the pattern being the later the age of acquisition of injury, the more obvious the deficits. This pattern is more evident with respect to naming rather than auditory comprehension deficits and is consistent with the reported finding that there is a relative preponderance of expressive to receptive disorders in acquired left hemisphere injuries" (Vargha-Khadem, et al., 1985, p. 692). Unfortunately, the authors provided no information on the status of academic achievement.

Vargha-Khadem et al. also found a robust association between side and age of lesion on hand preference. According to the authors, "....All patients with prenatal and early postnatal left hemisphere lesions (even those with minimal hemiparesis affecting only the lower limb and no motor or sensory impairments in the right hand) were strongly left-handed" (pg. 693) -- and probably pathological left-handers (Satz, 1972). The observed link between improvement in naming and increasing left hand dominance provides an additional clue about neural mechanism that might account for recovery. This issue will be discussed later under recovery mechanisms.

A final recent study reported aphasia, language and intellectual recovery in 37 carefully diagnosed children with unilateral vascular accidents, 17 left [10 prenatal, 7 post-natal (Mean age = 5.4)] and 20 right [9 prenatal, 7 post-natal (Mean age = 6.1)] (Riva, Pantaleoni, Milani and Devoti, 1990]. The authors found complete recovery from aphasia in both of the left-lesioned groups (virtually all of whom were left-handed and probably pathological left-handers). However, subtle linguistic and intellectual problems were observed in both of the left-lesioned groups, but only when compared to a small group of 13 control children whose mean WISC-R Full Scale IQ and PPVT-R IQ was 120 and 133, respectively. These extremely high IQs in the controls, compared to the patients (Mean IQs 102 and 103, respectively) may have spuriously inflated between group differences. Once again, no report of academic achievement was given, leaving the status of other functional domains unknown.

Comment. Although the weight of evidence clearly supports the characteristic pattern of non-fluency and faster recovery in childhood aphasia, the explanation for these distinctive outcomes has remained less clear.

Symptom Pattern: Possible Brain Mechanisms or Substrates

One clue to brain substrate comes from the recent study of young adult stroke (Ferro & Crespo, 1988). They found that localization of the lesion was largely responsible for the preponderance of nonfluent aphasias in this age group. Most of these lesions (infarcts) occurred in the prerolandic and subcortical regions. Interestingly, Hecaen, Perenin and Jennerod (1984) noted that most

of the symptom pattern in childhood aphasia, including the rarer forms of auditory and visual verbal comprehension, were more common following anterior, as compared to posterior lesions. According to the authors:

> One might speculate, on the basis of this evidence, that the anterior (fronto-rolandic) region of the left hemisphere is more engaged in diverse aspects of language in the child than in the adult, even though the specialization of this region is already fixed for verbal expression (p. 285).

Hypothesizing a more diffuse representation in the anterior fronto-rolandic structures of the left hemisphere in childhood, led Hecaen et al. (1984) to suggest that this type of organization might permit greater sparing or recovery of comprehension because the neural substrate for this later developing functional domain (i.e., left posterior cortex) was functionally immature and uncommitted at the time of insult. This explanation is quite similar to the concept of functional maturity of neural structures advanced by Patricia Goldman and her colleagues (Goldman and Galkin, 1978; Goldman, 1979; Goldman and Rosvold, 1972). According to this view, if neural structures are still functionally immature or uncommitted at the time of injury, this permits greater plasticity and reorganization to occur.

One might speculate that the speech nonfluency that typically characterizes the initial symptom picture in childhood aphasia, but which recovers dramatically over time, may parallel the initial impairment and later recovery on tests of delayed alteration seen after orbital prefrontal lesions in infant rhesus monkeys (Goldman et al., 1972). In both cases the symptoms are produced by injury to neural structures which reached functional maturity earlier. Conversely, recovery may have occurred via later developing structures that were functionally uncommitted at the time of injury. These later maturing structures, whether ipsilateral or contralateral to the lesion, probably represent equipotential substrates for intrahemispheric and/or interhemispehric reorganization in man.

According to Hecaen et al. (1984), the early functional specialization of the fronto-rolandic region of the left hemisphere for diverse aspects of speech activation may account, in part, for reports of initial speech arrest and later recovery. After injury to these anterior structures, recovery is accomplished by intrahemispheric reorganization in adjacent, more posterior structures that reach full maturation only later in childhood. However, it is unclear whether this delayed maturation of posterior structures, which are ultimately the substrate for more complex linguistic operations, accounts for the relative absence of or faster recovery of comprehension defects in the

childhood aphasias. Despite these ambiguities, it is reasonable to conclude that instances of sparing and/or recovery of speech after early unilateral brain injury in man may be explained in part by the degree of functional maturity. We now know that not all structures within the left or right hemisphere reach functional maturity at the same time and that mechanisms of reorganization are probably facilitated by structures, probably equipotential, in either hemisphere that are functionally immature or uncommitted at the time of the lesion (See Satz, Strauss & Whitaker, 1990 for review).

Recovery Course: Possible Brain Mechanisms or Substrates

The preceding comments suggest that instances of sparing or recovery of speech and language functions after left brain injury in children might be facilitated by mechanisms of intra- or interhemispheric reorganization that were functionally uncommitted at the time of insult.

The most direct support for an intrahemispheric mode of reorganization comes from the seminal report by Lamar Roberts (Penfield & Roberts, 1959). However, this study also provided indirect support for an interhemispheric mode of reorganization. The data have been retabulated to reflect both outcomes.

Table 1 presents the results of language status (aphasia) following left-sided lobectomy in nonaphasic adult epileptic patients whose lesion onset was after age 2 [right side]. Inspection of this table shows that approximately 70% of right-handed and left-handed patients became aphasic after operation on the left hemisphere.

Table 1 presents the same data for nonaphasic adult epileptics whose lesion onset was before age 2 [left side]. Inspection of this table shows that the incidence of aphasia after left-sided operation was much lower in both handedness groups (46% in right handers and only 12% in left handers). The absence of aphasia in the majority of these early lesion onset cases (55/71 = 78%) likely reflects the effects of speech reorganization in the

TABLE 1

Aphasia Onset After Left Lobectomy X Age of Lesion and Handedness

Early (<age 2)				Late (>age 2)			
Hand	N	#Aph	%	Hand	N	#Aph	%
Right	22	10	.46	Right	157	115	.73
Left	49	6	.12	Left	18	13	.72

From Penfield and Roberts, 1959.

right hemisphere after injury to the critical speech zones in the left hemisphere. The mechanism of reorganization is probably facilitated by latent and equipotential structures in the right hemisphere that are activated after injury to the dominant or leading hemisphere.

In contrast, the onset of aphasia, which was much less frequent in these early left-sided lesion cases (16/71 = 22%) probably reflects, in part, the effects of speech reorganization via other latent though equipotential structures within the injured left hemisphere. As noted above, these posterior structures are hypothesized to represent the substrate for ontogenetically later complex cognitive and linguistic operations (i.e., semantic).

The most direct support for an interhemispheric mode of reorganization is based on the results of the amobarbital procedure developed by John Wada (Wada & Rasmussen, 1960). The presence of speech arrest only in the right hemisphere (after right intracarotid infection of sodium amytal), points to an interhemispheric reorganization. However, the presence of speech arrest only in the left hemisphere could reflect an intrahemispheric maintenance or reorganization of speech. The latest review of this literature is by Satz, Strauss, Wada & Orsini, (1988). Four studies are reviewed which represent the few known sources that report parametric data on lesion focus, lesion type, age of onset of damage, handedness and hemispheric speech dominance using different assessment methods (amobarbital, temporal lobectomy and dichotic listening).

Two of the studies, both seminal investigations, have already been published. Rasmussen and Milner (1977) used the sodium amytal technique on a large series of left brain-impaired patients (N=396) where hemispheric dominance was based on the presence of speech arrest following unilateral carotid injection. Penfield and Roberts (1959) assessed the presence or absence of aphasia in patients (N=246) after left temporal lobectomy; patients were classified as left dominant for speech if aphasic and atypical (i.e. right) if not aphasic following surgery. Strauss et al. (in preparation) used the sodium amytal technique on 53 consecutive left brain-impaired cases in which age of onset of damage was estimated in months. Orsini (1984) used a test-retest dichotic verbal listening task (free recall) on 40 consecutive children and adolescents with verified left focal seizures in which age of onset of damage was also estimated in months. A ratio score (R - L)/(R +L), was used to determine the ear advantage which was then used to infer the probable contralateral side of hemispheric speech dominance.

The results are presented in the following tables. Table 2 shows the percentage of left versus atypical speech cases by age of lesion onset (Early < 6 years; Late > 6 years). The data from each of these studies (left lesions only) have been retabulated

and pooled for abbreviation and ease of presentation. Cases of bilateral and right-sided speech have also been pooled into a single group labeled atypical.

TABLE 2
Percentage speech type by lesion onset across studies

Onset	Left	Atypical
Early	107/262 (41%)	155/262 (56%)
Late	377/473 (80%)	96/473 (20%)

The results were remarkably consistent across studies. Early left focal brain injury (before age 6 years) is associated with a dramatic shift from the expected pattern of left hemisphere speech dominance observed in normal populations (approximately 96%) versus the present series (107/262 = 41%, $p < .001$). Within the present series, the incidence of left hemisphere speech dominance was also much lower in the early versus late onset groups (41% versus 80%, $p < .001$). Inspection of Table 2 shows that this difference was due to a dramatic increase in the percentage of atypical speech dominance in the early left brain injury cases (Early = 59%; Late = 20%, $p < .001$).

A similar association was noted for handedness (Table 3). Early left brain injury was associated with a dramatic increase in the incidence of manifest left-handedness (MLH) [162/262 = 62%] compared to non-brain-injured populations (approximately 10%, $p < .001$). Also, the incidence of MLH, after early brain injury, was almost threefold higher in the atypical speech dominant group (left = 33%, atypical = 82%, $p < .001$), suggesting that the interhemispheric shift in speech was associated with a corresponding shift or alteration in hand preference. However, the association is by no means invariant. There were cases of both unimodal (speech only) and bimodal (speech and hand) transfer.

TABLE 3
Percentage MLH by speech type by lesion onset across studies

Onset	Left	Atypical
Early	35/107 (33%)	127/155 (82%)
Late	102/377 (27%)	43/96 (45%)

When these different types of interhemispheric transfer (speech only vs. speech and hand) were examined more closely, it was found that in each of the studies, a bimodal shift was associated with a much earlier onset of damage (bimodal = 75%, unimodal = 35%, $p < .001$). When age of damage was reported in months, the bimodal group (speech and hand) showed a much earlier onset (bimodal = 22 months, unimodal = 59 months, $p < .05$) as well as a lower incidence of bilateral speech organization (bimodal = 10%, unimodal = 60%, $p < .05$).

When cases of interhemispheric versus intrahemispheric speech organization were compared across studies, it was shown that time of onset of damage was also associated with type of hemispheric speech organization after left brain injury. In each study, there was a much higher incidence of early injury in cases of interhemispheric reorganization of speech (62% vs. 22%, $p < .001$).

What conclusions, if any, can be drawn from these results? The most striking finding is that the earlier the left hemisphere perturbation (i.e., before 12 months), the greater the likelihood of a bimodal interhemispheric reorganization including a shift in both speech (right) and handedness (left). Lesions occurring between the ages of 12 and 72 months (6 years) are more likely to result in either a unimodal interhemispheric shift in speech, a bilateral or less complete form of hemispheric speech reorganization, or an intrahemispheric maintenance or reorganization of speech. In each of the latter three cases, the likelihood of a corresponding shift in handedness is lower than with lesions occurring before the first birthday, which are more likely to result in a bimodal hemispheric reorganization. It is hypothesized that this latter type of hemispheric reorganization probably accounts for instances of sparing following left hemisphere injury while the former modes of reorganization account for instances of recovery in speech. Unfortunately, it is unclear whether the rate and/or degree of recovery is related to the type of inter- or intrahemispheric reorganization. This question has yet to be tested empirically.

Recovery: Other Cognitive and Achievement Outcomes

It should be noted from the preceding review that the vast majority of studies on recovery in childhood aphasia have been addressed to speech restitution, with only recent focus on linguistic status (e.g., Woods and Carey, 1979; Vargha-Khadem et al., 1985; Riva et al., 1990). What has remained conspicuously unclear is the status of other nonlinguistic cognitive domains such as visuo-spatial performance as well as specific academic domains such as reading and math performance.

Reading and Math Performance. The status of academic performance after left focal brain injury in childhood has traditionally been relegated to observational or anecdotal report in studies of recovery of speech and language (Satz and Bullard-Bates, 1981). Nevertheless, several of these reports have suggested a poor outcome in reading and math despite the rapid recovery from aphasia in these children. Only recently has this problem been investigated empirically using appropriate selection and measurement procedures. The most informative studies have been conducted by Dorothy Aram and colleagues (Aram and Ekelman, 1988; Aram, Chapter ___, this volume). Because of Aram's contribution to the present volume, this outcome domain can be critically reviewed in her chapter. However, the thrust of her work can be summarized in a concluding paragraph from a recent study of early unilateral vascular injury (left and right) in children (Aram and Ekelman, 1988):

> Despite differences between lesioned and control children, the relatively high mean performance of the lesioned subjects is notable. The mean performance for lesioned subjects on all clusters examined here is at or above the 39th percentile, an impressively high level of performance for children with unilateral brain lesions. While the presentation of group percentile data masks individual performance, which in some instances is low among lesioned subjects and does not account for premorbid potential, the high overall performance evidences considerable presumed reorganization of cognitive abilities, scholastic aptitude, and academic achievement following unilateral lesions sustained in childhood (pg. 915).

Two important findings from this research are: (1) that the effects of the lesions are typically mild and insufficient to cause a major learning disability or developmental dyslexia and (2) in selected cases of school difficulty, the outcome is unrelated to lesion side, except for math (i.e., right hemisphere lesion). Both findings challenge the putative role of the left

hemisphere in developmental dyslexia (Dejerine, 1891; Geschwind and Galaburda, 1985) and lend support for a bilateral neural substrate in the etiology of this disorder (Satz, 1989).

Visuo-spatial Performance. Despite the dramatic recovery from aphasia in childhood, including the apparent sparing or recovery in reading and math performance following left hemisphere injury (Aram and Ekelman, 1988; Aram, Chapter ___), the status of other non-linguistic functions has remained problematic. According to Teuber (1974):

> "...it is less clear how complete such an escape or rapid recovery of language can be, after very early lesions, or whether other aspects of performance suffer when language seems to escape" (p. 73).

Teuber (1974) provided the first formal explanation for this apparent paradoxical outcome which eventually become known as the 'crowding hypothesis' (see Strauss, Satz, and Wada, in press for review). Teuber concluded as follows:

> "All in all, these findings suggest a definite hemisphere specialization at birth, with a curious greater vulnerability to early lesions for those capacities that depend, in the adult, on the right hemisphere--as if speech were relatively more resilient or simply earlier in getting established. Yet this resiliency is purchased at the expense of nonspeech functions or if one had to admit a factor of competition in the developing brain for terminal space, with consequent crowding when one hemisphere tries to do more than it had originally been meant to do" (p 73).

Despite the theoretical importance of this hypothesis, there have been few attempts to test it in the last 16 years. The reason is that such a study would require objective information on lesion onset, side of dysfunction (left-sided), cerebral speech pattern (left, right or bilateral), as well as a broad assessment of linguistic and non-verbal cognitive functions. Prior to a recent investigation by Strauss, Satz and Wada (in press), no study had yet satisfied all these criteria. The authors selected 27 young epileptic adults (Mean age 24) who had sustained early left brain injury (before age 6) and were being evaluated for possible surgical treatment. Carotid amytal testing revealed that in 14 patients, speech was exclusively mediated by the left hemisphere, and in 6 patients, speech was exclusively in the right hemisphere. Seven patients were shown to have bilateral speech representation. The mean age at which cerebral damage was incurred was 11.7 months (Range = birth - 6 years). While many of the left hemisphere speech group (9/14) had damage before 12 months, virtually all of

the atypical (right, bilateral) speech cases (12/13) had their onset of damage by the first birthday. Consistent with other studies reviewed in this chapter, virtually all of the left handers (based on demonstration preference) [8/9] were in the atypical speech hemisphere group (8/13 = 62%). This finding again lends support to the bimodal shift phenomenon (speech and hand) observed in cases of very early left brain injury (Satz, et al., 1988).

The primary test of the crowding hypothesis compared the left- and atypical hemisphere speech groups (both of whom had sustained early left brain injury) on a battery of linguistic (N = 9) and non-verbal cognitive tests (N = 10). To date, most studies, including the seminal studies of Woods and Carey (1979), Vargha-Khadem et al., (1985), and Aram and Ekelman (1988), have examined the status of verbal and non-verbal performance in patients without knowledge of their mode of hemispheric speech dominance. Consistent with the hypothesis, those patients with atypical speech showed significantly lower performance on most of the non-verbal measures (8/10) and on only two of the verbal measures (2/9). This relative pattern of dissociation comprised several domains of non-verbal ability including intelligence, memory and visuospatial function. We noted that although the atypical speech group was also lower on two of the nine verbal measures, the primary deficit pattern was clearly associated with skills which are typically mediated by the right hemisphere. But why the selective impairment in non-verbal cognitive skills? The most likely explanation is that the interhemispheric shift in speech, which facilitates language recovery, probably interferes with or 'crowds out' the acquisition of non-linguistic skills that would normally depend on this hemisphere. This explanation, according to Strauss et al. (in press), implicitly assumes two separate lesions, one structural (left) and one functional (right) to account for the paradoxical impairment in non-language domains following early injury to the left hemisphere.

Despite the relative dissociation in functional performance, it should be noted that verbal functions were not entirely within the normal range in the left hemisphere speech group when compared to an age-matched normal control group. However, this group performed as well as the control group on all measures of non-verbal ability. By contrast, patients with atypical speech patterns performed lower than the controls on almost all measures of verbal and non-verbal ability suggesting that there might be an intellectual price to pay for such plasticity.

Concluding Comments. One unsettled issue concerns why the right hemisphere permits this type of reorganization after early left brain injury. It has long been known that such plasticity is not evident in the left hemisphere (e.g., visual-spatial ability) after equivalently early right hemisphere lesions (Dennis, 1980; Lansdell, 1969). Is there something unique about the right

hemisphere that permits this type of interhemispheric and intrahemispheric reorganization to occur? As noted earlier, the interhemispheric reorganization of speech is probably facilitated by equipotential structures within the right hemisphere that are activated (released from inhibition) after early left brain injury. However, this explanation does not account for the relative resiliency of verbal functions in this type of intrahemispheric (right) competition. We have recently suggested (Satz, Strauss and Whitaker, 1990) that this reorganization may be facilitated by the more diffuse representation of functions that have been attributed to this hemisphere (Semmes et al., 1960) as well as to its slower rate of maturation. This latter explanation, which conflicts with those that hypothesize a much earlier maturation of the right hemisphere (Geschwind and Galaburda, 1985), rests on the fact that the left hemisphere is uniquely specialized from birth to provide the substrate for speech and language functions (Kinsbourne and Hiscock, 1983) and that this substrate represents a type of biological left-right maturational gradient that favors earlier maturation and resiliency of left hemisphere speech functions (Corballis and Morgan, 1978).

A second issue, still relatively understudied, concerns the role of the right hemisphere in more complex semantic and propositional speech. Although linguistic studies have traditionally focused on the special role of the left hemisphere in the analysis of phonological, syntactical and sequential processing of speech (Lieberman, 1963), it was Hughlings Jackson (1878) who much earlier noted the special role of the right hemisphere in more complex aspects of linguistic communication. Only recently has this position been subjected to experimental test. Some of the seminal studies have been conducted by Howard Gardner and associates (Gardner, Brownell, Wapner and Michelow, 1983) and Van Lancker and associates (Van Lancker and Kreiman, 1986; Van Lancker and Kempler, 1987). These studies have used complex familiar phrases, including idioms and proverbs as the unit of analysis and have shown that these phrases are processed like unitary, nonsyntactically analyzed elements that involve more inferential and holistic interpretation -- a process quite compatible with Jackson's (1878) earlier view of the right hemisphere. Interestingly, Van Lancker and Kempler (1987) have shown that despite the complex nature of these familiar phrases, aphasic patients showed better comprehension of these formulaic phrases than of novel (literal) expressions, the latter of which are analyzed at the level of the word.

Van Lancker and associates (personal communication) have also shown that the interpretation of familiar (non-literal) phrases show a slower and more gradual acquisition with age than novel (literal) phrases, and that performance does not asymptote until ages 12-14 years. This delayed acquisition of performance on familiar phrases seems quite compatible with some of the preceding

views concerning the slower rate of maturation of the right hemisphere which, in part may confer greater plasticity for reorganization after early left hemisphere insult. An alternative view, as discussed earlier, is that this type of complex inferential processing (i.e., holistic) is uniquely facilitated by the more diffuse representation of functions that have been attributed to this hemisphere (Semmes, et al., 1960).

Although the preceding views provide some explanation, in part, for the capacity of the right hemisphere to assume greater participation in more complex aspects of speech output (fluency) and input (comprehension) after early insult to the left hemisphere, it is still unclear whether the delayed acquisition of more complex comprehensional processing (e.g., familiar phrases) may be paradoxically compromised after such early left brain injury. This additional component of the 'crowding hypothesis' is currently under investigation.

This chapter, in summary, has attempted to evaluate the current status of symptom pattern and recovery in childhood aphasia, with special focus on recent European studies that are less accessible to North American investigators. The chapter also addressed the current status of other cognitive, motor and achievement domains in childhood aphasia, especially following early injury to the left hemisphere that have long remained controversial or unknown. Finally, an attempt was made to account for some of the different recovery outcomes in childhood aphasia, with special reference to intra- and interhemispheric mechanisms of reorganization.

References

Alajouanine, T.H. & Lhermitte, F. (1965). Acquired aphasia in children. Brain, 88, 653-662.

Aram, D.M., & Ekelman, B.L. (1988). Scholastic aptitude and achievement among children with unilateral brain lesions. Neuropsychologia, 26, 903-916.

Assal, G., & Campiche, R. (1973). Aphasie et troubles du language chez l'enfant apres contusion cerebrale. Neuro-Chirurgie, 19, 399-406.

Basser, L.S. (1962). Hemiplegia of early onset and the faculty of speech with special reference to the effects of hemispherectomy. Brain, 85, 427-460.

Benson, D.F.(1972). Language disturbances of childhood. Clinical Proceedings Children's Hospital of Washington, 28, 93-100.

Bernhardt, M. (1885). Ueber die spastiche cerebralparalyse im Kindesalter (Hemiplegia spastica infantilis), Nebst einem Excurse uber: "Aphasie bei Kindern". Archiv fur Pathologische Anatomie und Physiologie und fur Klinische Medecin, 102, 26-80.

Branco-Lefevre, A.F.(1950). Contribuicao para o estudo da psicopatologia da afasia en criancas. Archivos Neuro-Psiquiatria (Sao Paulo), 8, 345-393.

Byers, R.K., & McLean, W.T. (1962). Etiology and course of certain hemiplegias with aphasia in childhood. Pediatrics, 29, 376-383.

Clarus, A. 1874. Uber Aphasie bei Kindern. Jahresb.Kinderheilkd. 7, 369-400.

Collignon, R., Hecaen H., & Angelerques, G. (1968). A propos de 12 cas d' aphasie acquise chez l'enfant. Acta Neurologica et Psychiatrica Belgica, 68, 245-277.

Corballis, M.C. and Morgan, M.J. (1978). On the biological basis of human laterality. I. Evidence for a maturational left-right gradient. Behavioral and Brain Sciences, 1, 261-269.

Dejerine, J. (1892). Contribution a petude anatamopathologique et clinique des differentes varietes de ceicite verbale. Mem. Soc. Biol., 4, 61.

Dennis, M. (1980). Strokes in childhood I: Communicative intent, expression, and comprehension after left hemisphere arteriopathy in a right-handed nine-year-old. In R.W. Reiber (Ed.), Language development and aphasia in children. New York: Academic Press.

Ferro, J.M.& Crespo, M. (1988). Young adult stroke: Neuropsychological dysfunction and recovery. Stroke, 19, 982-986.

Freud, S., (1968). Infantile cerebral paralysis. (L.A. Russin, trans.). Coral Gables: University of Miami Press (Originally published,1897).

Gardner, H., Brownell, H.H., Wapner. W., & Michelow, D. (1983). Missing the point: The role of the right hemisphere in the processing of complex linguistic materials. In E. Pereeman (Ed.). Cognitive processing in the right hemisphere. New York: Academic Press.

Geschwind, N. (1973). Late changes in the nervous system: An overview. In D.G. Stein, J. J. Rosen, & N. Butters (Eds.). Plasticity and recovery of function in the central nervous system. New York: Academic Press.

Geschwind, N., and Galaburda, A.M. (1985a). Cerebral lateralization: Biological mechanisms, associations, and pathology. I. A hypothesis and a program for research. Archives of Neurology, 42, 428-459.

Goldman, P.S. (1972). Developmental determinants of cortical plasticity. Acta Neurobio. Exp., 32, 495-511.

Goldman, P.S. & Rosvold, H.E. (1972). The effects of selective caudate lesions in infant and juvenile rhesus monkeys. Brain Research, 43, 53-66.

Goldman, P.S. & Galkin, T.W. (1978). Prenatal removal of frontal association cortex in the fetal rhesus monkey. Anatomical and functional consequences in post-natal life. Brain Research, 152, 451-485.

Goldman, P.S. (1979). Neuronal plasticity in primate telencephalon. Anomolous projections induced by prenatal removal of frontal cortex. Science, 202, 768-770.

Guttmann, E. (1942). Aphasia in children. Brain, 65, 205-219.

Hecaen, H. (1976). Acquired aphasia in children and the autogenesis of hemispheric specialization. Brain and Language, 3, 114-134.

Hecaen, H., Perenin, M.T., & Jeannerod, H. (1984). The effects of cortical lesions in children: Language and visual functions. In C.R. Almli and S. Finger (Eds.). Behavioral biology of early damage (Vol.1, 277-299). New York: Academic Press.

Jackson, J.H. (1878). On affections of speech from disease of the brain. Brain, I, 304-330. (Reprinted in J. Taylor (Ed.). 1958. Selected writing of John Hughlings Jackson. New York: Basic Books. Vol. 2).

Kinsbourne, M. and Hiscock, M. (1983). Asymmetries of dual-task performance. In J.B. Hellige (Ed.), Cerebral hemisphere asymmetry: Method, theory, and application. New York: Praeger, 255-334.

Lansdell, H. (1969). Verbal and nonverbal factors in right-hemisphere speech: Relation to early neurological history. Journal of Physiological and Comparative Psychology, 69, 734-738.

Lenneberg, E. (1967). Biological foundations of language. New York: Wiley.

Lieberman, P. (1963). Some effects of semantic and grammatic context on the production and perception of speech. Language and Speech, 6, 172-187.

Loonen, M.B. and Van Dongen (1989). Acquired childhood aphasia. Paper presented at 12 European Conference of the International Neuropsychological Society, Antwerp, Belgium, July 7.

Martins, I.P. (1990). Type of aphasia and lesion localization. Paper presented at NATO Advanced Research Workshop on Acquired Aphasia in Children. Sintra, Portugal, September 13-15.

Martins, I.P. and Ferro, J. (1990). Recovery from acquired aphasia in children and lesion size in the temporal lobe. Paper presented at NATO Advanced Workshop on Acquired Aphasia in Children. Sintra, Portugal, September 13-15.

Paquier, P. (1990). Fluent aphasias in children. Paper presented at NATO Advanced Research Workshop on Acquired Aphasia in Children. Sintra, Portugal, September 13-15.

Penfield, W., & Roberts, L. (1959). Speech and brain mechanisms. Princeton: Princeton University Press.

Poetzl, T. (1926). Ueber sensorische Aphasie in Kindersalter. 2. Hals N.Ohrenklin, 14, 109-118.

Riva, D, Pantaleoni, C., Milani, N. and Devoti, M. (1990). Papers presented at NATO Advanced Research Workshop on Acquired Aphasia in Children. Sintra, Portugal, September 13-15.

Satz, P. (1972). Pathological left handedness; an explanation. Cortex, 8,121-135.

Satz, P., and Bullard-Bates, C. (1981). Acquired aphasia in children. In M.T. Sarno (Ed.), Acquired aphasia. New York: Academic Press.

Satz, P., Strauss, E., Wada, Jr., & Orsini, D.L. (1988). Some correlates of intra- and interhemispheric speech organization after left focal brain injury. Neuropsychologia, 26, 345-350.

Satz, P. (1989). Developmental dyslexia: An etiological reformulation. In G. Pavlidis (Ed.), Dyslexia: A neuropsychological and learning perspective, 1-26. New York: John Wiley.

Satz, P., Strauss, E. and Whitaker, H. (1990). The ontogeny of hemispheric specialization: Some old hypotheses revisited. Brain and Language, 38, 596-614.

Strauss, E., Satz, P. and Wada, J. (In Press). An examination of the crowding hypothesis in epileptic patients who have undergone the carotid amytal test. Neuropsychologia.

Semmes, J. (1968). Hemispheric speculation: A possible clue to mechanisms. Neuropsychologia, 6, 11-26.

Teuber, H.L. (1974). Why two brains? In The Neurosciences: Third Study Program. F.O. Schmitt and F.G. Worden (Eds.) Cambridge: MIT Press, 71-74.

van Dongen, H.R., & Loonen, M.C. (1979). Neurological factors related to prognosis of acquired aphasia in childhood. In Y. Lebrun and R. Hoops (Eds.), Recovery in aphasics. Amsterdam: Swets and Zeitlenger, B.V.

Van Lancker, D., & Kreiman, J. (1986). Preservation of familiar speaker recognition but not unfamiliar speaker discrimination in aphasic patients. In R.H. Brookshire (Ed.). Clinical aphasiology. Minneapolis: BRK Publishers.

Van Lancker, D.R. and Kempler, D. (1987). Comprehension of familiar phrases by left-but not by right-hemisphere hemisphere damaged patients. Brain and Language, 32, 265-277.

Vargha-Khadem, F., O'Gormon, A.M. and Watters, G.V. (1985). Aphasia and handedness in relation to hemispheric side, age of injury and severity of cerebral lesion during childhood. Brain, 108, 677-696.

Wada, J. and Rasmussen, T. (1960). Intracarotid injection of sodium amytal for the lateralization of cerebral speech dominance. Journal of Neurosurgery, 17, 266-282.

Woods, B.T., & Teuber, H.L. (1977). Changing patterns of childhood aphasia. Transactions of the American Neurological Association, 102, 36-38.

Woods, B.T., & Teuber, H.L. (1978). Changing patterns of childhood aphasia. Annals of Neurology, 3, 273-280.

Woods, B.T., and Carey, S. (1979). Language deficits after apparent recovery from childhood aphasia. Annals of Neurology, 5, 405-409.

V - CLINICAL FEATURES OF ACQUIRED APHASIA IN CHILDREN

CHARACTERISTICS OF LANGUAGE IN ACQUIRED APHASIA IN CHILDREN

A. VAN HOUT
Department of Neuropediatrics
U.C.L. 10/1303
St. Luc University Clinic
10, Avenue Hippocrate
1200 Brussels
Belgium

ABSTRACT. Until recently, the language symptoms in acquired aphasia in children were considered mainly expressive, with few of the characteristics of adult aphasia. Recent data, however, point to a number of aphasic symptoms in children more varied than previously thought and also often organized as in adult aphasias. In practice, this means that rehabilitation methods proven effective in adults could be used in children. Moreover, on a theoretical level, the traditional views of a progressive specification of the language areas in the left hemisphere are challenged, and analogical reasoning, often applied in child neuropsychology, is supported.

Introduction

The classic descriptions of acquired aphasia in childhood, focusing on its restricted, mainly negative symptoms, were at the basis of the hypothesis of a progressive localization of language within the left hemisphere (Lenneberg, 1967). This concept soon appeared to contradict behavioral and neurophysiological data pointing to an early specification of the left-brain areas for some sensorimotor dimensions of language (Kinsbourne, 1977).

In addition, the generally accepted views on symptoms in acquired childhood aphasia were in opposition with the current descriptions of symptoms of developmental dysphasias. How could one reconcile the fact that these later pathologies could be satisfactorily compared to adult aphasic profiles, as Rapin and Allen (1988) have done, and, by inference, their neurological basis proposed, when the actual occurrence of a lesion in children did not, according to the literature, give rise to symptoms similar to those of adult aphasia? Woods and Teuber (1978) suggested that a selection bias, the criterion of hemiplegia to assert the unilaterality of the lesion, could be the source of the alleged anterior type of aphasias in childhood.

In the 1980s, all those contradictory findings led to a reexamination of symptom data in acquired childhood aphasia in order to assess the validity of the classic descriptions.

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 117–124.

1. The Classic Descriptions

The classic signs reported in childhood aphasia were often called "motoric" and were said to occur whatever the lesion localization. Two main interpretations were in opposition: one regarded these "motoric" signs as similar to those of the anterior types of aphasia in adults, and another for which the expressive symptoms were considered a mere regression to earlier stages of language development.

The main features of this classical picture of acquired childhood aphasia were: 1) Mutism: the literature does not clearly differentiate mutism from hypospontaneity of speech, in which the paucity of speech initiation can be reduced by solicitation, or from non-aphasic mutism (Von Cramon, 1975), which may be due to a subcortical lesions. 2) Dysarthria is also differentially interpreted: some authors regard it as pathological, evocative of the "phonological desintegration" syndrome (Alajouanine and Lhermitte, 1965), while others consider it to be a return to a more infantile way of pronunciation (Jacobson, 1962). 3) Syntactic disorders are difficult to disentangle from hypospontaneity because these children are unwilling to initiate speech and because true agrammatism is not always observed since their mean length of utterance is short. The degree of alteration of the different language functions may vary, although the authors who have stressed the similarity of symptoms between the phonological disorders in acquired aphasia of childhood and in adult motor symptoms argue that syntactic troubles in children reflect a mere regression to less advanced stages of grammatical development (Alajouanine and Lhermitte, 1965). 4) Lexical problems have been described as true anomia, reduced with the help of phonemic cues, or as a "loss" or poverty of vocabulary, more similar to a reduction of use or an impoverishment of the memory stock. 5) However, the most salient feature of acquired aphasia in childhood was the lack of the common symptoms observed in adult posterior aphasic types. We proposed designating them as "positive signs" (Van Hout et al., 1975) in order to distinguish them from the classic symptoms of aphasia in children, which were mainly reductive or "negative". Thus, paraphasias, neologisms, perseverations, fluent speech, and long-lasting comprehension disorders were considered rare or, when occurring occasionally, to characterize the oldest children (for definitions of the aphasic terms, see Roch-Lecours, 1970).

Moreover, apart from a rare antero-posterior gradient found in some series (Hécaen, 1976), there was no link between symptoms and the localization of the lesions. This finding was interpreted as indicative of a lack of functional differentiation of the language areas within the left hemisphere. Symptoms similar to those in adults would only appear if the neuro-anatomical substrate for the sustained function had achieved its mature state.

2. The New Data

Recent findings resulting from examination of unselected series of acquired aphasia in childhood with age-adapted tests batteries (Van

Hout et al., 1985; Van Dongen et al., 1985) have fundamentally modified the conceptions of the symptoms in acquired childhood aphasia. We will briefly summarize some of our own findings in this area. We administered our tests battery adapted from the Spreen and Benton aphasic tests (1969), with modifications for comprehension and fluency measurements (Van Hout et al., 1985) as early as possible after the occurrence of the lesion or the emergence from coma. We were able to observe that all the signs said to be absent in childhood aphasia were, in fact, detectable, albeit in different percentages in the different children, and this well before the age of puberty. Those positive signs were fluent aphasias with severe and long-lasting comprehension disorders, stereotypes, perseverations, and logorrhea. The most prominent symptom was the presence of paraphasias, both verbal and phonemic, so, we decided to classify children's aphasias according to the measurement of the paraphasias during the acute stage. We scored, as did Goodglass and Kaplan (1972), as a percentage of the total number of words produced.

The paraphasias were greatly reduced after a few days in some children, in a few months in others, and they could persist, although attenuated, two years after the injury in those children where the other aphasic symptoms were also the most severe. Among the three groups of children defined by this importance of the initial paraphasias and their temporal evolution, there were no significant differences in etiology or lesion localization, or, in opposition to the classic view, in age at injury. For those children who had not recovered at long-term follow-up, the lesions were mostly posterior with some degree of heterolateral, although non-symmetrical, involvement.

Our use of an extensive examination battery permitted us to extract scores for the different language subcomponents--comprehension, repetition, naming, and fluency--and thus to demonstrate specific aphasic profiles. The expression of the scores in standard deviations allows direct comparisons of aphasic profiles for children of different ages. The specific aphasic types thus delineated were often evocative of the well-known adult aphasic syndromes. There were anomias, either category-specific (colours) or modality-specific (tactile). For some cases, repetition was specifically altered, which suggested a conduction aphasia, and, in some others, it was specifically spared, thus calling for a diagnosis of transcortical aphasia. Some of the children had severe comprehension disorders with a high percentage of paraphasias in ongoing speech, at times with neologisms, which evoked a sensory aphasia. Sometimes, these latter children presented episodes of fluent speech that even qualified as logorrhea, and that, in a few cases, was accompanied by anosognosia and could be classified as true Wernicke's aphasia.

The main parameter that seemed to differ in children and adults was fluency. We scored fluency as the mean number of words per minute during a given period of spontaneous speech (picture description and conversation). As a rule, except for some jargons, this parameter was altered, for several reasons: word-finding difficulties with pauses preceding content words; overall articulatory slowing, more marked in

younger children; and hypospontaneity, which occurred even in some jargonaphasias as the children rarely initiated speech.

Thus, with the exception of fluency, most of the classic data on symptoms in acquired childhood aphasia are challenged.

3. Implications for Language Organization in the Developing Brain

As for anatomo-clinical correlations, they, too, were very similar to what is found in the adult. Anomia occurred with left parietal lesions; repetition disorders with left parieto-temporal localization; transcortical sensory aphasia with left watershed types of lesions; and neologistic jargons with left postero-temporal location of the injury. These last correlations, in particular, call in question the traditional view that the occurrence of symptoms is directly correlated with age. Even in adults, it is often stated that Wernicke's aphasia characterizes the oldest patients, a conduction aphasia being more common, for the same lesion localisation, in younger adults. Those clinical data have been used to support the hypothesis of prolonged installation of left-hemisphere dominance for language, far beyond puberty (Brown and Jaffé, 1975).

Our recent description of a true Wernicke's aphasia in a ten-year-old boy strongly challenges this notion (Van Hout and Lyon, 1986). That the lesion in this case was caused by herpes simplex encephalitis argues for the severity of the lesion rather than age itself being responsible for the symptoms. Necrotic lesions due to herpes may well engender the same restriction of brain plasticity in a young organism as seen in elderly adults with a cerebro-vascular accident.

4. The Genesis of Neologisms

We explored these cases of severe jargons in-depth on a more cognitive level, particularly as regards the mechanism of production of the neologisms and mostly as single cases studies (Van Hout and Lyon, 1986; Van Hout and Evrard, 1987). The first case description is that of a boy, aged ten, who presented a herpes simplex encephalitis. He was successfully treated with antiviral agents, but he displayed a left temporal lesion. When the child regained consciousness, he produced limited stereotyped utterances at first. After a few days, these stereotypes began to break down and then to fuse together to form neologisms. His utterances were soon produced at a high speed and also became alliterative. The child was anosognosic for the inappropriateness of his productions and had severe comprehension disorders.

We analyzed the temporal evolution for some language parameters for one month, particularly for naming. A naming task is often useful for studying the genesis of neologisms, because the target word is known. The child was presented with the same set of pictures in succession in four different sessions, and his errors were analyzed. At first, he spontaneously produced only a stereotype. We then always offered a syllabic cue, which he blended with the last syllable of his stereo-

type. For instance, the French word "mouchoir" (handkerchief) was named by the stereotype "soleil" (sun), then, after the presentation of the cue "mou", he produced the blend "mouleil". After a while, he began to incorporate phonemes of the target between the cue and the last part of the stereotype. "Mouchoir" gave rise to the following production "moucheil", where the target phoneme "ch" was inserted between the cue and the end of the stereotype. Later on, he began to produce the target, but never immediately: other pictures had to be shown before the delayed appearance of a preceding target, and the correct answer was not given if the corresponding picture was shown after this production for which he appeared anosognosic. For instance, "mouchoir" was called "bouteille" (bottle), but then "mouchoir" was correctly produced after four or five other picture presentations. This seemed to be similar to a form of naming deficit described in adult aphasics that appears to bypass the semantic route to access directly the phonological word form (Ratcliff and Newcombe, 1982).

At about 14 days after the end of the perseverative stage, the child began to produce verbal paraphasias. They were limited in number for a given semantic category and usually presented a rather remote link with the target. The same phenomena--perseverations, blendings, delayed appearance of the target--were still present. Precisely when the verbal paraphasias appeared, true neologisms began to increase, probably as a result of the greater number of possible blendings with more available words.

The mechanism of blending could still explain the formation of neologisms, as in the following example. The picture of the moon ("lune" in French) was called "faleine", a neologistic production since more than half of the word's phonemes were substituted. A previous picture, a whale ("baleine" in French) had been called "petit oiseau" (little bird), a rather remote semantic paraphasia. Then the picture of a flower ("fleur" in French) had been correctly named, and, after this answer, the neologism was produced for "moon". "Faleine" was thus the result of the blending between the perseveration of the initial phoneme of the preceding word "fleur" and the delayed appearance of the target for "baleine". These findings support Buckingham's theory (1978) on the genesis of neologisms and oppose the "conduction hypothesis", which would have neologisms formed by numerous phonemic substitutions of either the target word or its semantic paraphasia. In our case, the number of phonemic paraphasias remained at a stable and low level, an additional argument in favour of Buckingham's theory.

The same mechanism of neologism formation was demonstrated in a three-year-old girl (Van Hout and Evrard, 1987) with a jargonaphasia due to herpes simplex encephalitis. The main difference with the previous case was that neologisms were fewer.

5. Conclusions

Our findings and those of the recent literature indicate that there is a positive semiology in acquired aphasia in childhood. All types of

adult-like aphasic forms have now been described in children (Cranberg et al., 1987; De Agostini and Kremin, 1986; Martins and Ferro, 1987; Van Dongen et al., 1985; Van Hout et al., 1985; Van Hout and Lyon, 1986; Van Hout and Evrard, 1987).

The localization of the lesion is similar to what is observed in adults, and this since at least the age of three years old.

Among the reasons for this discrepancy between recent findings and the previous literature is that, in the recent series, the children were examined earlier and with more age-appropriate tests batteries than in previous series. The difference in fluency between children and adults might also be a reason. In adults, the classic opposition between fluent and non-fluent aphasias corresponds rather neatly to the post and pre-rolandic localizations with Wernicke-like and Broca-like types. When altered, all the criteria used to define fluency (usually six in number according to Kerschensteiner, 1972) are homogeneous. In children, the alteration of the different parameters are frequently dissociated, and there are cases of aphasia without articulation problems and even cases of severe jargonaphasia accompanied by a reduced fluency, particularly in acute stages and in younger children. This may have obscured the picture and led to the assumption of a predominantly expressive character of symptoms in acquired childhood aphasia.

Thus, the differences between aphasias in children and adults now appear more quantitative than qualitative or to affect restricted parameters only, such as fluency.

Those findings have important therapeutic implications. The same symptoms, if deriving, as some cases indicate, from the same underlying cognitive mechanism, may benefit age adaptation of rehabilitation methods already proven effective in adults.

The similarity also observed in children and adults of the anatomic correlation between organized symptoms and defined lesion localization, suggests an early specification of the language areas in the left hemisphere, not only for sensori-motor features but also for the dynamic organization of the different aspects of language.

Finally, the similarities now apparent between aphasic symptoms of adults and children and their neuro-anatomic correlates support the analogical reasoning already applied in the field of developmental dysphasia and thus contributes to the foundations of a true neuropsychology of childhood.

REFERENCES

Alajouanine, Th. and Lhermitte, F. (1965) 'Acquired aphasia in children', Brain 88, 653-662.

Brown, R. and Jaffe, J.(1975) 'Hypothesis on cerebral dominance', Neuropsychologia 13, 107-110.

Buckingham, H., Avakian-Whitaker, H., and Whitaker, H. (1978) 'Alliteration and assonance in neologistic jargonaphasia', Cortex 14, 365-380.

Cranberg, L., Filley, C., Hart, E., and Alexander M. (1987) 'Acquired aphasia in childhood: Clinical and CT investigations', Neurology 37, 1165-1172.

De Agostini, M. and Kremin, H. (1986) 'Homogeneity of the syndrome of acquired aphasia in childhood revisited: case study of a child with transcortical aphasia', Journal of Neurolinguistics 2,1, 179-187.

Ferro, J.M., Martins, I.P., Pinto, F., and Castro-Caldas, A. (1982) 'Aphasia following right striato-insular infarction in a left-handed child: a clinico-radiological study', Developmental Medicine and Child Neurology 24, 173-182.

Goodglass, P.S. and Kaplan, E. (1972) The Assessment of Aphasia and Related Disorders, Lea & Febiger, Philadelphia.

Hécaen, H. (1976) 'Acquired aphasia in Children and the Ontogenesis of Hemispheric Functional Specialization', Brain and Language 3, 114-134,

Jakobson, R. (1962) Langage enfantin et aphasie, Editions de Minuit, Paris.

Kerschensteiner, M., Poeck, K., and Brunner, E. (1972) 'The fluency-non-fluency dimension in the classfication of aphasic speech', Cortex 8, 233-247.

Klein, S.K., Masur, D., Farber, K., Shinnar, S., and Rapin, I. 'Persistent fluent aphasia in a young child' (in press).

Lenneberg, E.H. (1967) Biological Foundations of Language, Wiley, New York.

Martins, I.P. and Ferro J.M. (1987) 'Acquired conduction aphasia in a child', Developmental Medicine and Child Neurology 29, 532-536.

Rapin, I. and Allen, D. (1988) 'Syndrome in developmental dysphasia and adult aphasia', in F. Plum (ed.), Language, Communication and the Brain, Raven, New York, pp. 57-75,

Ratcliff, G. and Newcombe, F. (1982) 'Object recognition: some deductions from clinical evidence', in: A. Ellis (ed.), Normality and Pathology in Cognitive Functions. Academic Press, London.

Roch-Lecours, A., Dordain, G., and Lhermitte, F. (1970) 'Recherches sur le langage des aphasiques', L'Encéphale 59, 520-546.

Spreen, O. and Benton, A.L. (1969) 'Neurosensory center comprehensive examination for aphasia', Neuropsychology Laboratory, University of Victoria.

Tanabe, H., Ikeda, M., Murasama, A., Yamada, K., Nakagawa, Y., Nishimura, Y., and Shiraishi, J. (1989) 'A case of acquired conduction aphasia in a child', Acta Neurologica Scandinavica 80, 314-318.

Van Dongen, H., Loonen, M., and Van Dongen, K. (1985) 'Anatomical basis for acquired fluent aphasia in children', Annals of Neurology 17, 306-309.

Van Dongen, H.R. and Visch-Brink, E.G. (1988) 'Naming in aphasic children, analysis of paraphasic errors', Neuropsychologia 26, 629-632.

Van Dongen, H.R. (1989) 'Acquired childhood aphasia, course and outcome', paper presented at the Meeting of the International Neuropsychological Society, Antwerp.

Van Hout, A. (1983) 'Importance of paraphasias in acquired aphasia in children', paper presented at the meeting of the International Neuropsychological Society, Lisbon.

Van Hout, A. and Seron, X. (1983) 'L'aphasie de l'enfant', Mardaga, Brussels.

Van Hout, A., Evrard, Ph., and Lyon, G. (1985) 'On the positive semiology of acquired aphasia in children', Developmental Medicine and Child Neurology 27, 231-241.

Van Hout, A. and Lyon, G. (1986) 'Wernicke's aphasia in a ten-year-old boy', Brain and Language 29, 268-285.

Van Hout, A. and Evrard, Ph. (1987) 'Aphasie sensorielle chez un enfant de moins de quatre ans', in J. Narbona and M. Poch-Olive (eds.), 'Neuropsychologie infantile: données récentes', Symposium international de Pampelune, 81, 294,

Von Cramon, D. (1975) 'Traumatic mutism and the subsequent reorganisation of speech functions', Neuropsychologia 19, 281-303.

Woods, M.T., Teuber, H.L. (1978) 'Changing pattern of childhood aphasia', Annals of Neurology 3, 273-280.

FLUENT APHASIA IN CHILDREN

H.R. VAN DONGEN
Department of Neurology
University Hospital Rotterdam-Dijkzigt
Room EE 2287
PO Box 1738
3000 DR Rotterdam
The Netherlands

P. PAQUIER
Division of Neurolinguistics
(Departments of Neurology and ENT)
Antwerp University Hospital
Wilrijkstraat 10
2650 Edegem
Belgium

ABSTRACT. It has been widely stated that acquired aphasia in children is invariably nonfluent. However, recent descriptions of different - including fluent- types of aphasia have modified considerably the traditional ideas on the standard clinical picture of acquired childhood aphasia. In a series of 42 children with an acquired aphasia, we observed seven cases in the peri-acute stage, which did not fit into the standard doctrine. By analyzing the main characteristics of their spontaneous speech, we wanted to investigate : (1) the frequency and degree in which the seven children show the fluency characteristics proposed by Kerschensteiner, Poeck and Brunner (1972); (2) whether a tendency for the speech/language variables to cluster according to adult types of aphasia exists; and (3) to what extent the heterogeneity of clinical pictures demonstrated in the recent literature would recur in these seven children. Our results confirm the heterogeneous clinical picture having been reported within fluent childhood aphasia.

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 125–141.

Introduction.

The traditional description of acquired childhood aphasia claims that : (a) verbal comprehension remains relatively preserved; (b) speech is invariably nonfluent and markedly reduced (ranging from mutism to articulatory problems); (c) aphasic children speak in a telegraphic style; (d) neologisms, jargon and logorrhea are absent; (e) recovery of language disorders is rapid and complete; and (f) acquired aphasia after right cerebral lesion is common (Clarus, 1874; Bernhardt, 1885; Freud, 1897; Guttmann, 1942; Basser, 1962; Alajouanine and Lhermitte, 1965; Lenneberg, 1967; Brown, 1976; Brown and Hécaen, 1976; Hécaen, 1976; Kertesz, 1985). In other words, the clinical picture resembles that of nonfluent aphasia.

Although the classical dichotomy between fluent and nonfluent aphasic speech (Jackson, 1915) has been updated by Goodglass, Quadfasel and Timberlake (1964), Howes (1964), and Howes and Geschwind (1964), the terms "fluent" and "nonfluent" do not always refer to uniform criteria. For instance, speech fluency has been defined by the phrase length, i.e. the number of words uttered between two pauses (Goodglass et al., 1964), but also by the speech rate, i.e. the number of words uttered per time unit (Howes, 1964). Other measures have also been introduced, such as inadequate pauses (Kerschensteiner, Poeck and Brunner, 1972), press of speech, i.e. the inability to stop speaking (Benson, 1967), or syntactic complexity and communicative capacity, i.e. the flexibility in using syntactic structures of varying complexity and the degree of aphasia (Wagenaar, Snow and Prins, 1975).

Definition problems arise even with the most widely accepted and apparently most simple criteria. For instance, according to Howes (1964), normal speech rate ranges from 100 to 175 words per minute (wpm), nonfluent aphasic patients uttering less than 100 and "hyperfluent" aphasic patients more than 175 wpm. Kreindler, Mihailescu and Fradis (1980) confirmed these values: normal speech rate ranges from 90 to 175 wpm. On the other hand, Benson (1967) found the nonfluent aphasics to speak less than 50 wpm and the "hyperfluent" aphasics more than 150 wpm, the intermediate area corresponding to the normal speech fluency. For Wagenaar et al. (1975) the cutoff values were respectively less than 300 words and more than 540 words in 6 minutes. Kerschensteiner et al. (1972) distinguished two nonfluent groups (from 0 to 50 and from 51 to 90 wpm), and one fluent group (more than 90 wpm).

Speech rate does not seem to be the only measure defining speech fluency. In one of the -to our opinion- methodologically best designed studies, Kerschensteiner et al. (1972) demonstrated that no less than 10 variables contribute to distinguish fluent and nonfluent clinical pictures, and that this classification of spontaneous aphasic speech reflects naturally occurring differences in language behavior. By using cluster analysis, they have ranked these 10 variables according to their greatest discriminating power between fluent and nonfluent groups, thus obtaining the following rank order: (1) phrase length; (2) pauses (i.e. hesitations); (3) prosody; (4) rate of speaking; (5) effort; (6) articulation; (7) word choice; (8) verbal paraphasias; (9) literal paraphasias; (10) perseveration. We refer to Monrad-Krohn

(1947), Benson (1967), and Kerschensteiner et al. (1972) for the definitions of these variables.

Although one incidentally encounters many of these 10 criteria in the various descriptions of acquired childhood aphasia published in the last decade, nevertheless all 10 have never been mentioned together[1]. After the observation of jargon aphasia in a 5-year-old boy reported by Woods and Teuber (1978), Van Hout, Evrard and Lyon (1985) subsequently described two cases of jargon aphasia with verbal and literal paraphasias in a series of 11 children; in addition, verbal perseverations as well as verbal comprehension deficits occurred, which, in association with repetition difficulties in the first case, suggest Wernicke's aphasia (see also Van Hout and Lyon, 1986), and associated with a normal repetition of spoken language in the second case, suggest transcortical sensory aphasia. Cranberg, Filley, Hart and Alexander (1987) documented another case of transcortical sensory aphasia in their series of eight children: two months after a traffic accident, the patient had verbal comprehension difficulties associated with an intact repetition of spoken language; verbal output was fluent, and contained literal and verbal paraphasias, as well as neologisms. Visch-Brink and Van de Sandt-Koenderman (1984) observed an 11-year-old girl whose verbal comprehension disorders, literal and verbal paraphasias, perseverations, neologisms, word-finding difficulties, paragrammatism, and repetition disorders evoked Wernicke's aphasia. Paquier, Saerens, Parizel, Van Dongen, De La Porte and de Moor (1989) described a 13-year-old girl presenting with alexia without agraphia, whose spontaneous speech was characterized by word-finding difficulties which occasionally resulted in verbal paraphasias, but whose syntax was normal. Hynd, Hern, Cuff and Semrud-Clikeman (1990) documented a case of anomic aphasia with fluent but empty and circumlocutory spontaneous speech in a 10-year-old girl. Martins and Ferro (1987) recorded acquired conduction aphasia in an 11-year-old girl showing many fluent characteristics: phrase length, syntaxis, prosody, and articulation were normal; some pauses and hesitations, as well as occasional literal paraphasias resulting from word-finding difficulties were noted. In reporting a second case of acquired conduction aphasia with incidental hesitations and literal paraphasias, but without syntactic or articulation disorders, Tanabe, Ikeda, Murasawa, Yamada, Yamamoto, Nakagawa, Nishimura, and Shiraishi (1989) underlined the diversity of the clinical picture of acquired childhood aphasia. Following Kerschensteiner et al.'s (1972) criteria, Van Dongen, Loonen and Van Dongen (1985) observed three children whose phrase length, hesitations, prosody, speech rate, effort of production and articulation corresponded to the fluent profile. The presence of neologisms and jargon, as well as the

---

[1] Although it seems that there was no need to introduce new criteria, modifications nevertheless had to be made. Van Hout et al. (1985), for instance, hold the view that the speech rate norms have to be halved in children. In their fluency scale from 0 to 7, the highest score corresponds to a rate of more than 45 wpm.

preservation of prosody have also been observed in cases of Acquired Aphasia with Convulsive Disorder (Landau and Kleffner, 1957).

These observations do not only prove that the standard description of acquired childhood aphasia needs modification, but they also demonstrate that a heterogeneity of clinical pictures within fluent aphasia types in children as well as in adults does exist, without the semiology of the former being necessarily identical to the latter in all respects.

In the period 1976-1988, we examined 42 children who became aphasic, and observed seven cases which did not fit into the standard description. By analyzing the main characteristics of their spontaneous speech, we wanted to investigate : (1) the frequency and degree in which the seven children show the fluency characteristics mentioned above (Kerschensteiner et al., 1972); (2) whether a tendency for the speech/language variables to cluster according to adult types of aphasia exists; and (3) to what extent the heterogeneity of clinical pictures demonstrated in the recent literature would recur in these seven children.

Patients and Methods

All children were examined at the University Hospital Rotterdam-Dijkzigt. Table 1 gives their main characteristics. All were right-handed and Dutch-speaking, and had had a normal psychomotor and language development up to the onset of the aphasia. None had shown any neurological symptoms before the onset of the disease. All children but one (patient 5 who was under age) attended normal primary school.

The initial language assessment was performed as soon as possible after the onset of the aphasia, i.e. in the peri-acute period. Spontaneous speech was elicited by means of standard questions, recorded in the presence of the mother, and rated according to Kerschensteiner et al. (1972)[2]. In order to allow for a more deepened aphasiological diagnosis, and to determine whether a further differentiation as to the aphasia type could be achieved within the fluent group, oral comprehension, oral repetition, and picture-naming tasks were also presented (see Van Dongen (1988) for detailed information).

[2] Another reason for choosing this method is the affinity between German and Dutch, besides the good methodological base.

Table 1: Patients' Characteristics

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7
Sex	F	F	F	F	M	F	M
Age at lesion onset (yrs, mths)	9.2	10.4	11.1	10.9	4.10	9.3	10.0
Etiology	Intracerebral hematoma	Cerebral trauma	Cerebral trauma	Acute general cerebral swelling	Hemophilus Influenza meningo-encephalitis	Intracerebral tumor	Herpes simplex encephalitis
Lesion location (CT-scan)	L temporal	L temporal	L temporal	L temporo-occipital (paramedian)*	L hemispheric R paraventricular	L temporal	L temporal
Test time post-onset (days)	30	3	7	21	12	26	23

* increased Technetium uptake on cerebral scintigraphy

MEDICAL OBSERVATIONS

Patient 1: this 9-year-old girl suffering from aplastic anemia was found to be dull and responded inadequately to questions. Upon admission, neurological examination revealed a nuchal rigidity and a right-sided hemiparesis. She was aphasic. The computed tomography (CT) scan showed a left temporal hyperdense space-occupying lesion, surrounded by a hyperdense halo. The density pattern and the lesion configuration suggested a hematoma with extravasation of blood in the subarachnoid space. The patient was treated with suspensions of thrombocytes and corticosteroids in high doses for 10 days. Seventeen days after the onset of the neurological signs, the extent of the hematoma was grossly reduced on CT-scan. The lateral ventricles were slightly enlarged. One month later, a bone-marrow transplantation was carried out. Two months later, the girl had a series of generalized tonic-clonic fits. Anti-epileptic treatment was successful. The hemiparesis gradually recovered. One year after the onset of the neurological signs, a third CT-scan showed a diffuse enlargement of the ventricles, and a peripheric hyperdensity suggesting cortical or subcortical calcifications.

Patient 2: this 10-year-old girl developed language difficulties without loss of consciousness after having been hit by a car. Upon admission, a large subcutaneous hematoma was palpable at the left temple. Apart from a slight right-sided facial paresis and aphasic disturbances, no other neurological signs were found. A CT-scan disclosed a left temporal impression fracture, a subgaleal hematoma in the vicinity of the fracture, and a mixed density lesion in the left temporal lobe indicating a cerebral contusion. Surgical correction of the impression fracture was performed six days after the accident. The postoperative course was uneventful, and the girl could be discharged two weeks after admission, showing only minimal aphasic signs. Two months after the accident, a control CT-scan showed local atrophy consecutive to the contusion in the left posterior temporal region.

Patient 3: this 11-year-old girl lost consciousness for some minutes consecutively to a traffic accident. After having been brought home, she talked confusedly, and was at times somnolent. One day later, she occasionally vomited and seemed drowsier. Upon admission, she was conscious, but gave inadequate answers and did not seem to understand instructions well. Neurological examination only showed a slight asymmetry of the reflexes, the knee and ankle jerks being brisker on the right. The girl seemed "confused" during the first three days after admission. On the fourth day, she had a focal convulsion followed by a post-ictal right-sided hemiparesis and hemi-inattention lasting for several hours. Soon thereafter, spontaneous recovery was observed, and 12 days after the accident she could be discharged with only slight word-finding difficulties. A CT-scan performed five days after the accident disclosed a small area of mixed density (pepper and salt appearance) in the left posterior temporal region.

Patient 4: this 10-year-old girl, intermittently hemodialyzed because of renal insufficiency, was admitted with headache, nausea, and restlessness at night. Neurological examination was normal, except for a refractory hypertension. Two months after admission, she went into a hypertensive crisis (290/190 mm Hg) followed by a coma. Neurological examination revealed nuchal rigidity, papilledema, and bilateral extensor plantar responses. An electro-encephalogram (EEG) showed diffuse bradyrhythmia, and theta and delta waves more marked in the left temporal region. Acute general brain swelling was diagnosed, and the girl was treated with corticosteroids, fluid restriction and antihypertensive drugs. She recovered gradually in the following days. Two weeks later, neurological examination showed a slight right-sided hemiparesis, a right homonymous hemianopia, and aphasia. Two months after the onset of the neurological signs, a cerebral scintigraphy revealed a small zone of increased Technetium uptake in the paramedian part of the left temporo-occipital region. The girl underwent renal transplantation 10 months after admission.

Patient 5: this 4-year-old boy was admitted because of a Hemophilus Influenza meningo-encephalitis. Initially, he was unconscious and had right-sided focal convulsions. When the coma subsided after eight days, the boy had a right-sided hemiparesis and was aphasic. A CT-scan was performed one week after the onset of the illness. Almost the whole left hemisphere was hypodense, and the median structures were displaced to the right. Moreover, a periventricular hypodensity was observed on the right. A cerebral angiography showed a pronounced capillary pattern in the distribution of both the left and the right carotid arteries. A second CT-scan two months post-onset revealed widening of the left lateral ventricle. The boy could be discharged three months after the onset of the illness, but had a right-sided hemiparesis, a right homonymous hemianopia, and an aphasia. In the following years, the right arm remained clumsy and became atrophic. In addition, the boy developed psychomotor epilepsy.

Patient 6: this 9-year-old girl was admitted because of lasting complaints of headache, nausea and tiredness. On neurological examination, the girl was conscious and well-orientated. No aphasic signs were present. Clinical investigation revealed papilledema, a right homonymous hemianopia, and a right-sided hypoesthesia. An EEG showed left temporal focal abnormalities. A space occupying tumoral mass in the left temporo-parietal area was disclosed by CT-scan, and confirmed by cerebral angiography. Fifteen days after admission, the tumor was extirpated. Neuropathological examination showed it to be a ganglioneuroma. After surgery, she developed grand-mal seizures and later on right-sided focal fits. She was aphasic and slightly hemiparetic. The patient's general condition improved steadily, and it was possible to discharge her one month after admission. However, she still had epilepsy, and presented a persisting right homonymous hemianopia.

Patient 7: this 10-year-old boy suddenly developed headache and high fever. Upon admission, neurological examination revealed a mild right-

sided hemiparesis and fluent aphasia (empty speech) with comprehension difficulties. A CT-scan disclosed a left temporal hyperdense area. Routine laboratory tests confirmed the diagnosis of herpes simplex encephalitis. In the following days focal epileptic attacks were observed. The boy was given anti-epileptic drugs and remained without convulsions. A repeated CT-scan one month later, showed a decrease of the lesion size. At that time, the boy was discharged. The hemiparesis had disappeared, and no further seizures occurred. However, the patient remained severely aphasic.

NEUROLINGUISTIC OBSERVATIONS

The patients' spontaneous speech characteristics are listed in table 2 and analysed in the Results.

Patient 1: 30 days after the onset of the aphasia, the girl was cheerful and had a high speech rate with paragrammatic errors. Although neither paraphasias nor neologisms were present in spontaneous speech, both were observed during the picture-naming and the oral repetition task. Naming difficulties were so severe that the girl stopped the test after 10 trials. She could repeat only three monosyllabic words correctly. On the Token Test (De Renzi and Vignolo, 1962), she could perform only six of the 10 items in the first (easiest) series. Oral comprehension difficulties were noted in conversational situation as well.

Patient 2: 3 days post-onset, the girl's spontaneous speech and picture-naming contained neologisms and literal paraphasias. Paragrammatism was present in conversational speech. Repetition of polysyllabic words was impossible, as was the repetition of three-word sentences. She scored poorly on the Token test (28/61 correct), and evidenced oral comprehension difficulties in spontaneous conversation.

Patient 3: 7 days post-onset, the girl's spontaneous speech was momentarily empty, and the range of information which could be exchanged was very limited (a.o. neologisms, paraphasias). She was paragrammatic and severely anomic (20/40 correct on the picture-naming task). The greatest part of polysyllabic words was repeated incorrectly. Oral comprehension difficulties were obvious, both in spontaneous conversation and during assessment (16/61 correct on the Token Test).

Patient 4: after the onset of the neurological signs, the girl's comprehension of spoken language was minimal. However, echolalia was present, and she was able to repeat single words. 21 days post-onset, severe word-finding difficulties in spontaneous speech resulted in many pauses and in occasional paraphasias. Paragrammatic errors were observed. Picture-naming was severely disturbed (5/13 correct), and contained paraphasias and neologisms. Repetition of three-syllabic words and of three-word sentences was normal. However, oral comprehension was defective in conversation and during assessment (21/61 correct on the Token Test).

Patient 5: 12 days post-onset, the boy jabbered phonemic jargon when reacting to verbal commands or to objects shown to him. Some pictures elicited a stream of unintelligible sounds. An attempt to administer a picture-naming test, a repetition test, and a simple oral comprehension test resulted in total failure.

Patient 6: 26 days post-onset, spontaneous speech was characterized by word-finding difficulties which resulted in conduites d'approche, generalizations, and in indefinite words. Paragrammatic errors were obvious. Paraphasias were noted during the picture-naming task (33/40 correct). The girl did not succeed in repeating complex polysyllabic words (conduites d'approche). Despite an apparently intact oral comprehension in conversation, mild receptive deficits were disclosed by the Token test (49/61 correct).

Patient 7: although a global aspontaneity was observed 23 days post-onset, fluent tachylalic outbursts were sometimes noted. Paraphasias, neologisms, and stereotyped utterances occurred. Paragrammatism was obvious. Neologisms and paraphasias also occurred during the picture-naming task (5/40 correct). Repetition of three-letter words was impossible. Oral comprehension deficits were noted during spontaneous conversation, and the boy did not succeed in performing one single command in the first series of the Token Test.

RESULTS

Table 2 gives the patients' spontaneous speech characteristics. The first 10 speech/language variables are ranked according to their power of discriminating between fluent and nonfluent aphasic speech, as was computed by Kerschensteiner et al. (1972). They could be rated in almost all children, but individual differences were observed.
Phrase length could be evaluated in six children. Four of them (patients 1, 2, 6, 7) produced phrases mainly containing more than four words or utterances (for patient 7 this was the case even outside the outbursts). Two children (patients 3 and 4) spoke in phrases mainly containing three or four words. One child (patient 5) could not be evaluated because he produced only incomprehensible phonemic jargon.
Pauses during conversation were judged to be normal in patients 1, 6 and 7, although patient 7 showed too few pauses during the tachylalic outbursts. Relatively many pauses were observed in two children (patients 3 and 4), and relatively moderate pauses in one child (patient 2).

Prosody was judged to be normal in all children but one (patient 5), in whom it was present but difficult to evaluate because of the phonemic jargon.

Speech rate ranged from 87 wpm (patient 4) to more than 200 wpm (patient 7), while no child showed any effort in producing speech, and articulation was normal in all subjects.

Word choice varied from not interpretable phonemic jargon (patient 5) to many clichés and relational words (patients 1, 2, 4, 7).

Table 2: Patients' Spontaneous Speech Characteristics

Speech/language variables''	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7
Phrase length: 1-2 w.	5%	9%	25%	30%	n.i.	10%	15%* 19%**
3-4 w.	29%	14%	42%	44%	n.i.	28%	25% 38%
> 4 w.	66%	77%	33%	26%	n.i.	62%	60% 43%
Pauses	normal	medium	many	many	n.i.	normal	normal**
Prosody	normal	normal	normal	normal	present~	normal	normal**
Rate of speaking (words per minute)	121	96	88	87	n.i.	110	206* 121**
Effort	none	none	none	none	none	none	none**
Articulation	normal	normal	normal	normal	normal	normal	normal**
Word choice: cliché	+	+	-	+	n.i.	+/-	+**
empty	+	-	+/-	+/-	n.i.	+/-	+**
Verbal paraphasias	none	none	some	some	n.i.	some	some**
Literal paraphasias	none	many (jargon +/-)	some	some	phonemic jargon	some	some**
Perseverations	moderate at sentence level	severe at word level	none	none	moderate at phonemic level	moderate at word/sentence level	severe at word/sentence level**
Neologisms	none	many	moderate	none	n.i.	moderate	moderate**
Others		palilalia & conduites d'approche		conduites d'approche		conduites d'approche	stereotypes**

n.i. : not interpretable; + : present; - : absent; +/- : momentarily present;
Patient 5 : ~: although not interpretable;
Patient 7 : *: during tachylalic outbursts; **: during conversation;
'': according to discrimination power between fluent and nonfluent aphasic speech (Kerschensteiner et al., 1972).

Some children obviously showed empty speech (patients 1 and 7), while others did not (patient 2). In some children (patients 3, 4, 6), speech was momentarily empty.

Verbal paraphasias occurred in four children (patients 3, 4, 6, 7). Literal paraphasias were absent in patient 1, and occurred moderately (patients 3, 4, 6, 7) to frequently (patient 2) in the other children.

In all but two children (patients 3 and 4), perseverations at phonemic, word, or sentence level occurred in varying degree.

Other "positive signs" (Van Hout et al., 1985), some of which were thought hardly to occur in acquired childhood aphasia according to the standard doctrine, were also found in our series: neologisms, conduites d'approche, stereotypes, and palilalia. We grouped these symptoms into two categories of characteristics, namely neologisms and others, which were added to Kerschensteiner et al.'s (1972) 10 variables. Neologisms were observed in four children (patients 2, 3, 6, 7), conduites d'approche in three children (patients 2, 4, 6), palilalia in one child (patient 2), and stereotypes in one child as well (patient 7).

As a first conclusion, we may state that all 10 fluency characteristics proposed by Kerschensteiner et al. (1972) were observed in varying frequency in our series.

The next step was to investigate whether some clustering between the various parameters, reflecting the different fluent clinical pictures already described in acquired childhood aphasia, could be found in our patients as well. As we did not include a group of nonfluent children in our study, we could not compute a rank order of speech/language variables with respect to their discriminating power between fluent and nonfluent aphasic speech in children. To investigate whether the rank order established by Kerschensteiner et al. (1972) would recur in our series, is consequently impossible. Some of our children illustrate this point.

Three children out of seven (patients 1, 6, 7) fell into the fluent range on the six characteristics having the greatest power of discrimination between fluent and nonfluent aphasic speech. However, their clinical pictures are very different. For instance, patient 7, who had the highest rate of speaking, did not produce the highest frequency of phrases longer than four words. Three children (patients 2, 3, 4) gave a fluent impression because of the normal prosody, the normal speech rate[3], the well preserved articulation, and the absence of effort. However, patient 2, who produced the highest frequency of phrases containing more than four words, showed moderate pauses due to word-finding difficulties. Patients 3 and 4, on the other hand, mainly produced three to four-word phrases with many pauses. Finally, the phonemic jargon gave the fluent clinical impression in patient 5.

---

[3] The rate of speaking in patients 3 and 4 border on the limit of 90 wpm, and can therefore be considered compatible with the normal fluency value.

Differences between the seven children were also found in the variables with lower discrimination power between fluent and nonfluent aphasic speech. Although patients 1, 6 and 7 are to be considered the children showing the most characteristic fluent features, differences in the clinical picture, which were underlined by the results on the additional language tests, nevertheless occurred.

Though patients 6 and 7 both produced literal and verbal paraphasias as well as neologisms, patient 6 showed a tendency to self-correction with conduites d'approche, whereas patient 7 appeared to be anosognosic for his verbal output problems. Taking into account the patients' performances on the additional language tests, the aphasia in patient 7 can reasonably be diagnosed as Wernicke's aphasia, whereas in patient 6 -the only child out of those three patients who showed conduites d'approche- the overall clinical picture is more similar to that of conduction aphasia. On the other hand, patient 1, who also had one of the most fluent profiles on the first six speech/language variables, only produced paraphasias and neologisms during picture-naming and oral repetition tasks, but not in conversational speech. Moreover, in patient 1 perseverations were observed moderately at sentence level, whereas they were severe at word and sentence level in patient 7. In addition, stereotypes were present in patient 7 but not in patient 1. Yet patient 1 also showed a Wernicke-like clinical picture, but without anosognosic signs.

In two children (patients 3 and 4) having very similar fluency profiles on the first six and even on the next four speech/language variables, the additional language tests, and in particular the oral repetition task, permit to differentiate Wernicke's aphasia in patient 3 and transcortical sensory aphasia in patient 4. Both patients momentarily showed empty speech, but clichés were only noted in patient 4. Patient 3 produced neologisms, while patient 4 had conduites d'approche. In other words, sensory aphasia can occur not only in children showing all speech/language variables with the highest fluency values (patients 1 and 7), but also in children with diminished speech rate and/or phrase length (patients 3 and 4). This clinical feature of fluent childhood aphasia is further illustrated by patient 2, whose overall aphasic profile also matches that of Wernicke's aphasia, although more pauses than normal were observed.

Finally, patient 5 demonstrates that phonemic jargon aphasia without clear morphemic boundaries can occur in children.

In summary, our results are comparable with Kerschensteiner et al.'s (1972) findings, in that many of the fluent speech characteristics they proposed in adults, also recur in our series of seven children having acquired an aphasia which did not fit into the standard doctrine. For each variable, individual quantitative and qualitative differences were observed: children with high speech rate did not necessarily produce the highest frequency of phrases longer than four words; some children producing many phrases containing more than four words did not necessarily show normal pauses; and some children with normal rates of speaking sometimes exhibited excessive pauses. Finally, various clinical pictures reflecting the heterogeneity observed in the recent literature

on acquired fluent childhood aphasia were found in our series. However, children with an adult-like Wernicke's aphasia did not all exhibit identical profiles. Consequently, we are of the opinion that more distinct types of Wernicke's aphasia can occur in children.

DISCUSSION

The clinical picture of acquired fluent aphasia in children.
Our findings demonstrate that, even when cases of Acquired Aphasia with Convulsive Disorder (Landau and Kleffner, 1957) are not taken into consideration, fluent (paraphasic) speech does occur in children, at least in the peri-acute stage, and that, as in adults, different clinical features also do exist within acquired fluent childhood aphasia. However, as Van Hout (1990) also pointed out, intermediate symptoms in opposition to adult fluency criteria can be observed. While nonfluency in adults has been defined as a reduction of speech rate and of the mean length of utterances, together with a reduction in articulatory agility, excessive pauses, marked effort, agrammatism, and dysprosody (Kerschensteiner et al., 1972), this study, as well as the recent literature on childhood aphasia (e.g. Visch-Brink and Van de Sandt-Koenderman, 1984) provide observations of fluent children producing well-articulated sentences reduced in length and/or in speech rate, but without effort and with normal prosody. Furthermore, pauses may not only be due to an articulation disorder, but also to word-finding difficulties. Moreover, Van Hout (1990) observed that excessive pauses reflecting an "unwillingness" or "unreadiness" to initiate speech and a parsimony in producing speech, may also fit in with an overall behavioral hypospontaneity related to psychological or emotional difficulties: "children often react to stressful situations by avoidance" (p 49). The present findings consequently corroborate Van Hout's (1990) and Van Hout et al.'s (1985) observations that acquired childhood aphasia appears as diversified as in adults but that its fluent characteristics may be more frequently dissociated.

Some remarks on the incidence of acquired fluent aphasia in children.
Martins and Ferro's (1982) observation of four fluent cases in a series of 14 children, and the present series of seven fluent patients are in contrast with the reported rarity as mentioned by Lenneberg (1967): "The so-called fluent aphasia (...) is rare or perhaps altogether absent among pediatric patients" (p 146). Several arguments to explain this discrepancy have been put forward (Van Dongen et al., 1985; Van Dongen, 1988). Paquier and Van Dongen (submitted) have summarized them as follows:

(a) In earlier studies of acquired aphasia in children, the suspicion of aphasia arose when a right-sided hemiparesis or hemiplegia was found on examination. In most cases an anterior lesion was probably present, causing a nonfluent aphasia. Moreover, as pointed out by Woods and Teuber (1978), the presence of a hemiplegia indicating a contralateral lesion does not preclude a diffuse bilateral encephalopa-

thy (for instance in cases of severe systemic infections). In addition, a selection of aphasic children based upon the presence of a hemisyndrome excludes the syndrome of Acquired Aphasia with Convulsive Disorder. Therefore, to rely upon hemiparesis as the neurological criterion for inclusion in studies of childhood aphasia might well have led to a methodological bias.

(b) If a (head) trauma is the underlying cause of aphasia, recovery may be observed within a short time (Van Dongen, 1988). The fluent characteristics of the aphasia may thus not have been recognized as such.

(c) When neurological deficits are inconspicuous, fluent aphasia can be misinterpreted as (post-contusional) confusion. However, the occurrence of paraphasias and of neologisms indicates the presence of language disturbances. Moreover, the cooperative behaviour of the children easily allows neuropsychological examination.

(d) The introduction of CT and Magnetic Resonance Imaging (MRI) plays an important role in the early detection of aphasias without other neurological symptoms. The threshold to perform these non-invasive examinations is much lower than is the case for angiography, pneumoencephalography or nuclear brain scanning. Moreover, cerebral contusions and infarctions are far more easily demonstrated on CT-scan and MRI. This could focus the attention of the neurologist on the possibility of fluent aphasia not yet detected or interpreted as such before the advent of modern neuroimaging techniques."

In addition, Van Hout (1990) has pointed out that the introduction of antiviral medication has reduced the mortality of severe infectious diseases, such as herpes simplex encephalitis. Consequently, and because the herpes virus has a tropism for the temporal lobes, posterior types of aphasia in these patients can actually be diagnosed more often than in the past.

Is the clinical picture of acquired childhood aphasia age-dependent ? Geschwind (1974) remarks that: "Perhaps the child's Broca area has no adequate practice in language to run as freely as that of adults" (p 472). Our findings do not only contradict this statement, but also Brown and Hécaen's (1976) hypothesis on the inter- and intrahemispheric language specification, as well as Brown's (1976) postulate that, in the neural organization of language, two trends can be discerned, namely "a sequence in younger children from mutism to agrammatism to phonemic-articulatory errors, and a sequence from adolescence to middle and perhaps into late life leading from phonemic paraphasia and word-finding difficulty to verbal or semantic paraphasia and, with increasing fluency, to semantic and neologistic jargon." (p 490). Evidence against the statement that aphasia is characterized by mutism and/or agrammatism in young children (Brown, 1976) is provided by patient 5 in the present series, by patient 6 (age at lesion 5,1 years) in Woods and Teuber's (1978) series, and by Van Hout et al.'s (1985) patient 10 (aged 4 years at onset), who all exhibited jargon aphasia. The theory of the age-specificity of the aphasic syndromes in children, which also states that aphasia in older children is characterized by phonemic-articulatory defects, has further been contradicted by the variety of

fluent types of acquired aphasia reported in the recent literature (see Introduction).

Acknowledgements.

The authors thank Mrs V. Serlet for revising the English text, and Mrs D. Tinnemans and Mrs H. Hilkemeijer for preparing the manuscript.

References.

Alajouanine, Th. and Lhermitte, F. (1965) 'Acquired aphasia in children', Brain 88, 653-662.

Basser, L.S. (1962) 'Hemiplegia of early onset and the faculty of speech with special reference to the effects of hemispherectomy', Brain 85, 427-460.

Benson, D.F. (1967) 'Fluency in aphasia: correlation with radioactive scan localization', Cortex 3, 373-394.

Bernhardt, M. (1885) 'Über die spastische Cerebralparalyse im Kindesalter (Hemiplegia Spastica Infantilis), nebst einem Excurse über: "Aphasie bei Kindern" ', Archiv für pathologische Anatomie und Physiologie und für klinische Medicin 102, 26-80.

Brown, J.W. (1976) 'The neural organization of language: aphasia and lateralization', Brain and Language 3, 482-494.

Brown, J.W., Hécaen, H. (1976) 'Lateralization and language representation: observations on aphasia in children, left-handers and "anomalous" dextrals', Neurology 26, 183-189.

Clarus, A. (1874) 'Über Aphasie bei Kindern', Jahresbuch für Kinderheilkunde 7, 369-400.

Cranberg, L.D., Filley, C.M., Hart, E.J. and Alexander, M.P. (1987) 'Acquired aphasia in childhood: clinical and CT investigations', Neurology 37, 1165-1172.

De Renzi, E. and Vignolo, L.A. (1962) 'The Token Test: a sensitive test to detect receptive disturbances in aphasics', Brain 85, 665-678.

Freud, S. (1897) 'Infantile Cerebrallähmung', translated by L.A. Russin, Infantile Cerebral Paralysis, University of Miami Press, Coral Gables, 1968.

Geschwind, N. (1974) 'Disorders of higher cortical function in children', in R. Cohen and W. Wartofsky (eds.), Selected Papers on Language and the Brain, Boston Studies in the Philosophy of Science, 16, Reidel Publishing Company, Dordrecht, pp. 467-481.

Goodglass, H., Quadfasel, F.A. and Timberlake, W.H. (1964) 'Phrase length and the type and severity of aphasia', Cortex 1, 133-153.

Guttmann, E. (1942) 'Aphasia in children', Brain 65, 205-219.

Hécaen, H. (1976) 'Acquired aphasia in children and the ontogenesis of hemispheric functional specialization', Brain and Language 3, 114-134.

Howes, D. (1964) 'Application of the word-frequency concept to aphasia', in A.V.S. de Reuck and M. O'Connor (eds), Disorders of Language, Churchill, London, pp. 47-75.

Howes, D. and Geschwind, N. (1964) 'Quantitative studies of aphasic language', in D.M. Rioch and E.A. Weinstein (eds), Disorders of Communication, Williams and Wilkins, Baltimore, pp. 229-244.

Hynd, G.W., Hern, K., Cuff, J.V. and Semrud-Clikeman, M. (1990) 'Anomic aphasia in childhood: charting recovery of function', Paper presented at the 13th European Conference of the International Neuropsychological Society, Innsbruck, Austria.

Jackson, H. (1915) 'On the physiology of language', Brain 38, 59-64.

Kerschensteiner, M., Poeck, K. and Brunner, E. (1972) 'The fluency-nonfluency dimension in the classification of aphasic speech', Cortex 8, 233-247.

Kertesz, A. (1985) 'Aphasia', in J.A.M. Frederiks (ed), Handbook of Clinical Neurology, Clinical Neuropsychology, 45, Elsevier Science Publishers, Amsterdam, pp. 287-331.

Kreindler, A., Mihailescu, L. and Fradis, A. (1980) 'Speech fluency in aphasics', Brain and Language 9, 199-205.

Landau, W.M. and Kleffner, F.R. (1957) 'Syndrome of acquired aphasia with convulsive disorder in children', Neurology 7, 523-530.

Lenneberg, E. (1967) 'Biological Foundations of Language, Wiley, New York.

Martins, I.P. and Ferro, J.M. (1982) 'Afasia adquirida em crianças. Estudo clinico e neuropsichologico longitudinal', Jornal da Sociedade das Ciências Médicas de Lisboa 146, 63-106.

Monrad-Krohn, G.H. (1947) 'Dysprosody or altered "melody of language"', Brain 70, 405-415.

Paquier, P., Saerens, J., Parizel, P.M., Van Dongen, H.R., De La Porte, C. and de Moor, J. (1989) 'Acquired reading disorder similar to pure alexia in a child with ruptured arteriovenous malformation', Aphasiology 3, 667-676.

Paquier, P. and Van Dongen, H.R. (submitted) 'Heterogeneity of fluent aphasia in childhood'.

Tanabe, H., Ikeda, M., Murasawa, A., Yamada, K., Yamamoto, H., Nakagawa, Y., Nishimura, T. and Shiraishi, J. (1989) 'A case of acquired conduction aphasia in a child', Acta Neurologica Scandinavica 80, 314-318.

Van Dongen, H.R., Loonen, M.C.B. and Van Dongen, K.J. (1985) 'Anatomical basis for acquired fluent aphasia in children', Annals of Neurology 17, 306-309.

Van Dongen, H.R. (1988) 'Clinical Aspects of Acquired Aphasia and Dysarthria in Childhood, Eburon Publisher, Delft.

Van Hout, A. (1990) 'Acquired Aphasia in Childhood. Its Impact on the Conception of Functional Maturation of the Brain and its Implication for Pediatric Neuropsychology', unpublished Ph.D. dissertation, Catholic University of Louvain, Belgium.

Van Hout, A., Evrard, Ph. and Lyon, G. (1985) 'On the positive semiology of acquired aphasia in children', Developmental Medicine and Child Neurology 27, 231-241.

Van Hout, A. and Lyon, G. (1986) 'Wernicke's aphasia in a 10-year-old boy', Brain and Language 29, 268-285.

Visch-Brink, E.G. and Van de Sandt-Koenderman, W.M.E. (1984) 'The occurrence of paraphasias in the spontaneous speech of children with an acquired aphasia', Brain and Language 23, 258-271.

Wagenaar, E, Snow, C. and Prins, R.S. (1975) 'Spontaneous speech of aphasic patients: a psycholinguistic analysis', Brain and Language 2, 281-303.

Woods, B.T. and Teuber, H.L. (1978) 'Changing patterns of childhood aphasia', Annals of Neurology 3, 273-280.

TYPE OF APHASIA AND LESIONS' LOCALIZATION

Isabel P. Martins, MD and J. M. Ferro, MD
Centro de Estudos Egas Moniz
Hospital de Santa Maria
1600 Lisboa
PORTUGAL

ABSTRACT. We studied a series of 33 children with acquired aphasia to determine the relation between type of aphasia and lesion site. There were 17 males and 16 females. Age of onset ranged from 1.8 to 15 years. Etiology was variable (14 trauma, 13 vascular, 4 infectious and 2 tumors). Mutism was very common in the acute phase, specially in vascular and traumatic cases and was associated predominantly with frontal or subcortical lesions. We found all types of aphasia that have been described in adults but younger children had predominantly non fluent types. Lesions localization was related to fluency, impaired comprehension and type of aphasia in the same way known for adults. This study suggests that the intrahemispheric specialization for language is established early in life.

INTRODUCTION

Acquired aphasia in children was traditionally described as monotonous in its clinical expression. Early authors, such as Freud (1897) and Bernhard (1885) stated that whatever the side or site of brain damage it consisted in an initial period of mutism, followed by a nonfluent speech without paraphasias and a normal verbal comprehension. This contrasted with the variety of aphasia types found in adults.

The explanation for such findings was either: 1) that language functions in children were not localized in different brain areas, or, alternatively, simply that 2) the limited verbal ability of the young child, or the lack of

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 143–159.

trainning of Broca's area did not permit a more complex and diverse presentation.

In the last 10 years, and specially since the advent of CT Scan, reports have emerged describing other types of aphasia in children, such as Wernicke's (Van Dongen et al, 1988; Van Hout et al, 1985 and Woods and Teuber, 1978) transcortical sensory (Cranberg et al, 1987; Martins et al, 1987) and conduction aphasia (Martins and Ferro 1987; Tanabe et al 1989) but there has been no systematic study of lesion localization in these types of aphasia.

We report our own series of children with acquired aphasia and we try to relate some of the clinical features to the lesions' localization and, also, to their etiology and age of onset.

INCLUSION CRITERIA

We included in this study all our cases of children with acquired aphasia, due the focal hemispheric lesions, sustained after some normal language acquisition and before age 15, who were clinically aphasic at the time of our first evaluation.

We did not include children who had been aphasic but who were fully recovered when first seen. We excluded cases of acquired aphasia with epilepsy, those with previous developmental delay, and children with lesions sustained at, or before, birth.

METHOD

Children were assessed with a language battery consisting of:

1. Rating for severity of aphasia by the Boston Diagnosis Severity Rating Scale (Goodglass and Kaplan, 1972).

2. Analysis of spontaneous speech (for fluency, syntax, paraphasias, anomia, dysarthria)

3. Language tests: a) children younger than 6 were assessed with the Reynnel Developmental Language Scales (Reynnel, 1977, Portuguese version), and with a repetition test; b) children older than 6 were evaluated with the Lisbon Aphasia Battery (with tests of naming, oral comprehension, word and sentence repetition) (Ferro, 1986) and a short version of the Token test (De Renzi and Vignolo, 1962) standardized for this age group (Ferro 1979). Children who attended school were also evaluated with reading and writing tests.

Lesions localization was determinated by CT or MRI scan in 20 cases and by clinical evaluation, EEG, angiography or surgery only, in 13 cases. In the 20 cases assessed by imaging techniques, lesion was located in the left hemisphere in 18 and in the right hemisphere in 2 (one child was a 6 year old

left handed girl (Ferro et al 1986) and the other a 15-yr-old boy with a crossed aphasia) (Martins et al 1987). There were no cases of bilateral lesions seen in the CT scan or MRI.

POPULATION

Our population consists of 33 children, 17 males and 16 females; 31 were right handed, 1 left handed and 1 young child had no hand preference. Age of onset ranged from 1.5 to 15 years (with a mean of 8.7 years)

Most children (24/33) were examined within the first month post onset, 4 others were seen during the first year, and 4 were late referals, i.e. they were seen by us more than 1 year after the onset but while still clinically aphasic.

The etiology was traumatic in 14, vascular in 13 (10 ischaemic and 3 haemorrhagic), infectious in 4 and tumoral in 2 cases.

RESULTS AND DISCUSSION

We shall now analyse a number of features separately:

1) In whats concerns an initial period of mutism; we were able to determine its presence or absence, either through direct clinical assessment or from medical records, in 23 cases. Sixty nine percent of children (15/23) were initially mute and this confirms the high prevalence of mutism in chilhood aphasia.

Mutism was most consistently found in younger children (table 1): 90% of those aged 7 or less had been mute, while in the older ones (8 yr. or more) this percentage falls to 50%.

Table 1

AGE OF ONSET AND MUTISM

AGE OF ONSET (YEARS)	MUTISM YES	NO
< 2	+	-
3	++	
4		
5		
6	+++	
7	+++	
8		-
9	++	
10		-
11	+	- -
12	+	
13	+	-
14		-
15	+	-
TOTAL	15	8

Therefore mutism seems to be a particular response of the young child to a sudden language disturbance. It decreases with age and is quite unusual in adults.

Also, mutism was only present in vascular (6/7) and traumatic (9/11) cases, but not in children with tumors (0/2) or infection (0/3). This suggests that mutism is more common after a sudden brain damage, or a sudden language disturbance than following a progressive lesion. A possible explanation might be that sudden lesions produce a more marked diaschisis than progressive ones.

In 16 chidren we were able to study the relation betwen mutism and lesion site (in CT scan) (table 2). Mutism ocurred predominantly when there was

a prerolandic or subcortical involvement; it was uncommon in isolated posterior (temporal, parietal) lesions.

Table 2

LESION SITE AND MUTISM (16 cases)

LESION SITE	MUTISM YES	MUTISM NO
PREROLANDIC/ SUBCORTICAL	+++ ++	
PREROLANDIC+ SUBCORTICAL+ POSTROLANDIC	+++	
PARIETAL	+	++
TEMPORAL	+	++++
TOTAL	10	6

It is well known that the frontal lobes and the subcortical structures are of part of cortico-subcortical networks subserving motor programming and initiation. These findings suggest that mutism represents a transient period of loss of verbal and motor initiative and not a psychological reaction of the child, as has been previously suggested. Our results also suggest that very young children depend more upon these structures, in order to initiate speech, than do adults.

I will illustrate this point with two examples:

a) A 7-yr.-old right handed girl (Martins et al, 1987) had a head trauma with a depressed posterior fracture. Both from medical records and her mother's information we know that she was mute for one entire year. She was not anarthric for she did not even attemp to speak, (she also had no signs of brainstem or right hemispheric dysfunction). We saw her 4 years later and by then she had a conduction aphasia. Her CT scan showed a very extensive lesion involving most of her left frontal lobe, but also Wernicke's area, the supramarginal gyrus and the insula. Therefore this child had a very long period of mutism associated with large frontal lesion.

b) A 6-yr.-old, right-handed girl suffered an ischaemic stroke. The lesion involved the lenticular nucleus, the posterior limb of the internal capsule the subcortical frontal white matter and the rolandic cortex of the left hemisphere. She was mute for four days and had a right sided hemiplegia. One month later she had a Broca's aphasia (with a nonfluent, slow, dysprosodic and dysarthric speech, poor word repetiton and a normal verbal comprehension). She recovered completely from aphasia in 45 days, although her speech remained dysarthric and dysprosodic.

2. In what regards aphasia type, (table 3), we could find all types of aphasia that have been described in adults, which dismisses the old theory that aphasia in children is always non fluent.However non fluent aphasias were slightly more common (in 3 cases we could not adequately classify the type of aphasia, for children were assessed at bedside during the acute period. As they were not tested with the full language battery, they were just classed, according to their spontaneous speech, as non fluent. We also know that they had a normal oral comprehension).

Table 3

ACQUIRED APHASIA IN CHILDREN

TYPE OF APHASIA	
NON FLUENT	N = 19
NONFLUENT	3
BROCA	10
TRANSCORTICAL MOTOR	2
GLOBAL	4
FLUENT	N = 14
CONDUCTION	1
ANOMIC	6
WERNICKE	3
TRANSCORTICAL SENSORY	4

However our results would be quite different had we considered only children with aphasia and hemiparesis (table 4).

Table 4

TYPE OF APHASIA AND HEMIPARESIS

TYPE OF APHASIA	N	HEMIPARESIS YES	NO
BROCA	9	++++++++	+
GLOBAL	4	++++	
NONFLUENT	3	+++	
TRANSCORTICAL MOTOR	2	++	
ANOMIC	5		+++++
WERNICKE	4	+	+++
TRANSC. SENSORY	3	+	++
CONDUCTION	1		+
TOTAL	31	19	12

If hemiparesis was an obligate inclusion criteria, we would have missed most cases of fluent aphasia. This might explain why old series that used the presence of a right hemiparesis as a major criteria for inclusion (Basser, 1962) found mainly children with nonfluent types of speech. Besides language was not tested in such a systematic and detailed way as it is nowdays. It is possible that children with transient fluent aphasias and no motor defect were considered as confusional states and not as aphasic.

3. Type of aphasia was related to the age of onset: All children younger than 7, except one, had nonfluent types of aphasia and fluent types ocurred mostly in older ones (table 5). Our only exception here is a girl who suffered a head trauma when she was 18 months; this was followed by a severe and longstanding aphasia. She was first seen by us when she was 5 years old and by then she had a transcortical sensory (fluent) aphasia. We do not know what was her aphasia type initially.

Table 5

TYPE OF APHASIA AND AGE OF ONSET

APHASIA TYPE (N)	AGE OF ONSET (YEARS)														
	<2	2	3	4	5	6	7	8	9	10	11	12	13	14	15
BROCA (10)	+	+	+		+	++			+		+	+			
						+									
GLOBAL (4)								+	+				+		+
NONFLUENT (3)		++			+										
TR.MOTOR (2)							+								+
ANOMIC (6)							+	+		++	++				
WERNICKE (4)								+				+	+	+	
TR.SENSORY (3)	+										+				+
CONDUCTION(1)							+								

We were able to corroborate these findings in the published literature. Reviewing other aphasia series (Cranberg, 1987; Van Hout, 1985; Van Dongen, 1988; Woods and Teuber, 1978) and excluding cases of Landau and Kleffner syndrome, we confirmed that fluent aphasias were rare in young children. In the series of Van Dongen (1988) there is a 5-yr-old child with a fluent aphasia and Van Hout (1990) includes a 3-yr-old child with fluent aphasia, but these cases are extremely rare.

It is not very clear why this happens: It could be that: (1) the non-fluency represented a continuum from mutism or a loss of verbal initiative, (2) that the speech of very young normal children is naturally non fluent, or that (3) aphasic children are unwilling to speak (what has been called the hypospontaneity of speech).

Table 6

TYPE OF APHASIA AND ETIOLOGY

TYPE OF APHASIA		ETIOLOGY			
	N	TRAUMA	STROKE	INFECTION	TUMOR
BROCA	(10)	++++	++++++		
GLOBAL	(4)	++	+	+	
NONFLUENT	(3)	+	++		
TRANSC. MOTOR	(2)	+	+		
ANOMIC	(6)	+++	++		+
WERNICKE	(4)	+		+++	
TRANSC. SENSORY	(3)	+	+		+
CONDUCTION	(1)	+			
TOTAL	33	14	13	4	2

While stroke mainly produced nonfluent aphasia types (Broca's, global, transcortical motor), and infection mainly caused fluent aphasias (Wernicke's Transcortical sensory), trauma was associated with all types. These associations are probably related to the fact that stroke in children affects mainly the subcortical areas (Zimmerman et al, 1983) and infection by herpes encephalites damages mostly the temporal cortex. However and since most series of acquired aphasia include traumatic cases, it is still not clear why there should be no fluent cases in younger children.

5. We studied the relation between type of aphasia and lesions localization (table 7).

Table 7

LESION SITE AND TYPE OF APHASIA

APHASIA		LESION SITE			
TYPE	N	PREROL.	SUBCORT.	PARIETAL	TEMPORAL
BROCA	(7)	++++	++++	+++++	++
GLOBAL	(3)	++	+		++
TR.MOTOR	(1)		+		
ANOMIC	(1)				+
WERNICKE	(3)				+++
T.SENSORY	(3)			+	++
CONDUCTION	(1)	+		+	+
TOTAL	(19)				

We found a strong association between site of lesion and aphasia type, the same relation that could be expected in adults: fluent aphasia, such as Wernicke's, anomic and transcortical sensory, were associated with temporal lobe lesions, conduction aphasia with a fronto-parietal lesion (sparing Broca's area); and non fluent aphasias (Broca's, global and transcortical motor) resulted from anterior lesions, although in some cases it also involved the temporal lobe. This suggests that the intrahemispheric specialization for language is already present early in life.

It is also important to mention at this point, that the type of aphasia, defined by the scores obtained in the different language tests (Castro-Caldas, 1979), varies during their follow-up time. As children recover different aspects of language (comprehension, repetition, fluency), so they change the initial aphasia type, as can be seen in the following examples:

a) A 9-yr-old boy, suffered a head trauma. He was in mutism for a few days. When assessed on the 10th day he had a severe global aphasia. His speech was nonfluent, reduced to a stereotype, and naming, comprehension and

repetition were very poor. One month later his comprehension and repetition were much improved and he was then classified as transcortical motor, a type of aphasia more commonly associated with subcortical or frontal lesions. Instead his CT scan showed a small fronto- temporal lesion involving Broca's area and the temporal lobe, which explains the initial global aphasia.

b) A 12-yr-old boy suffered a head trauma with a drepressed parietal fracture. He could only produce monossylables for 3/4 months.He was first examined 4 years later, and by then he had a Broca's aphasia. His comprehension of simple commands was normal but he still had a low score in the Token Test. Four years later (8 years post onset) we was classified as a conduction aphasic. His scan showed a very extensive lesion involving the rolandic cortex and the temporal lobe lesion sites that are not usually associated with conduction aphasia. It is therefore plausible that his verbal comprehension was initially impaired, although this was not recorded at the time.

As aphasia in children recovers quickly, one may be misled if the neuropsychological evaluation is performed too late.

6) Going back to the question of fluency, we studied the relation between fluency and lesion site in the 20 cases with scan (table 8).

Table 8

FLUENCY AND LESION SITE

FLUENCY	N	LESION SITE			
		PREROL.	SUBCORT.	PARIETAL	TEMPORAL
NONFLUENT	12	++++ +	++++ +++	++++ +	++++
FLUENT	8	+		+++	++++ ++
TOTAL	20				

The results here are similar to those just mentioned: non fluency is related to anterior lesions or subcortical lesions and fluent speech to isolated posterior lesions.

Therefore, the predominantly nonfluent speech of very young children may just be a result of their anterior lesions, as would be expected in adults, and it remains to be determined if it is indeed related to the child's age.

7. Another controversial point concerns the presence of paraphasias. We studied the relation between paraphasias and lesion site in 20 cases. Nine patients had paraphasias, and this was associated with different lesion sites but predominantly temporal (table 9). We did not make a seperate analysis of the type of paraphasias.

Table 9

LESION LOCALIZATION AND PARAPHASIAS

PARAPHASIAS		LESION SITE			
	N	PREROL.	SUBCORT.	PARIETAL	TEMPORAL
YES	9	++	+	++	++++ ++++
NO	11	++++	+++ +++	+++ ++	++
TOTAL	20				

8) Finally we studied the relation between lesion site and verbal comprehension (table 10). And, as has been established in adults, a poor comprehension was mostly associated with temporal lobe lesions.

Table 10

ORAL COMPREHENSION AND LESION SITE

COMPREHENSION		LESION SITE			
	N	PREROL.	SUBCORT.	PARIETAL	TEMPORAL
GOOD	11	++++	++++ ++	++++ +++	+++
POOR	9	++	+	+	++++ +++
TOTAL	20				

CONCLUSIONS

1) An inital period of mutism is common in acquired aphasia in children. It is more common in young children, in those with stroke or trauma and when the lesion affects the frontal or the subcortical structures. Possibly these structures are more important to iniciate speech in younger than older children, and are often disturbed by the inital diachisis.

2) All types of aphasia described in adults can be found in children. The clinical expression can no longer be considered monotonous, although nonfluent types of aphasia predominate.

Paraphasias do occur, and there may be an impairment of verbal comprehension, specially when the temporal lobe is involved.

3) Lesions localization is related to type of aphasia, fluency, and comprehension, in the same way that one would expect it in adults.

There are still some unknows at this level which need to be resolved. They concern the nonfluency of speech and mutism. A large, possibly multicentric, study including more homogeneous cases night possibly answer some of these questions.

Also, the inclusion of negative cases could be important in clarifying this issue of functional localization in children. If we find children with lesions in language areas, but without language disturbances, then we have to assume that these areas may not be crucial after all.

The study of cases with techniques of metabolic assessment such as SPECT or PET, will certainly improve our understanding about dysfunctional areas

However, this classical approach (studying the relation of specific syndromes to their lesion site), will not lead us much further in the understanding of acquired aphasia. A neurolinguistic approach is becoming crucial if we mean to characterize exactly how these children do speak. This sort of approach will eventually allow us to analyse the critical differences between child and adult aphasia.

REFERENCES

Basser, L. S. Hemiplegia of early onset and faculty of speech with special reference to the effects of hemispherectomy. Brain 85, 427-460. 1962.

Bernhard, T.M., Uber die spastiche cerebral paralyse im Kindesalter, Nebst einem Excurse ubes: "Aphasie bei Kindern". Archiv fur Pathologische Anatomie und Physiologie und fur Klinische Medicin 102, 26-80, 1885.

Castro Caldas, A. Diagnostico e Evolução das Afasias de causa vascular (Doctoral Thesis). Faculdade de Medicina de Lisboa, 1979.

Cranberg, L. D., Filley, C. M., Hant, E. J. and Alexander, M. P. Acquired Aphasia in childhood: clinical and CT investigations. Neurology, 37, 1165-1172, 1987.

De Renzi, E. and Vignolo, L. A. - The Token Test: a sensitive test to detect receptive disturbances in aphasics, Brain, 85: 665-678, 1962.

Ferro, J. M. Análise neuropsicológica do insucesso escolar. Actas do I Encontro Nacional de Educação Especial. Sociedade Portuguesa para o Estudo Científico da Dificiência Mental, pp. 154-158, 1979.

Ferro, J.M. Neurologia do comportamento (Doctoral thesis). Faculdade de Medicina de Lisboa, 1986

Ferro, J.M.; Martins, I.P.; Pinto, F.A and Castro-Caldas, A. Aphasia following right striato-insular infarction in a left-handed child: a clinico-radiological study. Developmental Medicine and Child Neurology, 24, 173-182, 1982.

Freud, S.; Infantile cerebral paralysis (L.A. Russin, trans.) Coral Gables: University of Miami Press (Original published, 1897), 1968.

Goodglass, H. and Kaplan, E. The assessment of Aphasia and Realated Disorders. Philadelphia Lea & Fabriger, 1972.

Martins, I.P. and Ferro, J.M.: Acquired conductions aphasia in a child. Developmental Medicine and Child. Neurology, 29, 532-536, 1987.

Martins, I.P.; Ferro, J.M. and Trindade, A. Crossed Aphasia in a Child. Developmental Medicine and Child Neurology, 29: 96-109, 1987.

Reynell, J. K. Manual for the Reynell Developmental Language Scales (Revised). NFER Publishing company Ltd. England, 1977.

Tanabe, H.; Ikeda, M.; Murasawa, A.; Yamada, K. et al. A case of acquired conduction aphasia in a child. Acta Neurol. Scand. 80: 314-318, 1989.

Van Dongen, H. R. Clinical Aspects of Acquired Aphasia and Dysarthria in Childhood. Ebunom Publisher, Delft, 1988.

Van Hout, A., Evrard, Ph. and Lyon, G. On the positive semiology of Acquired Aphasia in Children. Develop. Med. and Child Neurol., 27, 231-241. 1985.

Van Hout, A., Characteristics of language in Acquired Aphasia in children. Paper presented at NATO Advanced Research Workshop on Acquired Aphasia in children. Sintra, Portugal, Sep. 13-15, 1990.

Woods, B. T. and Teuber, H. L. changing patterns of Childood Aphasia. Ann. Neurol. 3, 273-280, 1978.

Zimmerman, R. A., Bilaniuk, L. T., Packer, R. J., Goldberg, H. I. and Grossman, R. I., computed Tomographic–Arteriographic correlates in Acute Basal Ganglionic Infarction in Childhood, Neuroradiology, 24, 241–248, 1983.

VI - PROGNOSIS FOR RECOVERY OF ACQUIRED APHASIA IN CHILDREN

OUTCOME OF ACQUIRED APHASIA IN CHILDHOOD: PROGNOSIS FACTORS

A. VAN HOUT
Department of Neuropediatrics
U.C.L. 10/1303
St. Luc University Clinic
10, Avenue Hippocrate
1200 Brussels
Belgium

ABSTRACT: Among the prognostic factors in acquired aphasia in children, age used to be said to play the major role. However, the literature is confused on the subject because of the small number of cases and the lack of homogeneity for etiologies, age at the lesion, follow-up duration, and even the recovery criteria themselves. A critical review of the literature indicates that younger children do not always have a favourable outcome and that other prognosis factors may intervene. The importance of follow-up duration is stressed since recovery processes can extend over many years and also since linguistic and non-linguistic deficits may develop later.

Introduction

The factors of a prognosis for acquired aphasia in childhood are often confounded because of the small number of cases in series and also because of their lack of homogeneity.

In addition to the constraints imposed by the nature of the lesion itself such as its etiology, extension, and localization, the age at the lesion was said to play a major role in recovery, which was often assumed to be more rapid and complete than in adults. Moreover, while recovery in adults may be defined as a restitution of the prelesional language status, in children, the whole sequence of developing language is abruptly disrupted, so recovery also implies the continuation of this interrupted sequence with the development of new skills at the expected times. In this respect, the prognosis in acquired childhood aphasia has to project into the future and to do this not just for what concerns language.

One of the supposed neural aphasia recovery mechanisms from aphasia in children is a taking over of language by uncommitted brain areas (Goldman, 1974). Those non-functional immature zones are often those initially intended to support later non-linguistic cognitive functions. When these areas are engaged in their suppletive language functions, they may fail to develop the functions for which they were originally intended, and thus secondary disorders also appear for non-language functions. Those late-occurring disorders, either linguistic

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 163–169.

or visuo-spatial may entail learning disorders (Van Hout and Seron, 1983).

We will briefly examine these different clinical aspects of recovery in acquired childhood aphasia.

1. The Effect of Age

The qualification "more rapid" to designate recovery in children relative to that in adults is overused and is usually attributed to Lenneberg in his book, "The Biological Foundations of Language" (1967). However, while Lenneberg does use "better" and "more complete" in reference to child aphasia, his data do not permit one to conclude that recovery is more "rapid" in children than in adults, on the contrary. The comparison of recovery from aphasia in children and in adults was conducted by Lenneberg on the basis of two large series from the literature: the childhood series was that of Basser (1962) to which Lenneberg added a few cases from his own practice, and the adult series was the traumatic series presented by Russel and Espir (quoted by Lenneberg, 1967). Three different recovery rhythms were observed in the adults, with a steady, long lasting one being the less frequent. In children, this mode of evolution dominated the picture, extending over years. This opposition thus contradicts the frequent misquotation of Lenneberg's findings. What Lenneberg did consider as more rapid, was in fact, some aspect of symptom recovery within the childhood series themselves, in favour of very young children. Lenneberg supposed that recovery was achieved sometimes around puberty or even before (he himself does not appear quite clear about this and speaks of the age of eight or ten or of "puberty" without further precision). This end of recovery potential marked the end of the "critical" period for language acquisition.

However, this last notion is contradicted by other reports that describe continuation of the aphasic resolution processes well beyond of puberty. For instance, Woods and Teuber (1978) described a retrospective case of jargonaphasia in a five-year-old boy. When the boy was re-examined in his early teens, he was still aphasic (now with anomia), but, when seen again in his twenties, his language problem had subsided. The extension of this evolution over many years is evocative of some descriptions of hemispherectomies for early lesions. Hemispherectomy can be regarded as an extreme form of unilateral cerebral lesion, and long evolution durations have been described in this condition (Smith and Sugar, 1975). It may be that similar recovery processes could occur in cases of aphasia acquired at an early age, which is why there should be long-term follow up in the evaluation of recovery.

Consequently, a comparison of recovery variables across series is difficult, as most of them have different durations of follow-up, with long-term follow up being rare. Thus, outcome evaluation is difficult to establish, and the moment of recovery for a given function is also difficult to ascertain, as the intervals separating evaluations are uneven. Observed recovery may correspond to the interval between two

consultations rather than to the precise time of the regression of the symptom.

The duration of recovery was said by Lenneberg to vary with age within the childhood series themselves. However, other authors, such as Woods and Teuber (1978) did not find any relation between this duration and age at injury. For the recent series of Van Hout et al. (1985) and of Van Dongen et al. (1989), young children, and at least those with severe lesions, do not appear to recover more quickly than older ones. The long-term follow up a girl, who was four years old at injury (Klein et al., in press), did not show any modification in language behaviour after four years. Thus, the time of recovery in childhood is long, extending in some cases until early adulthood, and its relation with age at injury is not straightforward.

The completeness of recovery was said by Lenneberg to be more frequent in childhood aphasia than in adult aphasia. However, besides the study of Lenneberg and some traumatic series, there are few direct comparisons in the nature of outcome between childhood and adult aphasia. Most studies have focused on the differences of evolution by age within the childhood series themselves. Alajouanine and Lhermitte (1965), for instance, compared aphasic outcome in children before and after their early teens (traumas and vascular causes were equally distributed in the two groups). Although there was an effect of age on the nature of the aphasic symptoms, there was no age influence on the degree of recovery. Neither Byers and MacLean (1962) in their vascular series nor Assal and Campiche (1973) with their traumatic cases found any effect of age on the extent of recovery. Woods and Teuber (1978) found that all the children aged under eight at the time of injury recovered. However, this series probably has longer follow-up durations than the others. Van Dongen et al. (1989) did not report a better prognosis in younger children on the sole criterion of age at injury. Van Hout et al. (1985) differentiate at least two stages of recovery: a regression or attenuation of the acute symptoms and then the more progressive retrocession of residual symptoms, which are mostly hypospontaneity or anomia, the former being the more frequent in younger children.

2. The Effect of the Different Lesional Parameters

For the reasons already stated (lack of homogeneity, too few cases), many contradictions appear.

2.1. ETIOLOGY

As regards etiology, the prognosis for vascular lesions is often more favourable than for adults, probably owing to the plasticity of the focal conditions, such as thrombotic repermeabilisation, or to readier establishment of collateral circulation in the child. The vascular cases described by Byers and MacLean (1962) and by Cranberg et al. (1987) have a better prognosis for oral language recovery than adults with similar lesions. However, for other authors, the prognosis is

severe or more dependent of the nature of the aphasia itself, usually of a motor type, indicative of a favourable prognosis (Van Dongen et al., 1979).

Head trauma has been said to have a favourable outcome, but the extent and nature of lesions vary greatly from case to case in this condition, so it difficult to consider this etiology as homogeneous. Although, for instance, Guttman (1942) reported that recovery was more complete and rapid in children with traumatic aphasia than for other causes, long-lasting disorders have been described in cases with apparent recovery of oral language (Hécaen, 1976). Assal and Campiche (1973) have described 18 children with post-traumatic aphasia. In the majority of them, the aphasia was of short duration, but recovery did not occur during the long-term follow up reported in four cases with more severe aphasic signs. Van Dongen et al. (1989) state that the degree of severity of the injury is the better determinant of outcome even for this supposedly more benign etiology. Van Hout et al. (1985) reported jargonaphasia after head trauma in the youngest case of their series (four years old). Aphasia was still present more than a year after the injury.

Infections may have a bad outcome (Van Dongen, 1989), except for some cases of abscesses. The present-day antiviral treatment of herpes simplex encephalitis, which enables most patients to survive, is probably at the origin of an increase of posterior types of aphasia, for which the ultimate prognosis is guarded (Van Hout and Lyon, 1986).

In opposition to epileptic aphasia, where the symptoms are usually transient and fluctuate with the seizures, in the Landau-Kleffner syndrome (1957), the language disturbance, although fluctuating, is of long duration. An inverse correlation between a young age at injury and a bad outcome is generally found (Toso et al., 1981; Bishop, 1985). Deonna et al. (1977) distinguish three forms of this syndrome, and general recovery occurs in no more than 30% of cases.

2.2. LESIONAL EXTENSION

Lesional extension plays a major role in prognosis, and Van Dongen et al. (1989), comparing effects of severity versus bilaterality of the lesion, found that the latter factor appeared to have the most influence. Van Hout et al. (1985) came to the same conclusion in comparing children with similar etiology (herpes encephalitis) and suggested that, at least for posterior lesions, an heterolateral involvement, even if less extensive and non-symmetrical, may impede the usual recovery mechanism through interhemispheric transfer.

2.3. NATURE OF THE APHASIC SYMPTOMS

At the least, the nature of the aphasic symptoms themselves has a role in recovery in children as in adults. Sensory aphasias recover less than anterior types of aphasia. As regards the evolution of paraphasias, they are more persistent in confrontation naming tasks (Van

Dongen et al., 1985), and verbal paraphasias are more persistent than phonemic paraphasias (Van Hout et al., 1985). In cases of Wernicke's aphasia, and in accordance with findings in adults, recovery is accompanied by a reduction in the percentage of neologisms with an increase of verbal paraphasias and of pauses preceding content words (Van Hout and Lyon, 1986).

3. Residual Symptoms and Learning Disabilities

Scholastic problems as sequelae in acquired childhood aphasia are the rule rather than the exception. There are attentional difficulties, memory problems and subtle comprehension disorders. Subsequent learning disabilities may affect written language as well as calculation. These deficits may be a sequelae of a dyslexia or dyscalculia produced by the lesion itself, or may develop secondarily, even in children who had not acquired these skills at the time of injury as a result of the problems enumerated above (Van Hout and Seron, 1983). Acquisition of new knowledge or application of anteriorly acquired reasoning schemes to new data are difficult.

The detection of memory or of subtle comprehension disorders will depend of the level of analysis used by the observers. This is another reason why most prognostic evaluations conflict in the different series: the recovery criterion some authors use is the ability of the child to engage in free-flow conversation, while, for others, it also includes improvement of metalinguistic skills (Woods and Carey, 1979) or normal scholastic progress. A recent study by Cooper and Flowers (1987) was devoted to the investigation of residual language impairment in a series of 15 children with a history of acquired aphasia. All the aphasic children's performances were systematically worse than those of the controls, both on language tests and in general academic achievement. Unfortunately, there was a wide scatter of variables such as etiology, age at onset and age at testing. As a result, the outcome profiles were heterogeneous, and no prognostic factor could be discerned.

4. Conclusions

To conclude, prognosis in acquired childhood aphasia appears now to be much less favourable than was asserted in the older series. The link with a younger lesional age, once held to be predictive of a better outcome, is not universally recognised. Disorders of oral language, either subtle or more overt, may persist in the long term, such as alteration of non-linguistic functions and learning disabilities.

From a methodological point of view, there is a need to define more precisely the setting of recovery criteria and also to provide long-term follow up, as time is crucial in evaluation, both to disclose new deficits or disruptions in the developmental language sequence and also to observe possible ultimate language recovery.

REFERENCES

Alajouanine, Th. and Lhermitte, F.(1965) 'Acquired aphasia in children', Brain 88, 653-662.

Assal, G. and Campiche, R. (1973) Aphasie et troubles du langage chez l'enfant après contusion cérébrale', Neurochirurgie 19, 399-406.

Basser, L.S. (1962) 'Hemiplegia of early onset and the faculty of speech with special reference to the effects of the hemispherectomy', Brain 85, 427-460.

Bishop, D.V. (1985) 'Age of onset and outcome in acquired aphasia with convulsive disorders', Developmental Medicine and Child Neurology 27, 705-712.

Brown, R. and Jaffe, J. (1975) 'Hypothesis on cerebral dominance', Neuropsychologia 13, 107-110.

Byers, R. and Mc Lean, W. (1962) 'Etiology and course of certain hemiplegias with aphasia in childhood', Pediatrics 29, 376-383.

Cooper, J.A. and Flowers, C.R. (1987) 'Children with a history of acquired aphasia: residual language and academic impairment', Journal of Speech and Hearing Disorders 52, 251-262.

Cranberg, L.D., Filley, C.M., Hart, E.J., and Alexander M.P. (1987) 'Acquired aphasia in childhood, clinical and CT investigations', Neurology 37, 1165-1172.

Deonna, T., Beaumanoir, A., Gaillard, F., and Assal, G. (1977) 'Acquired aphasia in childhood with seizure disorders: a heterogeneous syndrome', Neuropediatrie 8, 263-273.

Goldman, P.S. (1974) 'Plasticity of function in the CNS' in D. Stein, J. Rosen, and N. Butters, (eds.), Plasticity and Recovery of Function in the Central Nervous System, Academic Press, New York.

Guttmann, E. (1942) 'Aphasia in Children', Brain 65, 205-219.

Hécaen, H. (1976) 'Acquired aphasia in Children and the Ontogenesis of Hemispheric Functional Specialization', Brain and Language 3, 114-134.

Kinsbourne, M. and Hiscock, M. (1977) 'Does cerebral dominance develop?' in S. Segalowitz and F.A. Gruber (eds.) Language Development and Neurological Theory, Academic Press, New York.

Klein, S.K., Masur, D., Farber, K., Shinnar, S., and Rapin, I. 'Persistent fluent aphasia in a young child' (in press).

Landau, W.M. and Kleffner, F.R. (1957) 'Syndrome of acquired aphasia with convulsive disorder in children', Neurology 7, 523-530.

Lenneberg, E.H. (1967) Biological Foundations of Language, Wiley, New York.

Smith, A. and Sugar, O. (1975) 'Development of above normal language and intelligence: 21 years after left hemispherectomy', Neurology 25, 813-818.

Toso, V., Moschini, M., Gagnin, G., and Antonu, D. (1981) 'Aphasie acquise de l'enfant avec épilepsie; trois observations et revue de la littérature', Revue neurologique 137, 425-434.

Van Dongen, H.R., Loonen, M., and Van Dongen, K. (1985) 'Anatomical basis for acquired fluent aphasia in children', Annals of Neurology 17, 306-309.

Van Dongen, H.R. (1989) 'Acquired childhood aphasia, course and outcome' paper presented at the International Neuropsychological Society Meeting, Antwerp.

Van Hout, A. and Seron, X. (1983) 'L'aphasie de l'enfant', Mardaga, Brussels.

Van Hout, A., Evrard, Ph., and Lyon, G. (1985) 'On the positive semiology of acquired aphasia in children', Developmental Medicine and Child Neurology 27, 231-241.

Van Hout, A. and Lyon, G. (1986) 'Wernicke's aphasia in a ten-year-old boy', Brain and Language 29, 268-285.

Van Hout, A. and Evrard, Ph. (1987) 'Aphasie sensorielle chez un enfant de moins de quatre ans' in J. Narbona and M. Poch-Olive (eds.) 'Neuropsychologie infantile: données récentes', Symposium international de Pampelune 81, 294.

Woods, M.T. and Teuber, H.L. (1978) 'Changing pattern of childhood aphasia', Annals of Neurology 3, 273-280.

Woods, M.T. and Carey, S. (1979) 'Language deficits after apparent clinical recovery from childhood aphasia', Annals of Neurology 6, 405-409.

RECOVERY FROM APHASIA AND LESION SIZE IN THE TEMPORAL LOBE

Isabel Pavão Martins MD and José M. Ferro MD
Centro de Estudos Egas Moniz
Hospital de Santa Maria,
1600 Lisboa,
PORTUGAL

ABSTRACT. We studied the factors influencing the prognosis of acquired aphasia in a series of 29 children with focal hemispheric lesions sustained after language acquisition. Twenty two children (76%) recovered completely from aphasia. Vascular and traumatic cases recovered better than those with encephalitis. Although many older children also recovered fully, all that did not recover were older than 7. In what concerns lesion size and site, determined by CT scan in 19 cases, the involvement of the postrolandic cortex and the extension of Wernicke's area damage were associated with a poor outcome. This suggests that recovery from acquired aphasia is more dependent upon the intact areas the left hemisphere than upon language shift to the nondominant hemisphere.

INTRODUCTION

It has long been recognized that the outcome of acquired aphasia is better in children than in adults (Clarus 1874; Lenneberg 1967; Hécaen 1976). Although its clinical features are very much the same in both cases (Van Hout et al 1985; Van Dongen et al 1985; Cranberg et al 1987), in the case of children, recovery is faster and more complete than in adults (Alajouanine and Lhermitte 1965; Cranberg et al 1987; Hécaen 1983; Satz and Bullard-Bates 1981).

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 171–184.

However, there are few studies systematically evaluating the role of the different variables involved in the recovery (Guttman 1942; Van Dongen 1988).

In an attempt to isolate factors underlying the good prognosis of childhood aphasia , we studied a group of children with acquired aphasia taking into consideration the individual effects of a number of variables.

POPULATION

Of all 54 children with acquired hemispheric lesions evaluated in our laboratory, we selected those who fullfiled the following criteria: 1) Children with acquired aphasia due to focal, unilateral, nonprogressive hemispheric lesion; 2) lesion was sustained after some normal language development after 1.8 months of age and before age 15; 3) follow-up time of 6 months or more.

We excluded children with bilateral lesions (shown by neuroimaging tecniques), those with tumors, cases with the Landau and Kleffner syndrome and children with a previous developmental delay.

Our population consists of 29 children, 13 females and 16 males, with a mean age of onset of 8 years (ranging from 18 months to 15 years), who have been followed for a mean of 5 years. All were right-handed but one.

Twenty-eight children had left hemispheric lesions, and one (a left-handed girl) a right hemispheric lesion.

The etiology, was traumatic in 12 cases, infectious (encephalitis) in 4, and vascular in 13 (10 ischaemic, 3 haemorhagic).

Lesion site was determined by CT or MRI scan in 19 cases, and by clinical evaluation, angiography or surgery only, in 10. CT Scans were analysed according to standard brain maps (Hayward et al, 1977) for the presence or absence of lesion in reference areas.

METHOD

Children were assessed with a language battery consisting of:

1. Rating for severity of aphasia, with the rating scale of the Boston Diagnosic Aphasia Examination (Goodglass and Kaplan 1972).

2. Analysis of spontaneous speech - including fluency, syntax, prosody, anomia, paraphasias and dysarthria (the type of errors was not quantified).

3. Language assessment: Children younger than 6 yrs were evaluated with the Reynell Developmental Language Scales (Reynell 1977) and a repetition test; children older than 6 were assessed with the Lisbon aphasia battery (Ferro 1986) (with tests of naming, repetition, oral comprehension), a short version of the Token test, (De Renzi and Vignolo 1962) and reading and writing tests.

Diagnosis of the type of aphasia was determined as a function of the score obtained in the 4 cardinal language tests (fluency object naming, sentences comprension, and word repetition) (Castro-Caldas 1979).

Children were considered to be fully recovered when their spontaneous speech was normal and they had normal scores for age in the Reynell scales or a normal performance in the 3 cardinal tests of the aphasia battery and a normal score for age in the Token test (Ferro 1979).

RESULTS:

The overall outcome was quite good, which is consistent with the findings of other authors (Satz and Bullard-Bates 1981). 76% of children recovered completely from aphasia, and most of them did so within the 1st year of follow up. All 7 children who did not recover had more than 1 year of follow-up, which excludes the explanation of their lack of recovery in terms of a short follow up time (table 1).

Table 1 - RECOVERY

N	%	RECOVERY	RECOVERY TIME
22	76%	full	16/22-1st year
			6/22-> 1 year
7	24%	nil/partial	7-> 1 year

We performed an individual analysis of the different factors which could be related to the prognosis: etiology, type of aphasia, age of onset, and lesion site and size.

1. In what concerns etiology, our cases showed that vascular and traumatic cases have a better prognosis than the cases of encephalitis. All the children with stroke and 67% (8/12) of children with trauma recovered fully, compared to only 1 out of 4 cases of encephalitis.

Also, in the vascular cases, the duration of aphasia was shorter, lost (11/13) children recovered within the first 6 months (Table 2). It also noteworthy that, in children, recovery might continue in the chronic period (i.e. after 6 months) which is quite unusual in adults.

Table 2 - ETIOLOGY AND DURATION OF APHASIA

DURATION OF APHASIA	TRAUMA	ENCEPHALITIS	VASCULAR
< 1 week			2
< 1 month			2
2-3 months	1		5
4-6 months		1	2
6-12 months	3		
> 1 year	4+4*	3*	2
% full recoveries	67%	25%	100%
follow up (years)	6.9+3.3	3.2+1.9	5.1+4.0

* persisting aphasia

2. Concerning the effect of age of onset in the prognosis, we found that there were children of all ages in the recovered group. However, all those who did not recover were older than 7 years of age at the time of the brain lesion (Table 3). This might show some influence of age but,

on the other hand, those older children, who did not recover, were also in the traumatic and encephalitis groups, which tend to harbour a worse outcome.

Table 3 – AGE OF ONSET AND PROGNOSIS

AGE AT ONSET	RECOVERED			NOT RECOVERED		
	T	E	V	T	E	V
< 2	1*		1			
3						
4						
5	1		1			
6	1*		3			
7	2*			1		
8	1	1			1	
9	1					
10	1		1			
11			2			
12	1					
13			1*	1	1	
14					1	
15			1	2		

* aphasic > 1 year; T= traumatic; E= Encephalitis; V= vascular

3. Regarding the influence of the type of aphasia in the prognosis, we found that nonfluent aphasias, did better (17 out of 19 recovered) than fluent aphasias (where only 5 out of 10 recovered) (Table 4), which suggests that the lesion site (either pre or post rolandic) is important in the recovery.

Table 4 – TYPE OF APHASIA AND PROGNOSIS

TYPE OF APHASIA	RECOVERED	NOT RECOVERED
NONFLUENT	17	2 (11%)
BROCA'S	4	1
GLOBAL	3	1
TRANSCORTICAL	2	–
NOT CLASSIFED	8	–
FLUENT	5	5 (50%)
ANOMIC	4	–
CONDUCTION	–	1
WERNICKE'S	1	2
TRANSCORTICAL	–	2

4. To evaluate the role of lesion site, we conducted a separate study including only 19 patients whose lesion had been determined by CT or MRI scan (18 children had left hemispheric lesions and 1, a left handed girl, a right hemispheric lesion).

Indeed (Table 5), children with postrolandic lesions, either isolated or associated with a subcortical or a prerolandic involvement, had a worse recovery when compared with those with prerolandic or subcortical lesions only. However, vascular lesions affected mainly (7/9 cases) subcortical structures and infectious cases mainly (4/4) the cortical temporal areas. Therefore lesion site and etiology are so closely related that it was difficult to study their effect independently.

Table 5 - LESION SITE AND PROGNOSIS

LESION SITE	RECOVERY	
	YES	NO
PREROLANDIC	3	-
SUBCORTICAL	3	-
POSTROLANDIC	5	3
SUBCORTICAL + POSTROLANDIC	2	-
PRE + POSTROLANDIC	1	2

5. We subsequently determined, in those 13 case with post rolandic involvenent, which of the posterior areas were more important (Table 6). There was a significant difference in only 2 areas: the temporal isthmus and the Helsch gyrus. The involvement of these areas was associated with a worst outcome. This may show that an intact input to the Wernicke's area is a crucial factor for recovery.

Table 6 - RECOVERY AFTER POST ROLANDIC LESIONS

LESION SITE	RECOVERY	
	YES n=8	NO n=5
Wernicke's area	4	5
Temporal isthmus *	-	4
Helsch Gyrus **	1	5
Parietal Lobe	7	4
Occipital lobe	3	3
Basal Ganglia	2	-
Pre-rolandic	1	2

* $p < .005$ ** $p < .025$

In the same group of patients, we studied the relation between recovery and the extention of the lesion in Wernicke's area. This was determined in 2 CT slices (Hayward et al 1977), the B/W slice (where one can see both Broca's and Wernicke's area) and the W slice (where one sees most of Wernicke's area). The lesion size in each slice was graded into 5 points:

0. if there was no lesion of Wernicke's area in that slice; 1. if the lesion occupied less than 25% of the Wernicke's area, in that slice; 2. lesion occupied between 25–75%; 3. more than 75%; 4.If all the area was damaged in that slice.

Table 7 shows that the larger the extent of damage in Wernicke's area, the less likely recovery becomes.

Table 7 – RECOVERY AFTER LESIONS OF WERNICKE'S AREA

PATIENTS		% Wernicke's area damage				
	CT slice	0	< 25	25–75	> 75	100
RECOVERED n=8	B/W	5	3	–	–	–
	W	4	3	1	–	–
NOT RECOVERED n=5	B/W	–	–	–	1	4
	W	1	–	1	2	1

Therefore both Wernicke's area and its input need to be intact for language recovery to take a place.

DISCUSSION

One of the crucial aspects of acquired aphasia in children is its prognosis for the entire concept of brain plasticity is implicit to it.

Although some recent studies have found variable degrees of language impairement in sophisticated language tests in children who suffered acquired aphasia (Woods and Carey 1979; Varga Khadem et al 1985), the truth is that those children were no longer clinically aphasic.

Indeed one of the most widely accepted points concerning acquired aphasia in children is that it has a good immediate prognosis which has been attributed to the greater plasticy of the brain in the young child.

Our study shows that we can not simply explain the good prognosis in terms of a greater plasticity, for there are many factors influencing the outcome.

One of the most important factors is undoubtely the etiology. In our study children with stroke had a very good recovery compared to those with trauma and especially the infectious cases. This is in contradiction with other authors (Guttman 1988; Van Dongen 1988) whose traumatic cases did better than vascular ones. On the other hand most authors agree that infectious cases have a poor recovery.

The partial disagreement, concerning the vascular cases, may be related to the effect of lesions localization. It is well known that stroke in children affects mainly the subcortical areas (Zimmerman et al 1973). Indeed in our vascular cases 7/9 had subcortical lesions sites, while in Van Dongen series (Van Dongen 1988) 3 cases (cases 9, 11, 14) out of his 6 vascular cases had cortical lesions, and those children recovered less than those with subcortical damage. Therefore we may have here both an influence of etiology and lesions site.

On the other hand traumatic lesions are not good models to study recovery. Trauma can produce, further to focal lesions, a diffuse encephalopathy through increased intracranial pressure, hydrocephalus or associated vascular lesions. Both the size and localization of lesions is extremely variable, not following a predictable pattern like stroke. Besides, the area of dysfunction may be much larger than that shown in CT scan. Comparison of series of the trauma is therefore difficult unless on a case-to-case basis, and it is not surprising that different series might have different results.

The same difficulties apply to the analysis of the infectious cases. In our series the prognosis was poor. The same was found in other series (Van Hout et al 1985; Van Dongen 1988). Although in our cases the

imaging techniques showed a unilateral lesion, it is well known that herpes may cause bilateral lesions. Therefore the worse prognosis here could be due to the bilateral involvement or to a more extensive lesion than the one displayed on CT scan.

These first results, considering all the above mentioned limitations, just show that etiology influences the prognosis, and this factor cannot be neglected when discussing the outcome of acquired brain lesions in children.

In what concerns the effect of age of onset (and this is possibly the most crucial point of recovery discussion), we found that all those who did not recover were older than 7 years at the time of the brain lesion. This could be just a case of etiological bias, since those 7 children were also in the etiological groups harbouring the worst outcome. However, there were also younger children in the traumatic group who, although being aphasic for more than 1 year, eventually recovered fully. In the large series reported by Woods and Teuber, (Woods and Teuber 1978) including predominantly vascular cases but having no lesions localization, only the 4 older children (older than 8) did not recover, and there was an inverse relationship between age of onset and full recovery (Woods and Carey 1979). However other authors could not confirm these findings (Van Dongen 1988).

This point raises the question of "plasticity", which has been overenphasized since the first reports of acquired aphasia in children. Lenneberg, in 1967 postulated that up to a certain critical age (estimated from 9 to 11 yrs) the right hemisphere would take language function, for the cerebral hemispheres were equipotencial in the young child. However subsequent studies have discarded this view, by showing that language lateralization to the left hemisphere occurs early in life probably since birth, and therefore is biologically determined. All sorts of evidence corroborated this last theory (the invariance theory): anatomical studies of human fetal brains (Witelson 1973), electrophysiolgical studies (Molfese et al 1975), dichotic listening studies , and studies of acquired aphasia (Satz and Bullard-Bates 1981, Woods and Teuber 1978). However it is also known, from the studies of left hemispherectomy (Dennis and Whitaker 1976), and studies with the Wada test in epileptics, (Satz et al 1988) that language can be acquired with the right hemisphere.

Therefore "plasticity" must now be explained in another way: 1) the right hemisphere can take language function if the left is removed or damaged at an early age; 2) the undamaged areas of the left hemisphere can be reorganized to take over the lost function. Whatever the

case there must be some influence of age otherwise recovery would be identical to adults. Possibly the interaction between the specific lesion site or size and the stage of functional maturation of the brain at the time of injury determines not only the strategy but also the degree of recovery, these being responsible for the final outcome. It is also interesting to note that recovery in children continues for a long period (years) which is quite unusual in adults, showing that funtional/strutural reorganization is a ongoing process.

Concerning the influence of type of aphasia in the prognosis we found that non fluent aphasics recovered better than fluent aphasics. This is particularly important if we consider that in old series, one of the main reasons for including patients was the presence of a right hemiparesis, hence introducing a bias favoring prerolandic and subcortical lesion sites. Most of those series actually reported a predominance of non fluent speech, good verbal comprehension and good prognosis all consistent with those localizations.

If we look at more recent series we will find that fluent cases or those with impaired comprehension did less well (Woods and Teuber 1978, Van Hout et al 1985)

When we considered localization, we corroborated these results: there was a predominance of posterior (post-rolandic) lesions in patients with no recovery. Of course here, lesion site and etiology are closely related for vascular lesions affected mainly subcortical structures and infectious cases the temporal cortical areas.

We could confirm this findings when reviewing the literature: in all 3 published series of Acquired Aphasia where scans are either described or printed (Van Hout et al 1985; Cranberg et al 1987; Van Dongen 1988), there was a predominance of posterior lesions in those with a poor outcome.

In addition, in our study, only 2 postrolandic areas were associated with a different prognosis: the temporal isthmus and the Helsch gyrus. Besides there was a correlation between the extention of lesion in Wernicke's area and the outcome, suggesting that both Wernicke's area and its input need to be intact for language recovery to take place. This can be explained in different ways: 1) either these areas have an unique functional/structural organization to take language function that cannot be overcome by neighbouring areas nor transfered to the nondominant hemisphere even in the case of children, or 2) their lesion affects recovery by interfering with phonologic decoding and therefore the ability to provide verbal input to the rest of the brain.

In conclusion: 1) the immediate recovery of acquired aphasia in children depends on a number of factors in particular lesions etiology, size and site; 2) the auditory association areas and Wernicke's area are important in recovery or new acquisition of language; 3) the influence of age or cerebral "plasticity" can only be understood if all other variables are strictly controlled.

Some of the problems raised, could be overcome by a multicentric study including only patients with unilateral ischaemic lesions, demonstrated by imaging techinques and with acute as well as chronic standardized neuropsychological assessment.

Besides, the inclusion of cases studied with metabolic techniques (Pet/Spect) could clarify the problem of recovery by showing which recovery strategy is prefered following lesions at particular ages or brain sites.

REFERENCES

Alajouanine, TH. and Lhermitte, F. Acquired Aphasia in Children. Brain, 88, 653-662, 1965.

Castro Caldas, A. Diagnostico e Evolução das Afasias de causa vascular (Doctoral Thesis). Faculdade de Medicina de Lisboa, 1979.

Clarus, A. Uber Aphasie bei Kindern. Jahresb Kinderheilkd, 7, 369-400, 1874.

Cranberg, L. D., Filley, C. M., Hant, E. J. and Alexander, M. P. Acquired Aphasia in childhood: clinical and CT investigations. Neurology, 37, 1165-1172, 1987.

Dennis, M. and Whitaker, H. A. Language acquisition following hemidecortication: Linguistic superiority of the left over the right hemisphere. Brain and Language, 3, 404-433, 1976.

De Renzi, E. and Vignolo, L. A. The Token Test: a sensitive test to detect receptive disturbances in aphasics. Brain, 85, 665-678, 1962.

Ferro, J. M. Análise neuropsicológica do insucesso escolar. Actas do I Encontro Nacional de Educação Especial. Sociedade Portuguesa para o Estudo Científico da Deficiência Mental, 154-158, 1979.

Ferro, J. M. Neurologia do Comportamento (Doctoral Thesis) Faculdade de Medicina de Lisboa, 1986.

Goodglass, H. and Kaplan, E. The assessment of Aphasia and Related Disorders. Philadelphia Lea & Fabiger, 1972.

Guttman, E. Aphasia in Children. Brain, 65, 205-219, 1942.

Hayward RN, Naeser MA, and Zatz, LM Cranial computed tomography in aphasia. Correlation of anatomical lesions with functional deficits Radiology, 123, 653-660, 1977.

Hécaen, H. Acquired Aphasia in Children and the Ontogenesis of Hemispheric Functional Specialization. Brain Lang. 3, 114-134, 1976.

Hécaen, H. Acquired Aphasia in Children: Revisited. Neuropsychologia. 21, 581-587, 1983.

Lenneberg, E. Biological Foundations of Language. New York, John Wiley, 1967.

Molfese, D. L., Freeman, R. B. and Palermo D. S. The ontology of brain lateralization for speech and nonspeech stimuli. Brain and Language, 2, 356-368, 1975.

Reynell, J. K. Manual for the Reynell Developmental Language Scales (Revised). NFER Publishing company Ltd. England, 1977.

Satz, P. and Bullard-Bates, C. Acquired Aphasia in Children. In Sarno, MT (Ed) Acquired Aphasia. New York, Academic Press pp. 399-426, 1981.

Satz, P., Strauss, E., Wada, J. and Orsini, D. Some correlates of intra and interhemispheric speech organization after left focal brain injury. Neuropsychologia, 26, 345-350, 1988.

Van Dongen, H. R., Loonen M. C. and Van Dongen K. J. Anatomical basis for acquired fluent aphasia in children. Ann. Neurol., 17, 306–309, 1985.

Van Dongen, H. R. Clinical Aspects of Acquired Aphasia and Dysarthria in Childhood. Ebunom Publisher, Delft, 1988.

Van Hout, A., Evrard, Ph. and Lyon, G. On the positive semiology of Acquired Aphasia in Children. Develop. Med. and Child Neurol., 27, 231–241. 1985.

Vargha–Khadem, F., Gorman, A. M. and Watters, G. V. Aphasia and Handedness in Relation to Hemispheric side, age at injury and severity of cerebral lesion during childhood. Brain, 108, 677–696, 1985.

Witelson, S. F. and Pallie, W. Left hemisphere specialization for language in the newborn: Neuroanatomical evidence of asymmetry. Brain, 96, 641–646, 1973.

Woods, B. T. and Carey, S. Language Deficits after apparent Clinical Recovery from Childhood Aphasia. Ann. Neurol. 6, 405–409, 1979.

Woods, B. T. and Teuber, H. L. Changing patterns of Childhood Aphasia. Ann. Neurol. 3, 273–280, 1978.

Zimmerman, R. A., Bilaniuk, L. T., Packer, R. J., Goldberg, H. I. and Grossman, R. I., Computed Tomographic–Arteriographic correlates in Acute Basal Ganglionic Infarction in Childhood Neuroradiology, 24, 241–248, 1983.

Krashen, S. D. Lateralization, language learning and the critical period: some new evidence Language Learning. 23, 63–74, 1973.

ACQUIRED CHILDHOOD APHASIA : OUTCOME ONE YEAR AFTER ONSET

M.Christa B. Loonen and Hugo R. van Dongen
The Department of Neurology
University Hospital Rotterdam-Dijkzigt
The Netherlands.

ABSTRACT. The effects of the variables age at onset, cause, severity and bilaterality of lesion, and type of aphasia on course and outcome were investigated in a group of 28 aphasic children. Analysis of spontaneous speech and tests of auditory verbal comprehension were used to determine the presence of aphasia. The severity of the cerebral lesion was assessed using a rating scale for CT-scans. Most of the children had not recovered completely one year after-onset. Recovery was significantly different according to etiological categories. Complete recovery was seen in the majority of traumatic cases.

Introduction.

The standard doctrine about childhood aphasia claims that recovery of language functions is rapid and complete (1). However, this claim rests largely on quotations from the earlier literature (2,3) and is increasingly contradicted by more recent reports (4,5,6,7,8).

The main factors considered to be associated with language recovery are : age at onset, cause, severity and bilaterality of lesion, and type of aphasia.

This paper has been published in "Archives of Neurology",vol 47, 1324-1328,1990 (Copyright 1990,American Medical Association). Permission for its reproduction here was kindly granted by the American Medical Association.

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 185–200.

AGE AT ONSET.

Alajouanine and Lhermitte (9) found no difference in "the speed of recovery in children less than 10 years old versus children of 10 or more years old". According to Lenneberg (3) the prognosis of acquired aphasia in children is directly related to the age at onset of the aphasia. Aphasias that develop before puberty would clear up completely. In the series of Woods and Teuber (10) all children who became aphasic before the age of eight years regained speech but recovery time ranged from one month to more than two years. In the syndrome of acquired aphasia with convulsive disorder (Landau-Kleffner syndrome = LKS) an opposite relationship is found, ie, the older the child at onset the better the outcome (11,12,13).

CAUSE.

Children with a head trauma have been reported to improve more than those with vascular disease (6,14). However, Byers and McLean (15) reported a complete restitution of speech function in 10 aphasic children with persistent hemiplegia due to a cerebrovascular lesion at follow-up from 1 - 4 years. Longitudinal follow-up studies present evidence that the prognosis in children with the syndrome of acquired aphasia with convulsive disorder is poor (16,17,18).

SEVERITY AND BILATERALITY OF THE LESION.

Persistent aphasic symptoms have been linked with the severity of the lesion (14,19,20). Extensive CT-scan data are limited to the first reports of children with a subcortical aphasia (21,22), a fluent aphasia (23), a crossed aphasia (24), and a conduction aphasia (25). Studying the incidence of paraphasias, VanHout et al. (7) mention CT scan abnormalities, but do not assess the severity of the lesion in 11 children who demonstrated an aphasia arising from a range of causes. However, in children with acquired aphasia with convulsive disorder, normal CT-scans are consistently reported despite severe aphasic disturbances (for refs. see 26).

It is claimed that if in childhood the left hemisphere is damaged, language can develop in the corresponding area of the right hemisphere. Consequently bilateral cerebral damage is considered a bad prognostic sign (20,27,28).

TYPE OF APHASIA.

Guttmann (14) emphasized the good prognosis of purely motor (nonfluent) aphasia in young children: combined motor and sensory aphasia had a more serious prognosis. Assal and Campiche (28) confirmed, but Collignon et al. (27) contradicted this finding. Fluent aphasia in children has been considered rare: no firm statements concerning prognosis are available (23).

The aim of this study was to investigate the association of age at onset, cause of the lesion, severity and bilaterality of the lesion, and type of aphasia with recovery of acquired childhood aphasia.

Assuming a rapid initial recovery, language functions of the children were assessed at one year after onset.

Subjects and Methods.

SUBJECTS.

In the period 1977 through 1985 we studied 30 children with acquired aphasia. The children were referred to the neurology department of the University Hospital Rotterdam-Dijkzigt, the Netherlands. CT scan data were available. With the exception of two children, one or more follow-up examinations were carried out in all cases.

Table 1 shows the subjects characteristics.

Table 1.

AGE AT ONSET, CAUSE OF CEREBRAL LESIONS AND CLINICAL FINDINGS, CT-SCAN DATA AND TOKENTEST/ TMT** SCORES.

case no/ sex	age at onset*	etiological factors	severity of cerebral lesion	Tokentest/ TMT score at onset
HEAD INJURY				
1 M	9.1	contusion; unconscious for 15 min.	N	17
2 F	11.1	contusion; unconscious for five min. at most	NS	13
3 M	3.9	contusion; unconscious for one month; left temporal depressed fracture; persisting right hemiparesis	S	2**
4 M	9.3	contusion; unconscious for five days; transitory right hemiparesis	N	22
5 F	10.5	left temporal depressed fracture and local contusion	S	28

Table 1.

AGE AT ONSET, CAUSE OF CEREBRAL LESIONS AND CLINICAL FINDINGS, CT-SCAN DATA AND TOKENTEST/ TMT** SCORES.

case no/ sex	age at onset*	etiological factors	severity of cerebral lesion	Tokentest/ TMT score at onset
14 F	6.7	persisting right hemiparesis secondary to left middle cerebral artery infarction of unknown cause	S R+	8
15 M	3.5	transitory right hemiparesis following cerebral infarction in internal capsule and caudate nucleus secondary to "sick sinus" syndrome	S	18**
INFECTIOUS DISEASE				
16 M	4.10	persisting slight right hemiparesis following Haemophilus influenzae meningo-encephalitis; residual epilepsy	S R+	0**
17 M	12.7	left frontal and temporobasal subdural empyema; residual epilepsy	S R+	21
18 M	11.2	transitory right hemiparesis secondary to herpes simplex encephalitis	S	2
19 M	9.8	left fronto-temporal subdural empyema secondary to frontal and maxillary sinusitis	S R+	2
20 M	11.4	severe conduct disorder and residual epilepsy following acute demyelinating encephalomyelitis	NS R+	0

Table 1.

AGE AT ONSET, CAUSE OF CEREBRAL LESIONS AND CLINICAL FINDINGS, CT-SCAN DATA AND TOKENTEST/ TMT** SCORES.

case no/ sex	age at onset*	etiological factors	severity of cerebral lesion	Tokentest/ TMT score at onset
6 M	7.3	multiple skull fractures; left parietal depressed fracture; transitory slight right hemiparesis	S	39
7 M	7.6	left temporal contusion; unconscious for some days	S	13
8 M	7.2	contusion; unconscious for one month	S	0**
VASCULAR DISEASE				
9 F	9.2	left parieto-occipital intracerebral hematoma; persisting right hemi-paresis secondary to aplastic anemia	S R+	6
10 F	7.0	atonic attacks without disturbed consciousness from the age of four yrs. At 7 yrs. loss of speech and choreiform movements secondary to "Moya-Moya" disease	NS R+	26
11 M	13.4	persisting right hemi-paresis secondary to left internal carotid artery occlusion of unknown cause	S R+	39
12 F	8.8	transitory slight right hemiparesis secondary to left frontal sub-cortical hematoma	NS R+	28
13 M	3.5	transitory right hemi-paresis following embolism in left middle cerebral artery	S	0**

Table 1.

AGE AT ONSET, CAUSE OF CEREBRAL LESIONS AND CLINICAL FINDINGS, CT-SCAN DATA AND TOKENTEST/ TMT** SCORES.

case no/ sex	age at onset*	etiological factors	severity of cerebral lesion	Tokentest/ TMT score at onset
LANDAU-KLEFFNER SYNDROME				
21 F	6.8	Landau-Kleffner syndrome	N	5
22 F	4.0	Landau-Kleffner syndrome	N	0**
23 M	6.0	Landau-Kleffner syndrome	N	2
24 M	4.8	Landau-Kleffner syndrome	N	0**
25 F	5.11	Landau-Kleffner syndrome	N	7
26 F	4.7	Landau-Kleffner syndrome	N	4**
CEREBRAL TUMOR				
27 M	13.0	left temporal oligo-dendroglioma	S	40
28 F	13.6	left temporal grade I astrocytoma	S	41
29 F	9.5	large neuroblastoma in left hemisphere	S	38
30 F	13.6	left fronto-temporal metastasis of adrenal carcinoma	S	40

TMT indicates Tridimensional Matrix Test;
R+ = including enlargement of right ventricle and/or sulci of the right hemisphere.
N = normal,
NS = non-severe,
S = severe (according to Vargha- Khadem et al., see ref. 35).

* Age is given in years and months
** TMT score

CT SCANS.

With a few exceptions, the CT scans were performed with scanners of the second or third generation. The scanning was carried out according to standard techniques. Using a rating scale (Table 2) the severity of the lesion was rated by a child neurologist (MCBL) who was unaware of the results of the neuropsychological examinations. The selected CT scans were performed at least 4 weeks after onset of aphasia. From each patient with CT scan abnormalities one representative picture is shown in Figures 1 through 4. The numbers correspond to the case numbers in Table 1.

Table 2

Rating scale for CT-scans.

Moderate to marked ventricular dilatation and gross loss of substance seen on three or more cuts.

Moderate to marked ventricular dilatation with minimal to moderate loss of brain substance seen on less than three cuts.

Moderate cerebral atrophy indicated by moderate to marked ventricular dilatation seen on three or more cuts.

Minimal hemisphere atrophy indicated by minimal ventricular dilatation; asymmetry seen on less than three cuts.

Normal.

Data are adapted from Vargha-Khadem et al. (35).

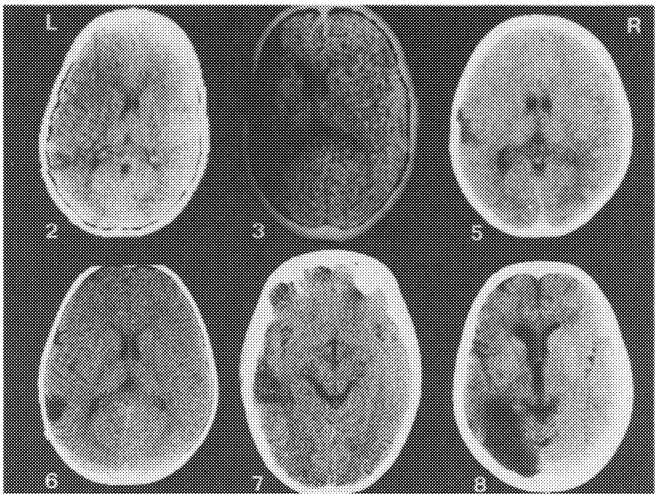


Figure 1. CT-scans of six patients with head injury. The numbers correspond to the case numbers in Table 1.

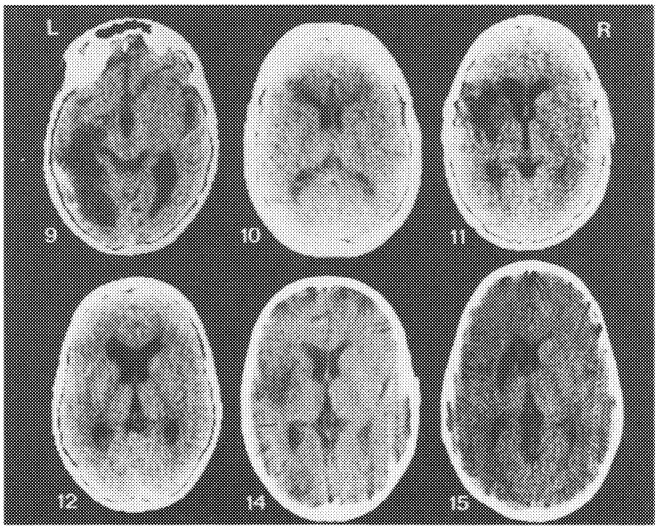


Figure 2. Ct-scans of six patients with vascular lesions. The numbers correspond to the case numbers in Table 1.

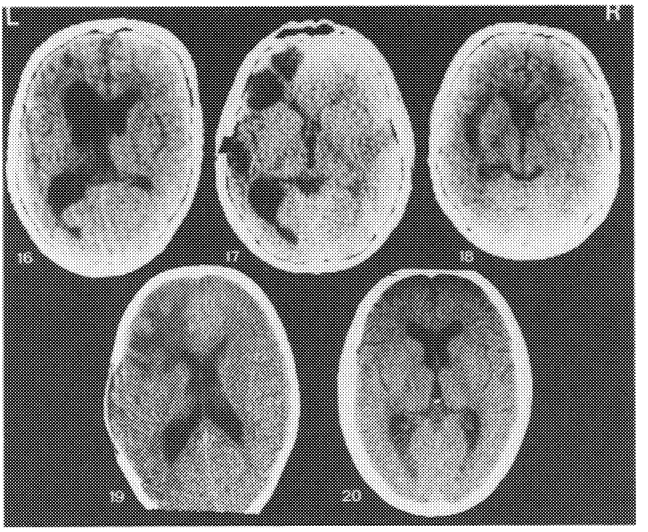


Figure 3. CT-scans of five patients with infectious disease. The numbers correspond to the case numbers in Table 1.

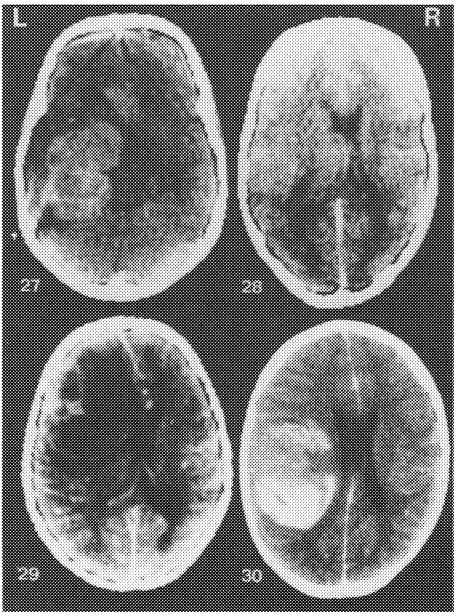


Figure 4. Ct-scans of four patients with a cerebral tumor. The numbers correspond to the case numbers in Table 1.

LANGUAGE TESTING.

The initial language testing was performed as soon as possible after the onset of the aphasia and follow-up examinations were carried out by the same investigator (HVD). Language measures, ie, analysis of spontaneous speech, object naming, repetition and auditory comprehension (Token Test, 29) and for toddlers the Tridimensional Matrix Test (TMT; 30) have been described elsewhere (23).

Children were considered as incompletely recovered when:
- aphasic signs were present in spontaneous language:
  a degree of 0 to 4 aphasia score on a severity rating scale
  (ref. 31).
- the performances on the Token Test or Tridimensional Matrix
  Test were one or more sd below the average
  for that child's age (32).

Results.

The data, summarized in Figure 5, reveal significant differences between the 6 LKS children and 4 children with a brain tumor on the one hand and the children of the other etiological groups on the other. The children with a tumor showed a worsening aphasia and died about one year after onset. The children with LKS differ with respect to age at onset (range 4 to 6 years), and CT-scan ratings (all normal).
The outcome in the 18 children with head injury or vascular, or infectious disease is reported together. In view of the small number of patients in the various groups the relationship between the recovery and the variables age at onset, cause, severity of lesion and type of aphasia are evaluated separately (tables 3 and 4).

EFFECTS OF AGE.

The assumption that the earlier in life a lesion is sustained the better the outcome was not confirmed (table 3). Our results indicate that there was no difference in recovery for those aged above or below 11 years. The very young children with LKS had a bad prognosis, as in accordance with the literature (11).

---

[1] Case 13 : reexamination was refused by the child's parents.
Case 12 : reexamination could be performed only onde month and five years after onset.

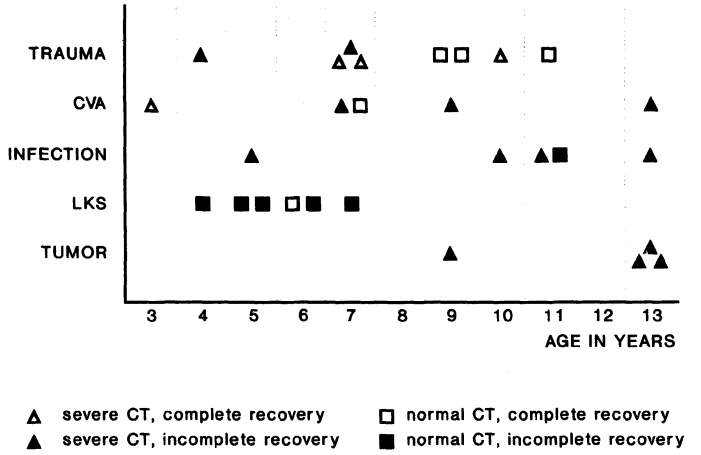


Figure 5. Severity of the lesions versus outcome one year after- onset in 28 patients. CVA indicates cerebrovascular accident; LKS, Laudau-Kleffner syndrome; and CT, computed tomographic.

Table 3.

OUTCOME IN 18 PATIENTS WITH HEAD INJURY, VASCULAR AND INFECTIOUS DISEASE RELATED TO AGE AT ONSET, CAUSE, SEVERITY OF LESION, AND TYPE OF APHASIA.

Variables	Recovered	Nonrecovered
Age at onset		
` 11 years	6	7
¨ 11 years	1	4
Cause*		
head injury	6	2
vascular disease/ infection	1	9
Severity of lesion		
CT severe	4	9
CT non-severe/ normal	3	2
Type of aphasia		
fluent	2	2
non-fluent	5	9

* P`.01 for one tailed Fisher's Exact Test.

CAUSE.

Six of the eight children with a head injury had a favorable outcome (table 3). This was in complete contrast with those with vascular or infectious disease. Furthermore, only one of the six children with LKS recovered completely.

SEVERITY AND BILATERALITY OF THE LESION.

In view of the small number of patients, we regrouped them on the basis of severity of lesion. Patients whose CT-scans were rated as 1 or 2 composed the group with a severe lesion and were compared with those patients with a non-severe lesion (categories 3, 4 or 5). Although recovery was frequently observed in children with non-severe/ normal CT scans, the association of outcome with severity of lesion was not significant (table 3).

Initially all children with a cerebral tumor showed a mild aphasia, although at admission the lesions on CT scan were categorized as severe. However, the aphasic disturbances increased very rapidly during their stay in the hospital. In contrast, all children with LKS demonstrated a severe aphasia at admission, despite normal CT scans.

In our series local lesions were limited to the left hemisphere. In eight patients, in addition to the lesion in the left hemisphere, a slight enlargement of the right ventricle and/or dilation of the right sulci was found. These children, except for one, had a poor outcome. However, this can also be explained by the severity of the lesion. As the severity and bilaterality of a hemispheric lesion are interrelated, we tested both the effect of severity when correcting for bilaterality, and the effect of bilaterality when correcting for severity (table 4).

Table 4.

OUTCOME IN 18 PATIENTS WITH HEAD INJURY, VASCULAR AND INFECTIOUS DISEASE RELATED TO SEVERITY, AND BILATERALITY OF HEMISPHERIC LESION*

Variables	Recovered	Nonrecovered
Bilateral hemispheric lesion		
8 (6 S + 2 NS)	1 (NS)	7 (6 S + 1 NS)
Left hemispheric lesion		
10 (7 S + 3 NS)	5 (3 S + 2 NS)	5 (4 S + 1 NS)

* Severe (S) and nonsevere (NS): category 1 or 2, respectively 3 and 4 or 5 of the rating scale for CT-scans (Vargha-Khadem et al., 35).

Neither the effect of severity when correcting for bilaterality, nor the effect of bilaterality when correcting for severity were significant (Fisher's Exact Test P= .19 resp. 0.13).

TYPE OF APHASIA.

Fluent aphasia was present in four children (patients 2, 5, 9 and 16). This type of aphasia was not significantly associated with recovery (table 3). It seems important that in the two children who recovered (patients 2 and 5), the aphasia was caused by head injury, a cause generally linked to a good prognosis. Conversely an incomplete recovery was present in the two children with an unfavorable cause ie, an infectious disease and vascular disease respectively.

Comment.

The present study contradicts the common clinical belief that acquired childhood aphasia is usually of short duration. Our results demonstrate that, with the exception of a traumatic cause, none of the other studied variables are unequivocally favorable or unfavorable for recovery. Often these variables are interrelated and the outcome will be determined by a complex interaction between a number of factors. These factors are not discriminable at present because the cited studies on childhood aphasia include only small etiological groups that are not comparable regarding the age at onset and severity of lesions. Despite these considerations the diverging opinions about outcome can be explained partially. In general a good outcome may be expected if (mild) head injury is the cause, even when the aphasia is severe at onset (28). On the contrary, a poor outcome is reported in a group limited to infectious disorders (33) or the Landau- Kleffner syndrome (16). In the present series we obtained similar findings.

Our data demonstrate a relationship between severity of lesion and cause. Normal CT-scans were found in all patients with LKS. However, in nearly all patients with vascular or infectious disease the CT-scan was severely abnormal, which would suggest that, in such cases, clinically manifested aphasia is observed only when the lesion is extensive.

The possibility that the intact right hemisphere has contributed to language recovery has to be considered. Bilateral lesions are frequently found with infectious and vascular diseases, which are associated with a poor outcome.

Disagreement in terms of favorable versus unfavorable outcome may be explained by the method of assessment. Until now recovery from aphasia has usually been estimated only in clinical terms (34), which can explain the different opinions. The need for a standardized aphasia examination has to be emphasized.

Finally the great difference in length of follow-up in the diverse series of aphasic children will hamper the prediction of outcome because the duration of aphasia in these series has not been related to the various prognostic variables.

In conclusion, despite the fact that this study includes 28 children and is one of the largest series in the literature, no firm prediction about the outcome can be made at present. At best, a certain trend can be suggested by our results: in contrast to head injury the other causes are linked to an unfavorable outcome.

The authors thank I. Rapin, MD, and D.Hasan, MD, for their helpful advice, and D. Bishop, DPhil, for revising the English-language text.

REFERENCES

1. Kertesz A. Aphasia. In: Frederiks JAM ed. Handbook of Clinical Neurology. Vol 45. Clinical Neuropsychology, Amsterdam, the Netherlands: Elsevier Science Publishers; 1985;45:287-331.

2. Basser LS. Hemiplegia of early onset and the faculty of speech with special reference to the effects of hemispherectomy. Brain 1962;85:427-466.

3. Lenneberg E. Biological foundations of language. New York, NY: John Wiley & Sons Inc; 1967.

4. Cooper JA, Flowers CR. Children with a history of acquired aphasia: residual language and academic impairments. J Speech Hear Disord 1987;52:251-262.

5. Satz P, Bullard-Bates C. Acquired aphasia in children. In: Taylor Sarno M, ed. Acquired aphasia. Orlando, Fla, Academic Press Inc; 1981:399-426

6. Van Dongen HR, Loonen MCB. Factors related to prognosis of acquired aphasia in children. Cortex 1977; 13,131-136

7. Van Hout A, Evrard Ph, Lyon G. On the positive semiology of acquired aphasia in children. Dev Med Child Neurol 1985; 27,231-241

8. Woods BT, Carey S. Language deficits after apparent clinical recovery from childhood aphasia. Ann Neurol 1979; 6,405-409

9. Alajouanine Th, Lhermitte F. Acquired aphasia in children. Brain 1965, 88:644-662

10. Woods BT, Teuber HL. Changing patterns of childhood aphasia. Ann Neurol 1978; 3,273-280

11. Bishop DVM. Age of onset and outcome in "acquired aphasia with convulsive disorder" (Landau-Kleffner syndrome). Dev Med Child Neurol 1985; 27,705-712

12. Dulac O, Billard C, Arthuis M. Aspects electro-cliniques et évolutifs de l'épilepsie dans le syndrome aphasie-épilepsie. Arch Franç Pediatr 1983; 40,299-308

13. Toso V, Moschini M, Gagnin G, et al. Aphasie acquise de l'enfant avec épilepsie; trois observations et revue de la literature. Rev Neurol (Paris) 1981; 137,425-434

14. Guttmann E. Aphasia in children. Brain 1942; 65,205-219

15. Byers RK, McLean WT. Etiology and course of certain hemiplegias with aphasia in childhood. Pediatrics 1962; 29,376-383

16. Dugas M, Masson M, Henzey MF, et al. Aphasie acquise de l'enfant avec épilepsie (syndrome de Landau et Kleffner): douze observations personelles. Rev Neurol (Paris) 1982; 138,755-780

17. Mantovani JF, Landau WM. Acquired aphasia with convulsive disorder: course and prognosis. Neurology 1980; 30,524-529

18. Van Harskamp F, Van Dongen HR, Loonen MCB. Acquired aphasia with convulsive disorder in children: case study with 7-year follow-up. Brain Lang 1978; 6,144-148

19. Gloning K, Hift E. Aphasie im Vorschulalter. W Z Nervenheilkd 1970; 28,20-28

20. Hécaen H. Acquired aphasia in children: revisited.
Neuropsychologia 1983; 21,581-587

21. Aram DM, Rose DF, Rekate HL, et al. Acquired capsular/striatal aphasia in childhood. Arch Neurol 1983; 40:614-617

22. Ferro JM, Martins IP, Pinto F, et al. Aphasia following right striato-insular infarction in a left-handed child: a clinico-radiological study. Dev. Med. Child Neurol 1982; 24,173-182

23. Van Dongen HR, Loonen MCB, Van Dongen KJ. Anatomical basis for acquired fluent aphasia in children. Ann Neurol 1985; 17,306-309

24. Martins IP, Ferro JM, Trindade A. Crossed aphasia in a child. Dev Med Child Neurol 1987; 29,96-109

25. Martins IP, Ferro JM. Acquired conduction aphasia in a child. Dev Med Child Neurol 1987; 29,532-536

26. Beaumanoir A. The Landau-Kleffner syndrome. In: Roger J, Dravet C, Bureau M, Dreifuss FE, Wolf P, eds. Epileptic syndromes in Infancy, Childhood and Adolescence. London, England: John Libbey Eurotext, 1985:181-191

27. Collignon R, Hécaen H, Angelergues R. A propos de 12 cas d'aphasie acquise de l'enfant. Acta Neurol Belg 1968; 68,245-277

28. Assal G, Campiche R. Aphasie et troubles du langage chez l'enfant après contusion cérébrale. Neurochirurgie (Paris) 1973; 19:399-406

29. De Renzi E, Vignolo LA. The Token Test: a sensitive test to detect receptive disturbances in aphasics. Brain 1962; 85,665-678

30. Kreindler A, Gheorghita N, Voinescu I. Analysis of verbal reception of a complex order with three elements in aphasics. Brain 1971; 94,375-386

31. Goodglass H, Kaplan E. The assessment of aphasia and related disorders. Philadelphia, Pa: Lea & Febiger, 1972

32. Blauw M, Ceyssens I, Paquier P, et al. De discriminatieve waarde van de Token Test voor taalpathologie bij kinderen. Log Fon 1987; 59,218-222

33. Van Hout A, Seron X. L'Aphasie de l'Enfant et les Bases Biologiques du Langage. Mardaga P, ed. Brussels, Belgium: Psychologie et Science Humaine; 1983

34. Cranberg LD, Filley CM, Hart EJ, et al. Acquired aphasia in childhood: clinical and CT investigations. Neurology 1987; 37,1165-1172

35. Vargha-Khadem F, O'Gorman AM, Watters GV. Aphasia and Handedness in relation to hemispheric side, age at injury and severity of cerebral lesion during childhood. Brain 1985; 108,677-696.

VII - LATE SEQUELAE OF EARLY BRAIN LESIONS

SCHOLASTIC ACHIEVEMENT AFTER EARLY BRAIN LESIONS

DOROTHY M. ARAM
Department of Pediatrics
Case Western Reserve University
2101 Adelbert Road
Cleveland, Ohio 44106

ABSTRACT. Data are summarized for three areas of scholastic achievement in children with lateralized brain lesions: reading; written language including spelling; and mathematics. In general, reading disorders appear to be more common following left than right hemisphere lesions, although the incidence varies widely. Written language deficits are the most common and persistent higher cognitive sequelae noted, yet very little systematic study characterizing these deficits has been undertaken. Spelling deficits have been described particularly following left, but also right brain lesions, although data relating type of spelling deficit to lesion laterality is equivocal. Although several groups of investigators have commented upon mathematics ability in their brain lesioned children, few generalizations emerge from these reports and it appears that no one has described the nature of the mathematical deficits presented. The scant data reported are related, where possible, to lesion laterality, site of lesion within a hemisphere, and age of lesion onset.

INTRODUCTION

Clinicians often comment that difficulties in academic achievement persist even when recovery of language and other aspects of cognitive behavior are felt to be normal. For example, Alajouanine and Lhermitte (1965) noted that none of their 32 children with acquired aphasia were capable of following normal progress in school. Despite the frequent observation that difficulties in academic achievement are a common sequelae of early brain lesions, very little research has detailed the deficits presented or factors related to variable levels of recovery. The present paper will address three areas of academic achievement: reading, written language including spelling, and mathematics. In each area literature reported from other clinicians and investigators will be reviewed, followed by a summary of studies that we have conducted in each of these areas. Any data relevant to variable outcome and the factors of lesion laterality, age of lesion onset and site of lesions will also be noted.

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 203–212.

Before entering a discussion of findings, a few words are in order concerning subject populations and contradictory findings. A recurring theme in this symposium is the highly diverse subject groups studied giving rise to very different findings. Outcome for academic achievement ranged from significant sequelae for all subjects (Alajouanine & Lhermitte, 1965) to essentially normal school performance (Kershner & King, 1974), and undoubtedly reflect to a large degree differences among the subjects studied. Rather than continue to belabor the major methodological problems in this area involving subject characteristics, the diverse subjects studied may provide a means for identifying factors variably related to outcome. Therefore, where possible I will attempt to interpret contradictory findings in reference to subject variables which may relate to outcome.

Because I will be drawing from several studies done by our group in Cleveland, I will first summarize the criteria for subjects entered into our studies. All children were identified as inpatients at Rainbow Babies and Childrens Hospital or from chart review of pediatric neurology or cardiology outpatients. To be entered into the study, each subject was required to meet the following criteria. First, for all subjects sustaining lesions after the perinatal period, documentation of normal neurologic, genetic and developmental status prior to lesion onset was required. Children with histories of premature births, neonatal complications other than congenital heart disorders, or genetic abnormalities were excluded. Second, the clinical neurological examination evidencing a unilateral vascular lesion needed to be confirmed initially by CT verification and subsequently by MRI scans wherever possible (not secured for patients with surgical clips or pacemakers) taken 6 months to 1 year after lesion onset. Children with suspected bilateral involvement were excluded including those with ongoing seizure disorders requiring anticonvulsants.

Currently, as of this month, we have 31 left, 16 right and 11 children with nonfocal lesions in our study. This latter group is comprised of two types of children: (1) children initially thought to have unilateral lesions on the basis of CT scans, and later discovered to have more widespread involvement on the basis of MRI findings; (2) or children who developed ongoing seizures requiring anticonvulsants during the course of the study. Although we initially excluded these nonfocal children from our study, we have begun retaining them in a separate group so as not to lose potentially informative data. Age of lesion onset ranges from prenatally to 16 years of age. Current age as of this month ranges from almost 1 year to 22 years. Left lesioned group incurred their lesions somewhat later than did the rights, and also on the average are approximately two years older. Fourteen left-lesioned and eight right-lesioned children sustained pre or perinatal insults. Many of the remaining subjects had underlying congenital heart disorders which gave rise to embolic events usually involving the middle cerebral artery. Four left-lesioned subjects had arteriovenous malformations; one left-lesioned subject had a stroke as a complication of a complex migraine; and one right lesioned subject

had an unknown basis for his lesion. All CT and MRI scans have been read and classified by the same neuroradiologist, blind to the child's clinical findings. Finally, all of our studies have compared the brain lesioned children to control children individually matched by chronological age, sex, race and social class. Since the majority of lesioned children had congenial heart disorders, control subjects were selected from chart review of pediatric cardiology patients who had experienced no neurological complications and were developing normally. An attempt was made to match lesioned and control children on the basis of arterial oxygen saturation levels; acyanotic control subjects were selected for lesioned subjects without heart disorders.

Reading

The three clinical group studies providing comments about reading status of children with acquired aphasia present conflicting clinical pictures. Alajouanine and Lhermitte (1965) report that 18 of the 32 children with predominantly left hemisphere lesions continued to have reading difficulties at follow-up with nine remaining severely alexic. In contrast, Hecaen (1976, 1983) reported that although reading difficulties were common acutely (occurring in 9/17 left lesioned children; 1/6 rights; and 1/3 bilaterally involved children), reading difficulties generally disappeared rapidly and completely, although it should be noted that reading difficulties persisted in three children, at least one of whom had bilateral involvement. Hecaen further stated that reading disorders were not related to site of lesion or to age of lesion onset. More recently, Cranberg, Filley, Hart and Alexander (1987) reported that at follow-up of at least one year post-lesion onset, 5/8 left lesioned aphasic children continued to be severely below grade level in reading, while 2/8 had mild residual reading problems. These investigators attributed the continued reading deficits to an "acquired handicap of new learning".

Case descriptions of aphasic children further illustrate the diversity of reading ability among children with acquired lesions. Dennis (1980) described a dissociation between age appropriate reading comprehension and a significant deficit in auditory comprehension in a 9-year old girl following a left temporoparietal infarct. Both Ferro, Martins, Pinto and Castro-Caldas (1982) and Martins, Ferro and Trindade (1987) have described reading deficits following right hemisphere lesions, the former, in a 6-year old girl following a right striato-insular infarction, and the latter in a 15-year old boy following a right occipito-temporal tumor.

I am aware of only three studies, other than our own, which have provided psychometric data for groups of children with lateralized brain lesions. The first, reported by Kershner and King (1974) involved seven left and seven right hemisphere lesioned children compared to controls on the Wide Range Achievement Test (WRAT) word recognition test. No difference was found between the groups. A second study presented in 1983 by Vargha-Khadem and her colleagues here in Portugal at the International Neuropsychology Society meetings, reported results on the Durrell Analysis of Reading

Difficulty which includes measures of reading speed and comprehension. The numbers of children below expected grade level for reading were as follows: for the prenatal left lesioned children, 13/17; postnatal lefts, 7/7; prenatal rights 5/15; and post-natal rights, 2/7. Vargha-Khadem concluded that although all patients were more likely to be reading retarded than were normal controls, reading deficits were more pronounced in left hemisphere lesioned patients, especially when lesions were sustained postnatally, where all continued to be performing below grade level. Cooper and Flowers (1987), on the other hand, documented a better prognosis on long-term follow-up for reading among a group of 14 children with acquired aphasia sustained between 3 and 12 years of age, with only three scoring below 1 standard deviation of the mean on the word recognition subtest of the Wide Range Achievement Test and 5/14 scoring below 1 standard deviation of the mean on the reading comprehension subtest of the Peabody Individual Achievement Test. Cooper and Flowers did not attempt to tie their outcome to either lesion laterality or age of onset. It should be noted, however, that although lesions in both studies were categorized as lateralized to predominantly the left or right hemisphere, many of the subjects in each study presumably had diffuse brain involvement given the inclusion of children with closed head injury, tumors, anoxic encephalopathy, and ongoing seizure disorders.

The limited and contradictory data available relative to prognosis for reading among children with unilateral brain lesions prompted us to undertake two studies with our school-aged children. The first was a part of a more general study of academic achievement, in which we administered the Woodcock-Johnson Psycho-Educational Battery. Contrary to our predictions, right but not left lesioned children scored significantly below control groups on the reading achievement cluster, a composite measure including word identification (i.e, single letter and word oral reading), word attack (application of grapheme-phoneme rules to nonwords, e.g. mibgus) and passage comprehension. We attributed the lack of significant differences on the Reading Achievement Cluster for left lesioned subjects to two factors: first, insensitivity of a screening test designed to assess a wide age range with few items at any one level; and second, masking of the considerable variability among lesioned subjects through reporting statistical results based predominantly on group mean performance. Therefore a follow-up study was undertaken aimed to provide more detailed and hopefully discriminating analyses of multiple components of reading and to focus upon individual difference in performance, rather than on group data alone. In this study we assessed 20 left and 10 right hemisphere lesioned children between 6 and 20 years of age, on a comprehensive battery of reading measures. Although group mean performance on all tasks was consistently below that of the control subjects matched for age, sex, race and social class, few differences reached statistical significance. These results demonstrated that the majority of children with unilateral brain lesions learned to read quite adequately; yet five left and two right lesioned children presented marked reading deficits in contrast to 1 of the 30 controls. We then attempted to identify variables

which may be differentially related to children with and without residual reading deficits. A strong family history for reading disorders was implicated in one left, one right and the only control subject, suggesting the contribution of genetic factors as the basis for these children's reading disorders, either alone or in combination with the neurological insult. Age of lesion onset was not found to be related to reading outcome. However, all left lesioned children with involvement of specific subcortical structures did present reading disorders along with other language problems, suggesting that these structures assumed a role in acquisition of higher cognitive functions which may be less amenable to reorganization following lesions sustained in early childhood.

What then do we know about reading achievement in children with lateralized brain lesions and how can we explain the variable findings reported? First, reading disorders appear to be more common following left than right hemisphere lesions, although clearly reading problems have been documented following right lesions. However, characteristic reading disorders related to lesion laterality or site among left in contrast to right lesioned patients have not been described (e.g. difficulty with phonetic decoding among lefts, or inferential comprehension among rights). Second, among the left-lesioned children a wide range in incidence of reading disorders has been reported, ranging from 100% following postnatally incurred left lesions (Vargha-Khadem, Frith, O'Gorman & Watters, 1983) to 25% of lesioned subjects (Aram, Gillespie & Yamashita, in press). Comparing the subjects included in these studies, it would appear that etiologies suggestive of nonfocal involvement (notably tumors, trauma and ongoing seizure disorders) are implicated among many children with significant reading disorders. Age of lesion onset, although suggested to be a variable related to reading prognosis (Vargha-Khadem et al., 1983) has failed to differentiate among children with and without reading problems in other studies addressing this variable (Aram and Ekelman, 1988; Aram et al., in press; Hecaen, 1983). Little attempt has been made to relate reading deficits to site of lesion within a single hemisphere, although reading deficits have been noted to follow both cortical (Martins et al., 1987) and subcortical (Ferro et al., 1982) lesions in the right hemisphere. Hecaen (1976; 1983) and Aram (Aram and Ekelman, 1988; Aram et al., in press) have failed to find a relationship between the occurrence or type of reading disorder encountered and site of lesion involving left cortical areas, although Aram et. al. (in press) have speculated that lesions involving subcortical structures in the left hemisphere are associated with more pronounced reading deficits than are cortical left lesions. These "conclusions", however, are based on as yet very few studies detailing either components of reading, or important subject characteristics.

Written Language and Spelling

Although most clinical reports suggest that written language deficits are the most common and typically the most persistent higher cognitive sequelae following brain lesions in children, I was unable to locate

any studies which detailed these children's written language characteristics. Alajouanine and Lhermitte (1965) reported that written language was always disturbed following left hemisphere lesions in children, and tended to be more severely involved than oral language in their 32 left-lesioned subjects. Hecaen (1976, 1983), who generally considered any impairments of higher cognitive functions observed in the acute period to resolve rapidly and completely, noted that writing disturbances were the most frequent and most variable disturbance observed during the acute period, tended to persist the longest, even permanently, and were particularly common among children with lesion onset prior to 10 years of age. Although he noted that writing disturbances were more common following left than right hemisphere lesions, Hecaen maintained that within a hemisphere, writing disturbances were of little localizing value. We, too, (Aram and Ekelman, 1988) found that written language, measured through the Written Language Cluster of the Woodcock-Johnson Psycho-Educational Battery (a composite measure involving spelling to dictation and proofing), was the one academic achievement area screened that was significantly lower for the left-lesioned as well as the right-lesioned groups of children in comparison to control subjects. Left-lesioned subjects with subcortical involvement presented the most severe deficits in written language. A relationship between age of lesion onset and written language was not found. Although there appears to be consensus that writing is the most frequent and most severe academic limitation following left lesions, as far as I have been able to determine, no one has characterized the nature of these written language deficits.

Spelling has been studied somewhat more thoroughly. Woods and Carey (1979) appear to have been the first to provide actual data documenting left-lesioned children's spelling measured by a series of eight words spelled orally. These investigators found spelling to be the only task, among the eight language tasks administered, to be significantly lower than controls for both the early and late left-lesioned group, irrespective of whether the subject was a recovered aphasic or had never been aphasic. Cooper and Flowers (1987) administered the WRAT: Spelling Subtest to the group of children with acquired aphasia. On follow-up at least one year post lesion onset, 8/14 subjects (53%) scored greater than 1 standard deviation below the mean on this test. Neither the Woods and Carey (1979) or Cooper and Flowers (1987) study attempted to analyze the nature of spelling deficits among their lesioned subjects. Vargha-Khadem et al., (1983) analyzed left and right lesioned subjects' ability to spell frequent and infrequent words. On the frequent word list, postnatal left lesioned subjects were significantly worse than all other subject groups; on the infrequent words all patient groups (pre and postnatal left and right lesioned groups) were significantly poorer than controls, with the postnatal left hemisphere group being significantly more impaired than the other three lesioned groups of children. Vargha-Khadem et al. (1983) also provided a qualitative error analysis, concluding that among postnatal left lesioned children, the morphemic and purely orthographic features (e.g. double letters,

silent letters) were poorly retained and that there appeared to be an impaired preservation of the sound frame of the word. Predicting that left-lesioned children would have greater difficulty spelling phonetically regular than nonregular words, and that the converse may be found for right lesioned subjects, we administered the Test of Written Spelling which includes a predictable word list (words that conform to predictable grapheme-phoneme rules and generalizations) and an unpredictable word list (words that do not conform to predictable rules and generalizations). Contrary to our predictions, the right-lesioned group alone was significantly lower than their controls on both word lists. Furthermore, no difference was apparent for either the left- or the right-lesioned groups on the predictable or unpredictable word lists, suggesting that these functions may not be differentially lateralized. However, our findings differ from those of Vargha-Khadem et al. (1983), and the basis for this difference is uncertain. Although Vargha-Khadem included children with more diffuse brain involvement than did we, her findings are more consistent with our initial predictions than are the findings of our study. A factor potentially contributing to our failure to differentiate between predictable and unpredictable words may be the measure used. A review of the words included on each list suggests that the two word lists may not be strictly "predictable" or "unpredictable", and in fact were quite similar.

Summarizing what we know about written language and spelling, we may conclude the following: (1) written language generally is found to be the most severe and persistent area of academic difficulty following brain lesions in children, particularly when the left hemisphere is involved; (2) the actual nature of the written language deficits presented have not been characterized in the literature; (3) spelling deficits are commonly reported following especially left, but also right hemisphere lesions; (4) data suggesting that specific types of spelling deficits relate to lesion lateralization is equivocal; (5) written language deficits have not been found to relate to cortical sites of lesions although, some limited evidence suggests that children with subcortical left lesions have especially pronounced deficits; (6) data attempting to relate severity of written language deficit to age of lesion onset is highly contradictory.

Mathematics

Although several groups of investigators have commented upon mathematics ability in their brain lesioned children, few generalizations emerge from these reports and no one has described the nature of mathematical deficits presented. Hecaen (1983) stated that acalculia is the only major neuropsychological deficit associated with impaired language in left-lesioned children, noting that 11/15 left-lesioned children were acalculic on follow-up. Similarly, Coopers and Flowers (1987) found a greater number of their children with acquired aphasia were greater than 1 standard deviation below the norms on the Math Subtest of the WRAT (12/14) than either of the other two WRAT subtests (reading and spelling), although these investigators did not

attempt to relate their findings to lesion lateralization. Cranberg et al. (1987) reported a lower incidence of mathematical problems, reporting math difficulties in 3/8 left-lesioned children.

In contrast, Kiessling et al. (1983) found that among hemiplegic children, left hand ability (suggestive of right hemisphere function) as measured by the Annett Pegboard was significantly correlated with math calculations, as measured by the WRAT. Similarly, we (Aram and Ekelman, 1988) reported that right-lesioned but not left-lesioned children were significantly poorer than controls on the Math Achievement Cluster (which included both calculations and applied problems subtests) of the Woodcock-Johnson Psycho-Educational Battery. In addition, unlike any of the other subject groups, right lesioned subjects were poorest on math achievement than any of the three areas examined (reading, math, and written language). Finally, Vargha-Khadem et al. (1983) reported that in her group the mental arithmetic subtest of the WISC did not discriminate among left-lesioned, right-lesioned or control subjects. At this point it appears that mathematical abilities may be deficient among some brain-lesioned children; but how such deficits relate to lesion laterality is anyone's guess. As with written language, no attempts have been reported to characterize the actual nature of mathematical difficulties presented following brain lesions in children.

SUMMARY AND CONCLUSIONS

The above review makes it clear that we have a long way to go before being able to characterize reading, written language or mathematics in children with brain lesions. While it appears that some of these children will have deficits in academic achievement, we do not yet have enough converging data to suggest trends let alone to prognosticate for an individual child. Indeed it is surprising given the common report of academic difficulties among brain-lesioned children, that so few investigators have addressed any of these areas.

In hopes of amassing a sufficient data base from which to generalize, I would like to encourage clinicians and researchers working with brain lesioned children to observe and provide data, either through clinical check lists similar to those advocated by Dr. Rapin, or through psychometric measures, at a minimum to the following areas:

Suggested Minimal Data

Reading:	single word recognition reading comprehension
Written Language:	written prose spelling
Mathematics:	calculations applied problems

Within each of these domains of academic achievement, numerous aspects could also be addressed, for example for word recognition the effect of regularity, frequency, word length and so forth. In addition, we are woefully lacking studies with brain lesioned children detailing deficits in any of these areas or tying observations to theoretical explanations of underlying processes and breakdowns, similar to studies that have appeared with hemispherectomized children or adults with acquired lesions. These areas beg for theoretically motivated studies which have the potential for furthering our understanding of early brain specialization for higher cognitive functions and for reorganization of these functions following lesions sustained early in development.

ACKNOWLEDGEMENT

Preparation of this manuscript was supported by a National Institute of Neurological Disorders and Stroke Grant NS 17366 to Dr. Aram.

REFERENCES

Alajouanine, T.H., & Lhermitte, F. (1965). Acquired aphasia in children. Brain, 88(4), 653-662.

Aram, D.M., & Ekelman, B.L. (1988). Scholastic aptitude and achievement among children with unilateral brain lesions. Neuropsychologia, 26, 903-916.

Aram, D.M., Gillespie, L.L., & Yamashita, T.S. (In press). Reading among children with left and right brain lesions. Developmental Neuropsychology.

Cooper, J.A., & Flowers, C.R. (1987). Children with a history of acquired aphasia: Residual language and academic impairments. Journal of Speech and Hearing Disorders, 52, 251-262.

Cranberg, L.D., Filley, C.M., Hart, E.J., & Alexander, M.P. (1987). Acquired aphasia in childhood: Clinical and CT investigations. Neurology, 37(7), 1165-1172.

Dennis, M. (1980). Strokes in childhood I: Communicative intent, expression, and comprehension after left hemisphere arteriopathy in a right-handed nine-year old. In R.W. Rieber (Ed.), Language Development and Aphasia in Children (pp. 45-67). New York: Academic Press.

Ferro, J.M., Martins, I.P., Pinto, F., & Castro-Caldas, A. (1982). Aphasia following right striato-insular infarction in a left-handed child: A clinico-radiological study. Developmental Medicine and Child Neurology, 24, 173-182.

Hecaen, H. (1976). Acquired aphasia in children and the ontogenesis of hemispheric functional specialization. Brain and Language, 3, 114-134.

Hecaen, H. (1983). Acquired aphasia in children: Revisited. Neuropsychologia, 21(6), 581-587.

Kershner, J.R., & King, A.J. (1974). Laterality of cognitive functions in achieving hemiplegic children. Perceptual and Motor Skills, 39, 1283-1289.

Kiessling, L., Denckla, M., & Carlton, M. (1983). Evidence for differential hemispheric function in children with hemiplegic cerebral palsy. Developmental Medicine and Child Neurology, 25, 724-734.

Martins, I.P., Ferro, J.M., & Trindade, A. (1987). Acquired crossed aphasia in a child. Developmental Medicine and Child Neurology, 29, 96-109.

Vargha-Khadem, F., Frith, U., O'Gorman, A.M., & Watters, G.V. (1983). Learning disabilities in children with unilateral brain damage. Paper presented at the International Neuropsychological Society meetings, Lisbon, Portugal.

Woods, B.T., & Carey, S. (1979). Language deficits after apparent clinical recovery from childhood aphasia. Annals of Neurology, 6(5), 405-409.

LATE SEQUELAE OF RIGHT VERSUS LEFT HEMISPHERIC LESIONS.

D.RIVA, C.PANTALEONI, N.MILANI, M.DEVOTI.
Istituto Neurologico "C.Besta"
Via Celoria 11
20133 - Milano
Italy

ABSTRACT

The problem of language impairment in children with focal brain lesion is complex. It involves many variables such as age and modality of onset, type and size of lesion and finally side and localization. According to this, 2 groups of children, who suffered from vascular accidents were selected. The time of acquisition was classified as "early" (before language acquisition) and "late" (after language acquisition). Results indicate that right as well left lesioned children perform worse than controls in lexical tests, while left lesioned ones perform worse in lexical and syntactic tasks. The influence of age at lesion, lesion size and epilepsy are discussed.

INTRODUCTION

The antiequipotentiality hypothesis claims that language can develop its most refined aspects only in the left hemisphere and that in case of early even congenital left brain lesion, right hemisphere can not develop the full range of language abilities.
Most of the developmental studies in this area have demonstrated in the last decade this theory (Varga-Khadem 1985; Aram 1988; Riva 1986). Only few of them contradict this position and provide data supporting the equipotentiality hypothesis (Woods 1979, 1980).
Particularly for the language it is difficult to select children with unilateral brain lesion, which is a rare condition per se, who share the same socio-cultural background and then who can be compared directly.
Furthermore many variables can affect the neuropsychological outcome: age at onset, epilepsy, extent and location of injury and acute or chronic modality of onset.

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 213–224.

AIM OF THIS STUDY

1. is to examine children with unilateral brain lesion both acquired before the language acquisition or later in life, but always after language acquisition, testing intelligence and some aspects of language
2. to verifie if the left specialization for language is already present in the very early stages of life.

SUBJECTS

Subjects were selected from a larger sample of children with unilateral brain lesion of different etiology, admitted to the Child Neurology Dept. of the Istituto Neurologico "C.Besta" of Milan. Only children with lesions caused by vascular accidents were included in the study.
The unilateral nature of the injury was determined by neurological examination, EEG and CT scan.
The exclusion criteria were: 1. history of severe seizures; 2. IQ<75; 3. age<7years; 4. prematurity; 5. suspicion or evidence of bilateral hemispheric involvement and 6. suspicion of genetic abnormalities.
On the basis of the above selection criteria the final sample was composed by 37 children.
The patient population was divided into 2 sub-groups, according to the age at injury. The first group was composed by prenatal or congenital lesions. The second group was composed by children who sustained brain lesion after the first year age. They were then classified as "early" right or left lesions (ER, EL) and "late" right or left (LR,LL). These four groups were distributed as follows:

SUBJECTS

GROUPS		N.	TEST-AGE (mean)	RANGE
EARLY LEFT LESIONS	= EL	10	12.6	7.2-16.4
EARLY RIGHT LESIONS	= ER	9	11.7	7.8-15.2
LATE LEFT LESIONS	= LL	7	12.4	8.1-15.8
LATE RIGHT LESIONS	= RL	11	11.2	7.9-15.7

		LESION ONSET AGE (mean)	RANGE
" " "	EL	0.06	0.0-1
" " "	ER	0.08	0.0-1
" " "	LL	5.4	2.5-12.5
" " "	RL	6.1	3.1-13.2

The etiology of the 4 selected groups is shown in the following table:

	VASCULAR LESION CAUSED BY	
EL	1.IPOXIA	6
ER	2.METABOLIC CAUSE	4
	3.MECHANIC CAUSE	6
	4.UNKNOWN CAUSE	3
LL	1.POST-IPOXIC INFARCTION	1
LR	2.INTRACEREBRAL HEMMORAGE	1
	3.EMBOLIC OCCLUSION	4
	4.ISCHEMIC LESION	6
	5.AVM (bledding)	1
	6.CAVERNOUS ANGIOMA (bledding)	5

MOTOR DEFICIT

All the children had laterizated motor deficit.
The extent of deficit was assessed not on strictly neurological parameters (hipotrophy or spasticity), but on neurofunctional parameters: 1. the use of the upper limb in playing activities and 2. autonomy in everiday activities like eating or dressing.
This motor handicap was quantified from normal=0, to severe=3. There is not a significant difference in the distribution of motor deficit severity in the four groups, as shown in this table.

MOTOR DEFICIT

GROUPS	DEGREE OF THE HANDICAP		
	1	2	3
EL	2	7	1
ER	4	4	1
LL	3	2	1
LR	7	3	1

HANDEDNESS

In order to provide a quantified measure of hand preference the Haed-Piaget questionnaire was administered to all subjects. Hand preference was estimated by asking to perform some common actions, some of them were bimanual. All the "late" patients were also investigated for handedness before onset of the disease, or for familiar left handedness.
All right patients were R-handed before lesion. The same is

true for the left ones. Only one left subject had a family history for sinistrality. This subject become ambidextrous after injury.
The distribution of the handedness at testing is shown in this table.

	Left	Right	Ambidextrous
EL	9	1	-
ER	-	9	-
LL	5	1	1
LR	-	11	-
C	-	13	-

EPILEPSY
All the subjects had epileptic seizures in their life. The total number was always <10. All the subjects were taking anticonvulsivant drugs at testing: the drug blood level was periodically checked, and it was kept in the normal range values.
EEG was then considered as an interffering variable on cognitive and language tests, and quantified as: normal=1, abnormal but not epileptic=2 and epileptic=3.
The distribution of the diffrent EEG types is tabulated in the following table. There is no difference between the 4 groups.

	EEG		
	NORMAL=1	ABNORMAL=2	EPILEPTIC=3
EL	1	7	2
ER	-	6	3
LL	-	4	3
LR	1	7	3

NEUROIMAGING
All lesions were studied by CT scan. In each case the scans were made with 8 mm slices from the base to the vertex. We used the 5 points rating scale, adapted by Varga-Khadem, which caracterized the severity of lesion by size (Varga-Khadem 1985). The scale is shown in this table.

RATING SCALE FOR CT SCAN (VARGA-KHADEM)

1. NORMAL
2. MINIMAL VENTRICULAR DILATATION WITH ASYMMETRY SEEN ON LESS THAN 4 CUTS.
3. MODERATE TO MARKED VENTRICULAR DILATATION SEEN ON OR MORE THAN 4 CUTS.
4. MODERATE TO MARKED VENTRICULAR DILATATION WITH MINIMAL TO MODERATE LOSS OF BRAIN SUBSTANCE SEEN ON LESS THAN 4 CUTS.
5. MODERATE TO MARKED VENTRICULAR DILATATION AND GROSS LOSS OF BRAIN SUBSTANCE SEEN ON LESS THAN 4 CUTS.

DISTRIBUTION FOR LESION EXTENT AND LOCATION

The distribution of the extents of the lesions don't differ in the 4 groups.

LESION EXTENT

	CT RATING SCALE				
	1	2	3	4	5
EL	-	1	-	5	3
ER	1	-	1	3	3
LL	-	1	-	2	3
LR	-	-	1	5	2

CONTROLS

13 children with similar age (mean age 12y; range 7y,4m-15y,6m) and schooling; with no history of neurological disease and no neo or perinatal distress even suspected, were chosen as controls. The control group was also chosen in respect to the social class, because numerous authors have found a significant correlation between cognitive development and sociocultural environment.

Since it does not exist in Italy a standardized social scale, we classified our subjects according to 3 social classes. The distribution is shown in the following table.

SOCIO-ECONOMIC STATUS: 3 LEVELS

	CLASS			
	LOW	MIDDLE	UPPER	TOT.
EL	2	7	1	10
ER	1	6	3	9
LL	2	3	2	7
LR	2	7	2	11
C	1	9	3	13

METHOD

All the children were examined by 2 examiners: none of the children with late lesion was tested during the acute recovery. Testing was performed at least 2 years after the lesion onset.
The subjects were tested with a battery of neuropsychological tests to assess intelligence and language abilities.
Intelligence was rated on the WISC old form, because the study started before than standardized version of the WISC-R for the Italian Children was completed.
The language assessment included:

1. tests to assess semantic aspects of language:
   a. Vocabulary test (WISC item) to test the expressive language
   b. Peabody Picture Vocabulary Test (PPVT) for scoring receptive language.
2. tests to assess syntactic aspects of language:
   a. Token Test for scoring the ability to manipulate abstract and propositional relations between linguistic elements. The Token test used is the adapted short form, composed by 36 items.

STATISTICS

Separete analysis of variance and post-hoc tests were carried out to compare all the groups together and the single groups with controls.

RESULTS AND DISCUSSION

INTELLIGENCE - VERBAL IQ:
1. The difference between all the groups taken together is significant (p=0.002).
2. Only congenital lesions perform significantly worse than controls at post-hoc tests.
3. There is no difference between left and right lesioned children on this test.

VERBAL IQ: POST HOC TEST

	MEAN C	MEAN S	F	P
C VS EL	120.6	99.8	4.8	0.04*
C VS ER	120.6	99.3	5.2	0.03*
C VS LL	120.6	102.5		NS
C VS LR	120.6	103.5		NS

INTELLIGENCE - PERFORMANCE IQ
1. The difference between all groups is very significant ($p<0.001$).
2. Post hoc tests demonstrate that only children with congenital lesion, both right and left, are significantly worse than controls, the right group being even worse than the left one.

PERFORMANCE IQ: POST HOC TEST

	MEAN C	MEAN S	F	P
C VS EL	115.9	92.4	3.02	0.04*
C VS ER	115.9	88.6	4.11	0.03*
C VS LL	115.9	102.1		NS
C VS LR	115.9	101.0		NS

INTELLIGENCE - TOTAL IQ
1. The difference between all groups is very significant once again ($p<0.001$).
2. Also in this case, applying the post hoc test the difference is significant between the congenital groups, both right and left, and controls.

TOTAL IQ: POST HOC TEST

	MEAN C	MEAN S	F	P
C VS EL	120.0	96.5	3.2	0.04*
C VS ER	120.0	93.6	4.4	0.03*
C VS LL	120.0	102.8		NS
C VS LR	120.0	102.8		NS

On cognitive tests all the groups perform worse than controls, but only early lesioned children, both left and right, in a significant way.
In conclusions, all the subjects perform within normal limits, but worse than controls: only early lesioned children, both left and right, in a significant way.
There is no difference in verbal abilities between right and left early groups.
Early lesions seem to have a more important impact on cognitive functioning, than later ones. The younger brain

seems to be more susceptible to injury than a more mature one. During development, maturation evolves from very diffuse systems to more definite and localized ones: this different distribution of higher functions on the neural network in different ages produce different effects in case of lesion. In the first case a more global impairment is found; in the second case a more selective deficit. So, congenital or very early lesions produce more important intellectual deficits; on the contrary, lesions sustained later cause deficit of the function related to a more definite area.
The v-p discrepancy calls for a different comment (see table).

V-P DISCREPANCY

	VIQ	PIQ	V-P diff.	P(t test)
EL	99.8	92.4	7.4	NS
ER	99.3	88.6	10.7	0.04
LL	102.5	102.1	0.4	NS
LR	103.5	101.0	2.5	NS
C	120.6	115.9	4.7	NS

The fact that this pattern is present only in the early lesion groups could mean that recovery fron intelligence deficits can be possible only at the expense of the sound hemisphere. The so called "crowded effect" claims that the price of left brain injury recovery is always paid by the right hemisphere, because the left hemisphere matures earlier than the right one. This theory is now under discussion, because recent anatomical studies have demostrated that different cerebral regions of the two hemispheres have different maturation times during development. Anyway there is not a conclusive evidence based on the differential rate of maturation for different brain regions and the "crowded effect" hypothesis can still explain our data.

SEMANTIC TESTS RESULTS
On the Vocabolary Test all the groups perform significantly worse than controls (p=0.03). The same is true for the PPVT (p=0.05). Both right and left lesioned children perform significantly worse in the lexical tests measuring expressive and receptive language (see table). This could mean that the elementary aspects of language have bilateral representation and that they can be disrupted in case of either right or left brain injury.

VOCABULARY: POST-HOC TEST

	MEAN C	MEAN S	F	P
C VS EL	13	8.8	11.6	.002*
C VS ER	13	10.2	5.5	.02 *
C VS LL	13	9.4	6.0	.02 *
C VS LR	13	10.3	2.9	.03 *

PPVT:POST-HOC TEST

	MEAN C	MEAN S	F	P
C VS EL	133.0	106.9	6.07	.02*
C VS ER	133.0	107.2	6.5	.01*
C VS LL	133.0	109.5	6.17	.02*
C VS LR	133.0	103.5	8.1	.01*

SYNTACTIC TESTS RESULTS (TOKEN TEST).
If we consider the total score of the test (score=1 for every correct item) only left hemisphere lesions perform worse than controls. The early group is significantly worse, while the late one show a trend towards significancy

TOKEN TEST (TOTAL SCORE): POST-HOC TEST

	MEAN C	MEAN S	F	P
C VS EL	33.3	31.7	4.6	.04*
C VS ER	33.3	32.4	2.5	.1
C VS LL	33.3	31.5	3.1	.09 !!
C VS LR	33.3	32.0	1.9	.1

If we consider the last part of the Token test, which is the most complex in linguistic elements, both syntactic and semantic, and if we score the items analitically, we can find a strong difference between left lesioned children, both early and late, and controls.

TOKEN TEST PART 6: POST HOC TEST

	MEAN C	MEAN S	F	P
C VS EL	77.3	73.2	4.7	.04*
C VS ER	77.3	74.4	3.0	NS
C VS LL	77.3	72.4	2.4	.03*
C VS LR	77.3	74.5	2.6	NS

Thus, on the Token test only children with left lesion, both congenital and acquired later in life, perform significantly worse than controls. This lends support for the hypothesis of an early specialization at least for certain types of verbal abilities.

CORRELATIONS: LESION EXTENT

We correlated verbal and cognitive tests with lesion size. None of these correlations were significant. Thus, despite the grading of the CT, and the equal distribution on the basis of severity of lesions in the 4 groups, the different neuro-radiological abnormalities do not correlate with the severity of language impairment as tested in this study. Usually in adult studies a significant correlation is found between lesion size and language deficits. The finding that lesion extensivness do not correlate with linguistic impairment strongly suggest more extensive neurobiological and behavioral plasticity, which means a more dinamic neuronal reorganization in children than in adults.

CORRELATION: EEG

In order to determine the relationship between age variable and test scores a one-way variance analysis was carried out. The EEG factor is significant for all tests.

TEST SCORES BY EEG

	F	P
VIQ	4.7	.002
PIQ	5.6	.02
TOT.IQ	6.2	.0006
VOCABULARY	6.7	.0007
PPVT	6.0	.001
TOKEN TEST	8.7	.0001

Furthermore, to determine the effects of the group variable and EEG abnormalities on the test scores, a 2-ways variance

analysis was carried out. The group-factor and the EEG factor have a significant main effect only on intelligence tests. The same variables have no effect on the linguistic tests. The interaction group per EEG is not significant too. This suggest that epileptic abnormalities can influence the global functioning of the brain, but not some specific verbal abilities, which seem to depend from other variables, probably from the localization.

TEST SCORES BY GROUPS and EEG

	variables		
	group	eeg	group per eeg
VIQ	.05	.07	.09
PIQ	.05	.05	.05
TOT.IQ	.04	.06	.05
VOCABULARY	NS	NS	NS
PPVT	NS	NS	NS
TOKEN TEST	NS	NS	NS

CONCLUSIONS

1. Unilateral brain lesions, both congenital or acquired later in life don't cause major intelligence and language deficits.
2. The immature brain seems to be more vulnerable to brain injury than the more mature one. This susceptibility causes more important cognitive deficits as measured by the WISC, but not more defective performances on the language test.
3. Lexical aspects of language are defective in both left and right lesions, confirming the bilateral representation of the basic structure of many functions.
4. Syntactic aspects of language are defective only in the left lesion groups, providing support for the hypothesis of a genetic programming for hemispheric specialization for certain verbal skills.
5. Variables:
   a) age at injury is a good predictor for cognitive impairment as measured on the WISC, but fails to be a significant predictor for linguistic impairments.
   b) lack of correlation between test scores and lesion extent suggests a more dynamic neurobehavioral recovery in children than in adults.
   c) EEG abnormalities influence the global cognitive power, but not specific language abilities tested in this study.

ACKNOWLEDGEMENTS

We thank dr Graziella Filippini for advice and help on statistical analysis and dr Ludovico D'Incerti for CT rating.

REFERENCES

1) Aram D M (1988) 'Language sequelae of unilateral brain lesion in children', in F. Plum (ed), Language, communication and the brain, ARMND Series, Raven press, New York, pp 171-97.
2) Riva D and Cazzaniga L (1986) 'Late effects of unilateral brain lesion sustained before and after age one', Neuropsychologia 24 (3), 423-28.
3) Riva D, Cazzaniga L, Pantaleoni C, Milani N (1986) 'Acute hemiplegia in childhood: the neuropsychological prognosis', J. of Pediatric Neurosciences 4, 239-250.
4) Varga-Khadem F, O'Gorman AM and Watters JV (1985) 'Aphasia and handedness in relation to hemispheric side, age at injury and severity of cerebral lesion during childhood', Brain 108, 677-96.
5) Woods BT (1980) 'The restricted effects of right hemisphere lesions after age one: Wechsler Test data', Neuropsychologia 18, 65-70.
6) Woods BT and Carey S (1979) 'Language deficits after apparent clinical recovery fron childhood aphasia', Annals of Neurology 6 (5), 405-409.

SELECTIVE DEFICITS IN LANGUAGE COMPREHENSION FOLLOWING EARLY LEFT AND RIGHT HEMISPHERE DAMAGE

Julie A. Eisele
Cornell University and
Department of Pediatrics
Case Western Reserve University
2101 Adelbert Road
Cleveland, Ohio 44106

ABSTRACT. Twenty-four children with discrete left or right hemisphere damage and 24 healthy controls were tested on their ability to judge the truth of embedded complements in sentences like 'Max remembered that he locked the door' and 'Max remembered to lock the door'. Experimental sentences varied according to the type of matrix verb (know, remember or forget), the presence or absence of syntactic and/or lexical negation, and the syntax of the embedded complement (tensed or untensed). The results of a sentence-question task revealed hemispheric differences in syntactic, semantic and pragmatic language competence. Subjects with left hemisphere damage failed to vary their truth-value judgments according to the syntax of the sentence. In contrast, subjects with right hemisphere damage demonstrated a selective deficit in processing certain types of lexical-semantic and pragmatic information. These results converge with the adult neurolinguistic literature and suggest an early specialization of both hemispheres for certain types of language knowledge.

1. Background

In the adult, the ability to process complex syntactic information appears to be an aspect of language particularly vulnerable to left hemisphere damage. Disorders of comprehension are most evident when a semantic interpretation of words and their underlying grammatical relations requires the ability to map between abstract levels of representation (Caplan and Futter, 1986; Caplan and Hildebrandt, 1988; Grodinsky, 1984; Schwartz, Linebarger, Saffran and Pate, 1987). For example, left hemisphere damage typically impairs the ability to interpret passive and relative clause structures where surface NPs must be 'mapped onto' or coindexed with thematic roles assigned to their original deep structure locations.

In the developing child, left hemisphere damage has also been shown to selectively disrupt the acquisition and subsequent recovery of syntactic knowledge. Adult-like aphasic symptoms in children as

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 225–238.

young as 3 to 4 years of age (Cranberg, Filley, Hart and Alexander, 1987; Van Dongen, Loonen and Van Dongen, 1985; Van Hout, Evrard,and Lyon, 1985) and the presence of residual deficits in syntactic ability following early left hemisphere damage (Aram, Ekleman and Whitaker, 1986; Aram and Ekleman, 1987; Dennis and Whitaker, 1976; Woods and Carey, 1979), has provided evidence for an adult-like neural representation of language at birth. Similar to the adult, residual deficits in syntactic ability in the child typically occur independent of more general cognitive deficits (Aram, 1988; Vargha-Khadem, O'Gorman and Watters, 1985), suggesting a modular representation of language throughout the course of development.

While these results support a continuous left hemisphere dominance for processing complex syntax, a growing body of research has revealed what appears to be a right hemisphere specialization for certain aspects of language knowledge. In particular, right hemisphere damage in the adult has impaired (i) semantic aspects of lexical knowledge (Gardner and Denes, 1973; Gazzaniga and Miller, 1989), (ii) pragmatic aspects of language such as the ability to draw inferences (Brownell, Potter, Bihrle and Gardner, 1986), appreciate metaphors (Winner and Gardner, 1977), verbal ambiguities and jokes (Bihrle, Brownell, Powelson and Gardner, 1984), and (iii) affective components of propositional language and speech (Ross and Mesulam, 1979). These findings are significant in that they provide evidence for an anatomical parcellation of what has been considered 'core' or syntactic language knowledge from certain semantic, pragmatic and cognitive aspects of language.

2. Statement Of The Problem

To date, little research has addressed the extent to which these subcomponents of language knowledge can be dissociated in the developing child. While a number of studies have reported residual deficits in syntactic knowledge following very early left hemisphere injury, few studies have focused on subtle aspects of language knowledge following early right hemisphere injury. It is not known for example, whether selective deficits in semantic or pragmatic aspects of language may be dissociated from syntactic ability following early left or right hemisphere damage.

3. The Present Study

In this paper we report the effects of early left or right hemisphere damage on children's comprehension of complex sentences which involve the integration of syntax as well as semantic and pragmatic language knowledge. Specifically, we test children on their ability to judge the truth-value of embedded complements like those exemplified on Table 1. Comprehension of these sentences requires the ability to map beyond a surface syntax representation to the level of logical form, a level of meaning which incorporates syntax with pragmatic and semantic aspects of logic and general cognition.

Table 1
Example Sentences

Complement Syntax	Truth-Value
Tensed Complements	**Presupposition**
1. Max **knew** [that he locked the door.]	TRUE
2. Max **did not know** [that he locked the door.]	TRUE
3. Max **remembered** [that he locked the door.]	TRUE
4. Max **did not remember** [that he locked the door.]	TRUE
5. Max **forgot** [that he locked the door.]	TRUE
6. Max **did not forget** [that he locked the door.]	TRUE
Untensed Complements	**Implication**
7. Max **remembered** [to lock the door.]	TRUE
8. Max **did not remember** [to lock the door.]	FALSE
9. Max **forgot** [to lock the door.]	FALSE
10. Max **did not forget** [to lock the door.]	TRUE

3.1 EXPERIMENTAL DESIGN

In certain instances, the truth or falsity of a sentence can be computed on the basis of simple assertion. For example determining the truth-value of the sentence 'The door is locked' involves deriving a sentence meaning based on words and their syntactic structure and evaluating this meaning against an immediate context.

In complex sentences like those in 1-6 on Table 1, however, the truth or falsity of the underlined complement is based not on a simple assertion, but rather on certain assumptions shared by both the speaker and the hearer. The truth of the underlined complement 'he locked the door' is **"presupposed"**, not asserted; it is shared background knowledge, which the speaker and the hearer assume to be true. For example in sentence 1, the speaker is 'asserting' the truth of the main clause, that Max knew something, but 'presupposing' the truth of the complement, namely that the door is locked. We can see that the truth of the complement is presupposed as background knowledge, because even when the main clause is syntactically negated as in 2 and 4 or lexically negated as in 5, the speaker/hearer still assumes the embedded complement to be true.

In contrast to sentences with tensed complements, sentences with untensed complements as in 7-10 no longer presuppose the truth of their complements. Rather in these sentences the truth of the complement is **"implied"** by the assertion expressed in the main clause predicate. For example the sentence 'Max did not remember to lock the door' no longer presupposes the fact that the door is locked, but rather implies that the door is unlocked.

Thus we can see from Table 1 that the syntax of the embedded complement is critically involved in determining the possible truth or falsity of the complement. In sentences with tensed complements as in 1-6, the truth of the complement remains constant despite the presence of syntactic or lexical negation. Here the tensed feature of the complement acts as a barrier, restricting the scope of syntactic negation to the matrix clause. In contrast, untensed complements impose no such constraint on the scope of matrix negation. In sentences 7-10 matrix negation has scope over the entire sentence.

Presuppositions as in 1-6 and implications as in 7-10 are influenced by lexical information as well. In sentences with untensed complements, implications of truth or falsity are directly determined by the conventional meanings of words in the main clause predicate. For example the falsity of an infinitive complement may be expressed by the negative implication of 'did not remember to' as in 8 or as in 9 by the semantically negative verb 'forget'

3.2 EXPERIMENTAL METHOD

Using a sentence-question task, subjects were asked to make two truth-value judgments for each of the 10 sentence types exemplified in Table 1. As shown in Table 2, one truth-value judgment pertained the matrix clause and one pertained to the truth of the embedded complement. For example, the sentence 'Max remembered that he locked the door' was

Table 2

Tensed Complements		Untensed Complements	
Example Questions	Correct Responses	Example Questions	Correct Responses
1. Max *knew* that he locked the door.		7. Max *remembered* to lock the door.	
Qm: Did Max know it?	Yes	Qm: Did Max remember?	Yes
Qc: Did he lock the door?	Yes	Qc: Did he lock the door?	Yes/Maybe
2. Max *did not know* that he locked the door.		8. Max *did not remember* to lock the door.	
Qm: Did Max know it?	No	Qm: Did Max remember?	No
Qc: Did he lock the door?	Yes	Qc: Did he lock the door?	No/Maybe
3. Max *remembered* that he locked the door.		9. Max *forgot* to lock the door.	
Qm: Did Max remember?	Yes	Qm: Did Max forget?	Yes
Qc: Did he lock the door?	Yes	Qc: Did he lock the door?	No/Maybe
4. Max *did not remember* that he locked the door.		10. Max **did** *not forget* to lock the door.	
Qm: Did Max remember?	No	Qm: Did Max forget?	No
Qc: Did he lock the door?	Yes	Qc: Did he lock the door?	Yes/Maybe
5. Max *forgot* that he locked the door.			
Qm: Did Max forget?	Yes		
Qc: Did he lock the door?	Yes		
6. Max *did not forget* that he locked the door.			
Qm: Did Max forget?	No		
Qc: Did he lock the door?	Yes		

followed by the matrix question 'Did Max remember' and the complement question 'Did he lock the door'. To each question subjects were permitted to respond 'yes', 'no' or 'maybe'.

For each sentence condition, subjects' responses were scored as correct when both matrix and complement questions were answered correctly. The appropriate 'yes', 'no' and 'maybe' responses are exemplified in Table 2.

The complete set of experimental sentences varied according to (i) the matrix verb ('know', 'remember' or 'forget'), (ii) the presence of syntactic negation and (iii) type of complementation; tensed sentential complements as in 1-6 or untensed infinitives as in 7-10. Two replications for each conditions resulted in a total of 20 sentences and 40 questions equally divided among 2 experimental batteries. For each condition, the order of questions (i.e. matrix or complement first) was systematically varied across the two replication items.

Prior to administration of the experimental batteries subjects underwent a pretraining session designed to ascertain (i) subjects' knowledge of cognitive verbs to be used in the task and (ii) subjects' comprehension of task methodology.

3.3 SUBJECTS

Twenty-four children with discrete left (N=14) or right (N=10) hemisphere injury and 24 normal healthy controls individually matched on age, sex and race served as subjects. See Tables 3 and 4 for a complete description of subject variables. Lesion subjects were selected by Dr. Dorothy Aram on the basis of discrete unilateral lesions as documented by acceptable CT or MRI.

Subjects ranged in age from 4;00 to 17;08 and age at lesion-onset ranged from the pre/perinatal period to 9;08 years. The majority of acquired lesions resulted from CVAs, with one left hemisphere lesion the result of an AVM and one due to meningitis. Prenatal lesions were the result of porencephalic cysts or of unknown etiology. At the time of testing, none of the lesion subjects were considered clinically aphasic and none scored more than one standard deviation below the mean on Full scale IQ.

3.4 INTELLIGENCE LEVELS

In order to compare the general intellectual ability of lesion subjects and their controls, each subject was tested on the Wechsler Vocabulary and Block Design subtests. Subtest scores are shown in Table 3. Planned comparisons between Left and Right lesion subjects and their respective controls revealed no significant group differences for either IQ subtest. (**Lefts**: Vocabulary $t(1,26) = 1.83$; $p<0.079$; Block Design $t(1,26) = 1.42$, $p<0.168$. **Rights**: Vocabulary $t(1,18) = 0.92$; $p<0.367$; Block Design $t(1,18) = 1.58$; $p<0.130$.

Table 3.

Left Lesion and Control Subjects

Subject I.D.	Age	Sex	IQ Vocab.	IQ Block D.	SES
LL01	4;00	F	10	10	3
LC01	4;00	F	17	13	1
LL02	5;11	F	12	11	3
LC02	5;09	F	14	11	1
LL03	8;08	M	9	7	4
LC03	8;06	M	12	10	3
LL04	8;11	M	10	13	3
LC04	9;01	M	12	17	2
LL05	10;02	F	15	15	2
LC05	10;06	F	12	19	1
LL06	10;08	M	17	9	3
LC06	10;10	M	16	10	2
LL07	11;01	M	11	10	2
LC07	10;11	M	11	10	2
LL08	11;03	M	10	13	1
LC08	11;05	M	14	11	1
LL09	11;08	M	11	8	2
LC09	11;08	M	15	12	2
LL10	11;09	M	9	12	1
LC10	11;11	M	13	12	2
LL11	13;03	M	11	5	3
LC11	13;04	M	9	11	1
LL12	14;05	M	16	15	2
LC12	14;06	M	16	12	2
LL13	15;07	F	10	16	2
LC13	15;07	F	10	11	1
LL14	17;03	M	10	13	3
LC14	17;08	M	14	18	1
LL					
Range	4;00-17;03		9-17	5-16	1-4
Mean	11;00		11.5	11.2	2.43
LC					
Range	4;00-17;08		9-17	10-19	1-3
Mean	11;01		13.2	12.6	1.57

Right Lesion and Control Subjects

Subject I.D.	Age	Sex	IQ Vocab.	IQ Block D.	SES
RL01	5;04	F	11	10	4
RC01	5;09	F	13	7	3
RL02	8;01	M	8	10	2
RC02	7;11	M	15	12	1
RL03	9;06	M	16	9	1
RC03	9;08	M	14	14	1
RL04	10;04	F	6	9	3
RC04	10;07	F	10	9	2
RL05	10;04	F	15	11	1
RC05	10;09	F	14	12	1
RL06	10;06	M	10	11	3
RC06	10;10	M	9	8	3
RL07	11;10	F	8	9	3
RC07	11;10	F	11	12	1
RL08	11;11	M	7	9	5
RC08	12;00	M	11	13	2
RL09	11;11	M	12	10	3
RC09	12;03	M	11	12	2
RL10	12;05	F	15	10	3
RC10	12;07	F	11	12	2
RL					
Range	5;04-12;05		6-16	9-11	1-5
Mean	10;03		10.8	9.8	3.00
RC					
Range	5;09-12;07		9-15	7-14	1-3
Mean	10;05		11.9	11.1	1.60

Table 4.

Left Hemisphere Lesion Subjects

Subject I.D.	*Test Age*	*Lesion Age*	*Post-Lesion Age*	*Etiology*[1]	*Lesion Location*[2]
LL01	4;00	PN	4;00	PN	Subcort.-BG
LL02	5;11	PN	5;11	PN	Pre-BG
LL03	8;08	PN	8;08	PN	Subcort.+BG
LL04	8;11	8 hrs.	8;11	CVA	Retro+BG
LL05	10;02	7;05	2;09	CVA	Pre+BG
LL06	10;08	0;04	10;04	AVM	Pre+Retro-BG
LL07	11;01	4;03	6;10	CVA	Subcort.+BG
LL08	11;03	PN	11;03	PN	Retro-BG
LL09	11;08	0;01	11;07	CVA	Pre-BG
LL10	11;09	7;07	4;02	CVA	Pre+BG
LL11	13;03	1;11	11;04	CVA	Retro-BG
LL12	14;05	8;00	6;05	CVA	Retro-BG
LL13	15;07	7;05	8;02	CVA	Subcort.+BG
LL14	17;03	PN	17;03	PN	Retro-BG
Mean	11;00				

Right Hemisphere Lesion Subjects

Subject I.D.	*Test Age*	*Lesion Age*	*Post-Lesion Age*	*Etiology*[1]	*Lesion Location*[2]
RL01	5;04	PN	5;04	PN	Subcort.-BG
RL02	8;01	4;06	3;07	CVA	Pre+Retro+BG
RL03	9;06	1;04	8;02	CVA	Pre+Retro-BG
RL04	10;04	0;06	9;10	CVA	Pre-BG
RL05	10;04	PN	10;04	PN	Subcort.-BG
RL06	10;06	5;02	5;04	CVA	Subcort.+BG
RL07	11;10	PN	11;10	PN	Subcort.-BG
RL08	11;11	3;04	8;05	CVA	Pre+Retro-BG
RL09	11;11	9;08	2;02	CVA	Subcort.+BG
RL10	12;05	2;07	9;10	CVA	Subcort.+BG
Mean	10;03				

1.
CVA= Cerebrovascular Accident
PN= Prenatal Injury
AVM= Arteriovenus Malformation

2.
BG= Basal Ganglia
Pre/Retro= Pre/Retro Rolandic Fissure
Subcort= Subcortical

3.5 RESULTS

Correct responses were then analyzed according to two separate factorial analyses: (i) a 2 (Matrix Verb; Remember/ Forget) by 2 (+/- Syntactic Negation) by 2 (Complement Type) factorial design and (ii) a 3 (Matrix Verb; Know/ Remember/Forget) by 2 (+/- Syntactic Negation) factorial design, each with repeated measures on Injury. Overall, both left and right lesion subjects performed significantly below their controls on both factorial analyses. However subsequent analyses revealed an interaction between hemispheric injury and each of the experimental factors we tested.

Concentrating on the verbs 'know' and 'remember', we report first the results for sentences with tensed complements followed by the results for sentences with untensed complements. We report last the results for the verb 'forget'.

3.6 SYNTACTIC NEGATION: TENSED COMPLEMENTS (KNOW/REMEMBER)

The results for sentences with tensed complements are shown in Figures 1a and 1b. As Figure 1a shows, LL subjects' performance was directly determined by the presence of syntactic negation. LL subjects correctly presupposed the truth of a tensed complement in affirmative sentences like 1 and 3, yet failed to do so when the matrix verb was syntactically negated as in sentences like 2 and 4. The predominant error strategy for LL subjects on sentences like 'Max did not remember that he locked the door' was to drop negation from the matrix verb or incorrectly negate the embedded complement.

In contrast to LL subjects, RL subjects performance on sentences with tensed complements was roughly equal to that of their controls. As figure 1b demonstrates, RL subjects did not differ significantly from their controls on sentences containing the matrix verb 'know' or 'remember' in either the affirmative or syntactically negative condition.

3.7 SYNTACTIC NEGATION: UNTENSED COMPLEMENTS (KNOW/REMEMBER)

The results for sentences with untensed complements are shown in Figures 2a and 2b. We can see from Figure 2a that LL subjects' performance on untensed complements was again directly related to the presence of syntactic negation in the matrix clause. That is, LL subjects were successful in making truth-value judgements for sentences like 'Max remembered to lock the door' but unsuccessful on sentences like 'Max did not remember to lock the door'. For both complement types then, LL subjects were particularly deficient in inferring truth of sentences with matrix negation.

As shown in Figure 2b, RL subjects also differed from their controls on untensed complements. Like LL subjects, they performed significantly below their controls when the matrix verb 'remember' was syntactically negated. In contrast to their success on tensed complements like 'Max did not remember that he locked the door', RL subjects were unsuccessful on untensed complements like 'Max did not

Tensed Complements

Figure 1a

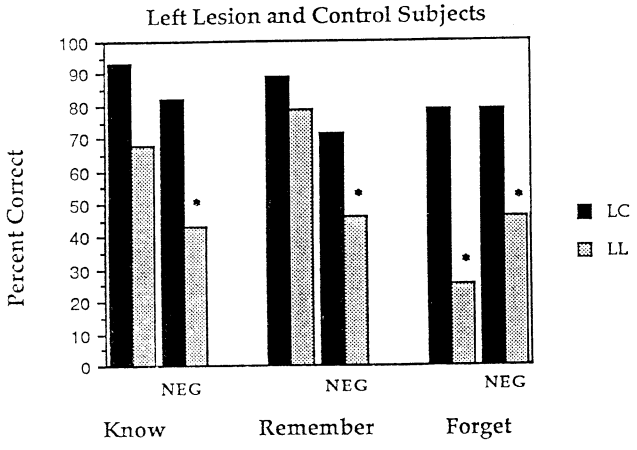
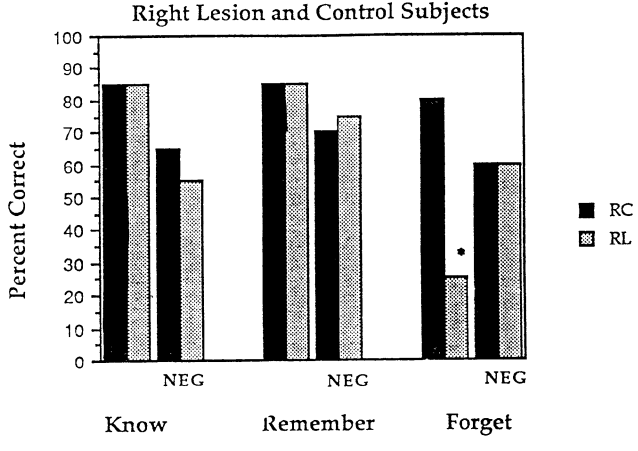


Figure 1b.



* Indicates significant difference between lesion and control group.

Untensed Complements

Figure 2a.

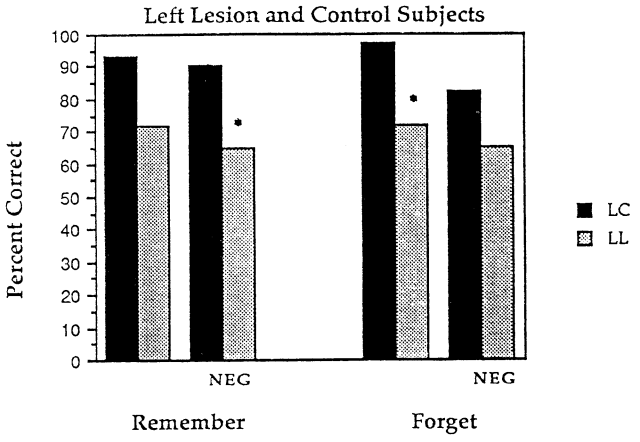
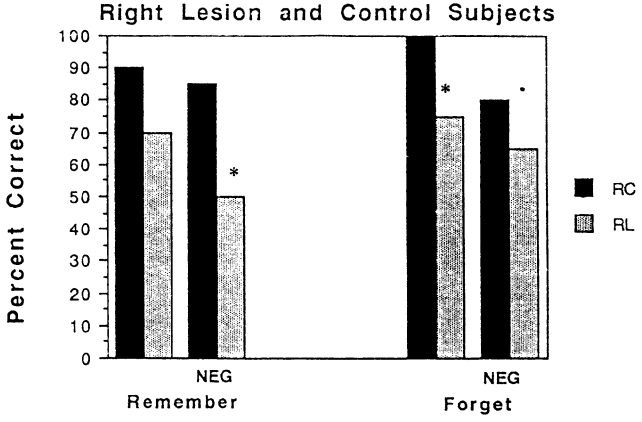


Figure 2b.



* Indicates significant difference between lesion and control group.

remember to look the door'.

3.8 LEXICAL NEGATION (FORGET): TENSED AND UNTENSED COMPLEMENTS

We turn now to a discussion of subjects' performance on the verb 'forget'. As Figures 1a and 1b show, for tensed complements, both groups performed poorer on the semantically negative verb 'forget' than they did on either the verb 'know' or 'remember'. Both groups also found the verb 'forgot' more difficult that the double negative verb phrase 'did not forget'. LL subjects performed significantly below their controls on tensed complements with the matrix verb 'forget' in both syntactically affirmative and negative conditions. RL subjects performed significantly below their controls only in the syntactically affirmative 'forget that' condition. In fact, this was the only tensed condition that discriminated RL subjects from their controls.

Looking at Figures 2a and 2b we can see that for untensed complements, performance on the verb 'forget' was not significantly poorer than performance on the verb 'remember'. However, both left and right lesion subjects performed significantly below their controls on the semantically negative 'forgot to' condition.

3.9 DISCUSSION

In our discussion above we demonstrated that the ability to infer the truth-value of complements like those in 1-10 involves not only pragmatic but syntactic knowledge as well. In particular, these inferences require the ability to represent the abstract syntactic marker of +/- tense and its constraint on negation scope.

Our results have shown that LL subjects can access pragmatic language knowledge and correctly infer presuppositions and implications in syntactically affirmative conditions. Critically however, LL subjects err in their truth-value judgments on sentences which require the ability to use the abstract feature of tense in their computation of negation scope. These results suggest that LL subjects have retained a certain degree of pragmatic language knowledge, yet are unable to integrate this pragmatic knowledge with syntactic constraints of the sentence. Similar to the adult then, disorders of comprehension in the child might also reflect an inability to map between abstract levels of language representation following left hemisphere damage.

We have also shown that left and right lesions subjects' ability to determine the truth-value of embedded complements varies according to the syntax of the complement. LL subjects are impaired on both tensed and untensed complements, whereas RL subjects experience difficulty only on untensed complements. As we discussed above, implications of truth as in 7-10 are determined largely by the conventional meaning of individual words expressed in the matrix and subordinate clause. We hypothesize that RL subjects' selective impairment on untensed complements may be due in part to a deficit in accessing semantic features of the lexicon. This hypothesis would

also explain RL subjects' selective deficit in processing the semantically negative verb 'forget'. Alternatively, RL subjects' success in determining the truth of presuppositions but not implications may reflect a partial deficit in pragmatic language knowledge.

Finally, we have shown that regardless of complement syntax, the semantically negative verb 'forget' significantly impairs both left and right lesion subjects' performance. Surprisingly, for left and right lesion subjects, performance was facilitated when the verb 'forget' was marked by syntactic negation. This result clearly demonstrates that negation as a property of the lexicon can be dissociated from syntactic negation. This result also demonstrates that subjects are not simply unable to process negation in general. Subjects perform better on the double negative 'did not forget' than they do when the verb 'forget' occurs on its own.

3.10 CONCLUSION

In summary, these results suggest that a complete acquisition of language depends on the normal functioning of both hemispheres throughout the course of development. Although the precise nature of neurolinguistic deficits in children remains undetermined, our results provide evidence for an adult-like pattern of language deficits following early left and right hemisphere damage.

3.11 ACKNOWLEDGEMENTS

I would like to thank the children and parents who participated in this study, Letitia Gillespie, Loretta Catalano, and Karen Nevin for their assistance in data collection and Janet Chien for her creative graphic skills. I thank Barbara Lust for her invaluable help and support during the creation of this manuscript and Dorothy Aram for graciously sharing her special population of children with me. This research was supported by an NIMH Predoctoral INRSA award (F31 MH09934-01) and a Cornell University College of Human Ecology Ph.D. thesis grant.

3.12 REFERENCES

Aram, D.M. (1988). Language Sequelae of Unilateral Brain Lesions in Children. In F. Plum (Ed.) Language, Communication and the Brain, Raven Press, New York.

Aram, D.M. and Ekelman, B.L. (1987). Unilateral brain lesions in childhood: Performance on the Revised Token Test. Brain and Language, 32, 137-158.

Aram, D.M., Ekelman, B.L., and Whitaker, H.A. (1986). Spoken syntax in children with acquired unilateral hemisphere lesions. Brain and Language, 31, 61-87.

Bihrle, A., Brownell, H., Powelson, J. and Gardner, H. (1984). Sensitivity to humorous and non-humorous narrative in organic patients: A special role for the right hemisphere. Paper presented at the Academy of Aphasia, Los Angeles, CA.

Brownell, H., Potter, H., A., and Gardner, H. (1986) Inference deficits in Right Brain-damaged Patients. Brain and Language, 27, 310-321.

Caplan, D. and Futter, C. (1986). Assignment of thematic roles to nouns in sentence comprehension by an agrammatic patient. Brain and Language, 27, 117-134.

Caplan, D. and Hildebrandt, N. (1988). Disorders of Syntactic Comprehension, MIT Press, Cambridge, Massachusetts.

Cranberg, L., Filley, C. Hart, E. and Alexander, M. (1987). Acquired Aphasia in Childhood: Clinical and CT investigations. Neurology, 37, 1165-1172.

Dennis, M. and Whitaker, H. (1976) Language acquisition following hemidecortication: Linguistic superiority of the left over the right hemisphere. Brain and Language, 3, 404-433.

Gardner, H. and Denes, G. (1973). Connotative judgments by aphasic patients on a pictorial adaptation of the semantic differential. Cortex, 9, 183-196.

Gazzaniga, M, and Miller, G. (1989). The recognition of antonymy by a language-enriched Right Hemisphere. Journal of Cognitive Neuroscience, 1 (2), 187-193.

Grodinsky, Y. (1984). The syntactic characterization of agrammatism. Cognition, 16, 99-120.

Ross, E. and Mesulam, M-M. (1979). Dominant Language Functions of the Right Hemisphere? Prosody and Emotional Gesturing. Archives of Neurology, 36, 144-148.

Schwartz, M., Linebarger, M., Saffran, E., and Pate, D. (1987). Syntactic transparency and sentence interpretation in aphasia. Language and Cognitive Processes, 2, 85-113.

van Dongen, H.R., Loonen, M.C.B., and van Dongen, K.J. (1985). Anatomical basis for acquired fluent aphasia in children. Annals of Neurology, 17, 306-309.

van Hout, A., Evrard, P., and Lyon, G. (1985). On the positive semiology of acquired aphasia in children. Developmental Medicine and Child Neurology, 27, 231-241.

Vargha-Khadem, F., O'Gorman, A.M. and Watters, G.V. (1985). Aphasia and handedness in relation to hemispheric side, age at injury and severity of cerebral lesion during childhood. Brain, 108, 677-696.

Winner, E. and Gardner, H. (1977). The comprehension of metaphors in brain-damaged patients. Brain, 100, 719-727.

Woods, B.T. and Carey, S. (1979). Language deficits after apparent clinical recovery from childhood aphasia. Annals of Neurology, 6(5), 405-409.

THE ROLE OF CENTRAL ACTIVATION (AROUSAL) IN THE RECOVERY OF COGNITIVE FUNCTIONS AFTER CEREBRAL INJURY IN CHILDREN

Hallgrim Kløve
Dept. of Biological and Medical Psychology
University of Bergen
Aarstadveien 21
N-5009 Bergen, Norway

ABSTRACT
The presentation will discuss theoretical and clinical aspects of hypo-arousal after cerebral injury. In addition to the structural changes, it is thought that changes in the neurotransmitter system play an important role. The empirical evidence and cases where pharmacological treatment as adjuvant in rehabilitation has been used will be presented.

In 1937 Charles Bradley published an article entitled The behavior of children receiving Benzedrine (Bradly 1937). This is the first report of the effects of central stimulants on behavior disorders in children. Contrary to common beliefs, Bradley conceived his idea of giving central stimulants to children on the basis of a number of reports describing the effect on the mood and other psychological reactions in adults. Most of these studies appeared in Lancet and some American journals during the 1930's. While the early papers did not specifically mention neurological considerations as a basis for using central stimulants, in the papers which have appeared more recently, the focus has been on the effects of central stimulants on stokes and in particular on post-traumatic conditions. In 1976 Lipper and Tuckman published a paper entitled Treatment of chronic post-traumatic organic brain syndrome with dextro-amphetamine: First reported case (Lipper & Tuckman 1976). Thus, while the earliest reports on the action of central stimulants on human behavior seem to be related to what at that time was considered to be basically psychiatric syndromes, more recent publications have discussed observations on the effects of neurologic sequelae. In this connection it is pertinent to mention the editorial in the first issue

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 239–243.

of the new rehabilitation journal entitled Brain Injury (Hayes, Stonnington & Lyeth, 1987). This editorial emphasized in particular the need to explore the psychopharmacological approach as an adjunct to rehabilitation of brain injury.

It is also interesting to look briefly at some highlights of Bradley's 1937 article. The article does, of course, reflect the state of the art in regard to the evaluation of drug effects on behavior at that time. Technically, we may have advanced considerably since 1937, but in reality, the new concerns related to clinical drug research and patients right make it seem that some aspects of drug research has not advanced very much during the last 50 years. While the use of central stimulants in children from 1937 and until fairly recently has been focused on alleviating symptoms of suspected congenital or developmental disorders, the observation of the behavioral modification properties of central stimulants in these children have led to speculations that central stimulants may also have a similar effect on aquired brain injury in children as well as in adults. This interest has been based on a number of clinical observations and an increasing number of experimental studies on animals. A large number of studies have demonstrated the beneficial effects on both behavior and cognitive performance in children with learning disabilities and hyperactive behavior (Barkley, 1977). The issue now is not if central stimulants are effective, but a more precice indications of their use.

Since in many patients, the target behavior improve with administration of centrally stimulating drugs, the drug is most likely acting upon a hypoactive central nervous system. The idea of a "paradoxical effect" of the centrally stimulating drugs has in recent research not gained much support. It appears that the pharmacological effect is related to a central hypoarousal state which is alliviated by use of a stimulant drug. Thus, it has been demonstrated that psychophysiolocal indicators of hypoarousal has been normalized as an effect of administration of centrally stimulating drugs (Kløve & Hole, 1979).

A number of studies during the last 10-15 years have adressed the observation that the improvement of cognitive functions and behavior using central stimulants is not limited to children with developmental problems, but it also seems to apply to

aquired brain injury both in children and adults (Kløve, 1987).

Bradley (1937) mentions as the most dominant change, the improvements in school performance. This occured in approximately half of the children. Bradley says: "To see a single daily dose of Benzedrine produce a greater improvement in school performance than the combined efforts of a capable staff working in a most favourable setting, this would be all but demoralizing to the teachers, had not the improvement been so gratifying from a practical viewpoint". - - "Possibly the most striking change in behavior during the week of Benzedrine therapy occured in school activities of many of the patients. 14 children responded in a spectacular fashion. There appeared a definite "drive" to accomplish as much as possible during the school period, and often to spend extra time completing additional work. Speed of comprehension and accuracy of performance were increased in most cases. - - It appeared promptly the first day Benzedrine was given and disappeared on the first day it was discontinued."

The question is to which extent these observations, relevant for children with supposed developmental problems, also are relevant for children with aquired brain injury. It is pertinent to review briefly a couple of animal studies. Feeney, Gonzales & Law (1982) reported that rats subjected to unilateral ablation of motor cortex, displayed transient contra lateral pareses. An immediate and enduring acceleration of recovery was produced by a single doze of dextroamphetamine given 24 hours after the injury. This effect was blocked by Haloperidol or by restraining the animals. There are a few other studies which report comparable results. Gage and Olton (1976) and Marotta, Logan, Potegal, Glusman & Gardner (1977) gave rats single doze of dextroamphetamine 24 after septal lesions. Recovery from hyperemotionality was accelerated in these animals. That amphetamine fascilities recovery from such dissimilar lesions (septum and motor cortex) and from such dissimilar behavior as reduction of emotionality and improvement of locomotion supports an hypothesis that catecholamines play a non-specific role in recovery from brain injury. This finding may have important implications for rehabilitation in patients with aquired brain damage due to stroke or trauma. Stimulation of catecholaminergic systems in conjunction with rehabilitation efforts may facilitate the often slow and frustrating recovery from such injuries.

Haloperidol and perhaps other drugs commonly used for their antipsychotic effects may be contra-indicated during recovery from brain injury because they block the catecholamine reseptors and thus retard the recovery of functions. Similar findings have been reported in human cerebral spinal fluid following cerebral infarctions as reported by Myers, Stoica, Pascu, Shimaza & Hartman (1973). If catecholamine levels contribute to the behavioral syndrom seen after cerebral injuries, it should be possible to revers some of the deficits by pharmacological manipulation of the catecholaminergic system. It was this idea that was the basis of Feeney's and associates' study. Amphetamine and Haloperidol which have potent oposing action on catecholamines and neurone activity were used to test this hypothesis. Motivated by the clinical effect of central stimulants in hyperactive children and the studies just cited, we have administered central stimulants to a number of selected patients.

In one striking case a 40 year old man was admitted to the emergency room of the University Hospital after a bicycle accident. A craniotomy was performed during which a subdural hematoma was evacuated in a very large contused portion of the left temporal lobe was removed. As the patient recovered from the accute effects of the injury, his behavior was disorganized. He was restless and his language was characterized by serious aphasic symptoms. He was given a small amount of Ritalin, and his behavior improved dramatically. His language functions improved immediately. It was possible to communicate with him, give him instructions and his expressive language was also greatly improved. The observation in this patient prompted us to study the issue in children. We have so far observed definite positive effects on language and behavior on 4 post-traumatic children all pre-schoolers. The effect of Ritalin is remarkable in that not only language functions but general behavior and non-language cognitive functions also improved.

REFERENCES

1. Barkley, R.A. (1977). A review of stimulant drug research with hyperactive children. Journal of Child Psychology and Psychiatry, 18, 137-165.
2. Bradley, C. (1937). The behavior of children receivingbenzedrine. American Journal of Psychiatry, 94, 577-585.
3. Feeney, D., Gonzales, A. & Law, W.A. (1982) Amphetamine, Haloperidol and experience interact to affect rate of recovery after motor cortex injury. Science, 217, 855-857.
4. Gage, F.H. & Olton, D.S. (1976). L-Dopa reduces hyperactivity induced by septal lesions in rats. Behavioral Biology, 17, 213-218.
5. Hayes, R.L., Stonnington, H.H. & Lyeth, B.G. (1987). Editorial: Pharmacological treatment of headinjury - a new challenge. Brain Injury, 1, 1-2.
6. Kløve, H. & Hole, K. (1979). The hyperkinetic syndrome: Criteria for diagnoses. In R.L.Trites (Ed.) Hyperactivity in children (pp. 121-136). Baltimore: University Park Press.
7. Kløve, H. (1987). Activation, Arousal and Neuropsychological Rehabilitation, 8, 297-309.
8. Lipper, S. & Tuchman, M.M. (1976). Treatment of chronic post-traumatic organic brain syndrome with dextroamphetamine: First reported case. Journal of Nervous and Mental Disease, 162, 366-371.
9. Marotta, R.F., Logan, N., Potegal, M., Glusman, M. & Gardner, E.L. (1977). Dopamine agonists induce recovery from surgically induced septal rage. Nature, 269, 513-515.
10. Meyers, J.S., Stoica, E., Pascu, I., Shimaza, K. & Hartman, A. (1973). Catecholamine concentration in CSF and plasma of patients with cerebral infarction and haemorrage. Brain, 96, 277-288.

VIII - LANDAU AND KLEFFNER SYNDROME

THE CONCEPTION AND EMBARRASSING BIRTH OF AN EPONYM

William M. Landau, M.D.
Department of Neurology and Neurological Surgery
Washington University School of Medicine
St. Louis, MO 63110
USA

Many years ago I received a courteous and deferential letter from a British pediatric neurologist who explained that there had been controversy in his hospital regarding interpretation of a test of motor coordination in infants called the Landau reflex, and would I settle the question? Although I was tempted to assume the role of authoritarian Herr Professor, I responded demurely that I must plead innocent because this reflex was invented and reported by a Berlin pediatrician the year before I was born. I have been embarrassed by that eponym ever since.

Frank Kleffner and I are, of course, pleased at the attention our little syndrome has reiceived for more than three decades. But as a presumptive expert in this field, I must confess humiliation at having learned so little about it in over half of my lifetime. Until this meeting, anyway.

In 1952 we moved from St. Louis to the National Institutes of Health in Bethesda, near Washington. There I worked in Marshall's laboratory of neuropsysiology and Kety's laboratory of cerebral circulation and metabolism. Marshall offered me an opportunity to remain permanently in basic neuroscience at NIH. But I recognized that I needed to be involved in neuroscience in relation to the clinical challenge and we returned to Washington University in 1954.

Although there are now many more big buildings than there were in 1954, then and now it was only a brief walk from McMillan Hospital Building where the Neurology Department is based to the Central Institute for the Deaf Residential School Building. I already knew Hallowell Davis, the great neurophysiologist who was Director of Research at the Central Institute. Davis is still very much alive at age 94. At that time he was a leader in the electrophysiology of hearing and already one of the fathers of clinical electroencephalography from his earlier career in Boston. His predecessor,

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 247–251.

who established the research division of the Central Institute, Rafel Lorente de No, a former student of Ramon y Cajal. Lorente de No had moved on the Rockefeller Institute in New York during the thirties.

I had the idea that there must be an interesting variety of neurological case material in this specialized setting. Richard Silverman, an audiologist, educator, and Director of the Institute was pleased with my interest and introduced me to the enthusiastic teaching faculty. The majority of the residencial students were afflicted with clearcut peripheral auditory deficiency. But a large minority were taught in a program directed by Frank Kleffner, called the aphasic division. This division was established by Mildred MgGinnis, a great teacher who simply considered that children whose language development was seriously impaired in relation to their general intelligence should be labelled aphasic. None were deaf, but later studies indicated that many with retarded speech were also afflicted with high frequency hearing impairment. Kleffner, Robert Goldstein, an audiologist, and I did surveys of general neurological examinations, EEGs, audiometry, and vestibular responses in the whole school population of 188 (Goldstein, Landau and Kleffner, 1958; Rosenblut, Goldstein and Landau, 1960; Goldstein, Landau and Kleffner, 1960).

One youngster whom we surveyed had congenital aphasia and congenital heart disease. He died unexpectedly at age 10 with complications of mumps. At autopsy we found bilateral old infarctions in the Sylvian regions with retrograde degeneration of the medial geniculate nuclei (Landau, Goldstein and Kleffner, 1960). Nevertheless, he had made very significant progress in both understanding and production of oral speech.

When I first presented to Kleffner the idea of a neurological survey of his students, the first thing he mentioned was the coherent and successful group of children with a history of language acquisition and then loss, along with benign convulsive disorder. He was quite surprised that clinical neurologists dind't know about those children. Of the six children in our original 1956 report (Landau and Kleffner, 1957), four were in residence at CID, and one, Case 2, was the elder sibling of Case 1.

As you know, over the next decade or more, new cases were identified in many centers. It seemed to take a while for Worster-Drought to discover that we had described the condition first. By 1978 it became clear that a major deficit of knowledge was what happens to these children when they grew up. I enlisted a young pedriatric neurologist, John Mantovani, into the follow-up study. To my embarrassment, our first discovery was that my original manuscript notes had been lost. Then with detective work worthy of Sherlock Holmes, we discovered all of the original protocols and school records, and were able to trace even the female

patients who had new married names. We also added three more patients with less than two decade follow-up (Mantovani and Landau, 1980).

The first of these, our case 7, was born in November 1954. Early development was normal in every way. At age 5 she was using complex sentences and her speech was considered normal by parents and teachers. At age 5 years, she was noted to be less responsive to verbal instruction and began to stutter. Within several months this progressed to almost total unresponsiveness to oral communication, and she could not speak intelligibly. Audiologic evaluation was normal. She was hospitalized at St. Louis Children's Hospital in August 1961.

She had no spontaneous speech and only occasional single-word responses to questions. She followed oral commands poorly but could imitate sounds with proper inflection. Except for moderately increased motor activity, examination was normal. Skull films and spinal fluid were normal. The EEG was markedly abnormal with bitemporal paroxsymal activity. She was treated with phenytoin, phenobarbital and paramethadione without significant improvement in language. In the next few months she became more hyperactive and impulsive and had frequent temper tantrums. She enrolled at CID and attended classes there on a full time basis for the next two years. Dr. Kleffner made sound motion pictures of her examinations in October 1961 and again in October 1962. These were transposed to VRC/TV tapes for display now.

In our first follow-up examination when she was 24, she was a right-handed second year college student majoring in liberal arts and doing particulary well in english and public speaking. Her only complaint was a slight problem with speech discrimination in a noisy background. Neurologic examination was normal. Her mother provided a recent photograph of the 35 year old mother with her two children. She continues to live a very normal life and her children have no neurological afflictions.

We all know that the most important ego satisfaction in science occurs when early intelligence proves to be correct with the tests of time and other scientific workers. Kleffnerr and I can only take pride in a reciprocal circumstance. We didn't understand the etiology or mechanism of the syndrome in 1956, and we have consistently sustained our ignorance for over three decades.

We did point out that dominant hemisphere focal paroxysmal activity, which might have been reasonably anticipated, was not characteristics of these patients. But the concept that language is normally organized only within the dominant hemisphere is a function only for our own simple minded inference from destructive focal brain lesions. In fact, the focal lesion truly identifies only the vulnerable site for given function. If one takes a larger view that language is, in the terms of Hughlings

Jackson, the most complex, most voluntary, least automatic of cerebral functions, then it may not be so surprising that generalized cortical dysfunction,too many lines busy with seizure, is associated with conspicuous deficit of highest order communication capacities. In recent years several authors, including some here, have emphasized that many of these patients, in addition to the language defect, have other important behavioral disturbances.

In regard to etiology, there is no necessary reason to believe that there is only one. But uniformity from case to case of the often self-limited syndrome does support the faith that when the evidence finally comes in, most cases will have related etiologies. My own guess is that a slow virus and/or autoimmune vasculopathy or encephalopathy will be identified.

Because the syndrome is so rare, even in major centers, it follows that we shall never be able to move beyond our present stupid level of case report speculation until we develop world class and world size collaborative case studies of both etiology and management. I think that such randomized studies are both ethical and necessary.

My old partner and I will be most gratified if this meeting gives rise to a sustaining international group of scientists who will engage and maintain organized efforts to understand and treat this condition successfully. The present generation knows that my generation's Schilder's disease is really adrenoleukodystrophy. Similarly we shall be especially pleased if our names can move into history as new knowledge dictates a label directly related to pathogenesis and pathopsysiology.

REFERENCES

Goldstein R, Landau W.M. and Kleffner F.R.: Neurological asessment of some deaf and aphasic children. Ann Otol Rhinol Laryngol 67: 468-479, 1958.

Goldstein R, Landau W.M. and Kleffner F.R.: Neurological observations on a population of deaf and aphasic children. Ann Otol Rhinol Laryngol 69: 756-767, 1960.

Landau W.M., Goldstein R. and Kleffner F.R.: Congenital aphasia: a clinicopathologic study. Neurolgy 10: 915-921, 1960.

Landau W.M. and Kleffner F.R.: Syndrome of acquired aphasia with convulsive disorder in children. Neurology 7: 523-530, 1957.

Mantovani J.F. and Landau W.M.: Acquired aphasia with convulsive disorder: course and prognosis. Neurolgy, 30: 524-529, 1980.

Rosenblut, B., Goldstein R. and Landau W.M.: Vestibular responses of some deaf and aphasic children. Ann Otol Rhinol Laryngol 69: 747-755, 1960.

THE LANDAU-KLEFFNER SYNDROME : DIAGNOSTIC CONSIDERATIONS

H.R. VAN DONGEN
Division of Neurology
Erasmus University Rotterdam
Room EE 2287
PO Box 1738
3000 DR Rotterdam
The Netherlands

E. DE WIJNGAERT
Royal Institute for Children
with Communication Disorders
Hasselt, Belgium

M.J. WENNEKES
De Wever Hospital
Heerlen, The Netherlands.

ABSTRACT. As an exact description of the early stage of the Landau-Kleffner Syndrome (LKS) is lacking, the present research has been undertaken in order to clarify this issue by studying data about sequence, duration and simultaneous appearance of the three characteristics of LKS : aphasia, epileptic manifestations and EEG-disturbances. Our results demonstrate a great variety in sequence and duration of the divers characteristics of LKS, indicating many pitfalls in diagnosing. The speech of the greater part of the children improved incompletely, while epileptic fits and EEG-abnormalities frequently disappeared in an earlier stage. Persistent severe language disorders can remain in failing to control clinical seizures and to normalize the EEG. The relation between seizure disorder (and EEG-abnormalities) and aphasia is not evident.

Introduction.

In 1957 Landau and Kleffner described in 6 previously normal children a language deficit associated with epileptic phenomenona : "The Syndrome of Acquired Aphasia with Convulsive Disorders". Later on this syndrome is frequently labeled as "The Syndrome of Landau-Kleffner" (LKS).
In the original study the syndrome has been mainly characterised by :
- aphasia : after passing the normal speech-milestones the children initially developed receptive language disturbances (word-deafness), later on followed by expressive difficulties.

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 253–261.

- clinical epileptic manifestations : "the children had a variety of convulsive manifestations : grand mal, partial, petit mal, and major clinic seizures."
- electro-encephalographic disturbances : "usually bilateral, often more prominent in the temporal lobes."
- With exception of aphasia, clinical examination did not demonstrate neurological abnormalities. The same applied to the laboratory investigations and skull X-rays.

Behavior problems were only observed in one child. Follow-up studies of these children at adult age did not reveal an obviously intellectual deterioration. (Mantovani & Landau, 1980).
Although the greater part of the children has been described before the introduction of imaging techniques as CT-scan and NMR, structural abnormalities are at present very rarely demonstrated. (Otero et al.,1989 ; Rapin, 1989). In addition to functional EEG-disturbances, Positron Emission Tomography (PET) showed " a relative hypometabolism limited to the left temporal region" (Maquet et al., 1988).

The above mentioned 3 characteristics are considered as obligatory upon diagnosing the Landau-Kleffner syndrome.

The syndrome remains ill described, however, concerning the sequence and duration of simultaneous (overlapping) occurrence of the main characteristics.

In the original study 2 cases are described in which epileptic fits were the first sign. Dugas et al. (1982) described 12 children: 5 of them started with epileptic abnormalities, 2 of them with aphasia. Evidently most of the children with epileptic manifestations - associated with or without EEG abnormalities - never become aphasic.
In the absence of this simultaneous occurrence, the diagnosis of LKS will remain doubtfully for a long time. For example in case 5 of the original study, the epileptic attacks appeared 2 years after the onset of the aphasia.

As an exact description of the early stage of the syndrome is lacking, the present research has been undertaken in order to clarify this issue by studying data about sequence, duration and simultaneous appearance of the characteristics.

Patients and methods.

Eight girls and three boys fulfilled the 3 characteristics as mentioned in the introduction. They were referred to the University Hospital Dijkzigt (Rotterdam ,The Netherlands) and the University Hospital Gasthuisberg (Louvain, Belgium). For special education, the Belgian children were admitted to the Royal Institute for Children with Communication Disorders. Table 1 shows individual data of these children. To all children a (non-verbal) intelligence test was at same point administered. The intelligence-quotients (IQ's) ranged from 85 - 125 (mean IQ = 100,5). Only one child (case 11) is left-handed.

The records of each child were reviewed. The following data were included :
- Aphasia : onset, severity, duration and course.
- Clinical epileptic manifestations : onset and duration.
- EEG disturbances : onset and duration.

The greater part of the children presented different types of clinical seizures. The variability of medication was striking. Therefore, the type of seizures and the drug treatment are not extensively described.

APHASIA SEVERITY RATING SCALE (0 - 5).

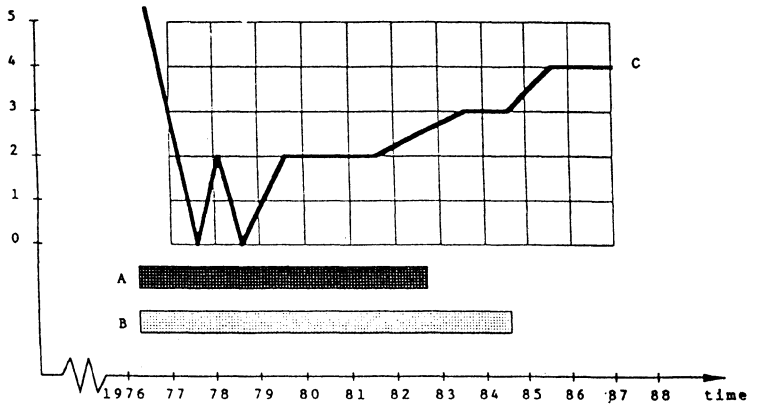


FIGURE 1 : COURSE OF APHASIA (C) AND DURATION OF CLINICAL EPILEPTIC MANIFESTATIONS (A) AND EEG - ABNORMALITIES (B).

Figure 1 illustrates the processing of this information : the course of aphasia (C) according the Aphasia Severity Rating Scale of Goodglass and Kaplan (see addendum), the duration of the clinical seizures (A) respectively the EEG abnormalities (B).

TABLE 1 : SUMMARY OF NEUROLOGICAL DATA AT FOLLOW UP (1987)

		ONSET		COURSE						FOLLOW UP (years)
PATIENT NO	SEX F/M	AGE	OCCURRENCE AND DURATION (m) OF FIRST SIGN	APHASIA		EPILEPSY		E.E.G.		
				COURSE	DURATION	COURSE	DURATION	PRESENSE & DURATION of ABNORMALITIES		
1.	F	3	APHASIA 3y3m	IMPROVING 0 - 2	9y	RECOVERED	2m	Normalized	09m	9y
2.	F	4	APHASIA 6m	FLUCTUATING 0 - 4	7y	RECOVERED	12m	Normalized	2y03m	8y
3.	M	4	APHASIA + EPILEPSY	IMPROVING 1 - 2	8y10m	IMPROVING	6y05m	Abnormal	9y	8y
4.	M	4	APHASIA 1y2m	WORSENING 2 - 0	7y	WORSENING	2y06m	Abnormal	6y	6y
5.	F	4	APHASIA 3m	IMPROVING 2 - 3	3y	RECOVERED	3m	Normalized	2y10m	4y
6.	F	4	EPILEPSY 1y4m	FLUCTUATING 0 - 4	2y06m	RECOVERED	2y09m	Normalized	2y	3y
7.	F	4	EPILEPSY 2y	FLUCTUATING 0 - 3	8y08m	RECOVERED	3y05m	Normalized	4y01m	10y
8.	F	5	APHASIA 5m	IMPROVING 0 - 4	17y	RECOVERED	16m	Normalized	3y03m	17y
9.	F	6	EPILEPSY 1y3m	FLUCTUATING 0 - 4	9y06m	IMPROVING	6y05m	Normalized	8y04m	11y
10.	F	6	EPILEPSY 10m	RECOVERED 3 - 5	1y09m	RECOVERED	8m	Normalized	1y	4y
11.	M	7	EPILEPSY 5m	FLUCTUATING 1 - 4	5y06m	RECOVERED	2y09m	Normalized	2y02m	5y

Results.

SEQUENCE OF SYMPTOMS.

Aphasia was the first sign in 5 cases, clinical seizures in 5 other cases, whereas in one child (case 3) the onset of aphasia and fits was simultaneously.

DURATION OF THE FIRST SYMPTOM.

The range in time in which no symptoms either aphasia or epilepsy developed was great : concerning aphasia as first and only sign the range was 3 months until 2 years ; concerning epilepsy 5 months till 2 years.

Table 1 specifies these chronological data and shows that in 5 children the duration of the first sign does not exceed one year. In contrast, in 2 children the first sign remains as only sign more than 2 years.

DURATION OF THE SIMULTANEOUS OCCURRENCE :

Table 2 shows the duration of the simultaneous occurrence. In the greater part of the children, this duration varies from 1 to 7 years. However, time span can be less than 3 months as cases 1, 5 and 10 illustrate. Our results demonstrate a great variety in sequence and duration of the characteristics of LKS. It seems difficult to predict

TABLE 2 : Duration of simultaneous occurrence of the characteristics of LKS.

duration	case no.
< 3 months	1 ; 5 ; 10
3 - 12 months	2 ; 6
12 - 84 months	3 ; 4 ; 7 ; 8 ; 9 ; 11

the long term outcome of aphasia: the speech of one child (case 10) recovered completely, the speech of three other children (cases 1, 5 and 8) improved considerably, in cases 2, 7 and 11 the speech had a fluctuating course. In the greater part of these children the epileptic fits as the EEG abnormalities had disappeared since years. However, severe language disorders persisted in failing to control clinical seizures. According to the literature the link between the recovery of aphasia and epilepsy is not direct. Our data confirm this view.

Discussion.

The syndrome of Landau and Kleffner has been considered as a rare disorder. Exact data about the incidence are completely lacking. Only Dugas et al. (1982) give a global assessment : in examining plus minus 1000 children a year, one child could be diagnosed as suffering from LKS. Moreover, the great number of case-studies emphasizes the impression of the rarity of the syndrome. As far as we know, there are only 2 studies including more than 10 children (Dugas et al.1982, Mantovani and Landau 1980).

However, pitfalls are present. Our study gives evidence that the epileptic attacks and EEG-abnormalities could disappear rapidly, while the aphasia persist: so high demands are made upon the reliability of the providing medical history-data. In addition, an EEG registration without epileptic abnormalities does not exclude previous abnormalities.

Other diagnostic problems could explain the low incidence of the syndrome :

a. If under 3 years of age a child acquired LKS, (mild degree of) aphasia could be diagnosed hardly.
b. The type of first sign i.e. aphasia cannot be recognized as a neurological symptom as the case can be when a child has been referred to otorhinolaryngologist, pediatrician, or childpsychiatrist unknown to the syndrome.
c. The fluctuating course of the aphasia as well as the epileptic manifestations could also complicate a correct diagnosis.
d. One has to consider that perhaps LKS can manifest itself in mild forms in which spontaneous recovery can occur rapidly.

In the literature there is a disagreement concerning several aspects of the syndrome :

Language disturbances. The initially predominant receptive disturbances "word deafness" are consistently recognized. However, exceptions are possible: "a few affected children have better comprehension than expression" (Gordon, 1990). There is less agreement concerning the expressive speech deficits. Some authors describe a non-fluent aphasia (Gascon et al., 1973), others describe a fluent aphasia with paraphasias and neologisms (Dugas et al., 1982, van de Sandt-Koenderman et al., 1984). The literature is not unanimous about the relation between nonverbal auditory agnosia and language deficits.(Rapin et al.,1977 ; Bishop,1985).

EEG - abnormalities. Our data give evidence that the EEG can normalize despite persisting aphasia which is in disagreement with the results of Gascon et al.(1973). Holmes et al. (1981) assumed that the EEG-abnormalities are an epiphenomenon of underlying pathology of brain arias concerned with speech, rather than a cause of the speech abnormality.
Because of the assumed prognostic significance for the aphasia, the ESES syndrome has been focused in the description of these children (Billard et al., 1982).
The phenomenon ESES is at present unclearly defined : there are great differences between the postulated frequency of spikes and waves during slow sleep (Patry et al., 1971 ; Dulac et al., 1983 ; Tassinari et al., 1982).

Behavioral disorders. Contrary to the opinion of Landau and Kleffner(1957), later studies have a different opinion on the frequency of behavior disorders. Dugas et al. (1982) describe severe behavioral problems in the greater part of the children out of his series. Deonna (1989) specifies this issue : "severe behavioral disturbances are only observed in early in het course of the disorder and stabilized with time". In agreement with this statement is the result of the follow-up study of Mantovani and Landau (1980) in which the good social adaptation is emphasized.

Types of clinical seizures. Finally, concerning the epileptic phenomena, most authors agree with the great variety of epileptic manifestations and the good effect of drug treatment. Dugas et al.(1982) emphasise the great frequency of generalised fits.

Course and prognosis. The long term fluctuating of aphasia in children ranging from age 4 to 7 years makes definite statements about the prognostic aspects of age at onset (Bishop, 1985) difficult. Deonna et al. (1977) describe three different forms of clinical course which have a different outcome too.

From our point of view, future research will be valuable if the diagnostic criteria are exactly specified. Consequently, only in this condition will strict statements be possible about prognostic signs and selection of children for special
treatment.
We therefore propose a more specific description of :

a. the premorbid language development and aphasic phenomena
   i.e. receptive and expressive difficulties : phonologic, morphologic, syntactic and pragmatic aspects.
b. the variety of clinical epileptic seizures in type, frequency and duration.
c. EEG-abnormalities, for example as described by Rodigues and Niedermeyer (1982).
d. Moreover, one has to demonstrate by imaging techniques that no structural cerebral abnormalities are present.
e. Finally, a strict selection of children without mental retardation and/or delayed language development is necessary.

Addendum.

Aphasia Severity Rating Scale (Goodglass & Kaplan, 1972).
- 0 : no usable speech or auditory comprehension.
- 1 : all communication is through fragmentary expression : great need for inference, questioning and guessing by the listener.
- 2 : conversation about familiar subjects is possible with help from the listener. There are frequent failures to confine the idea, but patient shares the burden of communication with the examiner.
- 3 : the patient can discuss almost all every day problems with little or no assistance. However, the reduction of speech and/or comprehension makes conversation about certain materials difficult or impossible.
- 4 : some obvious loss of fluency in speech or facility of comprehension, without significant limitation on ideas expressed or form of expression.
- 5 : minimal discernible speech handicaps : patient may have subjective difficulties which are not apparent to the listener.

References.

Billard C., Autret A., Laffont F., Lucas B., Degiovanni E.
Electrical status epilepticus during sleep in children :
a reappraisal from eight new cases. In : Sterman, MB. Shouse MM. and Passouant P.(eds.),Sleep and Epilepsy.
Academic Press, London, 1982 .

Bishop DVM. Age of onset and outcome in "acquired aphasia with convulsive disorder". Developmental Medicine and Child Neurology 27, 705-712, 1985.

Deonna T., Peter CL., Ziegler AL. Adult follow-up of the Acquired Aphasia. Epilepsy Syndrome in Childhood.
Report of 7 cases. Neuropediatrics 20, 132-138, 1989.

Dugas M., Masson M., Le Heuzey MF., Regnier N. Aphasie "acquise" de l'enfant avec épilepsie (syndrome de Landau-Kleffner), douze observations personnelles. Revue Neurologique (Paris) 138, 755-780, 1982.

Dulac O., Billard C. and Arthuis M., Aspects électro-cliniques et évolutifs de l' épilepsie dans le syndrome aphasie-épilepsie. Arch. Fr. Pediatr. 40, 299-308, 1983.

Gascon G., Victor D., Lombroso CT. and Goodglass H. Language Disorder, Convulsive Disorder, and Electroencephalographic Abnormalities. Arch. Neurol. 28, 156-162, 1973.

Goodglass H., Kaplan E. The assessment of aphasia and related disorders. Lea and Febriger, Philadelphia, 1972.

Gordon N. Acquired aphasia in childhood: the Landau-Kleffner syndrome. Dev. Med. Child Neurol. 32, 267-274, 1990.

Holmes GL., MCeever M., Saunders Z. Epileptiform activity in aphasia of childhood : an epiphenomenon ? Epilepsia 22, 631-639, 1981.

Landau WM., Kleffner FR. Syndrome of acquired aphasia with convulsive disorder in children. Neurology 7, 523-530, 1957.

Mantovani JF., Landau WM. Acquired aphasia with convulsive disorder : course and prognosis. Neurology 30, 524-529, 1980.

Maquet P., Hirsch E., Marescaux C., Franck G. Electrical status epilepticus during sleep (ESES) in children, epilepsy and acquired changes of higher functions. Pharmacological and position (PET) study lecture. One day multidisciplinary symposium on language disorders and epilepsy in children. CHUV Lausanne, june 1988.

Otero E., Cordova S., Diaz F., Garcia-Teruel I., and Del Brutto O. Acquired Epileptic Aphasia (the Landau-Kleffner Syndrome) Due to Neurocysticercosis. Epilepsia 30 (5), 569-572, 1989.

Patry G., Lyagoubi S., Tassinari C. Subclinical 'electrical status epilepticus' induced by sleep in children. Arch. Neurol. 24, 242-252, 1971.

Rapin I., Verbal auditory agnosia in children (Letter). Dev. Med. Child Neurol. 30, 685, 1989.

Rodrigues I., Niedermeyer E. The aphasia epilepsy syndrome in children : electroencephalographic aspects. Clinical Electroencephalography 13, 23-35, 1982.

Shoumaker RD., Bennet DR., Bray PF., Curless RG. Clinical an EEG manifestations of an unusual aphasic syndrome in children. Neurology 24, 10-16, 1974.

Tassinari C. A., Bureau M., Dravet C., Roger J. Electrical status epilepticus during sleep in children (ESES). in : Sleep and Epilepsy, Ed. by M.B. Sterman, AP, 1982.

Van Dongen HR., Meulstee J., Blauw-Vanmoerik M., Vanharskamp F. The Landau-Kleffner syndrome : a case study with a fourteen year follow-up. European Neurology 29, 109-114, 1989.

Van Der Sandt-Koenderman W., Smit I., Van Dongen H., Van Hest J. A case of acquired aphasia and convulsive disorder : some linguistic aspects of recovery and breakdown. Brain and Language 21, 174-183, 1984.

NATURAL HISTORY, COURSE AND PROGNOSIS OF THE LANDAU AND KLEFFNER SYNDROME

M. DUGAS, C. L. GERARD, S. FRANC, D. SAGAR
University of Paris VII
Lariboisière St Louis School of Medicine
Hospital ROBERT DEBRE
48 Bd Serurier
75019 PARIS

ABSTRACT. Since 1957, over 156 cases of Landau and Kleffner syndrome have been published. As yet, nevertheless, no general overview of the syndrome's natural history has been written. This is partly because of methodological difficulties that arise, and which are discussed. Our experience includes among 18 cases of Landau and Kleffner syndrome 12 subjects followed at least until the age of fourteen and during at least seven years. Among these, none have completely recovered ; the evolution was very unfavourable in one case. In seven cases it was unfavourable and in four mild sequels were noted (favourable evolution). The role of the age at onset of the language disorders and of the duration of follow-up, the interest of sleep EEG are discussed. This clinical experience shows that the pattern of evolution of Landau and Kleffner syndrome may not be as heterogeneous as has been supposed.

The Landau and Kleffner (L-K) syndrome already owes its fascination to the nature of the pathological phenomena which make it up (Landau and Kleffner (1957)). Moreover the evolution of the impaired subjects contributes to its uniqueness. "Variability" is the term which comes forward constantly : for the manner of presentation of the symptoms, for outcome of language disorders, but also for the sequels presented by patients having reached adolescence or adulthood.

The question of outcome is approached with very different objectives depending on the studies. The following may be distinguished : 1/ nosographic objectives : the description of varying evolutionary patterns permits to distinguish, as does Deonna et al. (1977), several "L.K. syndromes" ; 2/ etiopathogenic objectives : demonstrating the influence of the age of onset of language disorders on the prognosis is an argument employed by authors like Dulac et al. (1983), Bishop (1985), to positively state the "perceptual" nature of the primary disorder ; 3/ Therapeutic objectives : early markers of outcome would be useful in showing the need and the effect of rehabilitation and of pharmacological therapies. This has been proposed for certain aspects of the sleep E.E.G.

Many authors have dealt with the course of the L.K. syndrome by focussing on one or another of these objectives. Therefore no general description yet exists.

The methodological obstacles are numerous. By running through them, we would like to emphasize the external factors of the variability of L.K. syndrome, and weaken the position arguing for an heterogeneity of the syndrome.

First, we shall not return to the problems of definition and the presence of "impure" cases in the literature. Authors who have been involved with the question

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 263–277.

of the evolution of L.K. syndrome have generally been cautious in using restricted criteria of inclusion.

Secondly, the methodological obstacle is the rarity of the syndrome. This brings one who attempt to systematize the course of L.K. syndrome to blend his/her personal observations with those from the literature. It is difficult to give much credibility to the statistical data produced by these approximating practices. They risk to bring a lot of confusion to all future works of synthesis.

Thirdly, the particular course of language disorders in L.K. syndrome may provoke two types of bias in works on L.K. syndrome prognosis : 1/ if subjects whose sequels are being judged are not selected up to a certain minimum age and followed over a minimum length of time, one can risk being too pessimistic. Several studies (Deonna et al. (1989) ; Ansik et al. (1989)) underline the lateness of progress in language. This tendency can be counterbalanced by late relapses unknown for the same reasons. 2/ The dropped out are at the origin of an important bias for another type of study involving the very final outcome (at adult age). 3/ This bias is insufficiently uncovered in single-case studies or follow-up studies lasting 20 to 30 years.

The fourth type of methodological difficulties stems from the judgement criteria used for assessing the evolution. The parallel course between language, scholastic, social or psychological adaptation is not an evident fact. Satz (1990) underlines also this problem in his review about what becomes of acquired aphasias in children. Language disorders in aphasic children classically have a good prognosis but this contrasts with their difficulties in scholastic and social adaptation. To explain this fact, one can invoke the part played by psychosocial factors linked to the etiology but also that played by a developmental disruption secondary to durable privations of linguistic capacities. One can imagine that lasting developmental disruptions contribute to the late sequels of patients with L.K. syndrome in the areas of oral language, of academic achievement and of cognition. The complexity of the analysis of long term consequences is increased by two other facts : the actual role of epilepsy in cognition and the difficulty to determine the real extent over time of the functional and/or lesional damage, to the cerebral zones implicated in language.

The complexity of these dynamic factors explains that when there is an apparent stability of deficits, it is difficult to know what to assign to primary morbid processes and what to assign to secondary developmental consequences. When there is a "lessening" of the deficits, it is difficult to separate improvement from compensation.

To bypass these difficulties of evaluation, one should appreciate outcome in a multidimensional manner, distinguishing between the expressive and receptive aspects of oral language, the difficulties in scholastic acquisition (especially written language), the difficulties in psychological and social adaptation. This aspect remains the aim of cross-sectional studies of the prognosis. But these studies must be paralleled by longitudinal studies of single-cases. These alone allow us to assume complete recovery or lack of recovery found in cross-sectional studies. The historical course of this syndrome has still a lot to reveal to us. We therefore feel that the methodology in the study of the development of L.K. syndrome must dissociate the notions of evolution and prognosis.

We have found 83 cases of Landau and Kleffner syndrome published up to 1982 (Dugas et al. (1982)) and 156 up to 1989, . We selected 33 cases among these 156 for the prognosis review, with the following criteria :

1) regression of language after a period of normal development ;
2) existence of epileptic seizures and/or paroxystic E.E.G. abnormalities ;
3) sufficient information for a multidimensional approach to prognosis ;
4) follow-up till the minimum age of 14.

These 33 cases were selected from the following publications : Mantovani and Landau (1980) : nine cases ; Stein and Curry (1968) : one case ; Worster-Drought (1971) : five cases ; Gascon et al. (1973) : one case ; Brissaud and Richardet (1974) : one case ; Lou et al. (1977) : one case ; Deonna et al. (1989) : seven cases ; Lankriet (1980) : one case ; Noel (1980) : three cases ; Vidailhet et al. (1983) : one case ; Beaumanoir (1984) : one case ; Cole et al. : (1988) : one case ; Van Dongen et al. (1989) : one case.

The age of onset of the language disorders among these 33 cases is represented on figure 1.

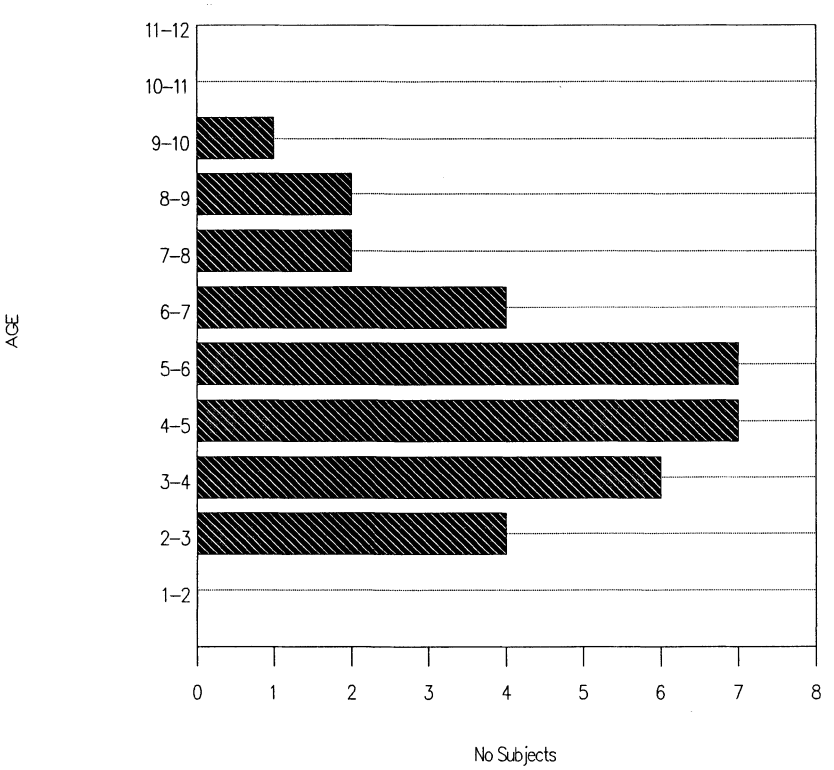


Figure 1 : Age at onset of language disorders (Years) for 33 published cases with Landau-Kleffner syndrome

We have defined 4 categories for globally evaluating the outcome of these patients.

P1/ Very unfavourable outcome (4 cases) :
- no social or professional autonomy ;
- massive comprehension disorder (even for simple command) ;
- no oral expression.

P2/ Unfavourable outcome (11 cases) :
- mediocre socioprofessional insertion ;
- communication disability with oral language unintelligible or significantly reduced.

P3/ Favourable outcome (11 cases) :
- good socioprofessional insertion ;
- persistance of oral or written language difficulties which do not impede communication.

P4/ Very favourable outcome (7 cases) :
- no difficulties with autonomous living ;
- no observable abnormality in oral or written language.

Among the 20 patients old enough to hold down a job (eighteen years or more) 15 do have one. Five of these have a job which exposes them considerably to oral and written communication. Among the 10 others who do have a job, two have no oral communication. Among the 13 school-age subjects, three follow a professional course (one is to become an engineer, another a carpenter, the last an agent of the community). Seven are in classes at special schools. In three cases we have not found any information.

The only factor widely accepted as an influence on the global prognosis of Landau and Kleffner syndrome is the age of onset of the language disorders (Bishop (1985)).

TABLE 1. Distribution of 33 Landau-Kleffner syndrome published cases in different classes of prognosis depending on the age at onset of language disorders

	Prognostic classes				
Age at onset of language disorders	P1 No	P2 No	P3 No	P4 No	TOTAL No
Before 5 years	3	8	5	1	17
5 years and more	1	3	6	6	16
TOTAL	4	11	11	7	33

In table 1 , we have represented the numbers for each category of prognosis whose onset of language disorders is before five years or five and more. For Bishop, this age corresponds to an age threshold which enabled her to show that the precocity of the onset of the illness is associated with a worse "recovery". No very unfavourable outcome can be found here when the onset of the disorders is after the age of six, but one case of unfavourable outcome is found where the onset of

disorders is after the age of seven. The results from the 33 cases do not seem to confirm those of Bishop who had used less restricting criteria of selection. The comparison of distributions with a corrected chi square does not allow to reach statistical significance.

We find it useful to confront these data taken from the literature, with the data concerning a population smaller quantitatively but nonetheless studied in a more homogeneous way and with better control of the duration of the follow-up. From 1959 to 1989, we have personally followed 18 cases of L-K syndrome. Twelve had already been published in 1982. (Dugas et al. (1982)). Among these one could not be contacted again and one had died aged 8 years. The group includes 13 boys and 5 girls. The age of onset of language disorders is represented on figure 2.

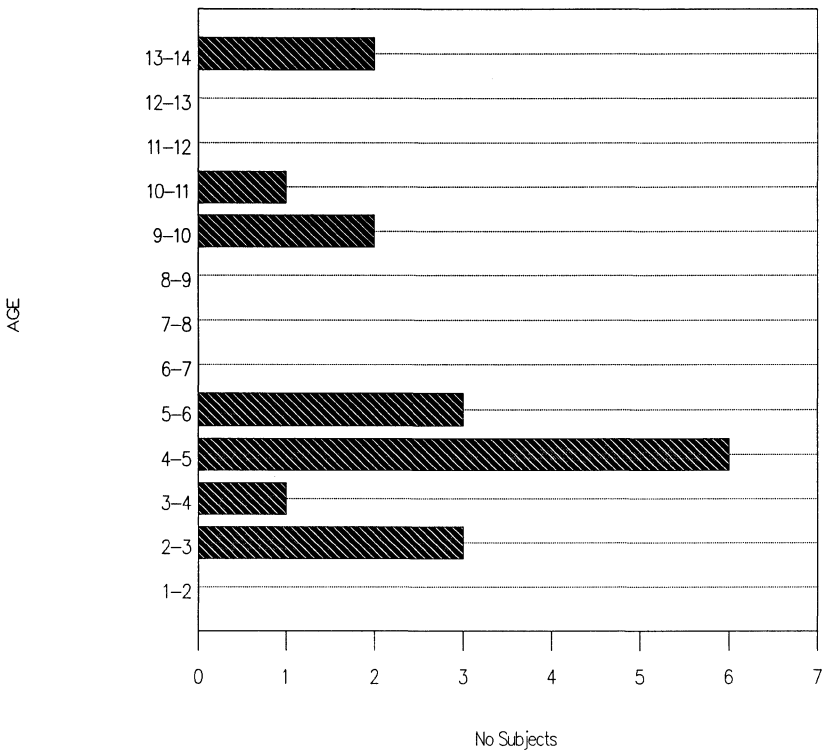


Figure 2. Age at onset of language disorders for 18 personal cases with Landau-Kleffner syndrome

We find the same grouping around the 4-6 years range as in the population of the 33 cases selected in the literature. For the evaluation of the global prognosis we have used the same categories P1-P4. With the concern for keeping results homogeneous, we have only applied our evaluation to the 12 subjects followed at least until the age of fourteen and during at least seven years. The distribution of the different categories of prognosis for the total population and according to age of onset of the language disorders is represented in table 2.

TABLE 2. Distribution of 12 personal cases of Landau-Kleffner syndrome in different classes of prognosis depending on the age at onset of language disorders

	Prognostic classes				
Age at onset of language disorders	P1 No	P2 No	P3 No	P4 No	TOTAL No
Before 5 years	1	6	2	0	9
5 years and more	0	1	2	0	3
TOTAL	1	7	4	0	12

The small number of subjects whose onset of language disorders is five years or more does not allow us to bring any more weight to our judgement of the part played by the precocity of the illness on its outcome. Nevertheless, we may underline the fact that one of our patients aged 9 years 6 months at onset of language disorders, still finds himself in category P2 unfavourable after seven years' follow-up, i.e. he still suffers from an important language disability. Our own data may give a more pessimistic outlook than those produced from the 33 cases of the literature. More than half of these had a "favourable outcome". Only four out of our 12 patients find themselves in this category of prognosis.

To explain this poor global prognosis, we cannot implicate some frequent or severe behavioral perturbation in our population. In seven of our cases, we observed long lasting minor behavioral disturbances combining hyperactivity, impulsivity and opposition. These perturbations were chronologically linked with language disorders and followed their fluctuations. However in these seven cases, they disappeared during adolescence.

In only one of our cases, which presented at the onset of the language disorders a stopping of intellectual development, we observed some major behavioral disturbances including motor stereotypies, impairment of visual contact and responsiveness, aggressiveness. These behavioral disturbances associated to the language disorders could evoke a disintegrative psychosis. However 17 years after onset these disturbances had progressively disapeared except for some intrafamilial paroxystic assaults. The language disorders and an intellectual deficit (PIQ 50 with the WAIS) still exist in this 22 years old male subject, classified in our P2 category.

The distribution of IQs in our population shows that the intellectual level factor played a role as we can see in table 3.

TABLE 3. Distribution of performance intellectuel quotients (PIQ) in three studies of long term outcome of Landau-Kleffner syndrome

PIQ	<50	50-69	70-89	90-110	>110	TOTAL
	Number of subjects					
Mantovani and Landau (1980)	0	0	0	3	6	9
Deonna et al. (1989)	0	1	2	2	2	7
Dugas et al.	0	3	5	2	2	12

They are lower than in the Mantovani and Landau (1980) series who report the best evolution, but are comparable to Deonna's et al. (1989) whose findings are more pessimistic.

As Dulac et al. (1983), we asked ourselves whether the length of the follow-up influences the rating of the prognosis and if differences at the level of this variable can lead to contradictions among the data from different origins.

TABLE 4. Distribution of 32 published cases of Landau-Kleffner syndrome in different prognostic classes depending on the duration of follow-up

Duration of follow-up	Prognostic classes				
	P1 No	P2 No	P3 No	P4 No	TOTAL No
Inf. to 10 years	0	4	1	0	5
Btw. 10 and 20 years	3	5	6	1	15
Sup. to 20 years	1	2	3	6	12
TOTAL	4	11	10	7	32

Presented in table 4 are the numbers in each category of prognosis, according to the duration of the follow-up for 32 cases from the literature previously studied by us. It appears that this variable has an influence (at least as great as the age of onset of the illness) on the category of prognosis : for example, six out of seven cases from the "very favourable outcome", category P4, have been followed-up over a period greater than 20 years.

TABLE 5. Distribution of 12 personal cases of Landau-Kleffner syndrome in different prognostic classes depending on the duration of follow-up

Duration of follow-up	Prognostic classes				
	P1 No	P2 No	P3 No	P4 No	TOTAL No
Inf. to 10 years	0	2	0	0	2
Btw. 10 and 20 years	1	4	3	0	8
Sup. to 20 years	0	1	1	0	2
TOTAL	1	7	4	0	12

The same study (see table 5) concerning our 12 patients subjected to prognosis evaluation show that our pessimistic appreciation may be linked to the small number of follow-up lasting more than 20 years. The average follow-up duration time for these patients is 16 years, with a range going from 7 to 33 years.

The "perception" theory (Bishop (1985), Dulac et al. (1983)) rests on the hypothesis of two different profiles of development according to the age of onset of the language disorders. This hypothesis is not based on a longitudinal study. With the data from the literature and from our own cases, we have found that the statment of different categories of prognosis only on the basis of a cross-sectional study may be the result of late development, linked either to factors of compensation or to a lessening in time of the illness'activity. To support these two possibilities it would have to be shown that the categories of prognosis P1-P4 are in fact developmental or evolutionary stages of one single process and one has to find actual signs of the illness'activity.

Concerning the first point, we have attempted to show the continuity of linguistic deficits on the scale, from category P1 through category P4, confronting our own data with those from our selected 33 cases of the literature at the time of their last assessment.

Category P1 : the four cases from the literature present with a massive receptive disorder, auditory agnosia, and a lack of oral expression. The only compensatory channels for communication and comprehension are through visual support, demonstrated in two cases : one uses the written mode, the other, sign language (Deonna et al. (1989)). Our only case from this category is in the same situation, but without any possibility of compensation.

Category P2 : for the 11 patients from the literature belonging to this group, comprehension is deficient even if variable comprehension of simple commands exists. Expression is reduced to a few words or agrammatic utterances. Intelligibility is very affected. Resorting to gesture is possible but hardly used. Written language is used in only one case to communicate. There seems to exist a parallel between the comprehension disorder and the unintelligibility, except in one case where the expressive disorder is predominant. The presentation of our seven personal patients belonging to this category of prognosis is more homogeneous. Even if in certain cases, verbal comprehension is lacking, some comprehension is possible from a context and from a visual support. We find in our observations a parallel between comprehension disorder and unintelligibility. Auditory agnosia is observed in a

manner which is inconstant and not total. The disorder of syntax is massive. Some of our patients have a few notions of written language (global approach to reading) but the comprehension of written sentences is poor and writing performances ressemble most often a jargon.

Category P3 : for the 11 patients from the literature belonging to this category, mild difficulties in comprehension persist (depending on the auditory context and the length of the utterances). There is no auditory agnosia. Expression is marked by a certain dysfluency (increased latency times) and the use of simplified syntax. There are no difficulties with intelligibility. The disorders of written language seem frequent : there were present in three cases out of five in which the written language was studied. In the only case where complex elaborated language was studied (Van Dongen et al. (1989)), this seemed deficient. The disorders of our own four patients belonging to this category are fairly similar. They remind the aphasic sequels observed in other contexts. These subjects seemed affected by lexical difficulties and especially by the high level aspects of language : when discourse needs to include abstract notions, metaphorical expressions ; when the subject must make a synthesis, bringing all facts together, or formulate a heard or read text in his own words. In three out of four cases, dyslexic and spelling difficulties exist ; one finds the highest rate of receptive difficulties at the written language level. But written language may be used to ease lexical access to understand and transfer information.

Category P4 : in this group, it is surprising to find patients who exerce a profession particularly exposed to communication. Their language is judged to be normal : but for the majority we don't know on which kind of test this judgement is based.

We present the synthesis of these linguistic aspects on table 6.

TABLE 6. Profiles of linguistic deficits corresponding to prognostic class

	Prognostic classes			
Linguistic deficits	P1	P2	P3	P4
Auditory agnosia	++	+	0	0
Comprehension disorder (simple commands)	++	+	0	0
Comprehension disorder (long sentences)	++	++	+	NQ
Fluency disorder	NQ	++	+	0
Phonological disorder	NQ	++	+	0
Syntactic disorder	NQ	++	+	0
Written language disorder	++	++	+	NQ
Disorder of high-level language	NQ	NQ	++	NQ

NQ not quoted, ++ constant or important, + moderate disorder, 0 absence of disorder

The result is similar to the developmental stages put forward by longitudinal studies of single cases, as the one reported by Van Dongen et al. (1989).

To confirm this impression, we have retrospectively categorized with the same rating P1-P4 the linguistic/language disabilities presented by our 12 patients at different stages in their development (table 7).

TABLE 7. Disability (classified P1 to P4) during evolution (12 personal cases of Landau-Kleffner syndrome)

Years*	Duration of follow-up (years)							
	O	5	10	15	20	25	30	35
4	P1	P2	P2					P3
2.6	P2	P1		P1				
4.8	P2 P1 P2 P1	P2		P2				
4.8	P1 P4 P2 P2 P3				P3			
9.6	P2	P2 P1 P2 P1 P2						
3.6	P2 P1	P1	P2					
9	P2 P3	P3	P3					
5.4	P2	P3		P3				
2.6	P1	P1	P2	P2				
4.6	P2 P1 P2 P2				P2			
2.5	P1 P2		P2					
4.6	P1	P1 P2						

*Age at Language disorder onset

In 11 out of 12 cases, the categorization in its final form is the result of a development which we would qualify as delayed because most of the time 10 years are needed to show it up. It is only with such a time scale that one can state an existing homogeneity in the developmental character of L.K. syndrome. The fluctuating forms appear only as a beginning variant. We have observed these forms in 4 out of 12 cases.

The existence of these fluctuations and the lateness of certain recoveries following a stable phase in the disorders bring us to distinguish two phases in the illness : an active one and a recovery one. The active phase can last more than five years : one of our female patients showed language regression eight years after the onset of the disorder. It is hard to give a superior limit or threshold to the recovery phase : we have observed some progress in two of our cases after 12 years of evolution. But these notions of "active" and "recovery" phases is fragile as long as we will not be able to associate them with physical or physiological signs.

Many studies point to the absence of a parallel between language disorders, epilepsy and standard E.E.G (Holmes et al. (1981), Toso et al. (1981), Dugas et al. (1982)). As for epilepsy, the absence of a parallel is explained by its inconstant character and the rarity of the seizures. In our experience over 18 patients, two have never had a seizure. But the reputation of epileptic phenomena being benign in L.K. syndrome should be accepted with caution, we feel, as we had one case of death occurring as a result of a convulsion at night, three years post-onset of the

disorder, at eight years and a half. Another of our patients suffered a grand mal attack 20 years after the onset of the disorder, during a trial decrease of treatment.

The main problem about the standard E.E.G. is that, in the absence of typical abnormalities, it does not lend itself to longitudinal studies. The advantage of the sleep E.E.G. is a greater accuracy in the search for abnormalities and the possibility for quantification. This explains the interest of Billard et al. (1981), Shoumaker et al. (1984), Picard et al. (in press) in iterative sleep recordings. Each one, for a small number of cases and during a short period of observation, showed a parallel between the development of E.E.G. abnormalities and language abnormalities ; moreover the sleep E.E.G. modifications could, following the authors, predict a few months in advance, certain modifications at the verbal level.

We dispose of 23 recordings taken at night from 11 patients. We have picked up, for each of these recordings, the evolution of the language disorders during the three months following the tests : stability, improvement, worsening.

TABLE 8. Relation between abnormalities on sleep-EEG and Language disorders in 11 personal cases of Landau-Kleffner syndrome

	No of sleep-EEG		
Pattern of evolution of the language disorders	Pathological	Non pathological	TOTAL
Stability	9	1	10
Aggravation	7	0	7
Improvement	1	5	6
Total	17	6	23

Table 8 shows the results. We have marked the pathological E.E.G. when the electrical abnormalities were bilateral and present almost continuously. Only in two cases out of 23 was the sleep E.E.G. not followed in a coherent fashion by the language disorders, but these results must be interpreted with caution because of the small number of E.E.G. judged non pathological.

It is possible that the new functional imagery techniques (SPECT, cerebral cartography) may produce physiological events better correlated and easier to interpret in anatomo-functional terms (Nakano et al. (1989), Picard et al. (in press)). Neither SPECT nor E.E.G. cartography actually have at present sufficiently reproducable and sensitive processing techniques to lend themselves to judgements on the evolution of activity recorded from single cases.

In which psychological terms can we describe this slow evolution of language disorders ? The semiological scheme which we have tried to trace on the basis of the evolution is somehow similar to the rare pictures of verbal deafness described in the adult (Buchman et al. (1986), Mendez et al. (1988)). We find the link with auditory agnosia which is also the most transitory and non compulsory element. The lasting character of language deficits is compatible, especially in the child, with the bilaterality of temporal lesions observed in adult cases. Visual supports

are less conspicuous than for adults (but even for adults, they are efficient only with isolated words and not for the comprehension of language) ; for the child, they cannot lean onIO semantic networks, which at this age, organise themselves in a privileged manner starting from auditive lexical forms.

Semantic damage seems to us a central element, even though it is masked during the active phase of the illness. It can also exist on its own, as shown by the semiology of Landau and Kleffner syndromes we have observed with an onset at nine years or more (Dugas et al. (1976)). The clinical picture presented by these subjects is in appearance different from younger subjects. Expressive and receptive disorders are associated at the same level. The condition is judged less severe because it does not have the spectacular aspect of auditory agnosia. At the acute phase, these patients are dysfluent because of word finding difficulties and problems in the selection of syntactic structures adapted to the semantic content. In the sequels'phase, one finds in these patients, as in patients affected younger but having then reached P2 stage, difficulties in building speech unlinked to a context of events or of environment serving them as a guide ; the semantic disorder is especially observable in a situation of definition when semantic links must be made explicit. One finds similar difficulties in written expression.

According to this semiological scheme, we believe that factors influencing the prognosis may act at two partly distinct levels. First level : the more or less permanent nature and character of the causal cerebral dysfunctionning. If one judges by the length of what we have called the active phase of the illness, this is determining in a number of cases, given the fact a lesional or structural character may be suspected. In four cases we have observed an intellectual deficit which has increased in two cases during the course of evolution. Everything happened in these two cases as if there had been at the onset of the disorder a stoppage of intellectual development : this is not without reminding one of what is observed in Lennox-Gastaut syndrome (1966). Second level : the importance of the aphasic disorder which demonstrates the topographical importance of the cerebral dysfunction. These two levels are of course linked : the duration of the active phase acts on the quality of the suppleances even more when it presents at a critical phase for the establishment of hemispheric dominance. This separation at two levels of the determinants of the prognosis presents the advantage of recognising specific areas for therapeutic action : at the first level chemotherapy ; at the second level rehabilitation.

Acknowledgements : We thank M.J. Filteau for her advices on the presentation of the english version and D. Sgarbossa and D. Cambuzat who realized the presentation of the text, figures and tables.

REFERENCES

Ansink, B.J.J., Sarphatie, H., Van Dongen, H.R. (1989) 'The Landau-Kleffner syndrome - Case report and theoretical considerations', Neuropediatrics 20, pp. 170-172.

Bishop, D.V.M., (1985) 'Age of onset and outcome in "acquired aphasia with convulsive disorder' (Landau-Kleffner syndrome). Dev. Med. Child. Neurol. 27, pp. 705-712.

Beaumanoir, A. (1984) 'Le syndrome de Landau-Kleffner. In "Les syndromes épileptiques de l'enfant et de l'adolescent'. Paris : John Libbey, pp. 185-195.

Billard, C., Autret, A., Laffont, F., De Giovanni, E., Lucan, B., Dulac, O., Plouin, P. (1981) 'Aphasie acquise de l'enfant avec épilepsie. A propos de 4 observations avec état de mal électrique infraclinique du sommeil' Rev. E.E.G. Neurophysiol. 11, pp. 457-467.

Brissaud, H.E., Richardet, J.M. (1974) 'Le syndrome de désintégration du langage et comitialité' Journées Parisiennes de Pédiatrie, pp. 441-445.

Buchman, A.S., Garron, D.C., Trost Cardamone, J.E., Witcher, M.D., Schwartz, M. (1986) 'Word deafness : one hundred years later' J. Neurol. Neurosurg. Psychiat. 49, pp. 489-499.

Cole, A.J., Andermann, F., Taylor, L., Olivier, A., Rasmussen, T., Robitaille, Y., Spire, J.P. (1988) 'The Landau-Kleffner syndrome of acquired epileptic aphasia : unusual clinical outcome, surgical experience and absence of encephalitis' Neurology 38, pp. 31-38.

Deonna, T., Beaumanoir, A., Gaillard, F., Assal, G. (1977) 'Acquired aphasia in childhood with seizure disorder : a heterogeneous syndrome' Neuropädiatrie 8, pp. 263-273.

Deonna, T., Peter, C., Ziegler, A.L. (1989) 'Adult follow-up of the acquired aphasia-epilepsy syndrome in childhood. Report of 7 cases' Neuropediatrics, 20, pp. 132-138.

Dugas, M., Grenet, P., Masson, M., Mialet, J.P., Jaquet, G. (1976) 'Aphasie de l'enfant avec épilepsie. Evolution régressive sous traitement anti-épileptique' Rev. Neurol. (Paris) 132, pp. 489-493.

Dugas, M., Masson, M., Le Heuzey, M.F., Regnier, M. (1982) 'Aphasie "acquise" de l'enfant avec épilepsie (syndrome de Landau et Kleffner). Douze observations personnelles' Rev. Neurol. (Paris) 138, pp. 755-780.

Dulac, O., Billard, C., Arthuis, M. (1983) 'Aspects électro-cliniques et évolutifs de l'épilepsie dans le syndrome aphasie-épilepsie' Arch. Fr. Pediatr. 40, pp. 299-308.

Gascon, G., Victor, D., Lombroso, C.T., Goodglass, H. (1973) 'Language disorder, convulsive disorder and electroencephalographic abnormalities' Arch. Neurol. 28, pp. 156-162.

Gastaut, H., Roger, J., Soulayrol, R., Tassinari, C., Régis, H., Dravet, C., Bernard, R., Pinsard, N., Saint-Jean, M. (1966) 'Childhood epileptic encephalopathy with diffuse slow spike-waves (otherwise known as 'petit mal variant') or Lennox syndrome' Epilepsia 7, pp. 139-179.

Holmes, G., Mc Keever, M., Saunders, Z. (1981) 'Epileptiform activity in aphasia in childhood : an epiphenomenous ?' Epilepsia 22, pp. 631-639.

Landau, W.M. and Kleffner, F.R. (1957) 'Syndrome of acquired aphasia with convulsive disorder in children'. Neurology 7, PP. 523-530.

Lankriet, C. (1980) 'Syndrome de Landau et Kleffner, désintégration du langge et comitialité' Thèse Médecine, Lille. (FRANCE)

Lou, H.C., Brandt, D., Bruhn, P. (1977) 'Aphasia and epilepsy in childhood'. Acta Neurol. Scand. 56, pp. 46-54.

Mantovani, J.F., Landau, W.M. (1980) 'Acquired aphasia with convulsive disorder : course and prognosis' Neurology 30, pp. 524-529.

Mendez, M.F. and Geehan, G.R. (1988) 'Cortical auditory disorders : clinical and psychoacoustic features' J. Neurol. Neurosurg. Psychiat. 51, pp. 1-9.

Nakano, S., Okuno, T., Mikawa, H. (1989) 'Landau-Kleffner Syndrome EEG topographic studies' Brain and Development, 11, pp. 43-50.

Noel, A. (1980) 'Le syndrome de Landau-Kleffner' Revue de littérature à propos de quatre observations personnelles. Thèse Médecine, Lyon.

Picard, A., Dulac, O., Billard, C., Raynaud, C., Lacert, P. (in press) 'Aphasie acquise avec épilepsie : syndrome de Landau-Kleffner (évolution à long terme et suivi de deux observations sur neuf et huit ans)'. Rev. Neurol. (Paris).

Satz, P. (1990) 'Developmental dyslexia : an etiological reformulation. In Pavlidis, G.T. (ed.) : Perspectives on dyslexia. Vol. 1, New-York : John Wiley and Sons, pp. 3-26.

Shoumaker, R.D., Bennett, D.R., Bray, P.F., Curless, R.G. (1974) 'Clinical and EEG manifestations of an unusual aphasic syndrome in children' Neurology 24, pp. 10-16.

Stein, L.K., Curry, F.K.W. (1968) 'Childhood auditory agnosia' J. Speech Hear. Disord. 33, pp. 361-370.

Toso, V., Moschini, M., Gagnin, G., Antoni, D. (1981) 'Aphasie acquise de l'enfant avec épilepsie. Trois observations et revue de la littérature'. Rev. Neurol. (Paris) 137, pp. 425-434.

Van Dongen, H.R., Meulstee, J., Blaw-Van-Mourik, M., Van Harskamp, F. (1989) 'Landau-Kleffner syndrome : a case study with a fourteen-year follow-up' Eur. Neurol. 29, pp. 109-114.

Vidailhet, C., Debruille, C., Hentz, P., Tridon, P. (1983) 'A propos du syndrome de Landau-Kleffner. Aphasie acquise-épilepsie chez l'enfant' Rev. Otoneuroophtalmol. 55, pp. 215-219.

Worster-Drought, C. (1971) 'An unusual form of acquired aphasia in children' Dev. Med. Child. Neurol. 13, pp. 563-571.

SPEECH THERAPY IN LANDAU AND KLEFFNER SYNDROME

C.L. GERARD, M. DUGAS, D. SAGAR
University of Paris VII
Lariboisière St Louis School of Medicine
Hospital ROBERT DEBRE
48 Bd Serurier
75019 PARIS

ABSTRACT. Few of the 150 publications or more dealing with Landau-Kleffner syndrome have properly addressed the issues concerning speech therapy. The place taken by speech therapy has in fact varied according to the explanatory model to report on the syndrome's particular evolutionary pattern. In our review of the speech therapy approach for our 18 Landau-Kleffner cases, the situation is revealed as unsatisfactory ; therapeutic treatment can rarely be programmed on a long term basis and in a systematized way. The choice between alternative systemes and a reconstruction of oral language seems to us an oversimplified approach to the discussion. A direct approach to receptive difficulties is not applicable to these children. We prefer a restoring approach, passing through the neurocognitive analysis of linguistic difficulties. We illustrate this choice with one particular case as an example.

1 - INTRODUCTION

The modern literature emphasizes the need to evaluate, as well as other therapies, the use of speech therapy in child language disorders and adult aphasia (Howard and Hatfied (1987), Holland and Wertz (1988)). This need is understandable when one considers the length of certain treatment procedures. But, the same authors underline the methodological difficulties which such efforts must inescapably meet with. The use of protocols with controls or sophisticated assessment protocols with single cases leads most often to a negative appreciation of the efficacity of speech therapy (Howlin (1987)).

In turn, the defenders of speech therapy criticize these accusing reviews by underlining the considerable variability in interindividual reactions to speech therapy. Added to this, the complexity of the content of most treatment programs, of which only a tiny part cannot be taken into account in therapy assessment : these two factors would explain that the efficacity of speech therapy may not be demonstrated by such experimental methods.

Nevertheless, concerning children, one may present certain minimal conditions under which speech therapy can hope to prove efficient (Howlin (1987)):

- treatment must be adapted to the child's level of linguistic competence ;
- it must provide gains which patients can benefit from immediately.

The difficulties surrounding evaluation seem to peak in the case of Landau-Kleffner (L-K) syndrome. Because of its particular course, it is difficult to show anything tangible from single case studies, other than over a short period. But here,

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 279–290.

promoters of assessment techniques are confronted with a second criticism : the problem is not to demonstrate that speech therapy can have a one-off effect, but rather that it has a place among the factors which can influence the syndrome's course. The use of control groups appears unrealistic to us because of the rare incidence of L-K syndrome. We shall therefore attempt to theoretically show, if not the utility of speech therapy, it's necessity ; We also wish to suggest, on the basis of a number of positive experiments, certain forms it may take.

Our clinical material constitute 18 cases of L-K Syndrome, of which 12 have been followed up till the age of 14 and over at least seven years. On the basis of these subjects, we shall envisage the following points :

1. How does the need for treatment appear, from the point of view of prognostic data and the semiological course of linguistic deficits ?

2. What has been the practical function of speech therapy for these patients ?

3. How can one orientate certain choices of modality ?

4. The practical framework which we propose.

2 - SPEECH THERAPY IN L-K SYNDROME : THE NEEDS

These can be defined on short term and long term bases.

- on a short term basis, it is the adaptation to the disability which prevails, independently of the outcome, in particular the prevention of behavioural disorders : intervention with the child, the family, the school environment.

- on a long term basis : when treatment has not been planned in a systematized manner, the place it takes can be determined from the observation of the sequels. But the published observations of the long-term outcome of L-K syndrome do not specify which treatment modalities should be used. Discussion focusses only on the interest of alternative systems (Rapin et al (1977) ; Worster-Drought (1971)) and the problems met in the scholastic adaptation of these subjects. At the most, Deonna et al. (1989) underline the need to be undogmatic and not to neglect the importance of early use of alternative systems of communication such as sign language. This lack of interest for speech therapy strategies stems partly from the variability of the syndrome's course ; which leads us to think that these strategies are not crucial factors in the end. Nevertheless, our appreciation of the development in our own series gives a less heterogeneous picture of this evolution (Dugas et al in this volume). Here is a reminder of the results of the prognostic evaluation for 12 cases. Globally, no one has a very favourable outcome. Four retain minimal linguistic difficulties, seven have an unfavourable outcome, one a very unfavourable outcome. Out of six patients old enough to hold down a job, three actually have one. Among the three others, two are in support working centres. Among the six school age children, only one has achieved a professional formation to become a mirror-maker : the five others are either in institutions for specialised education or are in school. Social interaction is considered to be normal (the patients have friends, extraprofessional or school

relationships) in five cases, poor in the seven other cases. Therefore, whatever action is taken at the level of language recovery, one can say that compensations are largely insufficient at the level of interaction. From this observation, several hypotheses for action of speech therapy treatment present themselves, according to one's interpretation of the outcome.

- first hypothesis (perceptual hypothesis) : the more or less complete but permanent damage to the auditory-verbal channel explains the lengthy and very incomplete character of linguistic recovery ; in this case the role of speech therapy should be minimal, limiting itself to contributing to the instalment of alternative systems of communication.

- Second hypothesis : the late pattern of evolution is the result of a slow and progressive process of refunctionning of the responsible cerebral structure for linguistic activities. The role of speech therapy would be to follow this recovery step by step, preventing (in the perspective of oral communication recovery) any consequences of lasting privations of linguistic capacities ; this should allow, at each stage of development, for academic achievements and a social role despite deficits.

- Third hypothesis : There is no functional recovery, and any progress can only be due to inter or intra-hemispheric transfers of linguistic competences. Here speech therapy has at once a more active and a more extended role similar to cognitive treatment developped for brain damaged patients. (Gray and Dean (1989)).

Such systematic action is in fact rarely possible. The choice between these hypotheses is in fact more dependent on practical conditions than theoretically based decisions. This is what our practical experience of treatment feasability has showed us.

3 - EXPERIENCE OF FEASABILITY

The exceptional character of L-K syndrome leads us no doubt, as a centre for national reference, to see and see again these children, to prescribe and follow pharmacological treatments through, but rarely :

- to find local solutions for school integration

- to direct the carrying-out of speech therapy treatment, even if we may have some ideas as to it's general orientations.

This leads to the impossibility in practice of having treatment plans and rehabilitation plans systematically carried out on a long term basis, from a centralized experience of this type of pathology. The obstacles met are linked :

- with difficulties in finding areas for integration adapted to these children.

- with finding speech therapists who accept to be confronted with so many unknown factors : the lack of early markers to the prognosis, difficulties in integrating the real nature of the disability.

- with the child's very frequent reluctance to face the treatment situation.

In fact, speech therapy is often accepted because it is the only way of trying something which has to do with rehabilitation, as is shown in school situations encountered by our children.

TABLE 1. Type of school placement for 18 Landau-Kleffner Syndrome cases

Normal scholastic placement	6 cases
Special classes for learning disabled children	5 cases
Placement in institution	7 cases

We will note remark that none of these children have been integrated in centres for the deaf. Institutions for special education in France are centres which integrate psychotic children or mentally retarded children. Small classes are reserved for learning disabled children who do not here receive speech therapy, which can only be given in parallel in other public services (hospitals, clinics) or more often as part of private practice. The only places where both schooling and treatment of severe language disorders can be given, are in fact in hospital units where the length of stay is not usually more than one year. Table 2 shows the distribution of children over all practice modes of speech therapy.

TABLE 2. Speech therapy practice types in 18 Landau-Kleffner syndrome cases

No speech therapy	5 cases
Speech therapy in an institution	1 case
Speech therapy in private practice	7 cases
Speech therapy in a University center	5 cases

It is difficult to obtain precisions from the speech therapists about the modalities of the therapy treatment. This is why we dispose of information only in nine cases.

TABLE 3. Supports used for speech therapy in 9 cases of Landau-Kleffner syndrome

Subject	Auditory Therapy	Phonemic Analysis	Modelling verbal Expression	Written Language	Symbols	Signs
1	-	+	+	+	-	-
2	+	+	-	-	-	-
3	+	+	+	+	-	-
4	+	+	+	+	-	-
5	-	+	+	+	-	-
6	+	+	+	+	-	+
7	-	+	+	+	-	-
8	-	+	+	+	+	-
9	-	+	+	+	-	-

The more or less spontaneous choices made, show.

- that these children are never treated with the use of coded/symbol systems : such attempts are refused by the child and the family who resent this alignment with deafness. Often the family is content with a system of idiosyncratic

communication blending noises, gesture and drawing, shared with the teacher or the speech therapist.

- direct strategies privilege auditory information processing and the approach to verbal discrimination. They are associated to modelling attempts of oral expressions, made in an empirical fashion without a coded facilitation system except inconsistantly by resorting to lip reading. Work on written language is frequent but passes often through an attempts at the acquisition of grapho-phonemic conversion rules on an analytical mode with gesture support. This written language work is rarely linked with oral language work, which is always privileged. Written language work is mostly motivated by the parents'desire not to abandon ambitions of scholastic integration for their children.

We also present in Table 4 the time span between the onset of language disorders and the start of speech therapy and its duration. If we count the children not having had speech therapy and those who started it after more than one year of evolution, we can see that the resort to speech therapy is far from systematic. On the other hand, when it is given it is difficult to find rational criteria for cessation. This practical experience therefore shows that there are obstacles to the organisation of systematized speech therapy treatment around theoretical objectives.

TABLE 4. Time span between start of speech therapy treatment and onset of language disorders. Duration of speech therapy in 13 cases of cases Landau-Kleffner syndrome.

Subjects	Time span	Duration	
1	5 yrs	5 yrs	
2	1 yr	3 mths	
3	1 mth	7 yrs	(ongoing)
4	5 yrs	2 yrs	(ongoing)
5	4 mth	5 yrs	
6	1 yrs 6	8 yrs	(ongoing)
7	1 yrs 6	10 yrs	
8	1 yrs	10 yrs 6	(ongoing)
9	1 mth	3 yrs	(deceased)
10	1 mth	4 yrs	(ongoing)
11	2 yrs 6	7 yrs	(ongoing)
12	1 mth	3 yrs	(ongoing)
13	1 yr	1 yr	(ongoing)

Which would the optimal modalities be ?

4 - THE CHOICE OF MODALITIES

It appears useful to reflect on these on the basis of a concrete experience.

4-1. A CASE REPORT

C.D. saw his language disappear within a few days and presented, at the age of five, with a picture of severe auditory agnosia with mutism. He stayed in a centre for treatment of psychotic children for one year. The diagnosis of L-K syndrome was then made.

Till the age of 13, he was maintained in a small class not dealing in special education.

In parallel to the efforts for integration, speech therapy treatment was followed privately on a twice-weekly basis.

During seven years his language has not evolved :

- verbal comprehension has remained nil
- a partial auditory agnosia has persisted
- expression was reduced, most often unintelligible, occasionally with an emergence of meaningful words and propositional sequences which the child cannot produce on command.
- his performance I.Q. on the W.I.S.C.-R was 120.

His integration in the small group was satisfactory : he communicated by gesture, accompanying his attempts with vocalisation and drawings. He had acquired elementary school arithmetic by using contextual comprehension and visually supported associations. He did not use any coding or lip reading. Speech therapy was conducted to teach him certain sign-syllable correspondances heard with the possibility of repeating isolated syllables. There was no acquisition in the area of written language.

We then took him into our care unit for nine months, with twice-daily speech therapy, schooling and educational stimulation which was to generalize the acquisitions obtained in speech therapy. It is this experiment of systematized intensive treatment following an 8 year period of deficit stability which we wish to illustrate.

The objectives were : to improve his oral communication capacities to allow him to reach a professional formation in a normal environment.

The means used were :

- modelling his spontaneous jargon in order to give him capacities of informativity and intelligibility sufficient for daily interaction in an un protected professional environment.
- work on decoding capacities over minimal (syllabic) structures using lip reading whilst repeating words. This work has not aimed at the improvement of verbal comprehension, but the fixation of lexical representations the child could use without the necessary passage through auditory analysis. This work on clue taking allowed for lexical access conditioned by an artificially created organisation of his lexicon. The phonetic clues were associated with meaningful structures, memorized by using the association of the spoken word-written word-image, then, once the meaning was fixed, the association of spoken word-written word.
- he had at his disposition a dictionary of written words on a miniaturized computer, the image of which was to remind him of the verbal representation of the word. Written work has not allowed the child to acquire the rules of

transcoding, permitting him to understand or write any words other than those he knew visually. Function words were learnt in a systematic way also, by association of written words with spoken words and symbolic representations.

Results are presented in Table 5 :

TABLE 5. Effects of a program of intensive speech therapy in one case of Landau-Kleffner syndrome

Skills	3 mths	Effects after 6 mths	9 mths
Phonemic analysis	-	-	+
Repetition of words	-	+	+
Oral comprehension of			
words	-	-	-
sentences	-	-	-
Informativity	+	+	++
Intelligibility	-	+	++
Naming	-	+	+
Use of syntax	-	+	++
Reading words	+	+	++
Reading sentences	-	-	+
Spelling	-	-	-

- non effect
+ gains in comparison with baseline assessment
++ gains after first improvements

- oral-verbal comprehension has remained nil, the child coping with the help of context and his own feedback from his present capacity to produce understandable sentences, and by trial and error.

- oral language has become easier, intelligible though reduced with the possibility of producing SVC type sentences with tensed verbs, articles and also prepositions. Phonemic paraphasias in isolated naming have disappeared leaving semantic paraphasias. Thanks to these acquisitions, he produces oral language content sufficiently informative for daily purposes.

- he now has access to some comprehension of read sentences. But written language comprehension as well as expressive capacities in oral language remain limited by his limited use of meaning ful units, systematically learnt by association mechanisms.

- the result from the point of view of autonomy is the same as that which could have been obtained with a symbolic communication system, except that here this was oral language which was used. We do not feel that sign language would have allowed for access to a more autonomous code given the weak resorting to visual clues (this being explained by the loss of function of linguistic communication in whatever shape or form). Taking Ellis and Young model (1988) developped in order to understand the acquired pathology of production and comprehension of isolated words, one can attempt to represent what we have done with the child.

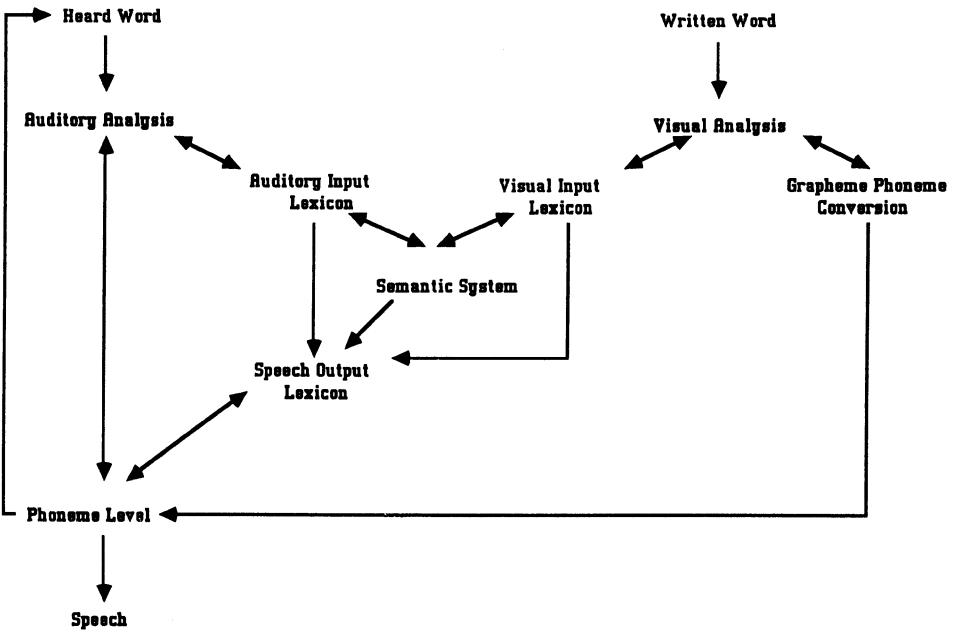


Figure 1: Model for recognition an d production for spoken words from Ellis and Young (1988)

By short-circuiting all channels leaving from the auditory analysis, we have recreated, from a visual input lexicon, links with the semantic system and with the speech output lexicon. This has been made potential by work at the phonemic analysis level, which has gone hand in hand with an improvement in phonemic discrimination, even if it has not had any bearing on comprehension. Thus we can see that this action has remained very focalized, since the capacities for grapheme-phoneme-conversion have not evolved.

4.2. IMPLICATIONS FOR SPEECH THERAPY PROGRAMS IN L-K SYNDROME

Not all L.K. children have such a durable disorganisation of these central functions. But in a child with mutism and total verbal agnosia, as in a child with only slight syntactic difficulties and anomia, one can find and interpret the limitations of linguistic competence. One understands by linguistic competence the group of implicit rules which allow to process language whatever it's context or it's form of presentation. It is important, we feel, to use the knowledge of models of cognitive neuropsychology even if they often only concern the isolated word, so as to build treatment programmes more logically. This is because these programmes cannot : either be based on any alternative which would allow natural, mediatized real communication or on spontaneous recovery.

We therefore place ourselves within the last theoretical model of development of linguistic capacities, which we have proposed to account for the syndrome's long-term outcome. This is the result of our personal experience of the course taken by our patients, but also of practical conditions of feasability.

This brings us to propose early, intensive, systematized treatment starting from a neuro-cognitive analysis, which indicates the different types of representation, their access channels make the representations available. This is to formulate hypotheses on the way children spontaneously make up for their difficulties, on the level of the structures which seem the most favourable for retention in learning. Often, one must face a reduction of linguistic competences which alternative systems do not permit to compensate for themselves. This is what Bishop (1982) demonstrated, on the difficulties to access comprehension of certain syntactic structures by the use of sign language. What does not come back spontaneously can only be compensated for by artificial means aiming at structures and not means. Visual supports are only one mean facilitating such acquisitions, but are not diverted means of activating these structures. In this way, we return to a phenomenon found in other frameworks than L-K syndrome, dyslexia (Lovett et al. (1989)). Comparisons of treatment strategies have shown in this pathological domain that speech therapy could not teach rules generating structures but that the most efficient mean concerned the associated learning of grapho-phonemic correspondences. In other terms speech therapy helps in fixing representations, not in creating linguistic competence which will then self generate progress.

Taking into account these limitations of speech therapy, the choices appear to us to be simple in the case of L-K syndrome.

- to always count on the use of residues of oral language
- when these are too insufficient, or when the intellectual potential does not allow one to be compelled to systematic, associative learning, creating a potential for lexical representation, then no more can be expected of sign language. In these cases, minimal systems of communication using gestural, iconic or pictographic symbols may be used. We prefer these as they seem more flexible. This flexibility allows to stick to the childs needs during the course of development, but also to follow a sequence going from the more iconic to the more symbolic, useful for preserving semantic and syntactic notions while waiting for oral or written language based code systems.

In the cases where oral language is minimal, even if comprehension is very deficient, systematic work must be done on the semantics and on the acquisition of syntactic structures, using multimodal learning which facilitates mnesic associations. Basic mathematics can be learnt visually. In all cases, treatment should follow a pragmatic line : this optimises the use of context in comprehension, the use of gesture as a help to comprehension ; it must teach the child again the principles of communication by caricaturing (turn-taking, state of mind awareness), put him in situations of optimal informativiting, by teaching him to recognise when, if not why, he has not reached his communication goal. It is through these macro-aspects of communication that the child can understand in a situation and often generalise his understanding, that he can revive the importance and the specificity of oral communication and specifically with it's temporal aspects. The temporal

organisation of linguistic information processing would be the primary deficit of verbal deafness in adults. (Buchman et al. (1986) ; Mender et al. (1988)).

One must not be contented with a good formal quality of spontaneous language in recovered patients ; this disappears once the subject is in a constraining situation of communication. This is all the more apparent when verbal or written expression involves a mastery of language which does not call on relating to a concrete situation : the expression of semantic links which proves the capacities to generalise, access to high level aspects of language which imply metalinguistic capacities, such as humour, figurative language which are so important at the time of adolescent transition.

We therefore have a strict approach to the notion of speech therapy, in the sense that it does not presume to act upon dysfunctions, but one counts on building communication from oral residues. This choice is also a direct intervention on expressive deficits, consequences in part of the sensory disorder judged to be primary. Another choice would be direct intervention on sensory deficits. Blanche DUCARNE (1986) promotes such an approach in verbal deafness and cortical deafness in adults, and by extension in L.K. syndrome.

This approach has also been proposed by Michel DUGAS (1972) for congenital agnosias. The principles of such treatment lie on a sequence passing through the following stages :

1/ auditory teaching focussing on non verbal sounds

2/ work on phonemic discrimination

3/ an approach to verbal comprehension through the construction of an oral input lexicon

4/ work on comprehension of connected language by teaching the child to segment heard language into meaningful units.

Such a sequence is difficult to teach to a child : this is because phonemic discrimination methods and their generalisation to verbal comprehension are only possible for the adult with verbal deafness? thanks to the great possibilities offered by preserved written language. The adult subjects have, contrary to the L-K syndrome cases, preserved linguistic competence registered in the written language, which allows eventual progress in discrimination to be efficient in comprehension. The child with an acquired auditory agnosia is all the more hampered in his progress in that language non longer means anything to him, and this creates a massive resentment towards treatment, which has no immediate and practical consequences on his daily life.

The other problem concerns the acquisition of written language : for the child whose receptive capacities remain limited, as in the case we have presented, this learning remains limited to the meaningful units learnt orally. One can hope to teach generative reading to others from the moment the subjects are capable of doing operations of segmentation on linguistic material. These are done using the first rebus system. The approach to grapho-phonemic rules should only be made as a secondary step. But, this is often only efficient in reading whereas in operations of transcription the persistent disruptions of the auditory analysis often make the realisation of spelling difficult. On the other hand, by systematic work on the

expression of short texts, one can obtain much more satisfying productions than those obtained in transcription.

5 - CONCLUSION

This presentation of our restoring conception of speech therapy has attempted to show that the problem often posed as central, of alternative systems, is a manner too simple and too short-sighted to approach the issue of rehabilitation of L.K. syndrome children. Having a deeper approach of the aphasic reality of the disability allows to hope for better adaptation, which calls for an important level of mediatic competence. It is a demanding view of the role of speech therapy which only makes it realistic in specialized centres familiar with neuropsychological reasoning. These treatments must also be totally integrated to pedagogical and educational strategies which should apply their principles. Without these conditions, speech therapy applied in an isolated way can only be an adapted contribution to a disability and cannot really act upon it. Nevertheless, one must not neglect the importance of this contribution for the child's behaviour and the help it supplies to the family.

Acknowledgements : We thank D. Sgarbossa and D. Cambuzat who realized the presentation of the text, figures and tables.

REFERENCES

Bishop, D.V.M. (1982) 'Comprehension of spoken, written and signed sentences in childhood language disorders' J. Child Psychol. Psychiat. 23, pp. 1-20.

Buchman, A.S., Garron, D.C., Trost Cardamone, J.E., Witcher, M.D., Schwartz, M. (1986) 'Word deafness : one hundred years later' J. Neurol. Neurosurg. Psychiat. 49, pp. 489-499.

Deonna, T., Peter, C. and Ziegler, A.L. (1989) 'Adult follow-up of the acquired aphasia-epilepsy syndrome in childhood. Report of 7 cases' Neuropediatrics 20, pp. 132-138.

Ducarne de Ribaucourt, B. (1988) 'Rééducation sémiologique de l'aphasie' Paris, Masson (2nd edition).

Dugas, M. et Moreau, N. (1972) 'Agnosie auditive congénitale ?' Rééducation orthophonique 68, pp. 211-220.

Ellis, A.W. and Young, A.W. (1988) 'Human cognitive neuropsychology' London, Laurence Erlbaum.

Gray, J.W. and Dean, R.S. (1989) 'Approaches to the cognitive rehabilitation of children with neuropsychological impairment'. In Reynolds, C.R. and Fletcher-

Janzen, E. (eds) Handbook of clinical child neuropsychology. New-York : Plenum Press, pp. 397-408.

Holland, A.L. and Wertz, R.T. (1988) 'Measuring aphasia treatment effects : large-group, small-group and single-subject studies'. In Plum, F. (ed) Language, Communication and the Brain. New-York, Raven Press, pp. 267-273.

Howard, D., Hatfield, F.M. (1987) 'Aphasia Therapy : historical and contemporary issues' London, Lawrence Erlbaum.

Howlin, P. (1987) 'Behavioural approaches to language' In : Yule, W. and Rutter, M. (eds) Language development and disorders. London, Mac Keith Press, pp. 367-390.

Lovett, M.W., Ransby, M.J., Hardwich, N., Johns, M.S., Donaldson, S.A. (1989) 'Can dyslexia be treated ? Treatment-specific and generalized treatment effects in dyslexic children's responses to remediation'. Brain and Language 37, pp. 90-121.

Mendez, M.F. and Geehan, G.R. (1988) 'Cortical auditory disorders : clinical and psychoacoustic features' J. Neurol. Neurosurg. Psychiat. 51, pp. 1-9.

Rapin, I., Mattis, S., Rowan, A.J. and Golden, G.G. (1977) 'Verbal auditory agnosia in children' Develop. Med. Child Neurol. 19, 192-207.

Worster-Drought, C. (1971) 'An unusual form of acquired aphasia in children' Develop. Med. Child Neurol. 13, 563-571.

THE LANDAU-KLEFFNER SYNDROME : REHABILITATION.

E. DE WIJNGAERT
Royal Institute for Children
with Communication Disorders
Borggravevijversstraat 9
3500 Hasselt
Belgium

ABSTRACT. Orthodidactics will be described as a dynamic process : written self-made lessons, based on the children's own experiences, are extended progressively in order to improve spoken as well as written language. Language therapy could only be effective if embedded in an atmosphere of enthusiasm and feeling at ease.

1. Introduction.

Regarding the characteristics of the syndrome, rehabilitation must be more than either medical treatment or language therapy. There is plenty of literature about medical treatment. In contrast, literature gives limited information only concerning language therapy or educational methods :

"a six year old boy was referred to psychiatry with severe agression and loss of speech..." WHITE et al., Can.J.Psychiatry, 32, 1987, p.599.

"Educational method may well influence the degree of linguistic attainment..." BISHOP, Developmental Medicine & Child Neurology, 27, 1985, p.705.

"...as well as intensive educational efforts in schools for deaf or aphasic children, currently provide the best opportunity for maximal recovery." MANTOVANI & LANDAU, Neurology, 30, 1980, p.529.

"...le traitement a associé : ...rééducation orthophonique ; psychothérapie ;... internat Médico-Pédagogique..." DUGAS et al., Rev. Neurol., 138, 1982, p. 755-780.

"...he subsequently underwent a six-week period of diagnostic language therapy employing the McGinnis association method and a psycholinguistic approach to language...(case2)" GASCON et al., Arch Neurol., 28, 1973, p.159-160.

Although this is not an exhaustive enumeration, more detailed information in the literature about the concepts 'rehabilitation',

I. P. Martins et al. (eds.), Acquired Aphasia in Children, 291–297.

'educational method' or 'speech therapy' is lacking.

Therefore, the purpose of this paper is : trying to define rehabilitation in a larger context. This implies educational (behaviour) and didactic aspects and individual speech therapy.

2. Educational approach.

Educational therapy will be emphasized, because - wrongly - almost no attention is paid to this subject in scientific literature. Speech therapy will be mentioned incidentally. Speech therapy is integrated in the class-program and frequency as well as moments of individual accompaniment are fixed in consultation with the teacher.

The educational approach we would like to propose is ORAL and it is based on 9-years experience with 6 LKS-children. All these children are now quite communicative and able to live a normal social live.

If the following approach amazes you, that may be due to the fact that there is no clear definition of the syndrome : what is real Landau-Kleffnersyndrome and what is not ?

Up to now our approach has been based on oral communication and pragmatic skills. Actually there is no reason for us to change our approach. It does not mean that we would not apply other forms of communication if necessary. But up to now we have never felt the need to do so !

Unfortunately, there is no such thing as a "ready to go" therapy program ! Such a program would not be possible because of the complexity of the problem. Speech therapy is not something like prescribing aspirins for a headache, a toothache or a fever etc...As a matter of fact, physicians often give advice about education, based on their own personal experiences only, which is an unfortunate approach !

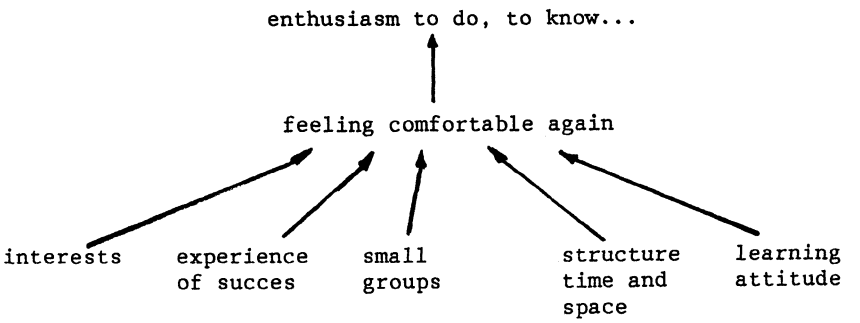
A rehabilitation program must change and be adapted continuously according to the child's ongoing development : an educational and a didactic approach is a DYNAMIC process !

It must be emphasized that rehabilitation concerns much more than language therapy, and language therapy could only be effective if embedded in a global approach. The key principle of this global approach is : creating a situation in which these children feel comfortable in spite of their (acquired) problems.

Assistance is similar to building a house : first of all, you need strong foundations : in our field this means that therapy can only be useful and effective if these children feel comfortable and happy...

The experience of failing to understand each other and/or no longer being understood makes clear that this "feeling comfortable" is not just a benefit, but a primary requirement.
This basic atmosphere of feeling at ease is a spontaneous breeding ground for the children's enthusiasm in learning, knowing and doing...It is like trying to excite appetite !
In France, this appetite is called " l'appetit". Hence, the expression "l'appetit de pouvoir parler" which is supposed to be of diagnostic importance for differing developmental dysphasia and autism.

How do you create this enthusiasm ?
In the first place through the involvement of the different therapists. It is not the method which is important but the people who work with it. ...Perhaps "it's just a matter of 'the right man in the right place.' !"



This atmosphere can be created by :
- Continuously starting from and reflecting on the interests of these children : there is no pre-structured training program that makes this possible or that has this flexibility. Of course this makes it very demanding for the teachers.
- Experiencing success : the children must experience that they can use language in a good receptive as well as expressive way. In the receptive field we try to realize this by adjusting our language to the receptive capacities of the children : short sentences ; speaking slowly and repeating often...so that the children do not have to live with the feeling that everything they hear is "Chinese"... In the expressive field we try to realize this by reacting with a "Yeessss...! to the answers of the child, even when they are wrong or incomplete.
- Working with small groups (2-5 children) and a high degree of individual accompaniment by a speech therapist (initially 7 to 10 times a week - gradually diminished).
- A structured environment is extremely important especially for young children (5 - 10 years) : these children have an avowed need for order, regularity, stability : they need to know what is going to happen next and when. Hence, visual presentation of **what** the children are going to do **when** and **where** is very important.
- Creating an atmosphere in which these children feel comfortable does not at all mean being overly tolerant. On the contrary with regard to behaviour and attitude, we are very demanding : being quiet, giving (even short) attention, taking other pupils into consideration ...are important conditions for learning.

3. Orthodidactics.

It is important to mention that orthodidactics as well as language therapy refer to children aged 6 and more. During the language stimulation in the classroom we distinguish between the spoken and written language skills.

3.1. ORAL LANGUAGE SKILLS.

By training the oral language skills we stimulate the use of language as a means of communication : by means of group sessions personal situations are being dealt with.
The emphasis is on :
- expressing oneself comprehensibly in short sentences or phrases,
- learning to express the essential ideas only, without getting lost in details,
- stimulating the children to use the right vocabulary through giving descriptions and looking for the right word. This is very important for children with naming difficulties.
- building up a logical and chronological story (picture - sentence),
- learning to hold on to a thought of their own while someone else is talking (waiting turns),
- learning to listen, to remember and repeat someone else's story (auditory memory),
- learning to ask questions....
- ...

The teacher plays the part of 'mediator' (cf. Feuerstein) : he leads the conversation, he makes sure that everybody participates and gets a chance to speak up...

The subjects for the conversation can be extracted from : weekend reports given by the parents, organised trips (markets, hospital...), incidental events.....

Such group sessions have a purpose of their own, but they can also be the beginning of the written language skills.

3.2. WRITTEN LANGUAGE SKILLS.

Between ages 6 and 14, the didactical approach is characterised by 3 consecutive levels :
- beginning
- middle
- end

3.2.1. *Beginning Stage*. The beginning stage is similar to the settling of a basic-camp at the start of a difficult hike.
At the start the emphasis lies on the learning of <u>technical skills</u>, such as reading abilities.
- the start usually consists of a small talk aroud a certain /WORD/

that the instructor wants to teach the children. This /WORD/ is generalised (ex. 'TREE' = for every tree possible ; not apple tree etc...)
- Around this /WORD/ a short and simple lesson is being drawn and written down on the blackboard. The /WORD/ is repeated several times in this lesson. Then everybody 'reads' this text (global reading).
- The letters are extracted from the /WORD/.
- In a /WORD/ are at the most 1 or 2 new letters. The new letter is used very often in the lesson and is also being placed in other words. For each new letter, 2 or 3 texts are used.
- The basic idea of this work method is to drill the children in a varied way. Obviously, these children will need more time before they are able to read...

3.2.2. *Middle Stage.* In the beginning stage, the texts were an aid to develop the children's technical reading skills. Now the lessons have a different goal namely to motivate and stimulate the children to use the language : the language they use must have a meaning ! Special attention is paid to the vocabulary and the sentence structure.

The fundamental idea behind this approach is that these children have a lot to tell even though it happens in an unstructured manner. Starting from a conversation (where the input from the teacher is important : he/she is the mediator) language is caught, reduced, adjusted and structured :
- At the start the texts are short and are read in turns : everybody reads his own sentences (that is why we put the child's name in front of every sentence).
- A second step is for each child to read the sentence of another student : do you understand ?
- The length of the sentences and lessons increases gradually and the story is being written without the names of the children in front of the sentences. Although the students are still the most important persons in the lesson, other proverbs than 'I' are used in the story. This process takes several years.

3.2.3. *Final Stage.* In the final stage - attained by a few students - the subjects of the lessons are no longer the students but they are chosen at random.
During this stage, we can start using the existing language books. Spelling-rules are not taught (they will never use them anyway). Understanding the content of a paper is important and not the reading speed. Emphasis is on problem-solving thinking and that is more than just language-teaching. Under the slogan "Just a minute - let me think..." we pay more attention to the development of correct thinking attitudes. Children should become more aware of their cognitive abilities (metacognitive awareness). We emphasize the children's learning to work systematically and to curb their egocentric thinking (what other students say may be equally correct...). Impulsive reacting is broken of. Children should be able to generalise from their experience.
We adjusted the Instrumental Enrichment Program of Reuven Feuerstein

taking into account the potentials of our children so that the positive results of this 'think training' are to us far more important than any theoretical foundation.

3.2.4. *Colour Pattern Scheme.* The Colour Pattern Scheme (CPS), developed by John Lea, Moor House School (G.B) will be used for children with severe receptive problems.
The Colour Pattern Scheme introduces concepts and sentence patterns. The functions of the different parts of speech are shown by writing each in a different colour : the colour red is associated with an object or person, the colour yellow with an action etc...

In our teaching methods the concept formation is used for LKS-children in periods of complete regression. The choice of vocabulary takes into account the interests of these children. The colours providing visual sentence patterns are used initially in the self-made lessons and in training key sentences.

3.2.5. *Key-Sentences.* The drilling of key sentences makes sense for children with serious utterance problems. Key sentences are taught especially to provide these children with a number of frequently occurring functional sentences ('I do not understand' ; 'What are we going to do ?' ; 'I am tired'...). The child has the feeling that "I can say something that makes sense and they understand me". Another advantage is the motivation of the parents. The family perceive the child's progress and they will react with enthusiasm on the child's utterances. Thus, from a pragmatic point of view, it can be said that key sentences are very useful therapeutic aids.

4. Summary.

The presented therapeutic approach is based on a combination of elements from different methods :
- a selection of the features of mediation, based on the Instrumentel Enrichment Program of R. Feuerstein (Israël),
- principles of the maternal reflective method of dr. Van Uden (the Netherlands),
- principles of the reduced language stimulation of dr. Goorhuis-Brouwer (the Netherlands),
- principles of the Colour Pattern Scheme of J. Lea, Moor House School. (Great Britain),
- pragmatics,
- last but not least : the unconditional dedication and motivation of the therapists...

The essence of the presented therapy consists in improving spoken and written language. This can be achieved by means of a gradual extension of self-constructed language lessons (which are the written but reduced reflection of a small conversation).

References.

FEUERSTEIN, R. Mediated Learning Experience. Jerusalem, 01/05/1986.

LEA,J. The Colour Pattern Scheme. A Method of Remedial Language Teaching. Moor House School, Oxted, Great Britain.

STERNBERG, R. J. Intelligence Applied. Understanding and increasing your intellectual skills. San Diego, HBJ, 1986.

VAN UDEN, A World of Language for Deaf Children. Part 1. Basic Principles. Amsterdam, Swets & Zeitlinger, 1977, 345 pp.

GOORHUIS-BROUWER, S. M. Niet Vanzelfsprekend.
Taalontwikkelingsstoornissen en afasie bij Kinderen. Orthovisies nr. 10. Groningen, Wolters-Noordhoff,1979, 108 pp.

GOORHUIS-BROUWER, S. M. ; DE BOER, G. H.A. Gezegd en Gezwegen. Uitgangspunten voor de therapie bij spraak- en taalgestoorde kinderen. Leuven, Acco, 1989, 120 pp.

INDEX

M

N