OTOLARYNGOLOGY HEAD and NECK SURGERY

Clinical Reference Guide

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SINGULAR
THOMSON LEARNING

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CONTENTS

CHAPTER

1

Rhinology and Paranasal Sinuses

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NASAL AND PARANASAL PHYSIOLOGY AND ANATOMY

Paranasal Sinus Anatomy

Lateral Nasal Wall (Fig. 1–1)

- **Uncinate Process:** sickle-shaped thin bone part of the ethmoid bone, covered by mucoperiosteum, medial to the ethmoid infundibulum and lateral to the middle turbinate (derived from the second ethmoidal turbinal)
- **Ethmoid Infundibulum:** pyramidal space that houses opening to the maxillary and anterior ethmoid sinuses; superior attachment determines spatial relationship of frontal sinus drainage (80% attach to the lamina papyracea resulting in frontal sinus drainage medial to the uncinate, 20% attach to the skull base or middle turbinate resulting in frontal sinus drainage lateral to the uncinate and into the infundibulum)
- **Semilunar Hiatus:** gap that empties the ethmoid infundibulum, located between the uncinate process and the ethmoid bulla
- **Sphenopalatine Foramen:** posterior to inferior attachment of the middle turbinate, contains sphenopalatine artery, sensory nerve fibers, and secretomotor fibers (parasympathetic fibers from vidian nerve to pterygopalatine ganglion)
- **Concha Bullosa:** an aerated middle turbinate, may result in nasal obstruction
- **Osteomeatal Complex:** region referring to the anterior ethmoids containing the ostia of the maxillary, frontal, and ethmoid sinuses, lateral to the middle turbinate

Frontal Sinus

- Embryology: does not appear until 5–6 years old
- Volume at Adult: 4–7 ml by 12–20 years old (5–10% underdeveloped)
- Drainage: frontal recess in anterior middle meatus either medial or lateral to the uncinate (posterior and medial to agar nasi cells), may also be lateral to agar nasi cells
- Vasculature: supraorbital and supratrochlear arteries, ophthalmic (cavernous sinus) and supraorbital (anterior facial) veins
- Innervation: supraorbital and supratrochlear nerves (V_1)
- Plain Film: lateral and Caldwell view
- **Foramina of Breschet:** small venules that drain the sinus mucosa into the dural veins

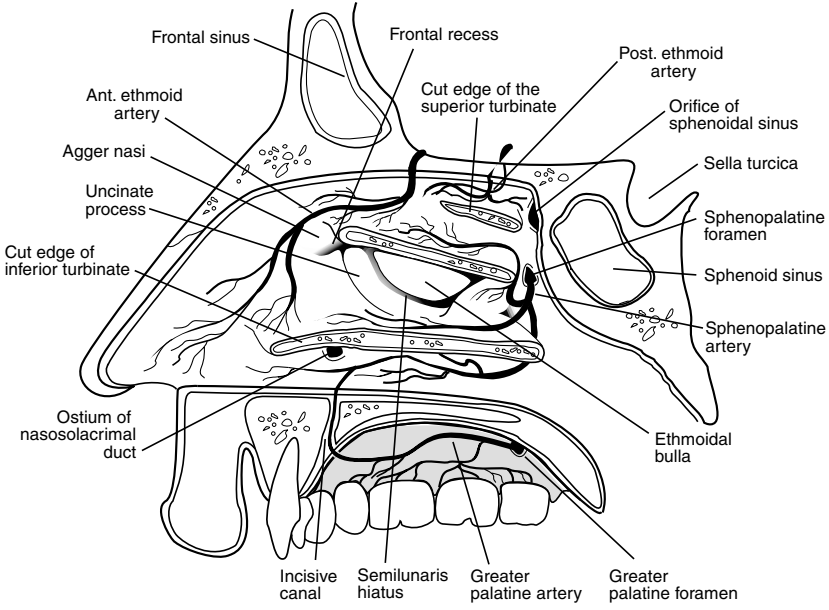

FIGURE 1–1. Anatomy of the lateral nasal wall including vascular supply.

Maxillary Sinus

- Embryology: first to develop in utero, biphasic growth at 3 and 7–18 years old
- Volume at Adult: typically 15 ml (largest paranasal sinus)
- Drainage: ethmoid infundibulum (middle meatus, 10–30% have accessory ostium)
- Vasculature: maxillary and facial artery, maxillary vein
- Innervation: infraorbital nerve (V_2)
- Plain Film: Water's view
- Adjacent Structures: lateral nasal wall, alveolar process of maxilla (contains second bicuspid and first and second molars), orbital floor, posterior maxillary wall (contains pterygopalatine fossa housing the maxillary artery, pterygopalatine ganglion, and branches of the trigeminal nerve)

Ethmoid Sinus

- Embryology: 3–4 cells at birth (**most developed paranasal sinus at birth**), formed from 5 ethmoid turbinals (1. agar nasi, 2. uncinate, 3. ethmoid bulla, 4. ground lamella, 5. posterior wall of the most posterior ethmoid cell)
- Volume at Adult: 10–15 aerated cells, total volume of 2–3 ml (adult size at 12–15 years old)
- Drainage: anterior cells drain into the ethmoid infundibulum, posterior cells drain into the spheno-ethmoid recess (superior meatus)
- Vasculature: anterior and posterior ethmoid arteries (from ophthalmic artery), maxillary and ethmoid veins (cavernous sinus)
- Innervation: anterior and posterior ethmoidal nerves (from nasociliary nerve, V_1)
- Plain Film: lateral and Caldwell view
- Adjacent Structures: skull base, anterior ethmoid artery (roof of anterior ethmoid cells), nasal cavity, orbit
- **Agger Nasi Cells:** most anterior of anterior ethmoid cells
- **Ground (Basal) Lamella:** posterior bony insertion of the middle turbinate which separates anterior and posterior ethmoid cells; posterior extension of the middle turbinate
- **Onodi Cells:** ethmoid cells that pneumatize lateral or posterior to anterior wall of the sphenoid, commonly mistaken as a sphenoid cell, optic nerve may indent into the lateral wall
- **Haller Cells:** ethmoid cells that extend into maxillary sinus above the ostium, pneumatize the medial and inferior orbital walls

- **Lamina Papyracea:** lateral thin bony wall of the ethmoid sinus, separates orbit from ethmoid cells as a part of the medial orbital wall
- **Fovea Ethmoidalis:** roof of ethmoid sinus

Sphenoid Sinus

- Embryology: evagination of nasal mucosa into sphenoid bone
- Volume at Adult: 0.5–8 ml (adult size at 12–18 years old)
- Drainage: sphenoethmoid recess in the superior meatus
- Vasculature: sphenopalatine artery (from maxillary artery), maxillary vein (pterygoid plexus)
- Innervation: sphenopalatine nerve (parasympathetic fibers and V_2)
- Plain Film: lateral and submentovertex (basal)
- Adjacent Structures: pons, pituitary (sella turcica), carotid artery (lateral wall), optic nerve (lateral wall), cavernous sinus (laterally), maxillary and abducens nerves, clivus

Anatomical Relationships of the Sphenoid Ostium

- adjacent to posterior border of nasal septum
- 6–8 cm posterior to the anterior nasal spine
- 30° angle from floor of nose (most reliable)
- typically 1.5 cm above the choanal floor
- 1/3 up from the choana to the base of skull

Nasal Anatomy

Nasal Cartilage

- **Upper Lateral Cartilage**
- **Lower Lateral (Alar) Cartilage:** paired cartilage, composed of lateral and medial crura
- **Sesamoid Cartilage**
- **Lesser Alar Cartilage**

Septal Anatomy (Fig. 1–2)

- **Quadrangular Cartilage:** septal cartilage
- **Perpendicular Plate of the Ethmoid:** projects from cribiform plate to septal cartilage
- **Vomer:** posterior and inferior to perpendicular plate
- **Maxillary Crest** (Palatine Bone): trough of bone that supports the septal cartilage
- **Anterior Nasal Spine:** bony projection anterior to pyriform aperture

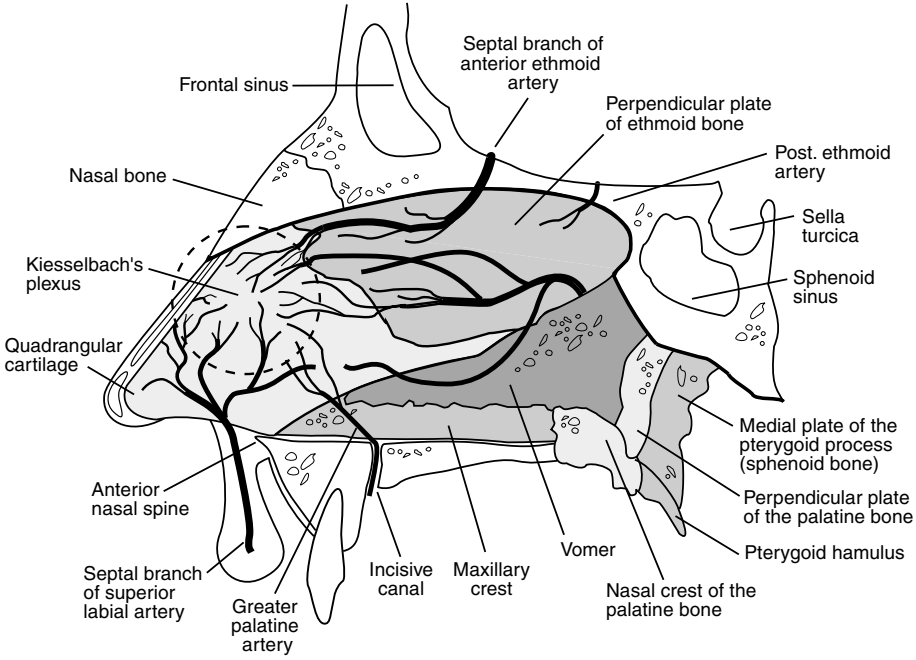

FIGURE 1–2. Anatomy of the septum including blood supply.

Sensory Innervation

External Innervation

- Supratrochlear and Infratrochlear Nerves (CN V_1): nasal dorsum
- External Nasal Branch of Anterior Ethmoid (CN V_1): nasal tip
- Infraorbital Nerve (CN V_2): malar, lateral nose, and subnasal regions

Internal Innervation

- Internal Nasal Branch of Anterior Ethmoid (CN V_1): anterosuperior nasal cavity
- Posterior Ethmoid Nerve (CN V_1): posterior nasal cavity
- Sphenopalatine Nerve (CN V_2): posterior and inferior nasal cavity
- Superior Alveolar Nerves (CN V_2)

Vascular Anatomy (Fig. 1–1 and 1–2)

External Carotid Artery Branches

Internal Maxillary Artery

- descending palatine artery → greater palatine and lesser palatine arteries
- sphenopalatine artery → sphenopalatine foramen (posterior to the middle turbinate) → medial (nasoseptal) and lateral nasal artery (middle and inferior turbinates)

Facial Artery

- superior labial artery → nasal septum and alar branches
- lateral nasal artery
- angular artery

Internal Carotid Artery Branches (branches of ophthalmic artery)

- anterior ethmoid artery (larger than the posterior ethmoid artery) → lateral nasal wall and septum
- posterior ethmoidal artery → superior concha and septum
- dorsal nasal artery → external nose

Venous System

- greater palatine vein drains into posterior facial vein (external jugular vein) and cavernous sinus
- septal vein drains into anterior facial vein (internal jugular vein)

- sphenopalatine vein drains into cavernous sinus and maxillary vein (internal jugular vein)
- anterior and posterior ethmoidal veins drain into ophthalmic veins (cavernous sinus)
- angular vein drains into anterior facial vein (internal jugular vein)

PHYSIOLOGY OF THE NOSE AND SINUSES

Physiology of the Nasal Airway

Nasal Cycles and Respiratory Airflow

- nasal airflow is regulated through the volume of the venous sinusoids (capacitance vessels) in the nasal erectile tissue (located primarily in the inferior turbinate and to a lesser extent in the anterior septum)
- the hypothalamus continuously stimulates a sympathetic tone (via the superior cervical sympathetic ganglia) to maintain a level of nasal vasoconstriction
- inspired air is warmed to body temperature and is humidified to almost 100% humidity
- **Sneeze Reflex:** induced by allergens, ammonia, viral infections, exercise, and other irritants which stimulates trigeminal afferents, complex efferent input results in a slow inspiratory phase, glottic and velopharyngeal closure (increases subglottic pressure), followed by a sudden glottic opening (sneeze)
- Regulation Response Types
  1. **Asymmetrical Congestive Response (The Nasal Cycle):** normal physiological congestion/decongestion cycle alternating between nasal sides every 2–7 hours
  2. **Symmetrical Congestive Response:** temporary bilateral congestion induced by exercise, changes in body position, hyperventilation, cold air, sulfur, histamine, and other irritants; lasts 15–30 minutes

Microvasculature

- regulates nasal volume, humidity, and heat exchange
- **Resistance Vessels:** arterioles and precapillary sphincters, regulate blood flow to the nasal mucosa
- **Subepithelial Capillaries:** fenestrated vessels allow for transport of solutes and fluids
- **Venous Sinusoids:** capacitance vessel, determines blood volume and nasal congestion

- **Arteriovenous Anastomoses (AVA):** regulate nasal blood flow by allowing blood to flow directly from the resistance vessels to the venous sinusoids

Regulation of Nasal Microvasculature

- Sympathetic Innervation: provides vasoconstrictor tone to arteries and capacitance veins, mediated through **Norepinephrine** (primary neurotransmitter), **Neuropeptide Y** (a weak vasoconstrictor, enhances effects of norepinephrine), and **Avian Pancreatic Polypeptide** (APP)
- Parasympathetic Innervation: controls secretions and dilates resistance vessels, mediated through **Acetylcholine** (primary neurotransmitter), **Vasoactive Intestinal Peptide** (VIP), and **Peptide Histamine Isoleucine** (PHI)

Nasal Valves

- **External Nasal Valve (Nasal Vestibule):** anterior nostril (nasal alar cartilage, columella, and nasal sill), potential cause of obstruction during inspiration
- **Internal Nasal Valve (Limen Nasi):** bordered by septum, anterior edge of the inferior turbinate, and caudal edge of upper lateral cartilage; narrowest segment (**50% of total nasal resistance**), potential site of obstruction secondary to Bernoulli's principle (narrowed segment accelerates nasal airflow resulting in a decrease in intraluminal pressure)

Mucociliary System

- Function: humidification, cleaning of inspired air, eliminating debris and excess secretion from paranasal sinus and nasal airway
- Mucociliary Flow: mass motion of mucous layer in the paranasal sinus of the mucous blanket at 1 cm/min (eg, migration in the maxillary sinus begins at the floor of maxillary sinus → natural ostium → nasal cavity → nasopharynx)
- Components
  1. **Ciliated, Pseudostratified Columnar Epithelium:** anterior border begins at limen nasi
  2. **Double Layered Mucous Blanket:** deep, less viscous, serous periciliary fluid (sol phase) and superficial, more viscous, mucous fluid (gel phase)
  3. **Mucous Producing Glands:** goblet cells (columnar cells, basal nucleus, secretory granules at lumen end), deep and superficial seromucinous glands (serous or mucous acini with cuboidal lined

duct complexes), and intraepithelial glands (20–50 mucous cells around a single duct)
- Major Composition of Nasal Mucus: 95% water, 3% glycoproteins (mucin), 2% salts, immunoglobulins (IgA), lysozymes (bacteriolytic), and lactoferrin (bacteriostatic)

Olfactory Physiology

- olfaction requires turbulent airflow from the anterior nares or the choanae
- pungent odors (vinegar, ammonia) may be perceived through **trigeminal nerve fibers** (via substance P pain fibers)
- Olfactory Epithelial Cell Types
  1. **Ciliated Olfactory Receptor Cells:** club shaped bipolar neurons with axons that synapse to the **olfactory bulb**
  2. **Microvillar Cells:** neuronal cells of unknown function
  3. **Supporting Cells:** sustenacular cells
  4. **Basal Cells:** allow capability of olfactory fiber **regeneration** (unlike other sensory cells)
- Olfactory Mechanism: odorant enters olfactory cleft → odorant dissolves in mucus → odorant binding proteins (OBP) concentrate the solubilized odorant → binds to olfactory receptor at the sensory cilia → receptor binding results → stimulates a specific G-protein (cAMP dependent) cascade for depolarization → synaptic connections form a complex network of secondary neurons (suggesting peripheral processing) before entering the brain (dentate and semilunate gyri)

EVALUATION OF NASAL OBSTRUCTION

History and Physical Exam

History

- Character of Nasal Obstruction: onset and duration, constant versus intermittent, unilateral (tumors, normal nasal cycle) versus bilateral obstruction, associated mouth breathing, snoring, anosmia/hyposmia/taste disturbances, tearing (nasolacrimal duct obstruction or allergy)
- Contributing Factors: potential toxin and allergen exposure, known drug allergies, medications (*see* Table 1–1), history of immunodeficiency, asthma, sinusitis, otitis media, allergy, sleep disturbances, facial trauma or surgery
- Associated Symptoms: allergic component (sneezing, itchy and watery eyes, clear rhinorrhea), sinus involvement (facial pain,

TABLE 1–1. Common Classifications of Drugs that Cause Rhinorrhea and Nasal Congestion

- **Antihypertensives**
- **Psychotropic Medications**
- **Oral Contraceptives**
- **Chronic Nasal Decongestants:** rhinitis medicamentosa
- **Cocaine:** local vasoconstriction
- **Tobacco:** irritates mucosa and impairs ciliary clearance
- **Antithyroid Medication**
- **Aspirin:** activates peripheral chemoreceptors
- **Marijuana**

headaches), acute infection (fevers, malaise, purulent or odorous nasal discharge, pain)
- Other Head and Neck (H&N) Symptoms: sore throat, postnasal drip, cough, ear complaints, halitosis, ocular pain, hoarseness
- Think "KITTENS" for differential diagnosis (*see* Table 1–2)

Physical Exam

- External Nasal Exam: external deformities (firmness, tenderness on palpation), nasal flaring, nasal airflow
- Anterior Rhinoscopy/Nasal Endoscopy: examine twice (with and without topical decongestion), quality of turbinates (hypertrophic, pale, blue), quality of nasal mucosa, nasal septum, osteomeatal complex obstruction, foreign bodies, nasal masses, choanal opening
- Quality of Nasal Secretions: purulent or thick (infectious), watery and clear (vasomotor rhinitis, allergy), salty and clear (CSF leak)
- H&N Exam: facial tenderness, tonsil and adenoid hypertrophy, cobblestoned posterior pharynx, cervical adenopathy, otologic exam

Ancillary Tests

- **Allergy Evaluation:** (*see below*)
- **Paranasal Plain Films:** may be considered for screening, largely been replaced by CT/MRI
- **CT/MRI of Paranasal Sinus:** indicated if obstruction may be secondary to nasal masses, polyps, or complicated sinusitis

TABLE 1–2. Differential Diagnosis of Nasal Obstruction: KITTENS Method

(K) Congenital	Infectious & Idiopathic	Toxins & Trauma	Tumor (Neoplasia)	Endocrine	Neurologic	Systemic
Neurogenic tumors	Infectious rhinitis	Nasal and septal fractures	Papillomas	Diabetes	Vasomotor rhinitis	Granulomatous diseases
Congenital nasopharyngeal cysts	Rhinoscleroma	Medication side effects (rhinitis medicamentosa)	Nasal Polyps	Hypothyroidism		Vasculitis
Teratoma	Chronic sinusitis	Synechia	Hemangiomas	Pregnancy		Allergy
Choanal atresia	Adenoid hyperplasia	Environmental irritants	Pyogenic granulomas			Cystic fibrosis
Nasoseptal deformities		Septal hematomas	Juvenile nasopharyngeal angiofibromas			
		Foreign bodies	Malignancy			

- **Biopsy:** indicated for any mass suspect for malignancy, avoid biopsy of vascular neoplasms (juvenile nasopharyngeal angiofibroma, sarcomas) or encephaloceles
- **Rhinomanometry:** provides an objective measurement of airway resistance, largely not utilized in clinical practice since highly time consuming, not cost effective, and inaccurate
- **Ciliary Biopsy and Mucociliary Clearance Tests:** electronmicroscopy and ciliary motility studies for ciliary defects
- **Nasal Secretion Protein and Glucose:** evaluate for CSF leak if suspected
- **Culture and Sensitivity:** surgically obtained cultures usually indicated for complicated acute rhinosinusitis and resistant chronic sinusitis
- **Pulmonary Function Tests:** suspect reactive airway disease component
- **Olfactometry:** qualitative and quantitative testing of olfactory substances

NASAL DISEASES

Congenital Nasal Disorders

Neurogenic Tumors

- **Fronticulus Frontalis:** embryologic space that normally fuses in the development of the frontal bones
- **Prenasal Space:** embryologic space between the nasal bone and nasal cartilage
- Pathophysiology: dura projects through the foramen cecum, through the fronticulus frontalis (**intranasally**), through the prenasal space into skin (**extranasally**) with failure of closure of neuropore
- Dx: MRI/CT to evaluate intracranial extension, do not biopsy

Encephalocele

- Pathophysiology: failed closure of neuropore or failed migration of neural crest cells results in ependymal-lined meninges herniation though the base of skull; communicates with subarachnoid space (CSF filled)
- more common in lumbar-sacral region
- Types by Contents
  1. **Meningocele:** contains meninges only
  2. **Meningoencephalocele:** contains meninges and brain elements
  3. **Meningoencephalocytocele:** contains meninges, brain, and a part of the ventricular system

- Types by Location
  1. **Occipital:** defect occurs over the occiput, most common
  2. **Sincipital (Frontoethmoid):** defect occurs between frontal and ethmoid bones at the foramen cecum; **Nasofrontal** (glabellar lesion), **Nasoethmoidal** (lateral nose lesion), and **Naso-orbital** (medial orbital wall lesion) **subtypes**
  3. **Basal:** transethmoidal, sphenoethmoidal, transsphenoidal, and sphenomaxillary subtypes
- SSx: soft, compressible masses that change with straining and crying, transilluminates, intranasal encephaloceles are often confused with nasal polyps
- Dx: CT or MRI reveals a bony defect, **Furstenburg test** (compression of the jugular vein causes increase in the size of the mass from increased CSF pressure)
- Complications: meningitis, nasal obstruction, cosmetic deformity, hydrocephalus
- Rx: surgical excision similar to gliomas, must also close the dural defect to prevent CSF leak and brain herniation (neurosurgical consultation)

Nasal Gliomas

- Pathophysiology: sequestered glial tissue or "pinched-off encephaloceles" results in unencapsulated collection of heterotrophic glial cells
- 60% extranasal, 30% intranasal
- 15% connect to dura
- SSx: intranasal or extranasal firm, nonpulsatile mass (typically not midline), skin covered, does **not** change in size with straining, may present as an intranasal polyp, broad nasal dorsum
- Dx: CT or MRI to evaluate for intracranial extension
- Complications: meningitis, nasal obstruction, cosmetic deformity
- Rx: surgical excision (intranasal approach for small tumors, extranasal approach for larger tumors), may require craniotomy for intracranial involvement (neurosurgical consultation)

Dermoids (Ectodermal Cysts, Nasal Dermal Sinus Cysts)

- Pathophysiology: defective obliteration of dural tissue in prenasal space or fronticulus frontalis, forms an epithelial lined cyst (may contain other ectodermal components including hair follicles and adnexal tissue)
- SSx: presents at birth; forms a fistulous tract, pit, or cyst on midline or off-midline of nasal dorsum or septum; tuft of hair may protrude from pit

- Dx: CT or MRI to evaluate for intracranial extension
- Complications: meningitis, CSF leak, infection, cosmetic deformity
- Rx: meticulous excision, must excise complete tract (usually subcutaneous although may dive deep into nasal bone or intracranially)

Rathke's Pouch Cyst

- Pathophysiology: persistent craniopharyngeal canal from failure of the obliteration of Rathke's pouch (a diverticulum of ectoderm which invaginates to form the anterior lobe of the hypophysis and pars intermedius)
- Location: nasopharynx
- SSx: typically asymptomatic, smooth mass in nasopharynx
- Dx: CT or MRI, biopsy
- Rx: antibiotics with marsupialization or excision for infected lesions

Nasolacrimal Duct Cyst

- Pathophysiology: failure of opening of the distal lacrimal duct
- Location: nasolacrimal duct
- SSx: usually asymptomatic, epiphora
- most spontaneously resolve
- Dx: MRI of paranasal sinus
- Rx: marsupialization for symptomatic cysts or antibiotics with excision for infected lesions

Thorwaldt's Cyst

- Pathophysiology: arises from a pharyngeal notochord remnant (pharyngeal bursa or pouch of Luschka)
- Location: nasopharynx (midline, surrounded by adenoid tissue)
- SSx: asymptomatic, smooth mass found in nasopharynx
- Dx: CT or MRI, biopsy
- Rx: observation, antibiotics with marsupialization or excision for infected lesions

Choanal Atresia

- Pathophysiology: persistence of the **bucconasal membrane** resulting in a complete or incomplete **bony** (90%) or **membranous** (10%) defect
- more common in females
- unilateral more common than bilateral atresia

- SSx: rhinorrhea, anosmia, nasal obstruction; bilateral involvement presents within first days of life with cycles of apnea and cyanosis followed by crying due to obligate nasal respiration in neonates
- Dx: passage of a 6F catheter, nasal endoscopy, CT of paranasal sinus
- **CHARGE Syndrome:** most common concurrent syndrome; **C**oloboma (iris keyhole defect), **H**eart disease, **A**tresia (choanal), **R**etardation (CNS), **G**enital hypoplasia, **E**ar abnormalities
- also associated with Apert, Treacher Collins, Trisomy D syndromes
- Rx: unilateral atresia may be managed on an elective basis via a transnasal approach for membranous defects or a transantral, transseptal, or transpalatal approach for bony defects; bilateral atresia must be addressed during first weeks of life (McGovern's nipple may be required for feeding initially for bilateral choanal atresia)

Inflammatory Nasal Masses

Nasal Folliculitis and Furuncles

- Pathogenesis: a pyodermia secondary to *Staphylococcus aureus* or *Streptococcus* bacterium, typically arises from a hair follicle (**folliculitis**), may organize to form pus with a central core (**faruncle**)
- SSx: intranasal tenderness, reddening and edema of nasal vestibule, sensation of tension at tip of nose, fever
- Dx: clinical exam
- Complications: septal abscess, septal chondritis, saddle-nose deformity, cavernous sinus thrombosis
- Rx: antibiotic ointment may be utilized initially for folliculitis, avoid manipulation, oral antibiotics with local antibiotic ointment for faruncles, incision and drainage for abscess formation

Septal Abscess

- Pathophysiology: commonly secondary to trauma (septal hematoma) or a faruncle
- SSx: widened septum, nasal obstruction, fever, erythema in nasal vestibule
- Complications: intracranial extension (cavernous sinus thrombosis, meningitis), septal chondritis, saddle-nose deformity
- Rx: aggressive management with incision and drainage and intravenous antibiotics

Rhinophyma

- Pathophysiology: massive hypertrophy of sebaceous glands (form of acne rosacea), associated with *Demodex folliculorum*

- SSx: begins with coarsening of nasal skin over cartilaginous portion of the nose, develops into a large protuberant lobular swelling of the nasal tip, nasal obstruction
- Dx: clinical exam
- Rx: surgical full thickness excision (laser, cold scalpel, or dermabrasion) until normal nasal contour, may require STSG

Rhinoliths/Nasal Foreign Bodies

- Pathophysiology: concretions secondary to encrustation of foreign body or longstanding nasal crusting may form **rhinoliths**
- SSx: unilateral, purulent rhinorrhea, pain, epistaxis
- Dx: anterior rhinoscopy, plain films, or nasal endoscopy
- Complications: secondary infection
- Rx: removal (may require general anesthetic, may trigger epistaxis)

Nasal (Sinonasal) Polyposis

- Pathophysiology: unclear, may be secondary to abnormal cellular homeostasis from chronic inflammation resulting in polypoidal degeneration, typically arises from lateral nasal wall
- associated with chronic sinusitis (approximately 50%), ASA intolerance, and asthma (**Samter's Triad**), allergy, fungal sinus infection, cystic fibrosis, trauma, and metabolic diseases
- SSx: smooth, pale, intranasal clustered grape-like masses (usually bilateral), nasal obstruction, anosmia, postnasal drip, rhinorrhea, hyposomia
- Dx: anterior rhinoscopy, nasal endoscopy, CT of paranasal sinus (nonenhancing nasal mass with partial or complete sinus opacification may reveal expansion of superior nasal fossa and ethmoid air cells in advanced cases, also required to evaluate for potential encephalocele, gliomas, or inverting papillomas)
- Histopathology Types: edematous (few inflammatory cells with edematous stroma), inflammatory (predominantly inflammatory cells), fibrous (collagen stoma)
- Complications: proptosis, diplopia, bone erosion, osteitis, meningitis

Management

- **Medical Management**: allergy desensitization, avoidance of aspirin or other allergens, nasal corticosteroid sprays or oral corticosteroids, hypertonic saline irrigations
- **Polypectomy**: effective in short term (high rate of recurrence), provides a biopsy specimen

- **Endoscopic Sinus Surgery**: treatment of choice, includes polypectomy, complete sphenoethmoidectomy, antrostomy for ventilation and drainage, polypoid specimen should be sent as specimen to evaluate for potential underlying tumor, recurrence common
- consider lipoxygenase pathway inhibitors

Neoplasms

Keratotic Papilloma (Benign Squamous Papilloma, Vestibular Wart)

- Pathophysiology: benign lesions arise from squamous or schneiderian epithelium (associated most commonly with **Human Papilloma Virus 6** and **11**)
- low malignant potential
- SSx: verrucous lesion, commonly on nasal vestibule, painless
- Dx: anterior rhinoscopy, nasal endoscopy
- Rx: simple excision or laser ablation; for septal keratotic papillomas a cuff of normal mucoperichondrium should be taken with lesion to avoid recurrence

Inverted Papilloma

- Pathophysiology: arise from proliferation of reserve cells in schneiderian mucosa (associated with human papilloma virus)
- more common in males
- often misdiagnosed as a nasal polyp (polyps are more translucent, bilateral, and bleed less)
- SSx: unilateral obstruction, sinusitis, epistaxis, rhinorrhea, diplopia, typically presents on the lateral nasal wall (rarely on the nasal septum), may be associated with a benign nasal polyp
- Dx: CT of paranasal sinus reveals erosion into lateral nasal wall or extension into maxillary sinus, may reveal calcifications; MRI may be considered for extensive involvement or for recurrence
- Histopathology: cristae-laden senescent mitochondria, inflammatory cells throughout epithelium, endophytic growth of epithelium
- Complications: 10% malignant degeneration from lateral wall lesions (rare from nasal septum), extension into sinuses, orbit (blindness, diplopia, proptosis), or intracranial and skull base
- Rx: adequate en bloc excision typically requires a medial maxillectomy, may require an ethmoidectomy or craniofacial

resection, endoscopic excision may be considered for select lesions (recurrence rate up to 20%)

Juvenile Nasopharyngeal Angiofibroma (JNA)

- most common vascular mass in nose
- **exclusive to adolescent males**
- Pathophysiology: benign vascular tumor, etiology unknown
- slowly growing, locally invasive, may spread intracranially, does not metastasize
- SSx: smooth **purplish** lobulated mass in nasopharynx or lateral nasal wall (posterior aspect of the middle turbinate), recurrent unilateral epistaxis (may be bilateral), rhinorrhea, nasal obstruction, anosmia
- Dx: CT/MRI/MRA (magnetic resonance arteriography) of paranasal sinuses (mass with extension into pterygomaxillary fissure), carotid angiography, avoid biopsy
- Complications: extension into sinuses, orbit (blindness, diplopia, proptosis), or intracranial and skull base
- Rx: surgical excision; consider preoperative embolization; radiation therapy reserved for residual tumors, intracranial extension, or inoperable candidates

Other Benign Tumors

- **Benign Salivary Gland Tumors**: rare, pleomorphic adenoma most common
- **Hemangiomas**: most often present at Little's area or inferior turbinate; Rx: excision with cuff of normal mucoperichondrium, may consider preoperative embolization
- **Pyogenic Granulomas**: friable polypoid lesion (usually on septum, may be secondary to trauma), difficult to distinguish from hemangiomas, presents with epistaxis and unilateral obstruction, may present during pregnancy ("pregnancy tumor"); Rx: excision although most resolve
- **Hemangiopericytomas**: arise from the pericyte, 10% malignant degeneration; Rx: excision, 50% recur
- **Osteomas**: common benign tumor, slow growing, usually asymptomatic, multiple lesions associated with Gardner syndrome (malignant degeneration of intestinal polyps); Rx: excision for symptomatic lesions otherwise may observe with serial radiographs
- **Chordomas**: arise from notochord of nasopharynx, may produce obstructive symptoms or involve cranial nerves; Rx: excision

Malignancy (*see* p. 259)

Systemic Diseases Affecting The Nose

Granulomatous Diseases (*see also* pp. 205–212)

- **Sarcoidosis**: cobblestoning of sinonasal mucosa from granulomatous inflammation, dryness, crusting, epistaxis, or septal perforation
- **Histocytosis X**: nasal mass, epistaxis, or septal perforation
- **Lethal Midline Granulomas**: clear to purulent rhinorrhea, septal perforation, epistaxis, facial destructive lesions

Vasculitic Diseases (*see also* p. 214)

- **Wegener's Granulomatosis**: inflamed friable mucosa, ulcerative septal perforation, saddle nose deformity, epistaxis
- **Periarteritis Nodosa**: nasal mucosal lesions
- **Lupus Erythematosis**: ulcerated nasal septum (nasal perforation)

Nasal Anatomical Abnormalities

Valvular Collapse

- Pathophysiology: weak structural support of nasal valve results in inspiratory obstruction
- Causes: congenital, trauma, iatrogenic (excessive cartilage removal at intercartilaginous junction), aging
- SSx: nasal airway obstruction on inspiration
- Dx: **Cottle maneuver** (demonstrates decreased nasal resistance by pulling superiorly on medial maxilla skin to open valve)
- Rx: mechanical cartilaginous spreader grafts, battons placed over the valve, traction sutures, or may consider nasal stent apparatus at night

Septal Deviations

- Types: traumatic and congenital
- Common Defects: spurs, crests, dislocation of quadrangular septal cartilage, buckling
- SSx: unilateral nasal obstruction (may be bilateral), hyposmia, epistaxis, recurrent sinusitis
- Dx: anterior rhinoscopy

Surgical Management

- **Submucous Resection**: obstructing cartilaginous and bony portion of the nasal septum is removed
- **Septoplasty**: removal of deviated cartilaginous and bony septum with reinsertion after remodeling and repositioning (preserves support system, less risk of perforation)
- Indications: nasal obstruction (deviated nasal septum), epistaxis, chronic sinusitis (when septum is obstructing), access for transseptal sphenoidotomy, headache from an impacted spur, septal neoplasia (rare)
- Complications: perforation, saddle nose deformity (over resecting cartilage anteriorly), cribriform plate fracture, septal hematomas, anosmia, septal abscess, bleeding

Septal Perforations

- Causes: septoplasties (most common cause, >50%), infections (tertiary syphilis), trauma (nose picking), neoplasms, granulomatous disease, vasculitis, cocaine abuse, corticosteroid nasal spray
- Dx: anterior rhinoscopy, consider biopsy of granulation tissue or abnormal mucosa to evaluate for malignancy, sarcoidosis, tuberculosis, and other granulomatous diseases
- SSx: crusting, epistaxis, whistling, obstructive sensation from turbulent flow, may be asymptomatic
- Rx
  1. saline irrigation, emollients
  2. consider sliding or rotating mucoperichondrial flaps with or without a fascial graft; contraindicated for large perforations (approximately >2 cm of vertical height), cocaine abusers, malignancy, granulomatous or vascular diseases
  3. silastic button

Septal Hematomas

- Pathophysiology: hemorrhage (usually from trauma) collects beneath mucoperichondrium and mucoperiosteum resulting in elevation of the mucosa off the cartilaginous septum (loss of vascular supply)
- SSx: unilateral obstruction (may be bilateral), septal swelling
- Complications: septal abscess, cavernous sinus thrombosis, saddle nose deformity

- Rx: immediate evacuation of hematoma, nasal packing, and antibiotic prophylaxis

Olfactory Dysfunction

Evaluation

History and Physical Exam

- Quality of Olfactory Dysfunction: anosmia, hyposmia (decreased olfactory acuity as seen in smokers, postmenopausal, elderly), phantosmia (sense odors that are not present), versus hyperosmia (heightened sense of smell as in hunger, as seen in cystic fibrosis, Addison disease), single-sided (obstructing, traumatic, infectious) versus bilateral olfactory loss
- Contributing Factors: history of upper respiratory infections, sinusitis, allergy, trauma, toxins, medications
- Associated Symptoms: changes in sense of taste (80% of flavor is appreciated from olfaction), other cranial nerve involvement (diplopia, hearing loss, hoarseness, etc.)
- Physical Exam: sinus and rhinology evaluation including nasal endoscopy to evaluate for obstruction, otologic examination to evaluate potential injury to the chorda tympani, full neurological work-up to determine other possible coexisting defects

Diagnostic Tests

- **CT of paranasal sinuses**: mainstay for complicated olfaction disorders or unclear etiology
- **MRI**: examine olfactory bulb and tracts
- **Olfactory Tests**: includes scratch and sniff identification tests, odor vials (including ammonia)
- **Taste Testing**

Causes

Obstructive Nasal and Paranasal Disease

- **most** common etiology of anosmia
- Pathophysiology: obstruction may compromise airflow to olfactory bulb
- Common Causes: mucosal edema, tumors, nasoseptal deformities, polyps
- Rx: address underlying cause, relieve obstruction

Upper Respiratory Infection

- **second** most common etiology of anosmia
- may cause parosmia (distorted perception of smell)
- Pathophysiology: may be secondary to viral-induced neuronal injury (**Essential Anosmia**), epithelial damage, or obstruction
- Rx: no effective treatment

Head Trauma

- **third** most common etiology of anosmia
- Pathophysiology: shearing forces injure the axons of olfactory neurons at cribiform plate (more common in occipital injuries)
- Rx: no effective treatment

Other Causes

- Congenital: familial dysautonomia, Kallmann's syndrome (autosomal dominant, hypogonadotrophic, anosmia secondary to incomplete olfactory bulb and stalk, hypothalamus, or olfactory epithelium)
- Tumors: frontal or temporal lobe lesions, esthesioneuroblastoma, meningiomas, pituitary adenomas
- Aging Effects: Parkinson's, Alzheimer's (typically causes parosmias)
- Medications and Toxins: smoke, sulfur dioxide, putrid gases, cocaine, cadmium, heavy metals, radiation, chemotherapy

Epistaxis

Introduction

- trauma (including nose picking and vigorous nose blowing) and mucosal dehydration are the most common cause of epistaxis
- hypertension, aspirin (and other platelet inhibiting medications), and alcohol abuse account for the most common causes of **refractory** epistaxis
- colder temperatures and dryness (winter seasons) increase risk of vascular injury
- nasoseptal deformities may result in epistaxis secondary to the drying effects of turbulent airflow

Kiesselbach's Plexus (Little's area)

- confluence of arterial vessels at the anterior nasal septum

- susceptible to bleeding due to the fragile mucosa and tightly adherent to underlying mucosa affording little resistance to mechanical stress
- Contributing Vessels: anterior ethmoidal, superior labial, greater palatine, and sphenopalatine arteries

Osler-Weber-Rendu Syndrome (Hereditary Hemorrhagic Telangiectasia)

- Pathophysiology: autosomal dominant → defect in contractile elements (elastic and muscular layers) of vessels, results in arteriovenous malformations
- SSx: friable mucosa, visceral and mucosal telangiectasia (tongue, oral mucosa, colon, lung), intermittent epistaxis, intracranial hemorrhage (neurological symptoms), hematemesis
- Rx: septoplasty or septodermoplasty (requires STSG, amniotic, or myocutaneous grafts for coverage after removing telangiectatic area), embolization, laser ablation

Evaluation and Management

History and Physical Exam

- after evaluating the ABCs (airway, breathing, intravenous access), ideally should perform a systematic evaluation of the patient prior to controlling the bleeding (may not be possible for heavy bleeding, may consider neosynephrine soaked cotton pledgets as a temporizing procedure)
- Characterize Epistaxis: estimate amount of blood loss, length of time of epistaxis, intermittent versus continuous bleeding, and side of bleeding; previous episodes, hospitalizations, packing, or other management for epistaxis
- Medical History and Blood Dyscrasias: hypertension, arteriosclerosis, leukemia, idiopathic thrombocytopenic purpura, von Willebrand's disease, renal and hepatic failure, anemia, hemophilia (higher risk of arterial, pulsatile bleeding)
- Medications: antiplatelet medications (ASA), anticoagulants (coumadin, heparin)
- Social History: cocaine abuse, alcoholism, smoking
- Toxin Exposure: ammonia, sulfuric acid, gasoline, phosphorus (associated with nasal dryness and crusting)
- Other Contributing Factors: previous septal or nasal surgery, recent trauma to nasal bone or septum, facial skeleton, dry environment, high altitude living (home CPAP ventilators or oxygen), symptoms of allergy, sinusitis, rhinitis, URI (typically short-lived bleeding)
- Think "KITTENS" for differential diagnosis (see Table 1–3)

TABLE 1–3. Differential Diagnosis of Epistaxis: KITTENS Method*

(K) Congenital	Infectious & Idiopathic	Toxins & Trauma	Tumor (Neoplasia)	Endocrine	Systemic
Nasoseptal deformities	Infectious rhinitis/sinusitis	Nasal and septal fractures	Juvenile nasopharyngeal angiofibromas	Pheochromocytomas (hypertensive crisis)	Granulomatous diseases
Osler-Weber-Rendu syndrome	Mucosal dehydration	Foreign bodies	Other benign or malignant sinonasal tumors		Vasculitis
	Escaped blood from gastrointestinal bleeding, hemoptysis, etc...	Medication			Blood dyscrasia (hemophilia, aspirin abuse, chronic renal failure)
		Environmental toxic agents			Hypertension
		Iatrogenic			
		Nasal picking			

*No Neurological causes of epistaxis

Physical Exam and Initial Ancillary Tests

- patient should sit up with body tilted forward to allow blood to be spit out and not swallowed
- initial attempt to stop bleeding by applying pressure to nasal alae for several minutes
- apply decongestant/anesthetic agents (4% cocaine or 0.25% phenylephrine HCL)
- acquire adequate lighting (head lamp), nasal speculum, bayonet forceps, frazier and Yankhauer suctions available to suction clot and attempt to localize active bleeding (examine for escoriations, foreign bodies, masses, nasoseptal deformities, etc.)
- for chronic or recurrent epistaxis without an obvious bleeding source patient should undergo an endoscopic exam
- Lab Tests: PT/PTT, bleeding time, liver function tests, creatinine, CBC, type and cross

Medical Management

- Acute Management: may need to correct hypovolemia (3:1 Rule: for every 100 cc of blood loss, replace with 300 cc of crystalloid fluid), hypertension (antihypertensive agents), or coagulopathy (fresh frozen plasma, platelets, cryoprecipitate)
- Chronic Management: hypertonic nasal spray and humidification; long-term medical management of hypertension; antimicrobial ointment to excoriated lesions; avoid excess straining, nose blowing, and digital manipulation

Cauterization

- may use silver nitrate, chromic acid pearls, electrocautery (for deeper penetration and posterior bleeding), cryotherapy
- laser cauterization may be considered for vascular malformations
- endoscopic instrumentation may be used for posterior and difficult to visualize bleeding
- Indications: minor bleeding, single bleeding points, easily visualized regions (Kiesselbach's plexus)
- Advantages: simple, quick, minimal tissue damage, no packing required
- Disadvantages: allows for coagulation of superficial vessel only, risk of perichondrial exposure, septal perforation (avoid cauterization of both sides of the septum at similar points), cartilage injury

Anterior Nasal Packing

- **Gelfoam, Cellulose, or Microfibrillar Collagen Packing**: provides a procoagulant effect, gentler packing, useful for coagulopathies,

dissolves (does not require removal), may be placed after initial coagulation
- **Nasal Tampons and Expandable Sponges**: expand with instillation of water to provide pressure against the nasal mucosa
- **Vaseline Strip-Gauze**: formal anterior packing placed to posterior choanae, controls most posterior bleeding
- keep packing in place for 3–5 days to allow vessel to develop a mature thrombus
- may supplement with thrombin, oxycellulose, or fibrin
- Indications: acute or recurrent epistaxis after failed medical management or cauterization
- Advantages: simple, proper formal strip-gauze pack controls most posterior bleeding, does not require inpatient monitoring
- Disadvantages: results in nasal obstruction, risk of pressure necrosis (nasal and septal cartilage), hypoxia, sinusitis, bacteremia, and epiphora; requires prophylactic antibiotics to reduce risk of otitis media, sinusitis, and toxic shock syndrome

Posterior Nasal Packing

- gauze, sponge pack, Foley catheter, pneumatic nasal catheters, or tonsillar packing is placed to close off the choana to prevent escape of bleeding into the nasopharynx
- requires a formal anterior pack for stability
- **Nasal Balloons**: catheter with two balloons (one placed in the nasopharynx and the other in the nasal cavity), designed for easier placement of a posterior pack, provides less trauma and is simple to adjust pressure
- Indications: failed anterior packing, skull base trauma, hemorrhage from a major branch of the sphenopalatine artery
- Advantages: may be inserted for severe bleed in the emergency room or office
- Disadvantages: risk of airway compromise (requires hospital monitoring), requires patient cooperation (painful), may require intubation or general anesthesia, eustachian tube dysfunction (hearing loss), other risks similar to anterior nasal packing

Embolization

- Indications: intractable nasal hemorrhage, surgically inaccessible sites, inoperable candidates
- Advantages: diagnostic (defines bleeding site) and therapeutic, may be repeated, can be done under local anesthesia

- Disadvantages: risk of embolic event (pulmonary emboli, stroke), requires active bleeding, facial pain

Vascular Ligation

- Indications: uncontrolled epistaxis (typically posterior bleeding), identifiable bleeding site
- Techniques
  1. **Anterior and Posterior Ethmoidal Artery Ligation:** approach from a Lynch incision, anterior ethmoid artery is located **14–18 mm** posterior to frontoethmoid suture line, posterior ethmoid artery is located **10 mm** posterior to anterior ethmoid foramen, the optic nerve is located **4–5 mm** posterior to the posterior ethmoid foramen
  2. **Maxillary Artery or Sphenopalatine Artery Ligation:** may be approached transantrally, transorally, or endoscopically
  3. **External Carotid Artery Ligation:** severe uncontrolled, life-threatening bleeding, ligate above the origin of the lingual artery
- Advantages: decreases pressure gradient in nasal vessels to allow clotting
- Disadvantages: periorbital ecchymosis, possible recurrence from collateral circulation, risk of retrobulbar hematoma, hemorrhage, optic nerve and infraorbital nerve injury, clip dislodging

Septoplasty/Submucous Resection

- Use: septal defects (unable to pack), Osler-Weber-Rendu Syndrome
- Advantages: removes diseased mucosa and replaces with STSG (**Saunder's Dermoplasty**), reduces drying effect by decreasing turbulent airflow
- Disadvantages: risk of septal perforation

ALLERGY AND RHINITIS

Allergic Rhinitis

The Allergic Response

Primary Reaction Phase

- Type I immediate hypersensitivity (*see* Table 1–4)
- occurs within 5 minutes of allergen exposure with maximum effect at 15 minutes
- allergen recognition by IgE antibodies stimulates mast cells and basophils (via the IgE Fc receptor)

TABLE 1–4. Hypersensitivity Types

	Type	Mediators	Reaction
I:	Anaphylactic	IgE	• immediate, self-limiting • IgE mediated, stimulates mast cells and basophils which release histamine and other inflammatory mediators
II:	Cytotoxic	IgG, IgM	• IgG, IgM multivalent binding to phagocyte or complement • eg, Transfusion reactions, Goodpasture's Syndrome, Bullous Pemphigoid
III:	Immune Complex	IgG, IgM, IgA	• antibody and complement complexes cause increased blood viscosity • removed by reticulo-endothelial system • eg, Renal Deposition, Arthritis, Glomerulonephritis, Serum Sickness
IV:	Cell-mediated	T-cells	• Delayed-type Hypersensitivity reaction (T-cell mediated) • eg, Graft Rejection, Contact Dermatitis
V:	Interference with Receptor	Ig	• antibody "resembles" a ligand and thus blocks or stimulates the receptor • pathophysiology of autoimmunity (eg, Hashimoto's Thyroiditis, Myasthenia Gravis, Graves' Disease)

- degranulation occurs after cross linking of the IgE (via a calcium influx trigger) releasing inflammatory mediators resulting in a net increase in local vascular permeability and proteolysis
- Mediators Released: histamine, serotonin, heparin sulfate peptide, arachidonic acid derivatives (leukotrienes, prostaglandins)
- rechallenged allergens stimulate mast cells more quickly and require less antigen load

Secondary (Late) Reaction Phase

- occurs 4–6 hours after acute phase
- migration of inflammatory cells (neutrophils and eosinophils) and continued activated basophils release a second phase of mediators (mast cells do not remain active)

Diagnosis of Allergy

History and Physical Exam

- Nasal SSx: sneezing, congestion, rhinorrhea
- Ocular SSx: redness, itchiness, watery, conjunctivitis, burning
- Otologic SSx: eustachian tube dysfunction, middle ear effusions
- Laryngeal SSx: scratchiness, dry, irritated, cough
- Other SSx: seasonal pattern (eg, in the upper Midwest tree pollen allergies occur between April–May, grass pollen May–July, weed pollen July–frost, molds year-round), food hypersensitivity, fatigue
- PE: clear rhinorrhea, congested turbinates, periorbital puffiness, nasal tip crease ("allergic salute"), open-mouthed breathing ("adenoid facies"), prominent pharyngeal lymphoid tissue, conjunctivitis
- Associated Disorders: chronic sinusitis (obstruction from mucosal edema), nasal polyps, asthma, otitis media with effusion

Adjunctive Testing

- **Nasal Endoscopy**: evaluate for nasal polyps, osteomeatal unit obstruction, adenoid tissue
- **Nasal Smear**: obtained from inferior turbinate mucosa, >25% eosinophils on nasal smear suggests allergy (neutrophils suggest infection)
- **Total Serum IgE**: not accurate or cost-effective

Skin Allergy Testing

- **Scratch Test**: scratch skin with placement of allergen or scratch with allergen, has largely been replaced for more objective and more reliable techniques
- **Prick Test**: series of allergens are inserted by needle into skin, positive "wheal-and-flare" reactions are compared to controls; rapid and safe test, risk of anaphylaxis, misses less sensitive allergy, grading is subjective
- **Intradermal Test**: similar to prick test except allergen is placed intradermally; more sensitive than prick test, however, more time-consuming and painful, risk of anaphylaxis, and subjective grading

- **Skin Endpoint Titration**: series of increasing concentrations of specific allergen are introduced intradermally to titrate to a positive response, useful for determining immunotherapy concentrations, highly sensitive and determines quantitative measurements, however, time-consuming

In Vitro Allergy Testing

- **Radioallergosorbent Test** (**RAST**): serum reacts with a series of known allergens, radiolabeled anti-IgE identifies specific antigen-IgE complexes
- **Enzyme-linked Immunosorbent Assay** (**ELISA**): similar to RAST except fluorescing agents are used for markers of antigen-IgE complexes
- Indications: equivocal skin test results, high risk of anaphylaxis (severe asthma, prior history), skin disorders (eczema), uncooperative patient (children and infants), failed immunotherapy (negative skin test is not an indication for in vitro allergy testing)
- Advantages: **highly specific**, no risk of anaphylaxis, no effect from skin condition (skin color, eczema, dermatographia) or medications (β-blockers, antihistamines, tricyclics)
- Disadvantages: **less sensitive**, requires up to 1–2 weeks for results, more expensive

Management

Anaphylaxis

1. **ABC's:** establish airway, oxygenation, and IV access
2. inject up to **0.3 ml** of epinephrine intramuscularly (IM)
3. consider **dopamine** for hypotension
4. if needed repeat injection of up to **0.3 ml** of epinephrine IM
5. add **diphenhydramine 50 mg, dexamethasone 4 mg,** and **cimetidine 300 mg** through IV
6. if needed repeat injection of up to **0.3 ml** of epinephrine IM

Level 1: Avoidance, Symptomatic Relief

Avoidance: based on **specific** allergy

- Dust: mattress cover, foam pillows, plastic case, low carpet or hardwood floors, frequent dusting and vacuuming, may apply benzyl benzoate to carpet to kill mites, consider synthetic carpets
- Molds: disinfect bathroom, clean furnace, dehumidify basement, clean refrigerator, avoid gardening, address potential sources of molds indoors (plants, old shoes, curtains)

- Pollens: air conditioning, air cleaners, keep windows closed, avoid cutting grass
- Animals: keep out of bedroom, use special shampoos
- high efficiency particulate air filters may be used for all airborne allergies
- home humidity prevents nasal dryness
- masks may be helpful in unavoidable allergy exposure

Symptomatic Medications

- **Nasal Saline Irrigations**: removes nasal mucus and crusts, aids in mucociliary clearance, thins tenacious mucus
- **First Generation Antihistamines** (H_1-receptor agonists): primarily for sneezing, itching, and rhinorrhea; side effects include sedation, dryness, confusion tolerance, and aggravation of prostatism (eg, diphenylhydramine, chlorpheniramine, promethazine, hydroxyzine)
- **Second Generation Antihistamines** (H_1-receptor agonists): lipophobic (does not cross blood-brain barrier) therefore less sedating, inhibit release of inflammatory mediators (eg, loratadine, astemizole, certirizine, fexofenadine, terfenadine)
- **Phenylamines** (α-adrenergic antagonist): vasoconstrictive effect decreases mucosal congestion; side effects include central nervous system stimulation (anxiety, anorexia), cardiac arrhythmias, hypertension, seizures, insomnia, psychosis
- **Imidazolines** (topical adrenergic antagonist) decreases mucosal congestion locally; risk of rebound nasal congestion (rhinitis medicamentosa)
- **Topical Glucocorticoids**: local reduction of inflammatory cells in nasal mucosa, decreased capillary permeability, reduces edema; minimal side effects (epistaxis, candidiasis, nasal dryness), decreases both acute and late phase reactions
- **Cromolyn**: stabilizes mast cells preventing release of mediators in acute and late phase reactions, effective only for prophylaxis; minimal side effects (sneezing, epistaxis, nasal irritation)

Level 2: Management of Complicating Factors

- must evaluate and treat potential concurrent disorders which may mimic allergy including vasomotor rhinitis, sinusitis, and rhinitis medicamentosa before changing treatment regimens

Level 3: Chronic Symptoms (Corticosteroids)

- most potent medication for symptomatic relief
- Mechanism of Action: decreases inflammatory migration, blocks arachidonic metabolites, decreases vascular permeability

- may be given orally, nasal aerosols, or via intraturbinal injections
- Side Effects of Oral Corticosteroids: increased gastric acid production (consider prophylactic concurrent H_2-blocker), hypertension, masks signs of infection, sodium retention, hypokalemia, posterior subcapsular cataracts, central nervous system stimulation (psychosis, seizures, insomnia), menstrual irregularities, aseptic necrosis of femoral head

Level 4: Immunotherapy

- Indications: unresponsive to medical therapy, severe symptoms, allergens that can not be avoided
- Advantages: suppresses allergy
- Disadvantages: patient must be reliable for multiple injections, requires a chronic regimen (3 years), risk of worsening symptoms and anaphylactic shock
- Contraindications: pregnancy (anaphylaxis risks hypoxia in fetus), autoimmune disorders, immunological compromised patients, β-blockers (increases sensitivity to allergens), easily avoidable allergens, noncompliant patients, food allergens
- Mechanism of Action: uncertain, initial small doses of allergen cause a rise in allergen-specific IgG which prevents binding of IgE, IgE may also become "exhausted"

Churg-Strauss Syndrome

- Pathophysiology: unknown etiology, causes angiitis and allergic granulomatosis
- Triad: hypereosinophilia, allergic rhinitis, asthma
- Other SSx: nasal polyposis, nasal obstruction, septal lesions, lung lesions, systemic vasculitis
- Rx: corticosteroids

Nonallergic Rhinitis

Infectious Rhinitis

Viral Rhinitis (Coryza, Common Cold)

- Pathogenesis: spread via infected droplets
- Common Viral Pathogens: rhinovirus (most common, >100 types), parainfluenza, adenovirus, enterovirus, respiratory syncytial virus
- SSx and Stages
  1. **Dry Prodromal Stage** (initial phase): nasal drying and irritation, low-grade fever, chills, general malaise, anorexia

2. **Catarrhal Stage** (second stage): watery clear rhinorrhea, anosmia, congestion, lacrimation, worsening of constitutional symptoms
3. **Mucous Stage:** thickened rhinorrhea (greenish and foul smelling if secondarily infected), improved constitutional symptoms

- Dx: clinical history and exam
- Rx: no cure for the common cold; antibiotics should be given for suspected bacterial infections only; symptomatic therapy includes decongestants (topical and systemic), antihistamines, ipratropium bromide sprays, hydration, humidification, nasal saline irrigations, analgesics, mucolytic agents

Bacterial Rhinitis

- typically secondarily infected viral rhinitis
- Common Bacterial Pathogens: *Pertussis*, *Diphtheria*, Group A *Streptococcus*, *Chlamydia*
- SSx and Stages: similar to above, however, rhinorrhea may be thickened, greenish, and foul smelling
- Dx: clinical history and exam
- Rx: antibiotic regimen, symptomatic therapy similar to viral rhinitis

Rhinoscleroma

- noncontagious
- Pathogen: ***Klebsiella rhinoscleromatis*** (Frisch's bacillus)
- Risks: endemic to East Europe, North Africa, South Asia, Central and South America
- SSx and Stages: (stages may last years)
  1. **Catarrhal:** persistent purulent rhinorrhea, nasal honeycomb-color crusting
  2. **Granulomatous:** small, painless granulomatous nodules in upper respiratory tract (including glottis and subglottis)
  3. **Sclerotic:** lesion heals with extensive scarring (dense fibrotic narrowing of nasal passage)
- Dx: biopsy and culture, serum antibodies
- Histopathology: **Mikulicz's cell** (foamy histocytes containing the bacteria, "moth eaten" cytoplasm), **Russell bodies** (bloated plasma cells with bifringent inclusions), pseudoepitheliomatous hyperplasia
- Rx: long-term antibiotics as dictated by culture and sensitivity, debridement, consider laser excision or cryotherapy

Rhinosporidiosis

- Pathogen: ***Rhinosporidium seebri*** (sporangium with a thick-walled cyst)

- Risks: endemic to Africa, Pakistan, India, Sri Lanka, spread from contaminated water (public bathing)
- SSx: **friable**, "strawberry" red (vascular) polypoid nasal lesion (epistaxis, obstruction)
- Dx: culture and biopsy
- Histopathology: pseudoepitheliomatous hyperplasia, submucosal cysts, fungal sporangia with chitinous shells
- Rx: surgical excision with cauterization of the base and oral antifungal agents, corticosteroid injections, may consider dapsone

***Rhinocerebral Mucormycosis* (see *Paranasal Sinus*)**

***Phaehyphomycosis* (see *Paranasal Sinus*)**

Nonallergic Chronic Rhinitis

- Causes: recurrent or chronic inflammation of the nose and sinus secondary to a variety of causes including vasomotor disease, irritant and toxin exposure, persistent environmental factors (changes in temperature and humidity), pregnancy, medications, endocrine disease (diabetes, thyroid disease), infections, granulomatous disease
- SSx: congestion, nasal obstruction, watery rhinorrhea, congested nasal mucosa, throat-clearing, fatigue, malaise
- Granulomatous Nasal Diseases: tuberculosis, leprosy, rhinoscleroma, syphilis, fungal, protozoan, sarcoidosis, Wegener's disease
- Dx: anterior rhinoscopy, biopsy for suspected granulomatous diseases
- Rx: address underlying cause, symptomatic medications may be used for temporary relief (decongestants, nasal sprays, etc.)

Nonallergic Rhinitis with Eosinophilia Syndrome (NARES)

- nasal eosinophilia without allergy
- SSx: symptoms of perennial rhinitis
- Dx: allergic symptoms with negative allergic tests
- Rx: symptomatic relief similar to allergic rhinitis (nasal corticosteroids, antihistamines, decongestants)

Rhinitis and Pregnancy

- Pathophysiology: unclear, may be multifactorial (cholinergic effects from increased estrogen may contribute)
- SSx: rhinitis, pale-blue mucosa, turbinate hypertrophy, manifests near the end of the first trimester, resolves after delivery

- Dx: avoid skin testing (risk of anaphylaxis), may use RAST testing and nasal cytology
- Rx: refractory to most regimens, conservative management (nasal saline irrigations, avoidance of allergens, may consider nasal steroids), avoid decongestants (may place fetus at risk), consult obstetrician for treatment

Rhinitis Sicca Anterior

- Pathophysiology: dry, raw nasal mucosa secondary to a variety of causes including changes in temperature and humidity, nose picking, dust, and other irritants
- SSx: dryness, nasal irritation, nasal crusting, epistaxis, septal perforation
- Dx: clinical history and exam
- Rx: saline irrigation, topical antibiotics, oil based nasal ointments

Atrophic Rhinitis (Ozena)

- Pathophysiology: mucosal glands and sensory nerve fibers degenerate, epithelium undergoes squamous metaplasia, destroyed mucociliary transport
- Causes: excess nasal surgery (excessive turbinectomy), suspected genetic component (more common in East Asia, Egypt, Greece), endocrine abnormalities, nutritional deficiencies (vitamin A or D, iron deficiency), chronic bacterial infections, trauma and irritant exposure
- SSx: mucosal and turbinate atrophy, wide nasal cavity, nasal crusting, offensive odor, epistaxis, anosmia, may have paradoxical sensation of nasal obstruction
- Dx: anterior rhinoscopy
- Complications: increased risk for secondary infection
- Rx: saline irrigation, oil based ointment impregnated nasal tampons, vitamin A and D and iron supplements, systemic or topical antibiotics (for secondary infections), consider nasal vestibuloplasty or periodic nostril closure for failed medical therapy

Anhidrotic Ectodermal Dysplasia

- Pathophysiology: X-linked genetic disorder resulting in scant mucus production and atrophic rhinitis
- SSx: atrophy of inferior and middle turbinates, fevers, recurrent otitis media, malodorous rhinorrhea, nasal crusting
- Triad

1. anhidrosis

2. hypotrichosis
3. anodontia

- Rx: pressure equalization tubes, saline irrigations, nasal hygiene, denture appliances

Rhinitis Medicamentosa

- Pathophysiology: semi-ischemic state secondary to any **topical nasal decongestants**, results in rebound congestion from decreased vasomotor tone, increased parasympathetic activity, increased vascular permeability (also results in decreased ciliary activity)
- may be irreversible if vagal tone becomes atonic
- SSx: mucosal edema, nasal obstruction, dryness, irritation
- Rx: discontinue topical decongestants, aggressive saline irrigation, oral decongestants, nasal steroid spray, may consider nasal stents, submucosal steroids, or short-term oral corticosteroids; avoid by limiting topical decongestants to 3–5 days

Vasomotor Rhinitis

- Pathophysiology: rhinitis secondary to overactive parasympathetic activity from a wide variety of triggers (*see below*)
- SSx: similar symptomatology to allergic rhinitis except with negative allergy evaluation, morning rhinorrhea, alternating sides, pale nasal mucosa
- Dx: diagnosis of exclusion, negative allergy work-up

Triggers

- Environmental: humidity and temperature changes, dust, smoke, pollution
- Endocrine and Metabolic: pregnancy, oral contraceptives (estrogen inhibits anticholinesterases), hypothyroidism
- Medications: antihypertensives, antipsychotics, cocaine
- Psychotropic: anxiety, stress, exercise

Management

- attempt elimination of irritants and address causal factors if possible
- Medical Management
  1. anticholenergic nasal sprays (ipratropium bromide)
  2. corticosteroid nasal sprays
  3. hypertonic saline nasal sprays
  4. may consider short course of oral and topical decongestants or antihistamines

- Surgical Management: indicated for refractory cases
  1. **Surface Turbinate Cautery:** may also consider cryotherapy or laser
  2. **Septoplasty:** removes mechanical points of irritation
  3. **Partial Turbinectomy:** reduction of lower and possible middle turbinates, total turbinectomy risks atrophic rhinitis
  4. **Division of Parasympathetic Fibers:** most commonly section the vidian nerve

PARANASAL SINUSES

Sinusitis

Introduction

Pathophysiology

- Anatomical (compromised patency of ostia causes hypooxygenation and impairment of sinus drainage)
- Dysfunction of Cilia Motility
- Change in Quality of Secretions
- Immune Dysfunction

Pathogens

Acute

- Viral: rhinovirus (most common)
- Bacterial: *S. pneumoniae* (most common bacterial agent), *H. influenzae, B. catarrhalis*, *S. aureus*, *S. pyogenes*
- Fulminant Fungal: *Aspergillosis*, *Phaehyphomycosis*, *Mucor, Rhizopus*

Chronic: anaerobes, *S. aureus, H. influenzae*, 1–2% fungal

Complicated Sinusitis

- Cystic Fibrosis: *P. aeruginosa, S. aureus*
- Nosocomial: *P. aeruginosa, Klebsiella, Enterobacter, Proteus*
- Immunocompromised: similar to nonimmunocompromised patients, however also susceptible to *Aspergillus, Rhizopus, Fusarium, P. aeruginosa, S. aureus*

Evaluation

Symptoms and Physical Exam Findings

- Acute Sinusitis Symptoms: facial pain and tenderness worse with straining of bending over, pressure headache (frontal, occipital headaches may arise from sphenoid sinusitis), nasal congestion,

postnasal drip, nasal obstruction, nasal discharge (mucopurulent, serous, mucoid), cough and halitosis (especially in pediatric patients)
- Chronic Sinusitis Symptoms: presents with more subtle symptoms of nasal obstruction, less fever and pain complaints
- Associated Symptoms: anosmia, loss of taste (chronic sinusitis), allergic components (sneezing, watery eyes), fever, malaise, lethargy, cough, eustachian tube dysfunction, anosmia/hyposmia
- Physical Exam: rhinoscopy/nasopharyngoscopy (osteomeatal unit obstruction, nasal masses and obstruction, purulence, hypertrophied adenoid tissue); decreased sinus transillumination (not reliable); frontal and maxillary tenderness (acute sinusitis); polyps and edema (chronic sinusitis)
- DDx of Facial Pain: rhinogenic, migraines and other headaches, dental disease, neuralgias, temporomandibular joint disease, ocular disease, tonsillitis, pharyngitis, otogenic disease, intracranial pathologies, hypertension, temporal arteritis

Evaluate for Causes and Risk Factors

- Anatomical: deviated septum, mucosal edema (rhinitis, allergic), hypertrophic adenoids, nasal masses, nasal foreign bodies, nasogastric tubes, nasal packing, facial fractures, concha bullosa, lateral deviated uncinate process, paradoxical middle turbinate, uncinate hypoplasia, nasal polyposis
- Ciliary Dysfunction: (*see below*)
- Medical Condition: immunocompromised (HIV, diabetes, malnutrition), cystic fibrosis, smoker, elderly
- Local Causes: apical dental infection, trauma, barotrauma

Imaging Studies

Plain Radiography

- Indications: screening study for acute sinusitis
- evaluates presence of air-fluid levels, opacification, and bone destruction
- high rate of false positives and false negatives to evaluate for chronic sinusitis

CT of Paranasal Sinuses

- Indications: severe acute sinusitis, medical failure of chronic sinusitis, diagnosis of epistaxis, nasal or sinus tumors, nasal polyps, CSF leak, trauma, preoperative films
- Evaluation
  1. examine distribution of mucosal disease (mucosal thickening, air-fluid levels suggest acute inflammatory process)

2. inspect development of sinus (symmetry, aeration of sinus cavities)
3. examine nasal structures, airway, and access
4. evaluate for underlying causes of disease (OMC patency, paradoxical turbinates, nasal septal defects, concha bullosas)
5. examine for anatomical variations and landmarks (cribiform plate, posterior ethmoidal height, thickness of skull base, optic nerve, orbital dehiscence, carotid artery)

MRI of Paranasal Sinuses

- improved soft tissue detail, poor bone resolution
- Indications: complicated sinusitis (intracranial and intraorbital extension), evaluation of soft tissue masses (neoplasms), fungal sinusitis (hypodensity in T2–weighted sequences from the presence of metallic proteinaceous material, magnesium, iron, and calcium)

Ancillary studies

- sinus cultures (may be obtained through an anterior maxillary puncture or from an endoscope) indicated for failed medical management, complicated sinusitis (sepsis, orbital infection, intracranial extension), immunocompromised patients
- immunological profile (qualitative immunoglobulins including IgG subclasses)
- ciliary biopsy
- allergy testing

Management

Acute Sinusitis (<1 month)

- Antibiotics: may treat empirically with first-line oral antibiotics with gram positive and gram negative coverage *(*amoxicillin, amoxicillin-clavulanate, trimethoprim-sulfate, cefuroxime, or azithromyocin) for 10–14 days, consider changing antibiotics if no improvement after 2–3 days, if no improvement after 1 week may consider sinus lavage for culture and sensitivity
- Improve Nasal Clearance: nasal saline irrigations, oral and topical decongestants, mucolytic agents, humidity
- Symptomatic Medications: analgesics, antipyretics
- Address Risk Factors: smoking cessation, septoplasty, remove nasogastric tube, antireflux regimen, etc

Acute Frontal Sinusitis

- frontal sinusitis is treated more aggressively to avoid intracranial complications

- identified by frontal pain and tenderness and frontal sinus air-fluid levels on x-ray
- parenteral antibiotics, observation for intracranial involvement (select patients may be followed with close follow-up on an outpatient basis)
- consider surgical management if no improvement after 24–48 hours of aggressive medical management

Chronic Sinusitis: *persistent sinus infection greater than 6 weeks*

- Antibiotics: 3–6 weeks regimen with broad spectrum agents (*eg,* amoxicillin-clavulanate, cefuroxime, ciprofloxacin, clarithromycin, cefpodoxime, cefprozil)
- nasal corticosteroid sprays are useful for chronic sinusitis
- nasal hypertonic saline irrigations, oral decongestants, and mucolytic agents
- medical management often fails with chronic sinus disease, surgical management is frequently required
- allergy management (*see above*)

Surgical Management: see below *for indications and techniques*

Pediatric Sinusitis

- usually involves the maxillary sinus and anterior ethmoids (the sphenoid and frontal sinuses are less developed)
- Most Common Pathogens: *Streptococcus, S. aureus, S. pneumoniae, M. catarrhalis, H. influenzae*
- SSx: similar presentations to URI, however tend to have cough, halitosis, persistent nasal obstruction, rhinorrhea, and fever
- W/U: similar to adults, however for persistent sinusitis must consider adenoid hypertrophy, cystic fibrosis, and immunodeficiencies

Management

- Antibiotics: antibiotics are the primary therapeutic agent in pediatric sinusitis, agents used are similar to adults
- Adjunctive Medical Agents: saline irrigations, nasal corticosteroids, oral decongestants, mucolytics
- Allergy Management: (*see above*)
- **Endoscopic Sinus Surgery**: indicated for select patients who fail extensive medical management and have significant effect on quality of life
- **Adenoidectomy**: controversial relationship with sinusitis (may harbor pathogens and block drainage), may be considered as an adjunctive therapeutic option

- **Antral Lavage**: controversial efficacy, addresses only the maxillary sinus, typically require multiple lavages

Complicated Sinusitis

Fungal Sinusitis

Fungus Ball

- Pathophysiology: noninvasive fungal infection (most commonly Aspergillus)
- SSx: unilateral chronic or recurrent sinusitis, unilateral proptosis, facial hypesthesia
- Dx: CT/MRI of paranasal sinus, biopsy with culture
- Aspergillosis Histology: septated 45 degree, Y-shaped (Sabouraud's agar stain)
- Rx: adequate surgical debridement, consider adjuvant less toxic antifungal medications

Allergic Fungal Sinusitis

- Pathophysiology: fungal infection, most commonly Aspergillus or demitaceous molds (*Alternaria, Bipolaris, Curvularia, Exophilia*), becomes the antigen for an allergic response
- Risks: atopic disease, young asthmatics
- SSx: sinusitis symptomatology with allergic component (sneezing, watery eyes, periorbital edema, etc.)
- Dx: allergic evaluation for fungal elements (RAST, skin testing, nasal eosinophilia), tissue stains reveal presence of allergic mucosa with fungal hyphae, CT/MRI of paranasal sinus
- Rx: surgical debridement with complete sphenoethmoidectomy, topical and oral corticosteroids, consider adjuvant antifungal medications or immunotherapy

Chronic Invasive Fungal Sinusitis

- Pathophysiology: pathogen invades soft tissue
- Pathogens: Aspergillosis, Saprophytics (*Mucor, Rhizopus, Absida*)
- SSx: chronic sinusitis with or without symptoms of local invasion (eg, blindness, cerebritis)
- Dx: biopsy and culture, MRI of paranasal sinus (image of choice, enhancement in T2-weighted images from fungal elements), CT of paranasal sinus
- Rx: surgical debridement and long-term amphotericin B and itraconazole (1 year)

Fulminant Fungal Sinusitis and Mucormycosis (Rhinocerebral Phycomycosis)

- Pathophysiology: pathogen invades soft tissue; in **mucormycosis** pathogen invades vessel walls causing local vascular occlusion, thrombosis, infarction, and tissue necrosis
- Pathogens: Aspergillosis (most common), Saprophytics (*Mucor, Rhizopus, Absida*)
- Risks: manifests almost exclusively in **immunocompromised host** (diabetic ketoacidosis, chemotherapy, HIV, bone marrow transplant)
- SSx: in **mucormycosis** may present with necrotic black turbinates and soft palate, epistaxis, cranial nerve involvement, progresses rapidly into obtundation and death
- Dx: biopsy and culture, MRI of paranasal sinus (image of choice, enhancement in T2-weighted images from fungal elements), CT of paranasal sinus
- Mucormycosis Histology: nonseptated, 90 degree broad branching hyphae
- Rx: urgent surgical debridement and long-term amphotericin B, address underlying derangements (eg, correct ketoacidosis)

Sinobronchial Syndrome

- association of chronic sinusitis with asthma, bronchiectasis, recurrent pneumonia, and chronic bronchitis
- Pathophysiology: controversial: may be from two separate manifestations with same underlying entity or postnasal drip with bronchial seeding which may result in bronchospasm
- Dx: clinical history and exam
- Rx: aggressive management of sinusitis may better control asthma

The HIV Patient with Sinusitis

- 75% of HIV patients develop sinusitis
- Pathophysiology: increased risk of sinusitis secondary to impaired immunity, mucociliary dysfunction, and atopy
- Pathogens: similar to nonimmunocompromised patients for CD_4 count >200, however for CD_4 count <200 high incidence of unusually and more virulent organisms (fungal, CMV, *Pseudomonas*, *Myobacterium*)
- Management of Sinusitis for CD_4 count <200 (CD_4 count >200 managed similar to a nonimmunocompromised patient)
  1. may consider initially one course of empiric therapy (broad spectrum antibiotics) and sinus regimen (decongestants, hypertonic saline irrigations, mucolytics, etc)

2. aggressive early work-up (CT/MRI of paranasal sinuses, nasal endoscopy)
3. low threshold for sinus aspirate for culture and sensitivity to avoid empiric therapy
4. early surgical management

Cystic Fibrosis and Sinusitis

- **Cystic Fibrosis (CF)**: autosomal recessive multisystem disorder characterized by abnormal exocrine gland function (chronic progressive pulmonary disorder with associated pancreatic, hepatobiliary, and genitourinary manifestations)
- universally develop chronic sinusitis
- 10% associated obstructive nasal polyps
- Dx: sweat chloride test >60–90 mmol per liter, genetic screening
- Pathogens: higher risk for pseudomonal infections (*Burkholderia capacia*) and methicillin resistant *S. aureus* (MRSA)
- Management of Sinusitis
  1. attempt initial conservative medical management with mucolytics, topical corticosteroids, and hypertonic saline irrigations
  2. avoid antibiotics (may develop resistant pathogens)
  3. avoid surgical management (high recurrence rate, increases nasal scarring, patients do not tolerate long-term general anesthesia due to retained pulmonary secretions)
  4. surgery may be considered for uncontrolled pain, nasal obstruction, mucocele, unresolved fevers, and fungal infections

Mucoceles

- **Mucocele**: obstructed sinus that undergoes expansion from mucous secretion
- frontal sinus most commonly involved
- Types
  1. **Primary:** arises de novo, mucous retention cyst (*see below*)
  2. **Secondary:** arises secondary to surgery, trauma, or tumor (or other nasal mass)
- Causes: trauma, chronic sinusitis, polyposis, sinus surgery, allergy, osteoma, hyperaeration of ethmoid
- SSx: asymptomatic, dull headache that localizes to involved sinus, periorbital swelling, ocular symptoms (proptosis, diplopia)
- Dx: CT of paranasal sinus reveals expansion of sinus with opacification, rounded process of a sinus cavity or air cell, bone remodeling (thinned sinus walls)
- Complications: pyoceles, may rupture (bacteremia), orbital and intracranial involvement, pituitary abnormalities, cosmetic deformity

- Rx: endoscopic sinus surgery, open procedures reserved for failed endoscopic approaches or rare lateral lesions in frontal sinus

Mucous Retention Cysts

- Pathophysiology: serous or mucinous submucosal collection of fluid secondary to blocked glands, may be infectious or allergic in origin
- SSx: typically asymptomatic, larger cysts may cause dental pain or symptoms from sinus obstruction
- Dx: CT of paranasal sinus, sinus films (10% incidental findings), most commonly found on floor of maxillary sinus
- DDx: dental radicular or follicular cysts
- Rx: observation if asymptomatic or nonobstructing, otherwise may consider surgical management

Kartagener's Syndrome

- Pathophysiology: deficient outer dynein arm results in primary ciliary dyskinesis
- Triad
  1. chronic rhinitis/sinusitis/otitis media
  2. bronchiectasis
  3. situs inversus
- SSx: recurrent sinusitis, otitis media, and rhinitis; male infertility (sperm dysmotility)
- Dx: ciliary biopsy with phase contrast microscopy or electronmicroscopy
- Management of Sinusitis
  1. aggressive antimicrobial therapy, consider prophylactic antibiotics
  2. mucolytics
  3. avoid surgical management with "standard functional" antrostomies (do not work since there is no normal mucociliary clearance)
  4. consider "gravity dependent" surgical inferior antrostomies for refractory sinus disease

Polyposis (*see* p. 17)

Complications of Sinusitis

Orbital Complications

- Intraorbital Pathways: direct extension (especially though thin walled lamina papyracea), thrombophlebitis (valveless veins), congenital dehiscence, trauma, direct lymphatics

- Dx: CT of paranasal sinus with contrast or MRI of paranasal sinus with gadolinium
- ophthalmology consultation should be obtained with any orbital complication from sinusitis
- urgent surgical intervention should be considered for orbital abscesses (or orbital cellulitis), changes in vision, progressive involvement of symptoms despite appropriate medical therapy, relapses, involvement of opposite eye
- concurrent aggressive sinusitis regimen (parenteral antibiotics, decongestants, mucolytics, saline nasal irrigations) is indicated for any complication of sinusitis

Stages of Orbital Complications

1. **Periorbital (Preseptal) Cellulitis:** eyelid edema, erythema, tenderness; no vision changes, chemosis, proptosis, or restriction of ocular muscles; Rx: parenteral antibiotics and concurrent aggressive sinusitis regimen (decongestants, mucolytics, saline nasal irrigations)
2. **Orbital Cellulitis:** proptosis, chemosis, may cause vision changes (anterior pupillary defect), may limit extraocular muscles; Rx: endoscopic sinus surgery to drain sinuses in all cases (*controversial*), parenteral antibiotics, vision acuity checks, aggressive sinusitis regimen (decongestants, mucolytics, saline nasal irrigations)
3. **Subperiosteal Abscess:** collection of pus between bone and periosteum, chemosis, may displace globe (proptosis), restrict extraocular motion, and affect vision; Rx: urgent surgical decompression
4. **Orbital Abscess:** collection of pus in orbital soft tissue, proptosis, chemosis, restricted extraocular motion, may have no light perception (may be reversible); Rx: urgent surgical decompression
5. **Cavernous Sinus Thrombosis** (*see below*)

Cavernous Sinus and Venous Sinus Thrombophlebitis

- Pathophysiology: perinasal sinus infection → orbital extension → mural thrombus forms in vessel wall (thrombophlebitis) → propagates distally as clot softens and begins to seed
- Pathogens: *S. aureus* (most common), hemolytic *Streptococcus* and Type III *Pneumococcus*
- SSx: "picket fence" spiking fevers, toxemia, papilledema, paralysis of extraocular muscles (CN III, IV, and V), proptosis, chemosis, eyelid edema
- Dx: CT/MRI may show intraluminal enhancement, positive blood cultures, **Tobey-Ayer or Queckenstedt's Test** (tests for obstruction;

external compression of jugular vein does not cause an increase in CSF pressure but compression on nonobstructed side does increase CSF pressure), CSF may reveal high cell and protein count
- Complications: meningitis, septic metastasis (pulmonary, blood)
- Rx: parenteral antibiotics, may require ligation of internal jugular vein if septic emboli suspected, anticoagulants (*controversial*), bed rest, sinus surgery once patient is stable

Intracranial Complications

- Intracranial Pathways: congenital dehiscence, trauma, direct extension (osteomyelitis), lymphatics, olfactory nerve sheath, venous system, foramina of Breschet
- Dx: initial CT of brain with contrast or MRI of brain with gadolinium (evaluate for mass effects that may risk herniation with a lumbar puncture), lumbar puncture for cells and culture

Epidural Abscess

- pus collection between skull and dura
- higher risk from frontal osteomyelitis (direct extension)
- Pathogens: *S. aureus, E. coli, Streptococcus, Pseudomonas, Proteus*
- SSx: headaches, low grade to spiking fevers, malaise, mental status changes (may be asymptomatic)
- Rx: parenteral antibiotics, neurosurgical consultation for possible drainage procedure, sinus drainage or obliteration procedure with wide exposure of dura until healthy tissue is exposed on all sides

Subdural Abscess

- pus collection between dura and arachnoid membrane (less resistance to spread, crescent shaped enhancement on CT/MRI, does not cross midline)
- Pathogens: similar to epidural abscess
- SSx: more neurological sequellae than extradural infections (seizures, delirium, hemiplegia, aphasia), mild increase in intracranial pressure (ICP) depending on the size
- Rx: high-dose parenteral antibiotics, neurosurgical consultation for possible drainage procedure

Brain Abscess

- Pathogens: *Streptococcus, Staphylococcus*, anaerobes
- Stages
  1. **Encephalitis**: (initial invasion) fevers, headache, nuchal rigidity
  2. **Latency**: (organization of abscess, liquification necrosis) minimal symptoms, may last weeks

3. **Expanding abscess**: intracranial hypertension, seizures, paralysis
4. **Termination**: rupture of abscess, often fatal

- Rx: parenteral antibiotics, neurosurgical consultation for possible drainage procedure, concurrent sinus surgery

Meningitis

- most common form of intracranial complication from sinusitis
- highest risk from sphenoid sinusitis
- Pathogens: *H. influenzae* (type B), *Pneumococcus,* hemolytic *Streptococcus*
- SSx: headache, lethargy, nuchal rigidity, fever, **Kernig's sign** (with hip in flexion, pain is elicited with leg extension), **Brudzinski's sign** (flexion at neck causes a reflexive flexion of the legs), seizures, photophobia
- Dx: initial CT/MRI with contrast or gadolinium (evaluate for mass effects that may risk herniation with a lumbar puncture), lumbar puncture for cells and culture
- Rx: parenteral antibiotics, sinus surgery with exposure of diseased dura (if present)

Other Complications

- **Osteitis**: diagnose initially with technetium bone scan (osteoblastic activity) and gallium bone scan (inflammation), follow with gallium scans; Rx: parenteral antibiotics, surgical debridement, sinus surgery
- **Pot's Puffy Tumor**: frontal bone osteomyelitis, soft doughy swelling of forehead, high risk of intracranial extension; Rx: parenteral antibiotics, trephination, may require surgical debridement
- **Superior Orbital Fissure Syndrome**: fixed globe, dilated pupil (CN III, IV, VI), ptosis, hypesthesia of upper eyelid (CN V_1); Rx: urgent surgical decompression
- **Orbital Apex Syndrome**: similar to Superior Orbital Fissure Syndrome with added involvement of optic nerve (papilledema, vision changes)
- **Sinocutaneous Fistula**: usually begins as a frontal osteomyelitis

Sinus Surgery

Functional Endoscopic Sinus Surgery (FESS)

- Advantages: superior visualization, better precision, preserves function (recognizes normal mucociliary flow pattern at the osteomeatal complex), completeness, no external scar

- Disadvantages: requires one-handed technique, monocular vision (difficulty with depth perception)
- Contraindications: osteomyelitis, no evidence of paranasal disease on CT, inaccessible lateral frontal sinus disease
- Steps of a FESS for Sinusitis: medialize middle turbinate, excise uncinate process, anterior then posterior ethmoidectomies, sphenoidotomy, frontal recess sinusectomy, create maxillary antrostomy
- Postoperative Care: sinus packing (2–6 days), oral antibiotics for a minimum of 2 weeks, aggressive nasal hygiene to prevent adhesions (saline irrigations), nasal steroids, nasal debridement at 1, 3, and 6 weeks

Indications for Endoscopic Sinus Surgery

- chronic sinusitis, complicated sinusitis, recurrent acute sinusitis, failed medical management of acute sinusitis, fungal sinusitis
- obstructive nasal polyposis
- sinus mucoceles
- remove foreign bodies
- tumor excision, transsphenoidal hypophysectomy
- orbital decompression, dacryocystorhinotomy, orbital nerve decompression, Grave's ophthalmopathy
- choanal atresia repair
- CSF leak repair
- control epistaxis
- septoplasty, turbinectomy

Ethmoid and Maxillary Open Sinus Procedures

Caldwell-Luc

- intraoral approach to anterior maxillary wall from canine fossa above gum line, the diseased mucosa is removed from the maxillary sinus, also allows for a middle meatal antrostomy, and ethmoidectomy (transantral ethmoidectomy)
- Indications: sinus disease not obtainable by endoscopic sinus surgery, inspissated secretions, neo-ossification, cystic fibrosis
- Advantages: allows adequate exposure of inside of maxillary sinus, favorable intraoral incision
- Disadvantages: nonfunctional, damages mucosa (decreased cilia count, increased fibrosis and bone growth), risk of infraorbital nerve and dental injury (hypoesthesia to teeth and lip), transantral ethmoidectomy does not allow exposure anterior to the ethmoidal bulla

Intranasal Ethmoidectomy (without Endoscopy)

- requires medialization of middle turbinate to gain access to the ethmoid cells, diseased mucosa is removed by piecemeal forceps dissection
- Indications: largely been replaced by endoscopic sinus surgery
- Advantages: no external scar
- Disadvantages: poor visualization (increased risk of bleeding and CSF leak), poor precision, excess mucosal damage, no visualization of frontal sinus recess or inside maxillary sinus

External Ethmoidectomy

- requires ligation of angular and anterior ethmoid arteries, access gained through lamina papyracea and lacrimal fossa
- Indications: inability to obtain transnasal exposure, subperiosteal abscess or orbital abscess
- Advantages: can access ethmoid sinus in all cases
- Disadvantages: poor visualization (especially anteriorly), poor precision, excess mucosal injury, external scar

Frontal Sinus Surgery

- **Frontal Sinus Trephination**: useful to relieve pain and obtain cultures for acute frontal sinusitis, may be used as an adjunctive procedure with endoscopic sinus surgery
- **Lynch Procedure** (**Fronto-ethmoidectomy**): consists of removal of the frontal sinus floor, middle turbinate, and anterior ethmoids, easiest and quickest technique, risk of recurrent mucocele formation from stenosis of nasofrontal duct
- **Riedel Method**: consists of removal of the frontal sinus floor and anterior wall (disfiguring), allows for complete obliteration
- **Killian Method**: modification of the Riedel by preserving a bridge at the supraorbital rim to reduce deformity
- **Lothrop Method**: creates a large drainage opening into the nasal cavity by removing bilateral anterior ethmoids, middle turbinate, and frontal septum

Osteoplastic Flap with Frontal Sinus Obliteration

- Technique: bicoronal flap for exposure, "trapdoor" access to the frontal sinus via a periosteal and bone flap (requires a template patterned from a Caldwell view film), remove mucosa, obliterate cavity and occlude frontonasal recess (may use fat, muscle, or bone)

- Indications: chronic or recurrent sinusitis, mucoceles (pyoceles), frontal bone osteomyelitis, benign tumors, frontal sinus fractures, orbital or intracranial complications
- Advantages: best view of entire frontal sinus and anterior base of skull, minimal deformity, direct approach, fail-safe method to eradicate frontal sinus disease (permanent and complete removal of diseased mucosa)
- Disadvantages: technically more difficult, time consuming, requires hospitalization, risk of mucocele formation and chronic pain
- Contraindicated: aplastic frontal sinus

Approaches to the Sphenoid

- **Transseptal Approaches** are useful for pituitary tumors and CSF leaks (**transsphenoidal hypophysectomy**)
- external transethmoidal and transantral routes are less popular approaches to the sphenoid sinus

Sublabial Transseptal Approach

- Indications: most common access for transphenoidal hypophysectomy (tumors), sinusitis, and mucoceles
- Advantages: wide midline exposure, no external scar
- Disadvantages: soft tissue trauma, sensory disturbances, difficult to displace distal lateral nasal mucosa with speculum, oral contamination

Transcolumellar Transseptal Approach (External Rhinoplasty)

- Indications: transsphenoidal hypophysectomy (tumors), sinusitis, and mucoceles
- Advantages: wide midline exposure, shorter access distance, no manipulation of upper lip (shorter recovery time)
- Disadvantages: small external scar, disrupts medial crura (major tip support), narrower exposure

Endonasal Approach

- Indications: transsphenoidal hypophysectomy (tumors), sinusitis, and mucoceles
- Advantages: limited septal incisions
- Disadvantages: compromised three-dimensional exposure

Endoscopic Approaches (Transnasal, Transethmoidal)

- Transnasal Indications: hypophysectomy or biopsy (tumors), sinusitis, and mucoceles
- Transethmoidal Indications: sinusitis, mucoceles, CSF leaks, biopsy, optic nerve decompression
- Advantages: limited septal incisions, visualization, better lateral exposure
- Disadvantages: one-handed surgery, difficulty with depth perception

Complications of Sinus Surgery

Ocular and Orbital Complications

- **Blindness**: may be secondary to an indirect injury (eg, retrobulbar hematoma, *see below*) or direct injury to the optic nerve
- **Intraoperative Orbital Fat Penetration**: increases risk of retrobulbar hematoma; Rx: recognize orbital fat (orbital fat floats); avoid further trauma; may complete the FESS; avoid tight nasal packing; observe for vision changes, proptosis, or restricted ocular gaze
- **Diplopia**: orbital muscle injury, most commonly from medial rectus and superior oblique muscles
- **Epiphora**: injury to lacrimal duct system, avoid operating anterior to the attachment of the uncinate; Rx: observation initially, if no resolution then dacryocystorhinostomy

Retrobulbar Hematoma

- Pathophysiology: most commonly from retraction injury of the anterior ethmoid artery which causes increased orbital pressure that compresses the vascular supply to the optic nerve, also may occur from venous injury near the lamina papyracea
- Avoidance: maintain orientation and operate under direct vision, examine CT for dehiscence, correct coagulopathies
- SSx: ecchymosis, proptosis, conjunctival changes (chemosis), pupillary changes (afferent pupillary defect)
- Rx
  1. if noticed intraoperatively terminate case
  2. Ophthalmology consult
  3. Mannitol (1–2 g/kg), consider high-dose steroids
  4. orbital massage and place ice pack
  5. lateral canthotomy, medial external (Lynch) procedure, or orbital decompression
  6. control hemorrhage

Intracranial Complications

- **CSF leak**: occurs near the frontal recess, anterior ethmoid artery, cribiform plate, and posterior ethmoid sinus; Rx: recognize leak (clear or swirling fluid), prepare site of leak, apply graft (mucosa, fascia, or muscle flap), stabilize graft (Gelfoam), consider neurosurgical consult, consider lumbar puncture, bed rest, avoid straining
- **Intracranial Infections**: meningitis, brain abscess
- **Intracranial Hemorrhage**: uncommon

Neural and Vascular Injury

- **Anosmia**: injury to the olfactory epithelium
- **Hemorrhage**: injury to sphenopalatine, carotid, and ethmoid arteries, carotid-cavernous fistulas; Rx: control bleeding with electrocoagulation or packing, consider embolization for carotid injuries
- **Paresthesias**: injury to infraorbital, supraorbital, supratrochlear nerves

Synechia

- most common complication of endoscopic sinus surgery
- scarring most commonly occurs between middle turbinates and nasal wall
- Rx: lysis of adhesions (endoscopic approach), may consider spacers (Telfa, Merocel, Gelfilm); prevent by minimizing trauma, reduce concha bullosas and symptomatic polypoidal middle turbinates, and good postoperative care (nasal hygiene, endoscopic debridements)

Other Complications

- residual disease
- hematomas and seromas (abdominal fat graft complication of osteoplastic flap surgery)
- facial edema (especially from Caldwell-Luc procedures)
- aspiration of packing material
- toxic shock syndrome
- osteomyelitis
- tooth numbness and pain (Caldwell-Luc)
- embossment (frontal sinus obliteration)

IMMUNOLOGY

Introduction

Cell-Mediated Immunity

- **Antigen Presenting Cells** (macrophages, dendritic cells, Langerhans' cells) phagocytize antigens then present a fragment of the antigen to the surface via Major Histocompatibility Complex (MHC) Type II receptor and secretes IL-1
- **Helper T-cells** recognize the antigen presented by the MHC Type II receptor complex and are activated by IL-1, resulting in secretion of IL-2 (IL-2 up-regulates other T-cells including killer T-cells, macrophages, and natural-killer cells)
- **Killer T-cells** attack the body's own cells that have been transformed from infection or malignancy via the MHC Type I receptor
- Major Histocompatibility Complex (MHC): surface receptors for antigenic determinants of foreign molecules
  1. **Type I**: found on all nucleated cells; encoded by gene complex, Human Leukocyte Antigen (HLA) A, B, and C
  2. **Type II**: found on antigen presenting cells and B-cells; encoded by gene complex HLA DR, DQ, and DP
- Common Clusters of Differentiation (CD) Markers
  1. **CD2** and **CD3**: all T-cells
  2. **CD4**: helper T-cells, associated with MHC Type II response, receptor for HIV
  3. **CD8**: killer T-cells, associated with MHC Type I response
  4. **CD56** and **CD16**: natural killer cells

Humoral Immunity

- B-cells are produced in bone marrow, migrate to lymph nodes and spleen, bare multiple receptors similar to the immunoglobulins they secrete
- B-cells are positive for CD19, 20, and 22, and carry MHC class I on their surface
- B-Cell Activation Types
  1. **T-cell Dependent Activation**: B-cell receptors internalize antigen, fraction of antigen presents on surface via MHC Type II receptor which recognizes helper T-cells, T-cell then stimulates B-cell (via IL-2 and IL-4) to mature to plasma cells which secrete immunoglobulins
  2. **T-cell Independent Activation**: large antigens (eg, carbohydrates on bacterial cell walls) bridge immunoglobulins on B-cell surfaces that activates the B-cell

Immunoglobulins

- glycoproteins produced by plasma cells that participate in antigen recognition, complement fixation, opsonization, and promotion of phagocytosis
- composed of two heavy chains (determines class: μ, γ, α, ε, δ) and two light chains (κ and λ), both heavy and light chains have a variable and a constant region
- antigen binds to the variable portion of heavy and light chains
- **Fab Fragment**: antigen binding portion of the immunoglobulin
- **Fc Fragment**: crystalizable fragment portion of the immunoglobulin that initiates other functions such as complement fixation
- kills bacterium by complement fixation via C1q (IgG and IgM) or antibody-dependent cellular cytotoxicity (**ADCC**) which attaches Fc to a cytotoxic cell
- Types
  1. **IgG** (γ): most abundant, involved in complement fixation and ADCC, may cross the placenta (provides protection in the newborn), divided into four subclasses (G1–4)
  2. **IgA** (α): predominantly found in external secretions, associated with dimeric "secretory piece" and a "J" chain
  3. **IgD** (δ): initial type of immunoglobulin secreted, trace amounts in serum
  4. **IgM** (μ): predominant antibody in early response phase (declines rapidly and replaced with IgG of same specificity), binds complement, pentamer arrangement
  5. **IgE** (ε): major contributor in allergy (Type I hypersensitivity), Fc fragment binds to mast cells and basophils

Nonspecific Immunity

- **Natural Killer Cells**: granular lymphocytes that participate in killing tumor cells and viral infected cells, do not depend on prior immunization, activated by interferon
- **Complement System**: system of plasma proteins that act with each other that causes lysis of cells and bacteria, stimulation of chemotaxis and cell activation, and opsonization
- **Monocytes and Macrophages**: produced in bone marrow, recognize and ingest foreign and damaged material
- **Polymorphonuclear Cells** (**PMNs**): granulocyte that accumulates in acute infections and participates in phagocytosis
- **Eosinophils**: granulocyte, active in allergic response and parasitic infections
- **Basophils and Mast Cells**: granulocytes that release histamine and other substances released with exposure to an allergen, IgE presents on cell surface

- Others: skin and mucosal lining, lysozymes, saliva, gastric acid, etc

Cytokines

- immunomodulatory peptides produced by mononuclear inflammatory cells that participate in paracrine cellular modulation
- see Table 1–5 for cytokines and their actions

Immunodeficiency

B-cell Disorders

- SSx: recurrent sinonasal and pulmonary infections, conjunctivitis, dermatitis, malabsorption, pyogenic bacterial infections
- Dx: quantitative immunoglobulins and subclasses, Schick test, serum protein, immunoelectrophoresis, in vitro specific antibody responses

TABLE 1–5. Cytokines and Their Actions

Cytokines	Source	Primary Action
IL-1	Mϕ and any nucleated cells, usually from stimulation by antigen MHC class II	activates other cells and stimulates IL-2 secretion, pyrogen
IL-2	activated T-cells	essential to stimulate T-cells, B-cells, and NK cells
IL-3	T-cells	proliferation of early hematopoietic cells
IL-4	T-cells	stimulates B-cells
IL-5	T-cells, mast cells	eosinophil proliferation, IgA production
TNF-α & β	lymphocytes, Mϕ, endothelium, keratinocytes	same as IL-1 but may be more cytotoxic to tumors
TGF-β	lymphocytes, Mϕ, platelets	inhibits cells (immunosuppressive)
IFN-α	leukocytes	anti-viral and anti-tumor effects, increases MHC cell surface proteins
IFN-β	fibroblasts, epithelial cells	similar to IFN-α
IFN-γ	T-cells, NK cells	direct cytotoxic effects

- **Hypogammaglobulinemia of Burton**: X-linked disorder causing a defect in tyrosine kinase, prevents pre-B cells maturation to B-cells (does not affect T-cells), does not manifest until 6 months of age (after maternal antibodies are gone)
- **Common Variable Immunodeficiency**: failure of B-cell maturation, associated with T-cell deficiencies and other autoimmune disorders, manifests in early adulthood; Rx: IVIgG
- **Isolated IgA Deficiency**: most common inherited B-cell defect, selective IgA B-cells do not mature to plasma cells, may be asymptomatic, associated with allergies, transfusion anaphylaxis, autoimmune disorders, and IgG subclass deficiency
- **Selective IgG Hypogammaglobulinemia**: may affect one or more than one subtype (G_{1-4})
  1. **IgG_3**: most common hypogammaglobulin deficiency in adults
  2. **IgG_2**: most common hypogammaglobulin deficiency in pediatrics

T-cell Disorders

- SSx: increased viral, fungal, protozoal, and bacterial infections, atrophic lymphoid tissue
- Dx: total lymphocyte count, T-cell count, skin tests (candidal, mumps controls, PPD), functional tests (proliferation to mitogens, alloantigens helper/suppresser function)
- **DiGeorge Syndrome**: disorder of third and fourth branchial arch development, thymic hypoplasia, also associated with hypoplastic parathyroids (hypocalcemia, tetany), aortic arch and facial abnormalities
- **Severe Combined Immunodeficiency Disease**: multiple genetic forms resulting in lack of T- and B-cell immunity, severe infections (pneumonia, diarrhea, thrush), higher risk of malignancy, variant associated with **adenosine deaminase** (**ADA**) **deficiency** (accumulation of deoxyadenosine, toxic to lymphocytes); Rx: bone marrow transplant
- **Wiskott-Aldrich Syndrome**: associated with thrombocytopenia (bleeding), eczema, and recurrent infections secondary to poor functional antibody response to polysaccharides (otitis media, pneumonia, and pyogenic organisms), increased risk of malignancy

Atopic Disease

- Pathophysiology: exaggerated immediate hypersensitivity response
- clinical features of asthma, urticaria, hay fever, eczema
- strong genetic disposition
- SSx: immediate wheal and flare skin reaction to common allergens

Human Immunodeficiency Virus (HIV) and Acquired Immune Deficiency Syndrome (AIDS)

- *see* Table 1–6: Head and Neck Manifestations of HIV by Anatomical Location
- Human Immunodeficiency Virus (HIV) infection results from inoculation of infected body fluid (blood, semen, saliva, etc)
- HIV is a retrovirus that attaches to the $CD4^+$ cell marker of T-helper cells, macrophages, and other immunological cells; proviral DNA is synthesized from reverse transcriptase which integrates into host DNA
- HIV results in a decrease in T-helper lymphocytes and impaired function of macrophages, neutrophils, B-lymphocytes, and complement activation
- HIV is also associated with abnormal immune regulation, atopy, and increased autoimmune disease
- Acquired Immune Deficiency Syndrome (AIDS) is defined when an HIV patient develops an AIDS-defining illness (candidiasis, cytomegalovirus disease, Kaposi's sarcoma, *Pneumocystis carinii*, others) or $CD4^+$ count <200 cells/μl
- the cause of death for most AIDS patients is from sepsis or disseminated neoplasms
- Risks: homosexuals, multiple sex partners, IV drug abuse, previous blood transfusions, health care workers (rare)
- Dx: anti-HIV antibodies detected by enzyme-linked immunosorbent assay (ELISA) and Western Block, polymerase chain reaction of viral genes, $CD4^+$ Count, $CD4^+/CD8^+$ Ratio

Other Immunological Disorders

- **Complement Disorders**: associated with autoimmune diseases, abnormal opsonization, and capsular organism infections
- **Chronic Granulomatous Disease**: multiple genetic forms results in dysfunction of intracellular hydrogen peroxide production (does not allow intracellular killing of organisms), susceptible to infection of catalase-positive organisms (*S. aureus*, *Aspergillus, Candida, Serratia*)

TABLE 1-6. Head and Neck Manifestations of HIV by Anatomical Location

Oral and Pharynx
- **Oral Candidiasis**
- **Oral Hairy Leukoplakia**
- **Herpes Stomatitis**
- **Thrombocytopenic Purpura**
- **Recurrent Aphthous Ulcers**
- **Bone Loss** (Bacillary Angiomatosis)
- **Gingivitis** (Acute Necrotizing Ulcerative Gingivitis, Necrotizing Stomatitis)
- **Kaposi's Sarcoma, Non-Hodgkin's Lymphoma, Squamous Cell Carcinoma**

Larynx
- **Epiglottitis**
- **Kaposi's Sarcoma, Non-Hodgkin's Lymphoma**
- **Laryngitis** (*Mycobacterium*, fungal, cytomegalovirus, Epstein-Barr virus, bacterial)

Neck
- **Deep-space Neck Abscess**
- **Infectious Lymphadenopathy** (*Mycobacterium, Pneumocystis*, cytomegalovirus, Epstein-Barr virus, *Toxoplasmosis*, Cat-scratch disease, bacterial)
- **Neoplastic Lymphadenopathy** (Hodgkin's and Non-Hodgkin's Lymphoma, Metastatic disease, Thyroid tumors)
- **Persistent Generalized Adenopathy**

Salivary Glands
- **Lymphoepithelial Cysts of the Parotid Gland**
- **Parotitis**
- **Salivary Gland Neoplasms**

Otologic
- **Acute and Chronic Otitis Media, Otitis Externa, Mastoiditis** (invasive *Aspergillosis*, *Pneumocystis*, *Mycobacterium*)
- **Malignant Otitis**
- **Sensorineural Hearing Loss** (Cryptococcal or Mycobacterial meningitis, Otosyphilis, Toxoplasmosis, autoimmune demyelination of the cochlear nerve, cerebellopontine angle tumors)
- **Tympanic Membrane Perforations**
- **Aural Polyps**
- **Facial Nerve Paralysis** (Herpes Zoster, Cytomegalovirus, Epstein-Barr, HIV, autoimmune demyelination, malignant otitis externa, meningitis and encephalitis)
- **Temporal Bone Neoplasms** (Hodgkin's and Non-Hodgkin's Lymphoma, Kaposi's sarcoma)

Paranasal Sinus
- **Rhinosinusitis** (*Mucor, Aspergillosis, Pseudomonas*)
- **Nasal Tumors** (Kaposi's Sarcoma, Nasal Lymphomas)

CHAPTER

2

Salivary Glands

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SALIVARY GLAND ANATOMY AND PHYSIOLOGY

Anatomy

Parotid Gland Anatomy (Figs. 2–1, 2–2)

- located between the ramus of the mandible and the external auditory canal and mastoid tip, overlies the masseter muscle (anteriorly) and sternocleidomastoid muscle (posteriorly)
- facial nerve divides the parotid gland artificially into deep and superficial lobes
- the superficial layer of the deep cervical fascia forms the parotid gland fascia which incompletely surrounds the gland
- Histological Cell Type: basophilic, **serous cells**
- **Stylomandibular Ligament**: formed by the fascial envelope between the styloid process and the mandible, separates the parotid gland from the submandibular gland
- **Stenson's Duct**: passes over masseter, through buccinator muscle, and opens opposite to the second upper molar (follows along plane from external auditory canal to columella and buccal branch of CN VII)

Venous Drainage

- superficial temporal vein + maxillary vein → retromandibular vein
- retromandibular vein → passes deep to the facial nerve → anterior and posterior branches
- anterior retromandibular vein + facial vein → common facial vein → internal jugular vein

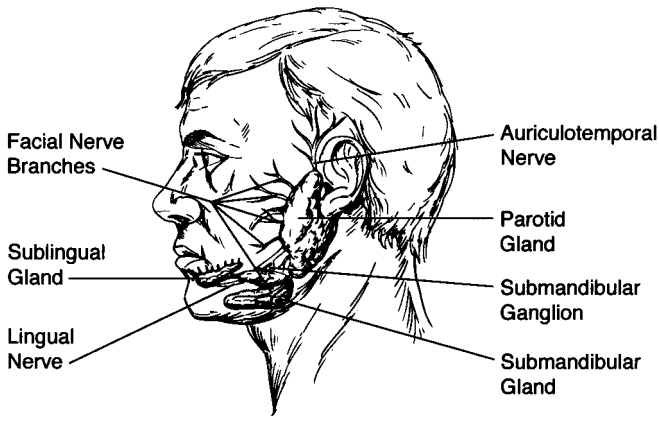

Figure 2–1. Positions of the major salivary glands and related nerves.

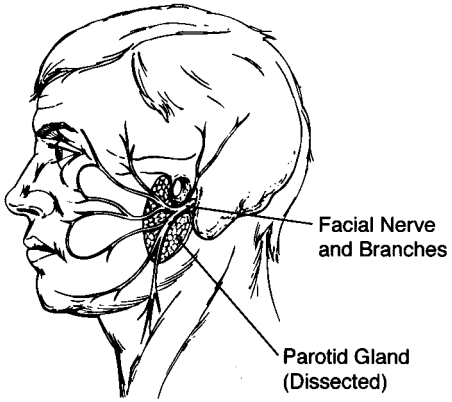

Figure 2–2. Position of the facial nerve, as illustrated following a superficial parotidectomy. The residual parotid gland illustrated above is the deep lobe.

- posterior retromandibular vein + posterior auricular vein (over SCM) → external jugular vein

Submandibular Gland Anatomy

- within the submandibular triangle (inferior to mylohyoid muscle, superior to the digastrics)
- superficial layer of the deep cervical fascia envelops the gland and contains the marginal mandibular nerve
- hypoglossal nerve runs deep to the digastric tendon and medial to the deep layer of the deep cervical fascia
- facial artery arises from the external carotid artery and courses medial to the posterior digastric muscle then hooks over the muscle to enter the gland and exits into the facial notch of the inferior mandible
- lingual artery runs along the lateral aspect of the middle constrictors, deep to the digastrics, and anteriorly and medially to the hyoglossus
- Histological Cell Type: **mixed cells** (serous and mucinous)
- **Wharton's Duct**: opens lateral to frenulum in the anterior portion of the floor of mouth, behind the incisors

Minor Glands and Sublingual Gland Anatomy

- **Sublingual Gland**: located within the submucosal layer of the floor of mouth
- **Minor Salivary Glands**: several hundred glands within the submucosal layer of the oral cavity, oropharynx, nasopharynx, and hypopharynx
- Histological Cell Type: **mucinous**
- **Ducts of Rivinus**: drain at the sublingual fold or plica of the floor of mouth

Embryology

- derived from the first pharyngeal pouch
- **4th** week: parotids formed from the posterior stomodeum (ectodermal) forming cords through the mesenchyme which later forms the capsule; parotid encapsulates late allowing entrapment of lymphoid tissue within the parotid fascia
- **6th** week: submandibular glands form as buds in the floor of mouth then grow into the submandibular triangle (endodermal)
- **9th** week: sublingual glands form as multiple buds in the floor of mouth (endodermal)
- Pathology: aberrant salivary gland tissue, accessory glands (most common in the parotid), diverticuli

Histology (Fig. 2–3)

- Secretory Unit: **acini cells** (contain abundant endoplasmic reticulum, golgi apparatus, and secretory granules; produces saliva) → **intercalated duct** → **striated duct** (contain abundant mitochondria for energy for water and electrolyte transport) → **excretory duct**
- **myoepithelial cells** surround acini and intercalated ducts

Physiology

Efferent Innervation of the Salivary Glands

Parasympathetic Innervation

- **inferior salivatory nucleus** (medulla)—**glossopharyngeal nerve** (**Jacobson's nerve**) — lesser (superficial) petrosal nerve → **otic ganglion**—*postganglionic parasympathetic fibers*—carried by auriculotemporal branch of CNV_3 → **parotid gland**

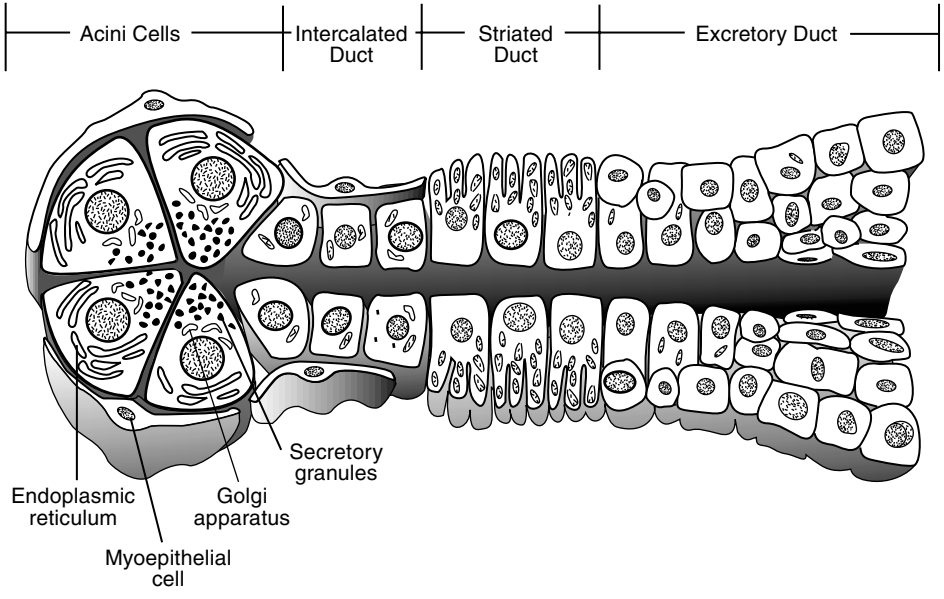

Figure 2-3. Schematic drawing of the salivary gland duct system.

- **superior salivatory nucleus** (pons) — nervus intermedius — **chorda tympani**—carried on lingual nerve → **submandibular ganglion**— *postganglionic parasympathetic fibers* → **submandibular** and **sublingual glands**

Sympathetic Innervation

- **superior thoracic nerves** → **superior cervical ganglion**— *postganglionic fibers via arterial plexus* → **submandibular** and **cutaneous vessels**

Salivation

- 1–1.5 pints of saliva/day
- Composition: >99% water, salts (calcium phosphate, calcium carbonate), organic compounds and enzymes (amylase, albumin, lysozyme, immunoglobulin A, ptyalin initiates the first phase of starch digestion, others)
- Function of Saliva: antibacterial (contains "secretory piece" needed for IgA, ABO isohemagglutinogens, perioxidases, and other immunological proteins), digestion (provides digestive enzymes and buffers), lubrication and moisturization, dental protection (prevents caries and promotes dental calcification), modulation of taste

PAROTID GLAND MASSES AND SALIVARY GLAND DYSFUNCTION

Evaluation of the Parotid Gland Mass

History and Physical Exam

- Character of Parotid Mass: onset and duration, rapid (inflammatory) versus slow growing (neoplastic), diffuse versus discrete mass (tumor), unilateral versus bilateral involvement (sialadenosis, mumps), associated pain, association with food ingestion (sialadenitis)
- Contributing Factors: history of smoking or alcohol abuse; exposure to radiation or toxins (lead or mercury); history of sarcoidosis, Sjögren's disease, tuberculosis, gout, amyloidosis; recent facial trauma or surgery
- Associated Symptoms: xerostomia, sialorrhea, weight loss, fever, trismus

- Physical Exam: palpation (mobility, size, consistency), bimanual palpation with duct inspection and saliva expression (or purulence), tenderness (inflammatory process), facial nerve function (malignancy), parapharyngeal space involvement (examine intraorally), cervical lymphadenopathy, complete head and neck history and physical exam

Imaging and Ancillary Tests

- **Fine Needle Aspirate (FNA)**: indicated for discrete nodules of the parotid gland, widely practiced although controversial (may not change management), differentiates cysts, inflammatory processes, lymphoma, and other neoplasms
- **CT/MRI**: indicated if suspect a tumor or for preoperative evaluation (*see* Table 2–1), ultrasound (U/S) differentiates cystic lesions
- **Superficial Parotidectomy**: diagnostically indicated for discrete nodules which require biopsy; incisional biopsy (enucleation) risks tumor seeding, recurrence, facial nerve injury, and violation of tumor margins
- **Technetium-99m Isotope Scan**: rarely utilized, may differentiate a Warthin's tumor or oncocytoma from other salivary gland neoplasms
- **Sialography**: visualizes ductal anatomy, indicated for ductal calculi, trauma, fistulas, Sjögren's disease, contraindicated in acute infections
- Lab Work: may consider mumps titers, complete blood count, autoimmune and Sjögren's Profile (SS-A, SS-B, ANA, ESR)

Salivary Gland Dysfunction

Types and Common Causes

- **Ptyalism** (drooling): neurological (Parkinson's disease, epilepsy), sialorrhea, swallowing disorders (relative ptyalism)
- **Xerostomia**: central (rare), primary salivary disorders (Sjögren's disease, radiation sialadenosis), dehydration, medications (psychotropics, general anesthesia, β-blockers), mouth breathing from nasal obstruction
- **Sialorrhea** (excessive saliva production): central, psychogenic, parasympathicomimetic medications (pilocarpine), diseased gland (tumor, inflammation)

TABLE 2–1. Computed Tomography versus Magnetic Resonance Imaging of the Parotid Gland

Computed Tomography	Magnetic Resonance Imaging
• better for bone imaging • less expensive • quicker image • less sensitive to patient motion • may differentiate deep tumors by identifying a fat strip • identifies calcific stones • distinguishes cystic nature of Warthin's tumors • contrast allows differentiation of vascular channels and abnormal lymph nodes	• better for soft tissue imaging (distinguish parotid tumors from parapharyngeal lesions, identifies capsule) • multiplanar views • no radiation required (less invasive) • facial nerve or retromandibular vessels may be used to distinguish deep and superficial lobes • cannot be used with pacemakers and metallic implants (aneurysm clips, cochlear implants) • better determines involvement of the facial nerve and parapharyngeal masses • Recall **T_1 weighted:** enhances fat, water appears dark, TR <1000, TE <25 ms **T_2 weighted:** enhances water, fat appears dark, TR >1000, TE >40 ms **Spin Density:** T_R >1000, T_E <25 ms

Management

- **Ptyalism/Sialorrhea**: chorda tympani transection (Jacobson's nerve neurectomy), ductal rerouting procedure to the posterior cavity, ligation of Stenson's duct, submandibular gland excision
- **Xerostomia**: artificial saliva, frequent small drinks, pilocarpine hydrochlorate drops, aggressive dental care

SALIVARY GLAND NONMALIGNANT PATHOLOGY

Salivary Gland Enlargement

Acute Suppurative Sialadenitis

- Pathogen: *S. aureus* (most common)
- Pathophysiology: salivary stasis or obstruction, retrograde migration of bacteria, postoperative parotiditis
- Risks: dehydration, postsurgical (GI procedures), radiation and chemotherapy, Sjögren's syndrome
- SSx: sudden onset of erythema, tenderness, warmth, and purulence at ductal orifice, auricle may protrude, trismus
- Dx: clinical history and exam, cultures
- Rx: rehydration, warm compresses, antimicrobial therapy (may require parenteral antibiotics for severe cases), sialogogues, parotid massage, oral irrigations, if no resolution after 2–3 days then consider CT or U/S to evaluate for abscess (may require I&D)

Mumps and Viral Infections

- presents in pediatric population (4-6 years old)
- Pathogen: paramyxoviruses
- SSx: 75% bilateral, painful parotid swelling (may involve submandibular and sublingual glands), malaise, fever, trismus
- Dx: mumps titers; hemoagglutination antigens; virus may be cultured from saliva, urine, or CSF
- Complications: sudden sensorineural hearing loss (SNHL) (CN VIII involvement), infertility (with orchitis), encephalitis, pancreatitis, and nephritis
- Rx: self limiting: requires only supportive care (hydration, analgesics), audiological evaluation, vaccine is available
- Other Viral Infections: CMV (higher mortality in neonates), Coxsackievirus, influenza A

Chronic Inflammation

- Types
  1. **Granulomatous:** tuberculosis, atypical mycobacterium, actinomycosis (poor oral hygiene, dental caries), cat scratch fever, sarcoidosis (*see* pp. 205–212.)
  2. **Secondary**: progression from acute sialadenitis
- SSx: xerostomia, slow painless enlargement, may be recurrent
- Dx: sialography ("tree-leaf" appearance), CT, PPD, serum markers, VDRL, FTA-ABS, cat-scratch antigen test, etc
- Complications: abscess, fistula, ductal destruction
- Rx
  1. **Medical**: sialogogues, antibiotics, warm compress, massage
  2. **Conservative Surgical Management**: ductal dilatation
  3. **Destructive Surgical Management**: gland excision, ductal ligation, gland irradiation, or neuronectomy

NOTE: incisional biopsy should be avoided because of high risk of fistula formation (especially with mycobacterial infections)

Uveoparotid Fever (Heerfordt's Syndrome)

- more common in women
- Pathophysiology: extrapulmonary form of sarcoidosis
- SSx: self-limited uveitis, parotid enlargement, SNHL, facial palsy, malaise, fever
- Dx: based on clinical history and exam and evidence of sarcoidosis in salivary gland tissue
- Rx: corticosteroids

Kuettner's Tumor (Chronic Sclerosing Sialadenitis of the Submandibular Gland)

- Pathophysiology: may be autoimmune mediated
- SSx: firm, enlargement of submandibular gland (similar to tumor)
- Dx: biopsy
- Histopathology: chronic inflammation with destruction of acinar cells, sclerosis, "cirrhotic" changes
- Rx: submandibular excision for diagnosis and treatment

Radiation Sialadenitis

- often permanent if exposed to >40–50 Gy
- SSx: xerostomia, hypogeusia, ageusia
- Dx: clinical exam and history of radiation exposure
- Histopathology: interstitial fibrosis
- Rx: symptomatic (pilocarpine drops, artificial saliva, frequent drinks), dental care (fluoride rinses)

Salivary Calculi (Sialolithiasis)

- most common in submandibular gland
- associated with gout (uric acid calculi)
- Pathophysiology: change in the viscosity of the saliva may cause mucous obstruction, calcium phosphate and calcium carbonate form around this center causing obstruction
- SSx: recurrent pain, swelling, worse with meals (salivary colic)
- Dx: stone may be palpable, sialography (**90% of submandibular calculi are radiopaque, 90% of parotid calculi are radiolucent**, may be multiple), CT, U/S
- Complications: fistulas, acute suppurative sialioadenitis, ductal strictures
- Rx: gland massage, bimanual expression, transoral incision, sialodochoplasty (reconstruct duct), gland excision if recurrent or if stone is lodged within substance of the gland, extracorporeal lithotripsy

Sjögren's Syndrome (Myoepithelial Sialadenitis, Benign Lymphepithelial Lesion)

- most common in middle-aged women
- Pathophysiology: systemic autoimmune destruction of exocrine glands, B-cell hyperactivity
- associated with **Non-Hodgkin's Lymphoma**
- Types
  1. **Primary**: exocrine gland involvement only
  2. **Secondary**: associated with other connective tissue disorders (most commonly rheumatoid arthritis)
- Other Sicca-like Causes: aging, medications (diuretics, anticholenergics, antihistamines, antidepressants), dehydration, hepatitis, other autoimmune disease
- SSx: **keratoconjunctiva sicca** (filamentary keratitis, sandy sensations in eyes), **xerostomia** (dental caries, dry mucosa), **intermittent bilateral parotid swelling** (atrophy at end-stage of disease), achlorhydria, Raynaud's phenomenon, pancreatitis, myositis, anemia, glomerulonephritis, hepatosplenomegaly
- Rx: artificial saliva, frequent small drinks, artificial tears, consider pilocarpine hydrochlorate drops and oral corticosteroids for severe acute exacerbation

Diagnosis

- Clinical History (must have 2 of 3): keratoconjunctiva sicca, xerostomia, or other connective tissue disease

- Biopsy: lip biopsy or minor salivary glands reveals lymphocytic infiltration with glandular atrophy
- Serology: ANA, RF, ESR, SS-A and SS-B (antibodies specific for primary Sjögren's syndrome), decreased IgM (suggest higher risk of progression to malignancy)
- Sialography: globular, multiple contrast collections throughout gland ("pine tree" appearance)
- Schirmer test: evaluates tear production

Mikulicz's Syndrome

- **Mikulicz's Disease**: all cases of recurrent parotid gland swelling that are nonautoimmune
- Common Causes: amyloidosis, duct stricture, bulimia, lymphadenitis, lead and mercury toxicity, chronic fatty infiltration from alcohol, and hypovitaminosis

Sialadenosis

- Pathophysiology: secondary to underlying endocrine or metabolic pathology (cirrhosis, diabetes, malnutrition, ovarian, thyroid, or pancreatic insufficiency) or medications (hypertensive medications, catecholamines, iodine-containing compounds)
- SSx: recurrent bilateral nontender parotid swelling (painful sialadenosis is associated with antihypertensive medications)
- Dx: clinical history and exam
- Rx: treat underlying disease or change medications

Salivary Gland Cysts

Introduction

- **most common mass in parotid**
- Risks: sialoadenitis, sialolithiasis, trauma
- W/U and Management
  1. CT scan or U/S to confirm cystic nature
  2. FNA to evaluate for malignant cells
  3. may observe or inject with sclerosing agents (tetracycline)
  4. surgical excision for cosmesis, recurrent lesions, solid lesions, abnormal cytology

Benign Lymphoepithelial Cysts

- increased incidence since **HIV** epidemic
- may progress to pseudolymphoma
- SSx: multiple, bilateral parotid cysts

- Dx: clinical history and exam, biopsy
- Histopathology: lymphoreticular infiltrate, clusters of lymphoid tissue (germinal centers), acinar atrophy, ductal metaplasia
- Rx: aspiration or excision (superficial parotidectomy)

Mucous Retention Cysts and Ranulas

- Pathophysiology: obstruction of minor salivatory glands
- **Mucous Retention Cysts**: true cysts of the minor salivary glands (lined with epithelial layer)
- **Ranulas**: mucous retention cyst of the floor of mouth
- **Plunging Ranula**: extension into hyoid muscles, may present as a neck mass
- SSx: cystic mass on floor of mouth
- Dx: clinical history and exam, biopsy
- Rx: excision or marsupialization

Other Cysts

- **Mucoceles**: not a true cyst, extravasation of mucus into soft tissue, usually from trauma to duct (**Post-traumatic Sialocele**)
- **Dermoid and Epidermoidal Cysts**
- **Branchial Apparatus Anomalies** (**First Branchial Cleft**): (*see* pp. 191–195.)

BENIGN SALIVARY GLAND TUMORS

Introduction

- Risk: radiation (latency of 7–30 yrs); alcohol and smoking not associated with most salivary neoplasms (except Warthin's tumors)
- Multicellular Theory: neoplastic cells originate from their counterparts (eg, oncocytic tumors from striated duct; acinar tumors from acinar cells)
- Biocellular Theory: all neoplastic cells differentiate from basal cells found in excretory and intercalated ducts
- **80%** of parotid tumors are benign; **80%** of salivary neoplasms are located in the parotid
- in general, the smaller the gland the more likely malignant

Pleomorphic Adenoma (Benign Mixed Tumor)

Introduction

- benign heterogeneous tumor composed of variable epithelial and myoepithelial components
- **most common tumor of each gland** (may also be found in the respiratory tract and nasal cavity)
- slightly more common in women
- SSx: slow growing (over years), unilateral, painless, firm mass (usually toward the tail of the parotid); rarely progresses to dysphagia (pharyngeal extension), dyspnea, or hoarseness (laryngeal involvement), or facial nerve palsies; deep lobe (10%) involvement may present with intra-oral swelling
- Recurrent SSx: multilobular nodules, not discrete; may arise in scar, subcutaneous tissue, deep lobe, or facial nerve sheath
- Complications: rare malignant transformation (Carcinoma Ex-Pleomorphic Adenoma and Sarcoma) or "benign" metastasizing (*see* pp. 78–79)
- Dx: biopsy specimen (parotidectomy), FNA
- Rx: surgical resection (superficial or total parotidectomy, *see below*) with wide margin for pseudopod extensions to prevent recurrence (>90% 10-year cure, approximately 30% recurrence rate for enucleation alone), radioresistant

Histopathology

- Cellular Components
  1. **myoepithelial component**: spindle shaped with hyperchromatic nuclei, may be more than one cell layer thick
  2. **epithelial components**: varied growth patterns (trabecular, solid, cystic, papillary)
  3. **stromal components**: product of myoepithelial cells: myxoid, chondroid, fibroid, or osteoid components
- fibrous pseudocapsule (except minor glands)
- micro-pseudopod extensions

Warthin's Tumor (Papillary Cystadenoma Lymphomatosum)

Introduction

- Pathophysiology: entrapped lymphoid tissue (parotid is the last gland embryologically to be encapsulated), ectopic ductal epithelium that develops within intraparotid lymph nodes, or hypersensitivity disease resulting in metaplasia of the duct
- second most common salivary gland tumor

- almost exclusively found in middle-aged to elderly men
- almost always involves the parotid
- **10% bilateral** (synchronous or metachronous); **10% multicentric**
- rare malignant transformation
- SSx: slow growing, painless, cystic, compressible mass
- Dx: surgical biopsy (parotidectomy), FNA (thick, turbid fluid), radiosialography (concentrates technetium-99m due to the presence of high mitochondrial content of oncocytes)
- Rx: superficial or deep parotidectomy (*see below*)

Histopathology

- Biphasic Layers
  1. **epithelial component**: lines papillary projections; double lining of oncocytes; inner or luminal cells, nonciliated, tall columnar nuclei at luminal aspect; outer or basal cells are round, cuboidal with vesicular nuclei
  2. **lymphoid component**: mature lymphocytes with germinal centers
  3. **mucous secreting cells**
- **Oncocytic Cell**: metaplasia (cytoplasmic alteration) of myo- or epithelial cells

Oncocytoma (Oxyphilic Adenoma)

Introduction

- rare, benign tumor exclusively of oncocytic cells (1% of salivary gland tumors)
- rare "malignant" low-grade tumor transformation
- SSx: slow growing, painless mass
- Dx: surgical biopsy (parotidectomy), radiosialography (concentrates technetium-99m due to the presence of high mitochondrial content of oncocytes), FNA
- Rx: superficial or deep parotidectomy (*see below*)

Histopathology

- encapsulated
- **oncocytic cells** (*see* Warthin's tumor)
- granularity from mitochondria

Monomorphic Adenoma

Introduction

- similar to pleomorphic except no mesenchymal stromal component: **predominantly an epithelial component** or (rarely) the myoepithelial component
- more common in the minor salivary glands (upper lip)
- 12% bilateral
- Dx: surgical biopsy (parotidectomy)
- Rx: superficial or deep parotidectomy (*see below*)

Types

Basal Cell Adenoma

- monomorphic adenoma that has predominately basaloid cells
- Subtypes: solid (commonest, solid nests of basal cells), trabecular (ribbon-like pattern), tubular, membranous
- Prognosis: rare malignant potential

Others

- **Myoepithelioma Adenoma**: monomorphic adenoma that has predominately myoepithelial cells
- **Clear Cell Adenoma**: must evaluate for metastatic renal primary
- **Membranous Adenoma**
- **Glycogen Rich Adenoma**

Pediatric Salivary Gland Neoplasms

Introduction

- hemangiomas and pleormorphic adenomas are the most common benign parotid tumors of childhood
- excluding hemangiomas, salivary gland infections, and lymphangiomas, >50% of parotid solid masses are malignant
- well-differentiated mucoepidermoid tumors are the most common salivary gland malignancy in children

Hemangiomas

- benign tumor of endothelial origin
- usually discovered at birth
- more common in Caucasian females

- 50% association with cutaneous hemangiomas

Capillary Hemangiomas

- **constant** shape, enlarges in proportion to growth of child
- may involve facial nerve
- SSx: lobulated, dark red and bluish mass overlying skin
- Histology: unencapsulated, capillary sized vessels, may invade facial nerve
- Rx: avoid surgery in childhood; tattooing, cryotherapy, laser obliteration

Cavernous Hemangiomas

- may enlarge rapidly
- **60%** spontaneously resolve by 4–6 years old
- less chance of regression than capillary hemangiomas
- Rx: corticosteroid therapy, may consider surgery if lesion can be completely removed, failure to involute, rapid growth threatens perioral, periorbital, or nasal tip regions

SALIVARY GLAND MALIGNANCY

Introduction

- SSx: malignancy suggested by the presence of a slow growing discrete mass (although may present as rapidly growing mass), facial nerve involvement, constant pain, cervical lymphadenopathy
- Evaluation: *see* Work-up of the Parotid Mass, pp. 66–68
- Dx: preoperative FNA is often utilized to counsel patient of risk of facial nerve involvement and malignancy, confirmation must be determined by a superficial parotidectomy (incisional biopsies are contraindicated due to the possibility of tumor seeding and violation of tumor margins)
- Poor Prognostic Indicators: submandibular gland involvement (parotid gland more favorable), parapharyngeal space involvement, high-grade tumors, larger size, facial nerve or skin involvement, painful tumors, recurrence, regional lymph nodes, distant metastasis (more common in adenoid cystic and undifferentiated tumors)

Malignant Salivary Gland Tumors and Their Management

Mucoepidermoid Carcinoma

- Features: components of epidermoid, mucinous, and intermediate cells, high- and low-grade tumors
- **most common salivary gland malignancy in children and adults** (adenoid cystic carcinoma is the most common in the submandibular gland)
- most commonly found in the parotid
- commonly induced by radiation
- 30–70% overall regional metastatic potential

Types

Low-Grade (Well-Differentiated)

- Histopathology: **more mucinous cystic elements**, aggregates of mucoid cells with strands of epithelial cells, positive keratin staining
- approximately 70% 5-year survival
- Rx: superficial or total parotidectomy (for deep lobe involvement), radical neck dissection for clinically positive nodes

High-Grade (Poorly-Differentiated)

- aggressive (<50% 5-year survival)
- Histopathology: **less mucinous elements, more solid nests of cells**, requires mucin staining to differentiate from squamous cell carcinoma, positive keratin staining
- Rx: superficial or total parotidectomy (for deep lobe involvement) with **elective neck dissection** (selective supraomohyoid neck dissection); radical neck dissection for clinically positive nodes; consider adjuvant radiation therapy for advanced tumors, regional disease, close or near surgical margins, or bone or neural involvement

Adenoid Cystic Carcinoma (Cylindroma)

- Features: high-grade tumor, aggressive, insidious growth (over several years), perineural spread (facial paralysis), local recurrence and distant metastasis (may present >5 years later)
- **most common submandibular and minor gland malignancy**
- Histopathology: low-grade have cribiform (nests of cells with round spaces, "Swiss cheese" appearance) or cylindromatous (tubular pattern) pattern, high-grade has more solid pattern (dense cellular pattern with few spaces)

- Prognosis: high-grade associated with poor prognosis (<20% 5-year survival), low-grade up to a 100% 5-year survival
- Rx: radical surgical resection (facial nerve resection if involved), consider adjunctive radiation therapy (or neutron beam); long-term follow-up required because of indolent course and possible distant metastasis, consider elective neck dissection versus postoperative radiation to the neck

Management

- Single-Modality Therapy: surgical excision of primary tumor for smaller tumors versus primary radiation for nonoperable candidates
- Multi-Modality Therapy: surgical excision of primary tumor with adjuvant radiation for advanced tumors, regional metastasis, close or near surgical margins, or bone or neural involvement
- Neck: radical neck dissection for clinical nodes only

Malignant Mixed

- Features: **high-grade tumor**, aggressive, explosive growth rate, poor prognosis (<50% 5-year survival)
- Rx: surgical excision with postoperative radiation therapy, may consider elective neck dissection (supraomohyoid)

Types

- **Carcinoma Ex-Pleomorphic Adenoma**: 2–3% malignant transformation from pleomorphic adenomas, carcinoma components only (arises from epithelial component)
- **Metastasizing Mixed Tumor**: distinct from carcinoma ex-pleomorphic, remains histologically benign
- **Carcinosarcoma**: contains components of both carcinomas and sarcomas
- **Noninvasive Carcinoma**: carcinoma in situ within a pleomorphic adenoma

Acinic Cell Carcinoma

- Features: **low-grade**, better prognosis (63–87% 10-year survival)
- **second most common salivary gland cancer in pediatrics**
- **3% bilateral**
- most commonly found in the parotid (serous acinar cells)
- Histopathology: **serous acinar cells** or clear cytoplasm cells, several configurations (microcystic, papillary, solid, follicular), lymphoid infiltrate

- Rx: surgical excision with wide margins, neck dissection for positive nodes only, adjuvant radiation therapy may be considered for advanced disease

Other Salivary Gland Malignancy Types

- **Squamous Cell Carcinoma**: high-grade, aggressive, often not the primary (must evaluate for primary)
- **Lymphomas**: rare as a primary site although may arise from intraglandular lymphoid tissue (from embryonic development), associated with Sjögren's Syndrome (see Head and Neck Cancer: Other Head and Neck Malignancy)
- **Adenocarcinoma**: high-grade, aggressive, originates from terminal tubules or intercalated ducts
- **Malignant Oncocytoma**: similar to the benign form with distant metastasis and local invasion
- **Epithelial-Myoepithelial Carcinoma** (**Clear Cell**): low-grade
- **Salivary Duct Carcinoma**: high-grade, similar to ductal carcinoma of the breast
- **Undifferentiated Carcinoma**: highly aggressive, worst prognosis, predominantly "small cell"

Parotidectomy

Superficial Parotidectomy

- Indication: diagnostic and therapeutic excision of benign or malignant tumors that involve the superficial lobe of the parotid only
- typically preserves facial nerve
- resects majority of parotid gland lateral to facial nerve (controversy on amount of parotid required for removal)

Facial Nerve Markers

1. **Tragal pointer**: the facial nerve may be located 1 cm medial, inferior, and deep from tragal cartilage
2. **Tympanomastoid Suture Line**: the facial nerve is 6–8 mm deep to the inferior end of the tympanomastoid suture line
3. **Digastric Attachment to Digastric Ridge**: identifies the plane of the facial nerve
4. **Retrograde Dissection from Distal Branches**: may be required in select cases
5. **Styloid Foramen**: may identify the main trunk

Total Parotidectomy

- Indications: high-grade malignancy or deep lobe or facial nerve involvement
- excision of facial nerve may be indicated for malignant tumors (encasement or invasion of facial nerve) and select cases of pleomorphic adenomas (recurrent)
- **Radical Parotidectomy**: includes possible mandibulectomy, petrosectomy, periglandular skin, or facial nerve, indicated for aggressive malignant disease

Parotidectomy Complications

- **Facial Nerve Paresis/Paralysis**: iatrogenic injury should be repaired immediately (*see* p. 360)
- **Hypesthesia of Greater Auricular Nerve**: usually resolves within 9 months
- **Salivary Fistulas**: uncommon, usually spontaneously resolve in 2–3 weeks; Rx: probe wound to release fluid (aspiration), pressure dressing, surgical closure for prolonged drainage (may consider tympanic neurectomy)
- also hematomas, infection, flap necrosis, trismus, seromas, and recurrence

Frey's Syndrome (Gustatory Sweating)

- Pathophysiology: injury to the auriculotemporal nerve (sympathetic fibers) results in aberrant innervation of cutaneous sweat glands (which share the same neurotransmitter) by postganglionic parasympathetic fibers
- may occur up to 5 years postoperatively
- less incidence with the use of "thick" skin flaps
- SSx: sweating and reddening of skin during meals
- Rx
  1. Medical Management: antiperspirant and anticholenergic preparations (scopolamine, glycopyrrolate, diphemanil methylsulfate)
  2. Surgical Management: tympanic neuronectomy (chorda tympani nerve section via tympanotomy approach), interpose a sheet of fascia lata or dermis between skin and parotid gland
  3. Radiation Therapy: reserved for failed management with severe symptoms

CHAPTER

3

Laryngology

R. Pasha, James P. Dworkin, and Robert J. Meleca

LARYNGEAL ANATOMY AND PHYSIOLOGY

Embryology

Development of the Respiratory Primordium

- at the fourth week of development the respiratory primordium appears as an outgrowth (**laryngotracheal groove**) from the ventral wall of the foregut (primitive pharynx)
- the laryngotracheal groove evaginates to form the **laryngotracheal diverticulum** dividing the foregut into a dorsal portion (esophagus) and ventral portion (larynx, trachea, and lung) separated by the **tracheoesophageal septum**
- the respiratory primordium (ventral portion) maintains open communication with the pharynx through the laryngeal orifice
- epithelial proliferation obliterates the laryngeal lumen, **recanalization** occurs by the tenth week (no recanalization results in stenosis)
- three tissue swellings (median swelling behind the hypobranchial eminence, which forms the epiglottis, and two lateral swellings that form the arytenoid cartilages) surround the laryngeal orifice

Branchial Arch Derivatives

II: lesser horn and upper portion of the hyoid bone
III: greater horn and lower portion of the hyoid bone
IV: supraglottic structures (thyroid cartilage), superior laryngeal nerve (SLN) structures (cricothyroid muscle and pharyngeal constrictors)
V/VI: glottic/subglottic structures (cricoid, cuneiform, corniculate, and arytenoid cartilages) and recurrent laryngeal nerve (RLN) structures (all intrinsic laryngeal muscles except the cricothyroid)

Anatomy

Laryngeal Neuromuscular Anatomy (Fig. 3–1)

- Extrinsic Depressors: (C1–C3) sternohyoid, sternothyroid, thyrohyoid, and omohyoid muscles
- Extrinsic Elevators: geniohyoid (C1), digastric (CN V and CN VII), mylohyoid (CN V), stylohyoid (CN VII) muscles
- **Posterior Cricoarytenoid** (**PCA**): (RLN) **only vocal fold ABductor**
- **Lateral Cricoarytenoid** (**LCA**): (RLN) vocal fold ADduction

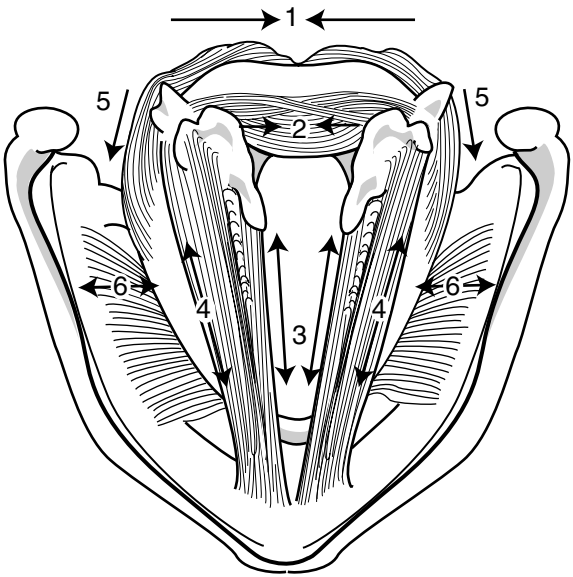

Figure 3–1. The intrinsic laryngeal muscles and their vector action: (**1**) posterior cricoarytenoid muscle; (**2**) interarytenoid muscle; (**3**) vocalis (medial aspect of thyroarytenoid); (**4**) thyroarytenoid muscle; (**5**) lateral cricoarytenoid muscle; (**6**) cricothyroid muscle. (copyright 1989 Novartis. Adapted with permission from *The Atlas of Human Anatomy*, illustrated by Frank H. Netter, MD. All rights reserved.)

- **Thyroarytenoid** (**TA**): (RLN) increases vocal fold tension, vocal fold ADduction, the medial aspect of the TA is called the **vocalis**
- **Cricothyroid**: (**external branch of SLN**) ADduction, increases vocal fold tension and length (**chief pitch-changing muscle**)
- **Interarytenoid**: (RLN) only **unpaired** muscle, ADduction

Laryngeal Cartilage

- **Thyroid, Cricoid, and Arytenoids**: **hyaline** cartilage (hyaline cartilage is the most common, found in most articular cartilage)
- **Epiglottis**: **fibroelastic** cartilage (less strength, elastin), attaches to thyroid cartilage
- **Corniculate Cartilage**: **fibroelastic** cartilage, **above** arytenoid cartilage, provides rigidity to A-E folds
- **Cuneiform Cartilage**: **fibroelastic** cartilage, **within** A-E folds, provides rigidity
- **Trieceous Cartilage**: sometimes found in **thyrohyoid ligament**, may be mistaken on x-ray as a foreign body when calcified

Laryngeal Joints

- **Cricothyroid Joints**: synovial, rocks (hinge)
- **Cricoarytenoid Joints**: synovial, rocking motion (anteromedially for vocal fold ADduction and posterolaterally for vocal fold ABduction)

Vocal Fold Layers (from superficial to deep, Fig. 3–2)

1. **Squamous Epithelium**: stratified, nonkeratinizing
2. **Superficial Lamina Propria** (**SLP**, **Reinke's Space**): loose fibrous matrix (few fibroblasts), gelatinous consistency permits fluency of vocal fold vibration (mucosal wave)
3. **Intermediate Lamina Propria**: elastin (some fibroblasts)
4. **Deep Lamina Propria**: fibroblasts and collagen (dense)
5. **Thyroarytenoid Muscle Complex**: thyromuscular bundle (thyroarytenoid muscle) and thyrovocalis bundle (vocalis muscle)

*NOTE: the squamous epithelium and superficial layer of the lamina propria form the **vocal fold cover**, the intermediate and deep lamina propria form the **vocal ligament (transition zone)**, the thyroarytenoid muscle complex forms the **vocal fold body***

*NOTE: The gelatinous consistency of the SLP allows for fluency of vibration of the cover over the body during voicing (cover-body concept of vocal fold vibration). This vibratory activity can be readily visualized using videostroboscopy and is referred to as the **mucosal wave**.*

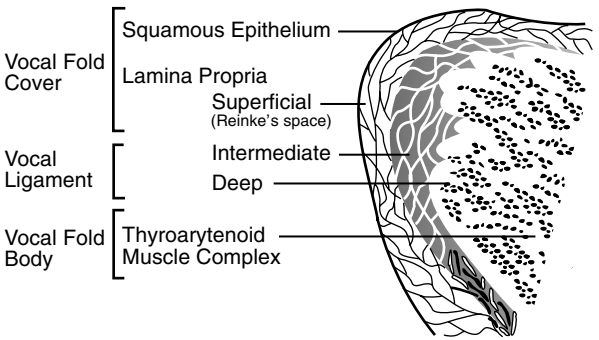

Figure 3-2. Histological layers of the true vocal fold.

Pediatric Airway Anatomy

- neonates and all non-humans have their epiglottis engage the soft palate forming a central tunnel for airway and the lateral sides for food to allow for simultaneous respiration and deglutition; in humans, the epiglottis descends during infancy
- >90% of neonates are obligate nasal breathers until 2 months because of lack of coordination of oral/respiratory functions and epiglottis location
- the epiglottis at birth is omega (Ω) shaped
- superior margin of the infant's larynx is at the level of cervical spine C1 and the cricoid cartilage at C4; as the larynx descends the cricoid cartilage aligns to C6–7 by puberty
- **1 mm** of laryngeal edema in the neonate can reduce airway by **60%**
- endotracheal tube size = (**age + 16**)**/4**; bronchoscope size = **age/4**

Physiology

Three Zones of Laryngeal Airway Protection

1. **Epiglottis and A-E Folds**
2. **False Folds**
3. **True Folds**: most significant

Laryngeal Sensory Innervation

- **Internal Branch of the SLN**: innervates the laryngeal mucosa above the glottis
- **RLN**: innervates the laryngeal mucosa inferior to the glottis
- **Negative Pressure Receptors**: SLN, maintains patency of airway during inspiration
- **Airflow Receptors**: cold receptors, stimulated from moving air
- **Drive Receptors**: proprioception
- **Laryngospasm**: exaggerated ADduction reflex caused by laryngeal irritation (reflux, foreign bodies, extubation, mucus); may be driven by the CNS, mediated by the superior laryngeal nerve
- **Cardiovascular Collapse**: bradycardia and hypotension caused by laryngeal irritation (intubation); uncertain mechanism

Voice Production

- **Requirements for Sound Production:** source of energy (eg, airflow) and source of vibration (eg, true vocal folds, neoglottis, articulator constrictions)

- **Myoelastic-Aerodynamic Theory:** true vocal folds are ADducted and tensed, subglottic pressure increases until level of pressure induces vocal fold vibration, which opens the glottis from an inferior to superior direction and closes from an inferior to superior direction
- **Bernoulli's effect:** forced air across a constricted zone produces negative pressure, allowing the true vocal folds to be "sucked" back together
- normal vocal fold vibration occurs vertically from inferior to superior, and horizontally along the superior surface of the vocal fold from medial to lateral

Components of Speech

- **Phonation:** production of voice, determined by vocal fold position, expiratory force, vibratory capacity of vocal folds, vocal fold length and tension
- **Resonation:** oral/nasal speech balance, determined by velopharyngeal musculature valving and by structure of the chest, nasopharynx, nasal cavity, and oral cavity
- **Articulation:** production of speech sounds, determined by actions of the lips, tongue, and jaw musculature activity
- **Respiration:** production of source of energy (airflow dynamics) for verbal speech, determined by inhalation/exhalation respiratory muscular activity
- **Prosody:** production of syllable stress, emphasis, and inflection patterns to provide affective speech tone, influenced by phonatory, articulatory, and respiratory forces

Voice Parameters

- **Pitch** (Hz): perceptual term, related to **frequency** of vocal fold vibration (Hz); determined by the length, tension, and speed of vibration of the vocal folds
- **Fundamental Frequency:** predominant pitch component of the speaking voice, **128 Hz** ("C" below middle "C" note) in males (long, thick vocal folds) and **256 Hz** (middle "C" note) in females (short, thin vocal folds)
- **Loudness** (decibels): the intensity of the voice; determined by subglottic pressure, glottal resistance, airflow rate, amplitude of vocal fold vibration, and force of vocal contact
- **Quality (Timbre):** determined by the synchronicity of vocal fold vibration and glottal competence

EVALUATION OF THE DYSPHONIC PATIENT

History and Physical Exam

History

- Character of Dysphonia: onset and duration, time course (acute versus chronic), periodicity (morning hoarseness associated with GERD, evening hoarseness associated with vocal abuse)
- Contributing Factors: recent upper respiratory infection, fever, sore throat, cough, congestion; history of voice, tobacco, or alcohol abuse; past medical history of peripheral nerve diseases and other neurological disorders, gastroesophageal reflux, hypothyroidism, psychological stressors; previous laryngeal trauma, surgery, or airway manipulation
- Associated Symptoms: odynophagia, dysphagia, aspiration, weight loss, hearing loss, heartburn
- Think "KITTENS" for differential diagnosis (see Table 3–1)

Physical Exam

- Assess Perceptual Quality of Voice: *see below*
- Indirect and Direct Laryngoscopy (Mirror, Flexible Nasopharyngoscopy, Videostroboscopy): assess vocal fold motion, examine laryngeal lesions and appearance of vocal folds (edematous, erythematous, atrophic, etc), assess glottic competence
- H&N Exam: neck masses, thyroid masses, complete neurological exam

Perceptual Voice Abnormalities

- **Abnormally High Fundamental Frequency:** may be due to tension producing vocal fold masses, muscle tension phenomenon, protracted pubescence
- **Abnormally Low Fundamental Frequency:** consider load producing vocal fold masses (Reinke's edema, growths) or hypothyroidism (laryngeal myxedema)
- **Abnormally Loud Voice:** may be secondary to sensorineural hearing loss, hyperfunction (muscle tension dysphonia), excessive respiratory efforts, or psychogenic
- **Abnormally Soft Voice:** may be secondary to conductive hearing loss, reduced respiratory efforts, glottic incompetence, vocal fold masses, vocal fold paralysis, vocal fold bowing defect, or psychogenic

TABLE 3–1. Differential Diagnosis of Dysphonia: KITTENS Method

(**K**) **C**ongenital	**I**nfectious & **I**diopathic	**T**oxins & **T**rauma	**T**umor (Neoplasia)	**E**ndocrine	**N**eurologic	**S**ystemic
Congenital webs	Laryngitis (viral, bacterial, and fungal)	Laryngeal cysts, nodules, polyps, and ulcers	Recurrent laryngeal papillomatosis	Hypothyroidism (laryngeal myxedema)	Cerebral palsy	GERD
Underdeveloped larynx	Vocal fold paralysis	Voice abuse	Laryngeal cancer	Adrenal, pituitary, and gonadic disorders	Multiple sclerosis	Connective tissue disorders (rheumatoid arthritis, SLE)
	Adductor spasmodic dysphonia	Reinke's edema	Benign laryngeal neoplasms (hemangiomas, cystic hygromas)	Pubescence	Extrapyramidal lesions (Parkinson's)	Psychogenic
	Muscle-tension disorders	Arytenoid dislocation			Stroke	
		Vocal fold granulomas			Guillain Barré	
		Caustic inhalation injuries			Myasthenia gravis	
					Other neurological disorders	

- **Hoarseness:** consider vocal fold mass, vocal fold paralysis, vocal fold bowing defect, muscle-tension abnormalities, vocal fold swelling, or psychogenic
- **Diplophonia:** two simultaneous pitches, may be due to recruitment of false vocal folds during phonation or the presence of an asymmetric mass (eg, unilateral vocal fold polyp or cyst)
- **Harshness:** strained or strangled; may be from upper motor neuron dysarthria (spastic), adductor spasmodic dysphonia, psychogenic, muscle-tension abnormalities, or a compensatory result from an underlying vocal fold mass
- **Tremorous:** suggests organic vocal tremor (extrapyramidal system), spasmodic dysphonia, or psychogenic
- **Breathy:** excessive airflow from a longer "open phase" or incomplete ADduction; may be from a glottal chink (mass effect), bowed deformity secondary to vocal fold paralysis, presbylaryngis, glottic lesions, muscle-tension dysfunction, abductor spasmodic dysphonia, or a psychogenic disorder
- **Arrest of Phonation:** sudden stops; consider spasmodic dysphonia, load bearing vocal fold mass, muscle-tension disorders, psychogenic
- **Aphonia:** complete absence of phonated sound; often a functional disorder (psychogenic), bilateral ADductor vocal fold paralysis, mass that results in an open glottis
- **Stridorous:** may be due to bilateral vocal fold paralysis, obstructing vocal fold mass, paroxysmal laryngospasms, or psychogenic
- **Hypernasality:** suggests velopharyngeal incompetence (flaccid or spastic dysarthria, anatomic defects)
- **Hyponasality:** may be secondary to hypertrophied tonsils or adenoids, sinonasal disease, nasal obstruction (septal deformities, nasal masses), or a nasopharyngeal mass

Ancillary Tests

- **Videostroboscopy**: examine vocal fold mucosa for general health (coloration, thickness, atrophy, etc), vocal fold anatomical defects (masses, glottal chinks, etc), vocal fold biomechanical disturbances (immobility or hypomobility, tremors, spasmodic or muscle-tension disorders), and vocal fold mucosal wave dynamics
- **Acoustic Analysis:** measures of fundamental frequency, pitch period fluctuations or **jitter** (normal = 0.40%), **amplitude** fluctuations or **shimmer** (normal = 0.50 dB), and harmonic-to-noise ratio (normal = 11 dB)

- **Speech Aerodynamic Studies:** measures of mean transglottal airflow rate (normal = 100 cc/sec), glottal resistance (normal = 30–50 cm H_2O/lps), subglottal pressure (normal 6–8 cm H_2O)
- **Perceptual Testing:** qualitative rating of voice features, utilizes numeric scoring system (1 = normal to 7 = most deviant from normal); GRBAS scale individually rates Grade of hoarseness, Roughness, Breathiness, Asthenic, and Strained quality of voice (0 = normal to 3 = severe)
- **Laryngeal EMG:** *see* Vocal Fold Paralysis, p. 115
- **Glottography:** measures light translumination during vibration and degree of vocal fold contact (eg, open quotient [duration vocal folds are open over the length of the glottic cycle], speed quotient [duration of glottal opening over glottal closure], and shift quotient [time of peak opening over duration of glottal opening])

UPPER AIRWAY OBSTRUCTION

Evaluation of the Stridorous Patient

Initial Management (ABCs)

- **Evaluate Airway:** quickly determine severity and stability of airway (acute versus chronic, progression of stridor, dyspnea at rest versus with exercise)
- **Establish Airway:** *see* pp. 443–447 for complete protocol in establishing an airway
- **Administer Oxygen:** masked ventilation may adequately improve oxygenation until able to secure airway; after establishing a secure airway, ease of ventilation and maintenance of oxygenation should be evaluated
- **Heliox:** may be considered for short-term oxygenation for stable airway obstructions; 80% helium, 20% oxygen (may increase O_2 concentration to 40%); helium has a lower molecular weight (decreased density) allowing passage past narrow obstruction
- consider humidification, corticosteroids, nebulized racemic epinephrine, and antibiotics

History and Physical Exam

- Character of Upper Airway Sounds: determine respiratory phase of stridor versus stertor *(see below),* onset and duration, constant versus intermittent

- Contributing Factors: complete perinatal history (maternal drug and alcohol abuse, venereal diseases, complicated delivery, neonatal intensive care unit stay); recent upper respiratory infection, fever, cough, sore throat, allergy; recent trauma, caustic ingestion, previous tracheotomy, intubations or airway manipulations, surgeries; medications (medicine allergies, ACE inhibitors); history of sarcoidosis, connective tissue disorders, granulomatous diseases (Wegener's, tuberculosis), asthma, cardiac and pulmonary problems
- Associated Symptoms: dysphagia, drooling, hoarseness, airway bleeding, weight loss, odynophagia, cough (barking cough), sleep pattern (snoring, daytime somnolence), choking (GERD, foreign body), feeding difficulties (regurgitation, worse with feeding)
- Nasopharyngoscopy/Indirect Mirror Exam: assess airway patency, vocal fold mobility, supraglottic structure, examine tracheal stoma (retroflex to access subglottis)
- Nasal Exam and Evaluation Nasopharyngeal Patency: may attempt to pass #6 catheter through both nares, nasal endoscopy, nasoseptal deformities, nasal masses, nasal congestion
- Complete Head and Neck Exam: oral cavity (macroglossia, tonsilar hypertrophy or infection), complete neurological exam (cranial nerves), evaluate for external compression (trachea midline, goiter, palpable laryngeal fractures), cutaneous lesions (hemangiomas)
- cardiac and pulmonary history and exam (wheezing, chest pain, retractions, cyanosis)
- Think "KITTENS" for differential diagnosis (*see* Table 3–2)

Description of Upper Airway Sound by Site of Obstruction

- Nasopharyngeal: **stertor** (snoring), no cough
- Oropharynx: gurgly
- Supraglottic: **inspiratory** stridor, throaty voice, **feeding problems**
- Glottic: **inspiratory** or **biphasic** stridor, hoarseness
- Subglottic: husky voice, **biphasic** stridor, **barking cough**
- Tracheobronchial: **expiratory** stridor, wheezing, suprasternal retraction indicate obstruction above thoracic inlet

Endoscopy

- Indications for Endoscopy: **(SPECS-R)** Severity of obstruction, **P**rogression of shortness of breath, **E**ating difficulties, **C**yanotic episodes, **S**leep apnea, **R**adiologic findings
- Direct Laryngoscopy: allows evaluation and instrumentation of the glottis and supraglottis

TABLE 3–2. Differential Diagnosis of Upper Airway Obstruction: KITTENS Method

	(**K**) **C**ongenital	**I**nfectious	**T**umor (Neoplasia)	**T**oxins & **T**rauma	**N**eurologic	**S**ystemic/ PSychiatric
Above larynx	Micrognathia Macroglossia Choanal atresia Lingual thyroid Nasoseptal deformity	Retropharyngeal abscess Peritonsillar abscess Mononucleosis Diphtheria	Juvenile nasopharyngeal angiofibromas Neurogenic nasal tumors	Facial fracture Retropharyngeal hematoma	Posteriorly displaced tongue Central sleep apnea	Allergic rhinitis Wegener's Obesity (obstructive sleep apnea)
Supraglottic	Laryngomalacia	Epiglottitis	Tumor	Intubation trauma		Sarcoidosis
Glottic	Glottic web Laryngeal atresia Vocal fold paralysis	TB laryngitis Laryngeal diphtheria	Respiratory papillomatosis	Laryngeal fracture Foreign body	Vocal fold paralysis	Hereditary angioedema

	(**K**) **C**ongenital	**I**nfectious	**T**umor (Neoplasia)	**T**oxins & **T**rauma	**N**eurologic	**S**ystemic/ PSychiatric
Subglottic	Vascular ring and aortic arch anomalies Tracheoesophageal fistula Subglottic stenosis	LTB (Croup)	Subglottic hemangioma	Subglottic stenosis Thyroid or neck masses (extrinsic compression)	Respiratory muscle paralysis	Wegener's Asthma
Tracheobronchial	Tracheomalacia Vascular rings	Tracheitis Bronchitis	Mediastinal, tracheal, or bronchial tumors	Foreign body		External compression (goiter)

- Bronchoscopy: flexible bronchoscopy allows identification of severity and location of stenosis, rigid bronchoscopy allows for instrumentation and for management for an emergent airway crisis

Ancillary Tests

- **Chest X-ray and Plain Neck Films:** screening films for laryngotracheal structural defects, intrinsic lung and mediastinal disease
- **CT/MRI of the Neck:** indicated to evaluate the location, extent, or compression of the stenosis; also evaluates destruction of local laryngeal structures
- **Modified Barium Swallow and Esophagram:** examine esophageal pathology, gastroesophageal reflux, aspiration, and vascular abnormalities
- **Pulmonary Function Tests and Flow Volume Loops:** identifies level of obstruction and assess for intrinsic lung disease
- **Magnified Airway (Fluoroscopy):** dynamic evaluation of airway, assess vocal fold motion
- **Arteriography:** indicated if vascular abnormalities are suspected
- **Labs:** arterial blood gas, complete blood count, electrolyte panel

Tracheotomies

Introduction

Tracheotomy Indications

- bypass upper airway obstruction (eg, sleep apnea, tumor)
- prevent complications from prolonged intubation (eg, mucosal ulceration, laryngeal stenosis, granulomas)
- assist with tracheal-bronchial suctioning (pulmonary toilet)
- provide protection from aspiration
- eliminate dead space (CNS depression)

The Pediatric Tracheotomy

- prolonged intubation generally preferred over tracheotomy in neonates up to 6 months (high risk of subglottic stenosis)
- may consider performing tracheotomy with a rigid bronchoscope to provide rigidity and facilitate dissection
- consider endoscopy every 3 months for pediatric tracheotomy to evaluate for stenosis

Tracheotomy Complications

- Intraoperative Complications: vessel injury (hemorrhage), pneumothorax, pneumomediastinum
- Immediate Postoperative Complications: postoperative pulmonary edema from release of pressure (Rx: positive pressure ventilation), acute obstruction (mucous plug), loss of airway (tracheotomy tube not secured)
- Long-term Complications: tracheal stenosis, granulation tissue, tracheal-innominate artery erosion, subglottic stenosis

Tracheotomy Management

Tracheotomy Care

- **Maintain Airway:** especially for first 48 hours to prevent accidental loss of airway; suture tracheotomy to neck skin, tight tracheotomy ties, clean inner cannula, first tracheotomy change may be considered after 3–5 days to allow the tract to form
- **Humidity:** prevents tracheal crusting and mucous plugs, initially saline should also be dropped into tube every 3–4 hours
- **Pulmonary Toilet (Aseptic Technique Suctioning):** tracheotomy tubes disrupt ciliary function, decrease subglottal pressure required for an adequate cough, and increase risk of microaspiration; requires regular suctioning of the tracheal airway, especially for the first few days
- **Skin Care:** cuff dressings may be considered to prevent skin breakdown
- **Check Cuff Pressure:** cuff pressure should be less than capillary pressure (**<25** cm H_2O) to prevent pressure necrosis (subglottic stenosis, tracheal-innominate artery erosion, tracheomalacia)
- **Feeding:** no solid food ingestion when cuff is inflated; capping tracheotomy tube facilitates swallowing

Decannulation

- tracheotomy tubes should be removed as soon as possible (especially in children) to prevent long-term sequelae such as tracheal ulceration, subglottic stenosis, tracheomalacia, etc
- prior to decannulation patient should undergo tracheotomy tube downsizing and a trial of capping (24 hours for 7 consecutive days without respiratory difficulties)
- original indication for tracheotomy must be resolved
- consider flexible nasopharyngoscopy to evaluate airway patency (evaluate subglottis with retroflexed look through the stoma)
- place airtight dressing to seal stoma after removal of tracheotomy tube

BENIGN LARYNGEAL PATHOLOGY

Congenital Laryngeal Defects

Congenital Webs

- most commonly **anteriorly** based
- Pathophysiology: incomplete recanalization at 8th week of embryological development
- Types: supraglottic (2%), glottic (**75%**), subglottic (7%)
- SSx: weak cry at birth, aphonia, variable degrees of respiratory obstruction (inspiratory stridor)
- Dx: flexible nasopharyngoscopy, direct laryngoscopy
- Rx: endoscopic lysis if >50% (laser excision), consider keel or open procedure for extensive involvement, may require tracheotomy

Congenital Subglottic Stenosis

- **<4 mm** in newborn
- third most common laryngeal anomaly
- Pathophysiology: incomplete recanalization, small diameter cricoid cartilage, or trapped first tracheal ring
- Types
  1. **Membranous:** circumferential, thickened mucous glands or fibrous tissue
  2. **Cartilaginous:** abnormal shelf on cricoid or a trapped 1st tracheal ring
  3. **Mixed**
- Grades: I. <70% obstruction, II. 70–90%, III. 91–99%, IV. complete obstruction
- SSx: **biphasic stridor** in first few months of life, may mimic croup or recurrent URI, failure to thrive
- Dx: endoscopy, chest x-ray, neck plain films, flexible nasopharyngoscopy to assess vocal fold motion

Management

- Secure Airway: may require tracheotomy (decannulation by 2–3 years old)
- Medical Management: reflux regimen, may consider corticosteroids (*controversial*)
- Grade I–II: consider **endoscopic management** (CO_2 or KTP laser excision with dilation); contraindications to endoscopic procedures include significant cartilage loss, concurrent laryngotracheal stenosis, posterior commissure involvement, circumferential cicatricial scar or thick scar (>1 cm), and infected tissue (chondritis)

Open Procedures: indicated for Grade III–IV or when endoscopic management is contraindicated

- **Anterior Cricoid Split**: no graft required, indicated to wean off of a ventilator before a tracheotomy is performed, indicated for a trapped first tracheal ring, requires adequate pulmonary reserve
- **Posterior Cricoid Split**: indicated for posterior stenosis, may use costal graft, usually requires a stent, may also be used concurrently with an anterior cricoid split
- **Anterior Laryngofissure with Anterior Lumen Augmentation**: requires an anterior graft
- **Laryngofissure with Division of Posterior Cricoid Lamina**: required if posterior glottis involvement, upper tracheal stenosis, or complete glottis stenosis
- **Laryngofissure with Division of Posterior Cricoid Lamina with anterior and posterior grafts**
- **Segmental Resection with End-to-End Anastomosis**
- **Hyoid Interposition**
- **Rotary Door Flap**
- **Epiglottic Reconstruction**

Laryngomalacia

- most common laryngeal anomaly
- most common cause of stridor in neonate and chronic pediatric stridor
- approximately 15% associated with secondary airway lesions
- Pathophysiology: immature cartilage, abnormal calcium metabolism
- SSx: **intermittent inspiratory stridor that improves in prone position**; worse with feeding, crying, or when placed on back; presents within weeks of birth; normal voice; usually self-limiting as cartilage stiffens with growth
- Most Common Laryngeal Findings
  1. inward collapse of A–E folds and cuneiform cartilage into laryngeal inlet during inspiration
  2. epiglottis collapses into laryngeal inlet
  3. short A–E folds
- Dx: clinical history and endoscopy
- Rx: observation (typically resolves with growth), epiglottoplasty (removes excess tissue), correct GERD if present, rarely requires tracheotomy or intubation

Tracheomalacia

- less common than laryngomalacia
- Pathophysiology: immature cartilage, abnormal calcium metabolism

- SSx: expiratory stridor, exacerbated with infection, usually self-limiting as cartilage stiffens with growth
- Dx: clinical history and endoscopy
- Rx: observation (typically resolves with growth), correct GERD if present, rarely requires tracheotomy or intubation

Vascular Rings

- Types
  1. **Double Aortic Arch**: **most common vascular anomaly to cause stridor**, right aortic arch persists, wraps around esophagus and trachea
  2. **Right Aortic Arch**
  3. **Anomalous Innominate**
  4. **Anomalous Left Common Carotid**
  5. **Pulmonary Artery Sling**: left pulmonary artery originates from right pulmonary artery, slings around right main stem bronchus, then between trachea and esophagus, 50% associated with distal bronchial hypoplasia
  6. **Retro-Esophageal Right Subclavian Artery** (**Dysphagia Lusoria**): most common vascular anomaly to compress aerodigestive tract (*see* p. 154)
- SSx: biphasic stridor (external compression of trachea), barking cough, dyspnea with feeding
- Dx: MRI, angiography, barium swallow, CT with contrast
- Rx: symptomatic compression requires surgical intervention (pexis of vessel or reimplantation)

Cri du Chat Syndrome

- Pathophysiology: deletion of short-arm of chromosome 5
- SSx: high pitched stridor, mental retardation, microcephaly
- Laryngeal Findings: narrowed endolarynx, diamond shaped, persistent interarytenoid cleft
- Dx: clinical findings

Posterior Laryngeal Clefts and Laryngotracheoesophageal Clefts

- Pathophysiology: posterior cricoid lamina does not fuse or tracheoesophageal septum does not develop
- associated with TE fistulas, laryngomalacia, congenital heart defects, cleft lip/palate, Down syndrome, others

- Syndromes
  1. **Opitz-Friass** (G-syndrome): hypertelorism, cleft lip/palate, hypospadias
  2. **Pallister-Hall**: hypothalamic hamarblastoma, hypopituitary, imperforate anus, postaxial polydactaly
- SSx: inspiratory stridor (like laryngomalacia), pneumonia, aspiration for Type II–IV
- Dx: endoscopy, esophagram
- Classification Types
  I. supraglottic arytenoid only
  II. extends past true vocal fold
  III. extends past cervical trachea
  IV. extends past thoracic trachea
- Rx: consider tracheotomy, Nissen fundoplication, endoscopic repair for Type I & II, anterior vs. lateral external approaches

Choanal Atresia (CHARGE Association): (*see* pp. 15–16)

Laryngitis

Acute Viral Laryngitis

- Pathogens: **rhinovirus** (most common), parainfluenza, respiratory syncytial virus, adenovirus, influenza virus, pertussis
- SSx: dysphonia, low-grade fever, hoarseness, cough, rhinitis, postnasal drip
- Dx: clinical history and exam
- Rx: conservative management required (hydration, pyretics, voice rest, decongestants, humidification, smoking cessation), antibiotics not indicated unless suspect secondary bacterial infection

Adult Supraglottitis

- Pathophysiology: typically secondary to purulent rhinosinusitis or tracheobronchitis
- Common Pathogens: ***H. influenzae*** (most common), *S. pneumoniae, S. aureus*, β-hemolytic *Streptococcus*
- SSx: fever, muffled voice, dysphagia, stridor (inspiratory), **obstructive symptoms may progress within hours**
- Dx: lateral neck plain films, flexible nasopharyngoscopy, indirect laryngoscopy

- Rx
  1. evaluate airway, severe and progressive symptoms may require fiberoptic intubation versus an urgent surgical airway
  2. humidification, hydration, corticosteroids, H_2-blocker
  3. parenteral antibiotics

Reflux-Induced Laryngitis

- Pathophysiology: inflammatory response of laryngeal mucosa from **Laryngopharyngeal Acid Reflux** (**LPR**)
- SSx: hoarseness (worse in the morning), choking spells at night, regurgitation, bitter taste in mouth, globus sensation, cough, chronic throat clearing, postprandial heartburn (seen <50% of the time in LPR)
- Laryngeal Findings: erythema and edema of the posterior commissure, arytenoids, superior surface of the vocal folds, and laryngeal surface of the epiglottis; diffuse supraglottic edema; laryngeal pachydermia (interarytenoid); granulomas of the vocal process
- for diagnosis and management *see* pp. 149–150

Acute Spasmodic Laryngitis (False Croup)

- common in toddlers
- Pathophysiology: uncertain noninfectious etiology, may be secondary to GERD or allergy
- Associated: anxiety, allergies
- SSx: **nocturnal** stridor and respiratory distress (not present during the day), otherwise normal head and neck exam aside from the stridor
- Dx: clinical history and exam
- Rx: supportive care (anti-reflux regimen), address allergies

Bacterial Tracheitis (Membranous Laryngotracheobronchitis—MLTB)

- Pathophysiology: bacterial superinfection
- SSx: thick secretions in airway, fibrinous membrane in trachea, high fever
- Dx: endoscopy with cultures
- Rx: aggressive pulmonary toilet, parenteral antibiotics, may require endotracheal intubation (pulmonary toilet)

Diphtheria

- uncommon since immunization (milder form may present despite immunization)
- Pathogen: *Corynebacterium diphtheria*

- Risks: nonimmunized children >6 years old
- SSx: sore throat, progressive airway obstruction, **thick**, **gray-green plaques, membranous, friable exudate on tonsils, pharynx, and larynx**, low grade fever, acetone breath
- Dx: flexible nasopharyngoscopy, culture and smears
- Complications: nephritis, airway obstruction, death (secondary to neurological toxin)
- Rx: establish airway via tracheotomy, avoid intubation (may dislodge exudative plaques causing an acute airway emergency), diphtheria antitoxins, antibiotics (penicillin or erythromycin), humidity

Croup (Acute Laryngotracheobronchitis, LTB) (see Table 3–3)

- most common cause of stridor in children
- primarily involves the subglottic region
- Pathogen: **parainfluenza 1** (**most common cause**), parainfluenza 3, influenza A, rhinovirus, respiratory syncytial virus
- Risks: children 1–5 years old during fall and winter seasons
- SSx: inspiratory or biphasic stridor, **gradual** onset (over days), long course (3–7 days), low grade fever, relief in the recumbent position, **brassy cough** (worse at night), hoarse, non-tender larynx, no dysphagia, no drooling

TABLE 3–3. Contrasting Acute Laryngotracheobronchitis (Croup) and Epiglottitis

	Acute Laryngotracheobronchitis	Acute Epiglottitis
Pathogen	Parainfluenza virus 1	Haemophilis influenze B
Age	<5 years old	2–6 years old
Location	subglottic	supraglottic
Onset	gradual (days)	sudden onset (hours)
Cough	barky	normal
Posture	supine	upright
Drooling	no	yes
Fever	low grade	high fevers
Radiographs	steeple sign	thumbprinting
Treatment	supportive	airway management and antibiotics

- Dx: clinical history and exam, plain neck films ("**steeple**" or "wine bottle" sign, narrowed subglottis), flexible nasopharyngoscopy
- Complications: pulmonary edema, pneumonia, membranous laryngotracheobronchitis

Management

- Assess Airway: intubation or tracheotomy rarely required unless there is a coexisting laryngeal abnormality (subglottic stenosis)
- Medical Management: humidified oxygen, parenteral fluids, nebulized racemic epinephrine, corticosteroids (*controversial*), antibiotics not required unless suspect bacterial superinfection
- Endoscopy: indicated if no resolution with conservative management or if intubation or tracheotomy required

Epiglottitis (*see* Table 3–3)

- Pathogen: **Haemophilis influenzae B**
- Risks: children >1 year old to adult (most common between 2–6 years of age)
- SSx: sudden onset (hours) and short course, **high fever**, dysphagia, **drooling**, dyspnea, "sniffing position" (neck flexed and head extended), no cough, normal voice, tender larynx
- Dx: clinical history and exam, plain neck films ("**thumbprint sign**"), serum HIB capsule antigen, cultures
- Complications: septicemia, acute airway obstruction (death)

Management

- **avoid aggravating patient** (do not examine airway with a tongue blade, draw blood, perform rectal temperatures, etc)
- Establish Emergent Airway: intubation performed in the operating room with preparation for a tracheotomy
- Endoscopy: examine and culture epiglottis
- Postoperative Care: monitored bed, parenteral antibiotics and corticosteroids for 7–10 days (consider ampicillin [20% resistance] with chloramphenicol or cefuroxime), consider extubation after 2–3 days

Chronic Laryngitis

- Common Etiologies: smoking, pollution, vocal abuse, sinusitis, rhinitis, laryngopharyngeal acid reflux
- SSx: hoarseness, pain, edema, dysphagia, respiratory compromise
- Dx: flexible nasopharyngoscopy, videostroboscopy, endoscopy (thick erythematous vocal folds) with biopsy to rule out malignancy

- Rx: address etiology (stop smoking, voice rehabilitation, treat rhinosinusitis, reflux regimen), humidification, mucolytics, consider short course of corticosteroids

TB Laryngitis

- see also pp. 209–210
- typically secondary to pulmonary TB
- Histopathology: cellular inflammation, granuloma in subepithelium, perichondritis
- Lesion: granulation and ulcerative tissue in **posterior glottis** (posterior interarytenoids most common, laryngeal surface of epiglottis, vocal folds)
- Rx: Isoniazid, Rifampin, voice rest, narcotics for pain

Syphilitic Laryngitis

- see also pp. 208–209
- rare manifestation of oropharyngeal syphilis
- Secondary Stage SSx: temporary mild edema, **painless**
- Tertiary Stage SSx: **gummas** may break down cartilage
- Rx: penicillin, tetracycline, erythromycin

Scleroma of the Larynx/Rhinoscleroma

- see also p. 34
- Pathogen: *Klebsiella rhinoscleromatis*
- Histopathology: pseudoepitheliomatous hyperplasia of the **larynx** (similar to blastomycosis)
- Rx: long-term antibiotics as dictated by culture and sensitivity

Leprosy (Hansen's Disease)

- see also p. 212
- Pathogen: *Mycobacterium leprae*
- Lesion: ulcerative lesions in the supraglottis
- Dx: biopsy (foamy leprous cells containing the bacillus), nasal smear
- Rx: Dapsone (diaminodiphenylsulfone), corticosteroids

Perichondritis of the Larynx/Polychondritis

- Causes: infection (TB, syphilis, septic laryngitis), trauma, tracheotomy, radiation effect, malignancy
- most commonly involves the thyroid cartilage, rarely involves epiglottis since fibroelastic cartilage (adherent perichondrium) protects from infection

- SSx: insidious onset, fever, odynophagia, tenderness, hoarseness, cough, dyspnea
- Dx: endoscopy reveals pale mucosal edema, CT of neck
- Complication: subperichondrial abscess, stenosis (respiratory compromise)
- Rx: establish airway, aggressive antibiotic regimen, consider incision and drainage of abscess, consider surgical debridement of necrotic or exposed cartilage
- NOTE: **Relapsing Polychondritis** has 50% laryngeal involvement (*see also* p. 317)

Fungal Laryngitis

- Risks: immunocompromised (uncontrolled diabetes, AIDS, chronic corticosteroids, etc), radiation, poor nutrition status, debilitating illnesses, long-term antibiotics
- SSx: odynophagia, mucositis, dysphonia, cough, dyspnea, aspiration
- Dx: endoscopy and biopsy (may be confused with malignancy)
- Rx: establish airway, antifungal regimen

Pathogens

- **Candidiasis** (**Moniliasis**): adherent, friable, cheesy, white plaques; spread from oral cavity (*see also* p. 174)
- **Aspergillosis**: allergic, noninvasive, or invasive forms (*see also* p. 42)
- **Blastomycosis**: **red laryngeal ulcers** or **miliary nodules on vocal fold** (*see also* p. 211)
- **Histoplasmosis**: ulcerative lesions in larynx (anterior larynx and epiglottis) (*see also* pp. 210–211)
- **Coccidiomycosis**: nodular laryngeal mass (*see also* p. 211)

Benign Laryngeal Neoplasms

Recurrent Respiratory Papillomatosis

- second most common cause of hoarseness in children
- 2/3 present before age 15, usually regresses by puberty
- extremely rare malignant change (higher risk in adults and **HPV types 16 and 18**)
- Pathophysiology: **HPV type 6 and 11** (similar to genital warts) → abnormal growth
- Risks: younger, first-time mothers (longer second stage of delivery in the birth canal), lower socioeconomic status; **50%** born from mothers with maternal condyloma acuminata, oral sex, multiple sexual partners

- Lesion: wart-like, irregular, exophytic (cluster of grapes), between any **junction of ciliated and squamous epithelium** (limen vestibuli, midzone epiglottis, ventricle margin, and undersurface of true vocal fold), nasopharyngeal, tracheal, and bronchial lesions are usually from contamination
- Types
  1. **Juvenile**: children, multiple sites of involvement, recurrent, may resolve spontaneously
  2. **Senile**: may involve single sites, recurrence less common, identical histology
- SSx: first presents with hoarseness then inspiratory or biphasic stridor, dyspnea, dysphagia
- Dx: flexible nasopharyngoscopy, videostroboscopy, endoscopy with biopsy
- Complications: pulmonary involvement (rare), may cause hemorrhage and abscess formation, respiratory compromise

Management

- Microlaryngoscopy with Laser Excision: frequent conservative endoscopic CO_2 laser ablation (biopsy to evaluate for malignancy)
- **avoid tracheotomy**, may seed lower airway or stoma
- Postoperative Management: yearly chest x-rays to evaluate pulmonary involvement, close follow-up with interval endoscopies (2–4 weeks after initial treatment)
- Adjunctive Therapy: (*controversial*) αINF (induces an antiviral and antiproliferative effect), acyclovir, Indole-3-carbinol

Chondromas

- more common in men
- Pathophysiology: most commonly arise from internal **posterior cricoid cartilage** (hyaline cartilage), may also arise from thyroid, arytenoid, epiglottic cartilage (fibroelastic)
- SSx: insidious hoarseness from vocal fold restriction, dyspnea for subglottic lesions, dysphagia for posterior cricoid lesions, globus sensation
- Lesion: smooth, firm, fixed tumor, normal mucosa
- Dx: endoscopic wedge biopsy, CT of neck (calcification)
- Rx: complete excision via an endoscopic or external approach (depending on the size of lesion)

Granular Cell Tumor

- 3% risk of malignant degeneration
- Pathophysiology: arise from Schwann cells in the posterior aspect of true vocal fold or arytenoids (originally believed to arise from myoblasts), may also be found on tongue, skin, breast, and subcutaneous tissue
- SSx: insidious hoarseness
- Lesion: small, sessile, gray mass
- Dx: endoscopy with biopsy
- Histopathology: may induce **pseudoepitheliomatous hyperplasia** near epithelial borders (**often confused with SSC**), polygonal uniform cells with vesicular nucleus, coarsely cytoplasmic eosinophilic granules, PAS and S-100 positive
- Rx; complete excision via an endoscopic or external approach (depending on the size of lesion)

Lymphangiomas (Cystic Hygromas)

- 90% present before 3 years old (65% present at birth), may persist in adult
- associated with venous malformations (lymphatics and venous system develop concurrently)
- SSx: **soft solitary painless compressible mass** (lymphatic dilation), dysphagia, dyspnea, may remain dormant
- Dx: clinical exam, endoscopy and biopsy
- Histopathology: lymphendothelial hyperplasia, during proliferative phase there is an increase in mast cells, during the involutional stage there are few mast cells
- Complications: respiratory distress, infection, disfigurement
- Rx: early conservative excision when symptomatic (spare vital structures), low rate of recurrence if completely removed (only 50% if gross tumor remains)

Hemangioma

- **most common head and neck neoplasm in children**
- typically presents by 6 months old then involutes by 2 years of age
- most common laryngeal site **left posterior lateral quadrant of subglottis** (although may appear anywhere in upper respiratory tract)
- 50% of subglottic hemangiomas associated with cutaneous involvement
- Pathophysiology: abnormal blood vessel growth
- Types: **Infantile** (typically subglottic), **Adult Onset**, **Compact** or **Capillary** (more common in infantile type, typically resolves), **Cavernous** (may enlarge rapidly, less chance of regression)

- SSx: polypoid or sessile lesions (left > right), biphasic stridor, worse with crying (hemangiomas become engorged with blood), dysphonia, dysphagia, seldom causes bleeding in the larynx
- Dx: endoscopy (avoid biopsy)

Management

- may observe if asymptomatic and nonprogressive
- embolization
- corticosteroids or interferon α-2A
- endoscopic CO_2 or argon laser excision (YAG lasers, although used for vascular tumors, do not work well with hemangiomas) or open excision for large lesions
- radiation therapy may be considered although increases risk of malignant degeneration

Systemic Diseases Affecting the Larynx

Sarcoidosis

- *see also* p. 207
- Laryngeal SSx: supraglottic submucosal mass (epiglottis most common), dysphonia, globus sensation, dyspnea
- Rx: endoscopic removal for symptomatic lesions (hoarseness or airway obstruction), may consider corticosteroids for significant exacerbations

Wegener's Granulomatosis

- *see also* p. 214
- most commonly involves the subglottis
- Laryngeal SSx: subglottic mass, dyspnea, biphasic stridor
- Rx: endoscopic removal for symptomatic lesions, medical management

Amyloidosis

- *see* p. 178
- Laryngeal SSx: anterior subglottic mass (polypoidal covered with smooth mucosa)
- Rx: endoscopic removal for symptomatic lesions (hoarseness or airway obstruction)

Arthritis of Cricoarytenoid Joint

- cricoarytenoid joint mostly commonly affected
- Rheumatoid Arthritis most common etiology

- SSx: hoarseness, stridor, dysphagia, pain with swallowing
- Rx: corticosteroids, antireflux regimen

Acquired Laryngeal Stenosis

Glottic and Supraglottic Stenosis

- Causes: blunt trauma, endotracheal tube trauma, infection, caustic ingestion, infection, foreign body
- SSx: inspiratory or biphasic stridor, dyspnea, cough
- Dx: flexible nasopharyngoscopy, endoscopy, or videostroboscopy, CT of neck
- Rx: endoscopic excision (laser) or dilation for thin webs; thicker webs require external approaches

Subglottic Stenosis

- typically more severe and more common than congenital
- Causes
  1. **Endotracheal Intubation**: pressure necrosis results in ulceration and cartilage exposure, healing occurs by secondary intention causing fibrosis
  2. **Postoperative**: pressure necrosis from a high tracheotomy or from a cricothyroidotomy
  3. **Granulomatous Disease**: tuberculosis (most common granulomatous disease of larynx), sarcoidosis, rhinoscleroma (*Klebsiella*), Wegener's granulomatosis
  4. **Infectious**: leprosy (epiglottic and vocal fold ulceration), syphilis, blastomycosis, coccidiomycosis, histoplasmosis
  5. **Idiopathic**: amyloidosis
  6. **Trauma**: foreign body, caustic ingestion, blunt trauma, hematoma, thermal injury
  7. **Systemic**: connective tissue disorders, GERD, radiation effects
  8. **Neoplasia**: chondroma, fibroma, malignancy
- SSx: dyspnea, biphasic stridor, cough, dysphagia
- Dx: endoscopy, CT of neck
- Rx: similar to Congenital Subglottic Stenosis

Other Laryngeal Lesions

Laryngeal Edema

Angioedema

- Types
  1. **Acquired Angioedema**: histamine mediated inflammation (Urticaria) secondary to a variety of substances
  2. **Congenital** (**Hereditary**) **Angioedema**: deficiency in **C1 esterase inhibitor** (controls the complement pathway)
- Common Causes of Acquired Angioedema: medications (**ACE inhibitors**, ASA, antibiotics, NSAIDs), food allergies (eggs, peanuts), insect bites, transfusions, infections (Hepatitis B, viral infections), emotional, other allergens
- SSx; rapid onset of facial, oropharyngeal, or laryngeal edema, "hot potato voice," stertor or stridor, pruritis, hoarseness
- Dx: clinical history, flexible nasopharyngoscopy, C1 esterase inhibitor serum levels
- Rx
  1. evaluate airway, severe and progressive symptoms may require fiberoptic intubation versus an urgent surgical airway
  2. epinephrine, parenteral corticosteroids, H_1 and H_2 blockers, aminophylline
  3. consider prophylactic **Danazol** for Hereditary Angioedema (increases C1 esterase inhibitor)

Reinke's Space Edema (Polypoid Degeneration)

- **Reinke's space**: superficial layer of the lamina propria, loose connective tissue (susceptible to fluid accumulation)
- Risks: GERD, smoking, hypothyroidism, vocal abuse, chronic throat clearing, chronic cough
- not associated with increased risk of laryngeal cancer
- Dx: indirect mirror exam, flexible nasopharyngoscopy, endoscopy, videostroboscopy
- Rx
  1. evaluate and manage for hypothyroidism and GERD (antireflux medications)
  2. voice rest, smoking cessation
  3. consider microlaryngoscopy and excision with removal of gelatinous material in Reinke's space

Other Causes of Laryngeal Edema

- infection (*see* pp. 101–102)

- trauma (intubation)
- venous obstruction, obstructed lymphatics
- hypoproteinemia
- increased permeability secondary to connective tissue disorders, leukemia, hypothyroid myxedema

Laryngeal Cysts

- **Vocal Fold Cyst**: subepithelial cyst, may open to free margin of vocal fold resulting in a sulcus, associated with reactive changes of opposite vocal fold; Rx: voice therapy (usually does not resolve), microflap excision (opposite vocal fold reactive changes typically resolve after removal of cyst)
- **Mucus-Retention Cyst**: minor salivary glands cyst, **supraglottic lesion**, may be obstructive (especially in children); Rx: endoscopic laser excision or marsupialization
- **Branchial Cleft Cyst**: (*see* pp. 194–195)
- **Ventricular Prolapse**: not a cyst, laryngeal ventricle protrudes between true and false vocal folds, associated with chronic bronchitis; Rx: endoscopic excision

Acquired Subglottic Cysts

- Pathophysiology: obstruction of mucous glands by endotracheal tube (requires less trauma than intubation granulomas)
- SSx: **post-extubation stridor occurring after short-term intubation**, cyst in lateral subglottis
- Dx: flexible nasopharyngoscopy, endoscopy, or videostroboscopy
- Rx: endoscopic excision (laser)

Laryngocele and Saccular Cysts

- **Laryngocele**: air filled dilation of the appendix of the ventricle, communicates with laryngeal lumen
- **Laryngopyocele**: infected pus-filled laryngocele
- **Saccular Cyst**: fluid filled dilation of the saccule without communication with the laryngeal lumen
- Pathophysiology: congenital or acquired expansion from increased intraglottic pressure of laryngeal saccule (blind sac in ventricle)
- Types
  1. **External**: laryngocele sac protrudes through thyrohyoid membrane presenting as a neck mass
  2. **Internal**: laryngocele sac remains within thyroid cartilage, less common
  3. **Combined**

- SSx: lateral compressible mass that increases in size with intralaryngeal pressure (external); cough and hoarseness (internal)
- Dx: indirect mirror exam, flexible nasopharyngoscopy, endoscopy, or videostroboscopy, CT of neck
- Complications: secondarily infected, malignant potential, airway obstruction
- Rx: marsupialization, complete endoscopic removal (laser) for internal laryngoceles, open approach for external laryngoceles

Presbylaryngis/Bowing Defects of the Vocal Folds

- Pathophysiology: aging results in ossification of the laryngeal skeleton, arthritis of the cricoarytenoid and cricothyroid joints, degeneration of the vocal fold layers (resulting in a **bowing defect**)
- Causes: screaming, coughing, throat clearing, toxic fumes, smoking, gastroesophageal reflux, endotracheal tube, allergy, rhinosinusitis, laryngitis
- SSx: hoarse-breathy voice, higher than normal pitch, voice fatigue, may have voice tremors, potential risk of aspiration
- Dx: indirect mirror exam, flexible nasopharyngoscopy, endoscopy, or videostroboscopy
- Rx: voice therapy, address aspiration if present

Vocal Fold Lesions Secondary to Vocal Abuse and Trauma

- Pathophysiology: trauma induced dilation of vessels on superior surface of vocal folds results in reactive changes (hyperemia, edema, submucosal hemorrhage, or possible scarring)
- Causes: screaming, coughing, throat clearing, toxic fumes, smoking, gastroesophageal reflux, endotracheal tube, allergy, rhinosinusitis, laryngitis
- SSx: hoarseness, odynophagia, odynophonia
- Dx: indirect mirror exam, flexible nasopharyngoscopy, endoscopy, or videostroboscopy
- Rx: voice rest, speech therapy, humidification, antireflux regimen, smoking cessation

Contact Ulcers

- ulceration on medial aspects of vocal processes of arytenoids
- Rx: conservative management (as above), antibiotics

Vocal Fold Granulomas (Intubation Granulomas)

- granulation tissue on the **medial aspect of the arytenoids near the vocal process**, posterior commissure region

- typically self-limiting
- Rx: remove or place smaller sized endotracheal tube, reflux regimen, antibiotics, speech therapy, may consider corticosteroids, rarely surgical intervention required (excision may be considered if symptomatic and pedunculated lesion, avoid removal of sessile lesions)

Vocal Nodules (Singer's Nodules)

- more common in young women and children
- **bilateral** inflammatory tissue at **junction of anterior and middle third of vocal fold** (the middle portion of the membranous vocal fold has the greatest amplitude in the mucosal wave making it susceptible to injury with voice abuse)
- Types
  1. **Acute**: edematous, soft, erythematous, vascular vocal fold lesion
  2. **Chronic**: more organized fibrosis, hard, white, thickened vocal fold lesion
- Rx: conservative management (as above), surgical endoscopic excision for failed voice therapy (avoid excision in children, high recurrence rate)

Vocal Fold Polyps

- typically unilateral, middle and anterior vocal folds at **free edge**
- Types
  1. **Mucoid**: translucent, broad-based vocal fold lesion, stems from inflammation in Reinke's space
  2. **Angiomatous**: hemorrhagic (erythematous), protuberant, multinodular vocal fold lesions
- Rx: conservative management (as above), microflap excision for symptomatic lesions

NEUROGENIC AND OTHER VOCAL PATHOLOGIES

Vocal Fold Paralysis

Evaluation of Vocal Fold Paralysis

History and Physical Exam

- Unilateral Vocal Fold Paralysis SSx: hoarse-breathy dysphonia, aspiration, stridor in children, dysphagia, limited phonation time, vocal fatigue, may be asymptomatic
- Bilateral Vocal Fold Paralysis SSx: inspiratory or biphasic stridor, weak cry, aspiration, hoarseness (voice may be normal)

- Contributing Factors: recent upper respiratory infection, fever, cough; previous neck trauma, toxin exposure, surgery (cardiothoracic, thyroid), or airway manipulation; history of tobacco or alcohol abuse; past medical history of cancer, cardiopulmonary disease, peripheral nerve disease and other neurological disorders, diabetes; neonatal history (complications, birth trauma, maternal infections, congenital defects)
- Physical Exam: neck masses (thyroid masses), complete neurological exam
- Indirect Mirror Exam or Flexible Nasopharyngoscopy: evaluate vocal fold movement, positioning during phonation, symmetry, pooling of secretions, movement of the arytenoids, **PPP rule** (**P**osterior commissure **P**oints to **P**aralyzed side in unilateral superior laryngeal nerve paralysis)

Ancillary Tests

- **Direct Laryngoscopy**: allows for arytenoid palpation to differentiate paralysis versus fixation
- **Videostroboscopy**: provides documentation, mucosal wave characteristics
- **Chest X-ray**: evaluates thoracic etiology
- **CT from Base of Skull to Aortic Triangle**: evaluate lesions along course of vagus nerve
- **Modified Barium Swallow/Esophagram**: evaluate for aspiration, esophageal lesions, vascular abnormalities
- **Laryngeal Electromyography**: determine vocal fold paralysis versus fixation, superior laryngeal nerve versus recurrent laryngeal nerve injury, myopathies (normal frequency, lower amplitude), neuropathies (lower frequency, normal amplitude), and reinnervation (polyphasic potentials)
- **Labs**: complete blood count with differential, trepemonal studies (FTA-ABS), Lyme titers, thyroid function tests, toxin screen (lead and arsenic levels), fasting blood sugar

Vocal Fold Positioning

- vocal fold position does not necessarily predict site of lesion
- **RLN Paralysis**: **paramedian vocal folds**, preserves partial ADduction from the action of the cricothyroid muscle (the only extrinsic muscle innervated by the SLN)
- **SLN Paralysis**: loss of cricothyroid innervation results in loss of vocal fold tension (lowers pitch of voice), bowing deformity, and loss of laryngeal sensory innervation (increased risk of aspiration), selective SLN injuries are rare (except thyroid surgery)
- **RLN and SLN Paralysis**: "**cadaveric**," intermediate vocal folds

- **Bilateral Vocal Fold Paralysis**: typically near midline (stridor, dyspnea, near normal voice) or abducted (breathy dysphonia, vocal fatigue, limited phonation time)

Causes of Vocal Fold Paralysis in Adults

- **Neoplastic**: (most common cause) bronchogenic and pulmonary neoplasia, skull base lesions, esophageal, laryngeal, thyroid tumors, mediastinal disease
- **Iatrogenic Injury**: thyroidectomy, neck surgery, cardiothoracic, vascular, and neurosurgical procedures; post-intubation neurapraxia (increased risk with cuff placed in proximal subglottis resulting in a pressure induced neurapraxia of RLN)
- **Idiopathic**: usually self limiting (may take up to 12 months to resolve)
- **Trauma**
- **Neurological**: multiple etiologies that affect nucleus ambiguus in the medulla including poliomyelitis, pseudobulbar palsy, myasthenia gravis, amyotrophic lateral sclerosis, brainstem stroke, multiple sclerosis, **Wallenberg Syndrome** (infarct of the posterior-inferior cerebellar artery [PICA] results in a brainstem nuclei infarct)
- **Infectious**: Lyme disease, syphilis, EBV, tuberculosis, viral
- **Systemic Diseases**: sarcoidosis, diabetes mellitus, cardiomegaly
- **Toxins**: lead, arsenic, quinine, streptomyocin

Causes of Vocal Fold Paralysis in Pediatrics

- **Arnold-Chiari Malformation and Meningomyelocele**: must **always** be considered in the neonate, compression of the 4th ventricle prevents flow of CSF resulting in compression of brainstem or increased intracranial pressure (ICP) forces brain stem herniation causing traction against vagal rootlets in foramen magnum; Rx: return of vocal fold function with relief of ICP
- **Other Neurological Conditions**
- **Idiopathic**
- **Birth Trauma**: increased risk with complicated deliveries (C-section, traction proposed mechanism), unilateral paralysis more common
- **Iatrogenic Injury**: high risk with tracheoesophageal fistula and congenital heart (ductus repair) repairs
- **Infection**: Syphilis
- **Vascular Abnormalities**

Unilateral Vocal Fold Paralysis Management

- must determine if self limiting or permanent paralysis
- left vocal fold paralysis more common in adults because left RLN is longer (greater risk of injury); however, congenital anomalies affect the

right vocal fold because of its shorter length (more susceptible to traction forces)
- may not require surgical management if other vocal fold compensates to provide adequate voice quality and no aspiration (observation with voice therapy)
- goal of unilateral surgical procedures is to **medialize** vocal fold without compromising the airway

Vocal Fold Injections

- Indications: elliptical (bowed) defects, may be used as a temporizing procedure
- Advantages: easy, immediate improvement, may be completed in the office (although most often managed in the operating room under local or general anesthesia)
- Disadvantages: irreversible if Teflon used, changes mucosal wave, does not correct large posterior defects, contraindicated if vocal folds are not at the same level
- Complications: granuloma formation (with superficial injections or migration of Teflon), under- or over-injection (risk of airway compromise), acute hypersensitivity
- Injection Materials
  1. **Gelfoam**: indicated if recovery is expected (neurapraxia), 30% overcorrection to compensate for saline resorption, effective for 4–6 weeks
  2. **Fat**: effective for 6 months (or longer), no foreign body reaction, up to 50% resorption after 6 months, 40% overcorrection to compensate for resorption
  3. **Collagen**: autologous or bovine collagen (risk of host reaction), effective for up to 3 years, collagen eventually is resorbed and replaced with host tissue
  4. **Teflon Paste** (tetrafluoroethylene): largely been replaced due to risk of granuloma formation with superficial injection, risk of migration, stiffening of the mucosal wave, and difficulty with removal

Thyroplasty

- Types
  - **I**: indicated for membranous vocal fold defects resulting in breathy dysphonia or aspiration, medializes vocal fold by inward lateral compression with an implant (silastic, gortex, hydroxyapatite) placed via a window in the thyroid cartilage
  - **II**: lateral expansion (thyroid cartilage split with graft placed to widen anteriorly by lateralizing the vocal folds)
  - **III**: indicated to lower vocal pitch or address adductor spasmodic dysphonia by shortening and relaxing vocal folds

IV: indicated to increase vocal pitch by lengthening and tensing vocal folds

- Advantages: reversible, preserves mucosal wave, immediate results, may be done under local anesthesia to allow for voice and visual feedback
- Disadvantages: requires a neck incision, technically more difficult to perform, may not be adequate for posterior defects
- Complications: morbidity from neck incision (hematoma, wound infection, bleeding), under- or over-compensation (risk of airway compromise), implant extrusion or migration

Arytenoid Adduction

- Indications: large posterior glottic chinks (triangular defects) or uneven vocal folds
- Advantages: may be used in combination with a Type I thyroplasty
- Disadvantages: irreversible
- Complications: hematoma, airway compromise, slipped sutures, pharyngocutaneous fistula

Reinnervation Procedures

- Indications: unilateral permanent vocal fold paralysis
- Advantages: maintains muscle tone, no foreign body reaction, best preservation of mucosal wave, most physiologic
- Disadvantage: does not result in active ADduction and ABduction, long operative time, delayed results (up to 6 months), high technical skill, may require a Gelfoam injection until reinnervation becomes effective
- ansa cervicalis nerve has similar fiber composition (myelinated versus unmyelinated) to the RLN making it compatible for RLN grafting
- Types
  1. end-to-end anastomosis of RLN
  2. ansa cervicalis to main trunk of RLN
  3. anastomosis of ansa cervicalis to adductor branch of RLN
  4. ansa cervicalis neuromusclar pedicle (omohyoid) to TA muscle
  5. vagus, ansa, or phrenic nerve fibers or motor end plate insertion into thyroarytenoid

Tracheotomy

- indicated for respiratory distress or chronic aspiration (for pulmonary toilet)
- *see also* pp. 96–97

Bilateral Vocal Fold Paralysis Management

- goal is to **lateralize** vocal fold for airway without compromising the voice and causing aspiration

- **Tracheotomy**: gold standard treatment for stabilizing airway, must undergo lateralizing procedure (at least 4 mm) before decannulation
- **Cordotomy** (**Laser**): transect between vocal process and vocal fold
- **External or Endoscopic Arytenoidectomy**: arytenoid excised (total or partial), risk of posterior glottic scarring, may use laser with endoscopy
- **Arytenoidopexy**: lateralize vocal fold by plicating vocal process to external laryngeal architecture
- **Reinnervation Procedures**: attach neuromusclar pedicle to PCA for ABduction (*see above*)

Other Neurogenic Voice Pathologies

Adductor Spasmodic Dysphonia

- more common than ABductor spasmodic dysphonia
- Pathophysiology: focal laryngeal dystonia (may be psychogenic origin), causes vocal fold **hyper-ADduction** (phonation against a closed glottis), exacerbated by stress
- SSx: strained or strangled voice, glottic stammering (phonation breaks); may be associated with other dystonias, tremors, or difficulty in breathing
- typically patients are able to whisper normally
- Dx: clinical exam, history, and voice profile
- Rx: voice therapy, **botulinum toxin injections of thyroarytenoid and lateral cricoarytenoid muscles** (effective for an average of 3 months)

Abductor Spasmodic Dysphonia

- Pathophysiology: focal laryngeal dystonia (may be psychogenic), causes vocal fold **hyper-ABduction** during voicing; exacerbated by stress
- SSx: abnormal whispered or breathy breaks during phonation, especially during voice onset
- Dx: clinical exam, history, and voice profile
- Rx: voice therapy, **botulinum toxin injections** of **posterior cricoarytenoid muscles** (effective for an average of 3 months)

Spastic Dysarthria

- Pathophysiology: bilateral pyramidal tract disorder resulting in weakness, paresis, hypertonicity, and hyper-ADduction of the vocal folds
- SSx: strained-strangled voice, periodic arrests of phonation
- Dx: indirect laryngeal exam (videostroboscopy, mirror, or fiberoptic nasopharyngoscopy)
- Rx: voice therapy

Flaccid Dysarthria (*see* Vocal Fold Paralysis above)

Hypokinetic Dysarthria

- Pathophysiology: degenerative disease of the upper brain stem (**Parkinson's disease**) results in hypertonicity and rigidity of laryngeal muscles
- SSx: hoarse-harsh voice, limited volume and pitch
- Dx: indirect laryngeal exam (videostroboscopy, mirror, or fiberoptic nasopharyngoscopy)
- Rx: voice therapy

Hyperkinetic Dysarthria

- *see also* Spasmodic dysphonia (*above*)
- Pathophysiology: disorder of the extrapyramidal system resulting in an organic voice tremor
- SSx: tremorous voice
- Dx: indirect laryngeal exam (videostroboscopy, mirror, or fiberoptic nasopharyngoscopy)
- Rx: voice therapy or botulinum toxin injections of thyroarytenoid

Ataxic Dysarthria

- Pathophysiology: cerebellar disorder resulting in incoordinated, clumsy, tremulous laryngeal muscular contraction during voicing (**not at rest**)
- SSx: uncontrolled loudness and pitch outbursts, mild hoarseness
- Dx: indirect laryngeal exam (videostroboscopy, mirror, or fiberoptic nasopharyngoscopy)
- Rx: voice therapy

Other Voice Disorders

Puberphonia

- Pathophysiology: occurs during puberty, difficulty adjusting to the larger, more mature larynx
- SSx: recurrent "cracked," shrill, high-pitched voice
- Dx: clinical exam, history, and voice profile
- Rx: voice therapy

Psychological Dysphonias

- **Conversion Disorders**: usually breathy-hoarse dysphonia with a normal laryngeal exam, not under voluntary control, secondary psychological need; Rx: voice therapy, psychological counseling
- **Plica Ventricularis**: faulty use of false vocal folds (hyper-ADduction), must rule out organic etiology

CHAPTER

4

General Otolaryngology

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THYROID AND PARATHYROIDS

Anatomy and Physiology

Embryology and Anatomy

- Thyroid Gland: two pear-shaped lobes, isthmus (connect the left and right lobes), and a pyramidal lobe (may present as a superior extension of the embryological thyroid duct)
- each parathyroid weighs 20–40 mg, the normal thyroid gland weighs approximately 20 gms
- **Superior Parathyroids**: most commonly located in the posterolateral aspect of the superior pole, 1 cm above the intersection of the recurrent laryngeal nerve and the inferior thyroid artery
- **Inferior Parathyroids**: more variable location, most commonly located 1–2 cm from the entrance of the inferior thyroid artery into the lower thyroid pole (may also be associated with the superior thymus)
- **Berry's ligament** (lateral suspensory ligament): attaches thyroid lobes to the trachea
- **C-cells** (parafollicular cells): cells within thyroid gland that secrete calcitonin

Embryology

- Thyroid Embryology: endoderm between the first and second branchial arch on the floor of pharynx (**foramen cecum**) invaginates around the fourth week and descends into mesenchymal tissue along the path of the thyroid duct (anterior to the hyoid bone) forming a ventral diverticulum that differentiates at the distal end into the thyroid anlage, the proximal portion typically atrophies by the sixth week
- Parathyroid Embryology: the **third** dorsal branchial pouch → inferior parathyroids and thymus; the **fourth** dorsal branchial pouch → superior parathyroids and C-cells of the thyroid
- Embryological Pathology: **Athyreosis** (rare), **Ectopic Thyroid** (may be found anywhere along the thyroid duct from the tongue as a **lingual thyroid** to the sternal notch), **Thyroglossal Duct Cyst** (*see* p. 195), **Supernumery Parathyroids**, **Aberrant Parathyroids** (most common location at the **anterior superior mediastinum**)

Vasculature

- external carotid artery → superior thyroid artery → superior pole of the thyroid
- subclavian artery → thyrocervical trunk → inferior thyroid artery → lateral lobes of the thyroid and the **inferior and superior parathyroid arteries** (superior parathyroid arteries may also arise from superior thyroid artery)

- aortic arch or innominate artery → **thyroidea ima artery** → the thyroid isthmus
- venous drainage from the superior, middle, inferior (largest) veins into the internal jugular and innominate veins

Nerves

- **Superior Laryngeal Nerve** (SLN): the external branch (motor fibers) parallels the superior thyroid artery and descends to innervate the cricothyroid muscle; the internal branch parallels the superior thyroid artery then pierces the thyrohyoid membrane (may anastomose with the sensory branch of the RLN to form the **loop of Galen**)
- **Recurrent Laryngeal Nerve** (RLN): ascends 1 cm lateral to the tracheoesophageal groove, closely associates with the inferior thyroid artery, near the middle 1/3 of the gland the nerve crosses posterior or superficial to the inferior thyroid artery, continues superior and medial along the posterior thyroid capsule, then enters the larynx between the cricoid cartilage and the inferior cornu of the thyroid cartilage (at the articulation)

Lymphatics

- isthmus and median lateral lobes → upward to the **delphian** (prelaryngeal), and digastric nodes
- inferior lateral lobes → pretracheal and cervical nodes

Thyroid Hormone Physiology

Thyroid Hormone (TH) Synthesis and Release

- anterior pituitary secretes TSH → increases thyroid iodide uptake (stored in lumen)
- iodination of thyroglobulin forms monoiodotyrosine (MIT) and diiodotyrosine (DIT) molecules (**organification**)
- MIT and DIT molecules link together to form **triiodothyronine** (T_3) or **thyroxine** (T_4), stored within the "colloid"
- released into blood after endocytosis and fusion with lysosome to release T_3 and T_4 (**T_4 is 90% of thyroid output**)
- Thyroid Hormone Transport: **Thyroxine-Binding Globulin** (TBG, made from the liver, increased with increased estrogen and pregnancy, binds 75% of T_4); **Thyroxine Binding Pre-Albumin** (TBPA, binds 15% of T_4); and **Albumin** (binds 5% of T_4)
- liver, kidneys, muscle, and anterior pituitary convert T_4 to T_3 via 5' monodeiodinase (**T_3 is four times more active than T_4 and binds with higher affinity to TBG**) and reverse T_3 (inactive form)

Regulation

- Thyrotropin-Releasing Hormone (TRH): released from supraoptic and paraventricular nuclei of hypothalamus (**not** affected by TH)
- Thyroid Stimulating Hormone (TSH): secreted by anterior pituitary, stimulates iodide trapping, increases release of thyroid hormone, activates growth of thyroid gland (inhibited by TH for negative feedback)
- **Wolff-Chaikoff Effect**: excess iodine inhibits thyroid hormone (usually temporary)

Thyroid Hormone Effects

- elevates metabolic rate (thermogenesis, increases oxygen consumption)
- essential for normal neural and skeletal development (stimulates chondrocytes, bone reabsorption, growth of neuronal tissue)
- increases sympathetic activity (increases heart rate and contractility)
- releases steroid hormones
- stimulates erythropoiesis

Pharmacology

- **Thinamides**: **Propylthiouracil** (PTU), **Methimazole** (Tapazole), inhibits T_3 conversion and the oxidation and organification of iodine; may cause hepatitis, agranulocytosis, parotiditis; Methimazole is contraindicated in **pregnancy**
- **Iodine, Lugol's solution**: excess iodine inhibits thyroid hormone (**Wolff-Chaikoff Effect**); contraindicated in rheumatoid arthritis
- **Glucocorticoids**: suppress the hypothalamic–pituitary–thyroid axis
- **Lithium**: inhibits thyroid hormone release; contraindicated in renal failure and cardiovascular disease
- **Propanolol, Metoprolol**: (β-blockers) used to control the peripheral manifestation of sympathetic overactivity (inhibits thyrotoxicosis)

Thyroid Function Tests (TFTs, *see* Table 4–1)

- **Total T_4**: radioimmunoassay measures free and bound T_4
- **Free T_4**: measures unbound T_4, more specific for hypo- and hyperthyroidism
- **TSH**: radioimmunoassay measures TSH, most sensitive test for primary hypo- and hyperthyroidism
- **Total T_3**: radioimmunoassay measures free and bound T_3, useful for toxic nodules and toxic multinodular goiters (higher increase in T_3 than T_4)
- **TRH Stimulation Test**: measures TSH after infusion of TRH, tests pituitary secretion of TSH and hypothalamic response

TABLE 4–1. Thyroid Function Test Results

Condition	TBG	Total T_4	RT_3U	Free T_4
Normal	Normal	Normal	Normal	Normal
Pregnant	↑	↑	↓	Normal
Liver/Renal disease	↓	↓	↑	Normal
Hyperthyroid	Normal	↑	↑	↑
Hypothyroid	Normal	↓	↓	↓

- **Radioactive Iodide Uptake** (**RAIU**): measures the percentage of radiolabeled iodine taken up by the thyroid, assess metabolic status
- **Calcitonin**: elevated in medullary thyroid carcinoma
- **Resin T_3 Uptake** (**RT_3U**): measures the binding capacity of existing TBG, indirect measurement of TBG, an increased RT_3U suggests a decreased total TBG (pregnancy or estrogen from oral contraceptives increases TBP and therefore increases total T_4, however will have a normal euthyroid state (normal free T_4)

Calcium Physiology

- calcium is 46% ionized and 46% bound to albumin (H^+ competes with protein binding of calcium, therefore acidosis increases more ionized calcium)
- 99% of calcium is stored in bone
- **Vitamin D**: stimulates calcium and phosphate absorption in the intestine (minor effect of bone reabsorption and kidney reabsorption)
- **Parathyroid Hormone** (**PTH**): increases serum calcium and decreases serum phosphate by stimulating osteoclastic reabsorption of bone, calcium absorption in the kidney, vitamin D production, and phosphate excretion
- **Calcitonin**: produced by parafollicular cells (C-cells), inhibits calcium reabsorption from bone and increases kidney clearance of calcium and phosphate

Thyroid Nodules and Cysts

Evaluation of the Thyroid Nodule (Fig. 4–1)

History and Physical Exam

- Associated Symptoms: rate of growth of nodule, hoarseness, pain, dysphagia, symptoms of hypo- or hyperthyroidism (*see below*)

Figure 4–1. Flow Diagram of the evaluation of the solitary nodule. Reprinted with permission from Gates GA. *Current Therapy in Otolaryngology Head and Neck Surgery.* 6th ed. St. Louis, MO: Mosby; 1997.

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- Contributing Factors: radiation exposure, family history of thyroid disorders
- Physical Exam: palpation of nodule (consistency, mobility, size, tenderness), cervical adenopathy, vocal fold mobility (nasopharyngoscope), stridor

Fine Needle Aspiration (FNA)

- indicated to identify benign nodules, malignant nodules, metastasis, and lymphoma
- 1–10% false-negative, depending on size of needle, experience of pathologist and technician
- fine needle biopsies are preferred over large-bore needles to reduce the risk of malignant seeding and to allow for multiple biopsies
- larger needle biopsies may be considered for failed fine needle aspiration

Interpretation and Management

- requires adequate specimen and a well-trained pathologist
- cysts tend to reduce after aspiration (may be cystic degeneration in malignancy)
- Benign: consider observation
- Uniform Follicular Epithelium and Abundant Colloid: suggests nodular or adenomatous goiters
- Inflammatory Cells: suggests thyroiditis
- Papillary Cells (psammoma bodies, giant cells): suggests papillary carcinoma since adenoma is exceedingly rare
- Follicular Cells: may be adenoma or carcinoma, requires a hemithyroidectomy to examine architecture for differentiation (extracapsular spread, lymph or vascular invasion, or metastasis indicate carcinoma)
- Amyloid Deposits (stained with Congo red): suggests medullary carcinoma
- Undifferentiated, Bizarre Cells: suggests anaplastic carcinoma

Thyroid Radionucleotide Studies/Scintigraphy

- Indications: to determine function of thyroid gland or nodule, identify ectopic thyroid tissue (including retrosternal goiter, lingual thyroid, and metastasis), determine size, shape, and symmetry of gland

Radionucleotides

- ^{131}**I**: high radiation burden, results available in 48–72 hours, tracer of choice to evaluate **metastasis**, also used to induce local tissue damage for hyperthyroidism
- ^{123}**I**: expensive, must be delivered daily, testing requires 2 visits at 4 and 24 hours (shorter half-life)

- **^{99m}Tc pertechnetate**: trapped by follicular cells, does not measure uptake, low dose radiation, less expensive, image obtained in single visit within 30 minutes

Interpretation and Management

- hypofunctioning ("**cold**") nodules have a 5–20% malignancy rate, consider lobectomy and isthmusectomy, decision of completion of thyroidectomy based on accuracy of pathology (may consider a frozen section)
- hyperfunctioning ("**hot**") nodules may be observed, 4% malignancy rate

Other Tests

- Screening Thyroid Function Tests: TSH, free T_4
- Ultrasound: indicated to distinguish cysts, guide FNAs, and identify nonpalpable lesions
- CT/MRI: evaluates substernal goiter, nodal involvement, airway and vascular displacement, tumor invasion
- Chest X-ray: metastasis work-up, tracheal displacement

Thyroid Cysts

- Pathophysiology: typically arises from degenerated nodules, may result from cystic degeneration of malignancy
- SSx: smooth, round mass
- Dx: FNA, ultrasound, CT
- Rx: may observe for regression, fine needle aspirate relieves pain and acquires cells for cytology, may consider lobectomy for recurrent cysts that are recalcitrant to drainage or bloody aspirates

Euthyroid Goiter

Diffuse Colloid Goiter (Adenomatous Goiter, Multinodular Colloid Goiter)

- more common in women
- Pathophysiology: iodine deficiency → TSH hypersecretion → stimulates chronic thyroid hyperplasia and involution → multinodularity
- **Toxic Nodular Goiter**: variation of diffuse colloid goiter in which one nodule is hyperfunctional resulting in hyperthyroidism
- Types
  1. **Endemic Goiter**: iodine deficiency, extrinsic goitrogens (soybeans, lithium, iodides, etc.)
  2. **Sporadic**: uncertain etiology

- SSx: multiple nodules of varying size, may present with compressive symptoms (stridor, dysphagia)
- Dx: FNA of a prominent nodule, TFT to confirm euthyroid state (may be hypothyroid), consider radionuclear scanning to evaluate functional status
- Rx: iodine replacement (reverses goiter), hormonal suppression (*controversial*), radioactive iodine therapy or surgical excision (subtotal thyroidectomy) may be considered for cosmesis, decompression, concern of malignancy, or toxicosis

Mediastinal Goiter

- Pathophysiology: intrathoracic extension of thyroid gland (inferior extension is the path of least resistance)
- SSx: dyspnea, stridor, dysphagia, choking, superior vena cava syndrome
- Dx: CT/MRI of neck and chest, radionuclear scanning
- Rx: surgical excision, typically accessed from collar incision although a sternotomy may be required

Hyperthyroid Disease

Symptoms

- Elevated Metabolic Rate: weight loss, fatigue, sweating, heat intolerance
- Increased Sympathetic Activity: palpitations, tachycardia, tremor
- Increased Protein Degradation: weakness, fine hair
- Neurologic Effects: increased deep tendon reflex, nervousness
- Reproductive Effects: abnormal menstrual cycle, decreased libido

Most Common Causes

- Graves' Disease
- Multinodular Toxic Goiter
- Subacute Thyroiditis
- Exogenous from medications, iodine induced
- Uninodular Toxic Goiter
- Thyroid Cancer
- Pituitary tumors

Graves' Disease

- Pathophysiology: thyroid receptor autoantibody (IgG) → stimulates glandular hyperplasia via TSH receptor → goiter and increased T_3 and T_4 secretion

- Risks: radiation exposure, **women** (adolescence or 30-40), genetic disposition
- Histopathology: hyperplasia, increased colloid material, papillary projections
- SSx: diffuse goiter, hyperthyroid symptoms (*see above*), infiltrate dermopathy, **exophthalmos** (autoimmune → extraocular muscle deposition), blindness from optic neuropathy, pre-tibial myxedema
- Dx: thyroid-stimulating autoantibodies; elevated T_3, T_4, RAIU, and thyroglobulins; decreased serum TSH; radioactive iodine scan reveals diffuse uptake

Treatment

- Propylthiouracil and Methimazole: (*see above*) may be considered for small goiter and moderate disease
- Propanolol: (*see above*) may be supplemented for severe symptoms
- Radioactive Iodine (^{131}I): indicated for more severe symptoms or failed medical therapy; there is risk of hypothyroidism and is contraindicated in pregnancy
- Subtotal Thyroidectomy: indicated for failed medical therapy, pregnancy (especially in second trimester), noncompliance, suspicious nonfunctioning ("cold") nodule, compressive symptoms

Management for Exophthalmos and Optic Neuropathy

- Ophthalmology evaluation
- artificial tears, taping retracted lids, protective eyewear
- if optic neuropathy persists despite medical therapy, a trial of corticosteroids is used for 2 weeks, if no improvement then surgical decompression is performed
- if exophthalmos persists after 6 months despite adequate therapy, radiation therapy or surgical correction (orbital decompression, eyelid retraction release, and strabismus surgery) is performed

Hypothyroid Disease

Symptoms

- Reduced Metabolic Rate: weight gain, cold intolerance, lethargy
- Decreased Sympathetic Activity: bradycardia, constipation
- Decreased Protein Degradation: weakness, fine hair, hoarseness
- Decreased Neurologic Response: slowed deep tendon reflex, depression
- **Myxedema**: nonpitting edema
- **Creatinism**: hypothyroidism in children; mental retardation, impaired physical growth, macroglossia, protruberant abdomen

- **Myxedema Coma**: severe hypothyroidism; hypothermia, hypoglycemia, hypoventilation, ileus, death; Rx: parenteral levothyroxine, corticosteroids

Causes

- Radiation Therapy
- Chronic Thyroiditis
- Idiopathic Atrophy
- Hashimoto's Thyroiditis
- Surgical or Radioiodine Ablation Therapies
- Secondary Hypothyroidism (pituitary or hypothalamic dysfunction)
- Iodine Deficiency
- **Congenital Hypothyroidism**: from maternal iodine expectorants, anti-thyroid medications, and anti-thyroid antibodies

Thyroiditis (see Table 4–2)

Subacute Thyroiditis (de Quervain's)

- Pathophysiology: viral (mumps, Coxsackivirus) → decreased iodine uptake (unlike Grave's)
- SSx: **painful enlarged thyroid**, self limiting, malaise, associated upper respiratory infection
- Dx: clinical history, TFT (may be transient hyperthyroid)
- Rx: symptomatic therapy, observation

Hashimoto's (Chronic Autoimmune Thyroiditis)

- associated with lymphoma, neoplasms, other autoimmune disease
- Pathophysiology: **anti-thyroglobulin & anti-microsomal Ab** → anti-TSH receptor → transient hyperthyroid then hypothyroidism
- Risks: women, genetic susceptibility (HLA-DR3), Sjögren's, DM, pernicious anemia
- Histopathology: fibrosis, lymphocytic infiltration
- SSx: slowly enlarging goiter, painless
- Dx: antimicrosomal antibodies, ESR, TFT (may have elevated, normal, or low serum levels of T_4 and TSH), FNA only for prominent nodules suspicious of carcinoma or lymphoma that do not resolve with medical therapy)
- Rx: long term thyroxine therapy with TFT monitoring; surgical excision for compressive symptoms, suspicious nonfunctioning ("cold") nodule, and pregnancy

TABLE 4–2. Clinical Differences Between Thyroiditis Conditions

Condition	Subacute	Hashimoto's	Riedel's	Suppurative
Incidence	common	common	rare	rare
Thyroid hormone status	hyper- then hypothyroid	hyper- then hypothyroid	hypothyroid	—
Onset	acute	gradual	gradual	rapid
Pain	common	none	rare	common
Goiter	rare	common	hard gland	rare

Other Thyroiditis

- **Painless Thyroiditis**: like subacute (self-limiting hyperthyroid) but **painless**, common in women postpartum
- **Riedel's Thyroiditis**: thyroid fibrosis of unknown origin, "rock hard" thyroid, produces local pressure and hypothyroidism; Rx: hormone replacement, may consider surgical release at isthmus
- **Acute Supporative Thyroiditis**: uncommon; Rx: systemic antibiotics, consider drainage for abscess formation

Thyroid Neoplasia

Introduction

- High Risks (**AMES**, *Surgery*. 1988; 104:951)
  1. Age: males ≥41 years old, females ≥51 years old
  2. Metastasis: presence of metastasis suggests malignancy
  3. Extent: extrathyroidal, major capsular involvement
  4. Size: nodule ≥5 cm
  5. Other: radiation therapy, autoimmune thyroiditis, more common in women (**children and males have a higher risk of malignancy if presents with a thyroid nodule**)
- Staging: I: thyroid only; II: nonfixed cervical node; III: fixed cervical node; IV: metastasis

Adenomas

- 70% of solitary nodules
- follicular adenomas are the most common
- papillary adenomas are exceedingly rare

Papillary Carcinoma

- most common
- more common in young females
- Risks: **radiation exposure, Gardner's syndrome**
- 30% palpable regional nodes (70% occult nodes)
- Dx: FNA (*see above*)
- Histopathology: papillary and follicular structures, **psammoma bodies** (calcific), intranuclear vacuoles ("Orphan Annie" eyes), **multicentric**
- Prognosis: **95%** 5-year survival; poor prognostic indicators include tumors >1.5 cm or extracapsular spread (cervical metastases have increased cervical recurrence rates without affecting survival)

- Rx
  1. total or near total thyroidectomy (multicentric)
  2. lobectomy and isthmusectomy (hemithyroidectomy) with radioactive iodine (I^{131})may be considered if <1.5 cm or in younger patients
  3. modified neck dissection (MND) for palpable nodes only (elective neck dissection [END] has not been shown to improve survival)
  4. thyroid suppression therapy

Follicular Carcinoma

- more common in the elderly and in females
- 50% **hematogenous** spread with distant metastasis (lymphatic spread rare)
- Dx: requires open biopsy (unable to distinguish adenoma from carcinoma from FNA)
- Histopathology: **unifocal**; extracapsular spread, invasion of lymphatics or vasculature, or metastasis required for diagnosis
- Prognosis: **70–85%** 5-year survival (20% with distant metastasis), worse prognosis for angioinvasion, extracapsular spread
- Rx
  1. lobectomy and isthmusectomy (hemithyroidectomy) for low risk groups (may consider intraoperative frozen section with completion total thyroidectomy (or subtotal) for carcinoma)
  2. subtotal or total thyroidectomy with radioactive iodine (I^{131}) for high-risk groups (**angioinvasion**, extracapsular spread)
  3. lateral neck dissection for clinical nodes
  4. thyroid suppression therapy

Hürthle Cell Tumors

- variation of follicular carcinoma (Hürthle cells predominate)
- difficult to distinguish adenoma from carcinoma (assume malignancy)
- Histopathology: Hürthle cells (large granular eosinophilic cells, trabecular pattern)
- Rx: same as follicular carcinoma

Medullary Thyroid Carcinoma

- Pathophysiology: derived from parafollicular or C-cells (produce calcitonin)
- Types
  1. **Familial**: 20%, **Multiple Endocrine Neoplasia IIA/B** (*see* Table 4–3), **multicentric, bilateral**
  2. **Sporadic**: 80%, **unifocal, unilateral**, worse prognosis
- 50–60% lymph node involvement (late vascular involvement), approximately 8% distant metastasis

TABLE 4–3. Multiple Endrocrine Neoplasms

MEN I (Werner's Syndrome)	**MEN II (Sipple Syndrome)**	**MEN IIB/III**
Parathyroid hyperplasia	Medullary thyroid carcinoma	Medullary thyroid carcinoma
Pancreatic tumors (insulinomas, gastrinomas)	Pheochromocytoma	Pheochromocytoma
Pituitary adenomas	Parathyroid hyperplasia	Mucosal neuromas
		Marfanoid habitus

- secretory granules of malignant C-cells release calcitonin and also secrete gastrin, adrenocorticotropin hormone (ACTH), substance P, carcinoembryonic antigen (CEA), and others
- Prognosis: 50–80% survival; worse prognosis if unilateral, sporadic type, younger patient, or with metastasis
- Dx: FNA or biopsy, elevated serum calcitonin, elevated CEA, presence of Ret-3 oncogene
- Histopathology: small round cells, amyloid stroma, may have calcification and fibrotic strands
- Rx
  1. patient and family should be screened for MEN syndromes (*see* Table 4–3), examine for mucosal neuromas, marfanoid habitus, serum calcium levels, urine catecholamines and metabolites (vanillylmandelic acid, metanephrine)
  2. total thyroidectomy with elective or modified neck dissection and medical thyroid suppression therapy
  3. screen with calcitonin levels to monitor for recurrence

NOTE: radioactive iodine (I^{131}) is not effective because parafollicular cells do not take up I^{131}

Anaplastic Carcinoma

- most commonly seen in the elderly
- Pathophysiology: may be from transformation of a well-differentiated carcinoma (may find coexistent follicular or papillary carcinoma)
- uniformly fatal (5-year survival), often found with metastatic disease
- Histopathology: giant and spindle cells variation, undifferentiated "bizarre cells"
- Rx: no adequate therapy, tracheotomy (protect airway), consider radiation and chemotherapy, surgical resection may be considered (*controversial*)

Lymphoma

- associated with chronic lymphocytic thyroiditis and Hashimoto's thyroiditis
- Dx: difficult to distinguish lymphoma from chronic lymphocytic thyroiditis by FNA alone, therefore a conformational open biopsy is required along with lymphoma staging work-up (*see* Head and Neck , pp. 274–280)
- Rx: radiotherapy to neck and superior mediastinum, surgical excision considered if tumor is confined to the thyroid

Thyroidectomy

Introduction

- preferable for patient to be euthyroid at time of surgery to avoid thyroid storm (*see below*)
- may consider preoperative potassium iodine to reduce vascularity of thyroid gland

Indications

- suspicion of malignancy
- compression symptoms (airway compromise, dysphagia)
- extension into mediastinum
- cosmesis
- failed medical management for Graves' disease or hyperthyroidism
- pregnancy in Graves' disease or Hashimoto's Thyroiditis
- documented metastasis from thyroid carcinoma

Postoperative Complications

- **Hematoma**: may present as respiratory compromise from compressive effect; Rx: for respiratory distress immediately remove sutures and open wound, for large hematomas control bleeding in operating room and place suction drains; for smaller hematomas may be observed (place on antibiotics) and aspirate contents after liquification (7–10 days)
- **Vocal Fold Paralysis**: RLN or SLN injury (SLN injury more common), prevent by identifying nerve and avoiding ligation of inferior thyroid artery; Rx: if discovered interoperatively may consider primary anastomosis (*see also* pp. 114–119 for further management)
- **Transient or Permanent Hypocalcemia**: if parathyroid is removed interoperatively, cut into fragments and replace into adjacent muscle bed (sternocleidomastoid or brachioradialis muscles), prevent by identifying glands and avoid ligation of inferior thyroid arteries
- **Thoracic Duct Injury**: rare, results in chyle leak or fistula (*see* p. 239 for management

Thyroid Storm

- Etiology: surgery, trauma, childbirth, infection
- 50% mortality
- SSx: fever, profuse sweating, tachycardia, nausea, abdominal pain, tremors, restless, psychosis, coma, stupor
- Dx: clinical history and exam

- Rx:
  1. immediate administration of inorganic iodine, propylthiouracil, propanolol, and corticosteroids
  2. supportive measures (glucose-containing IV fluids, cooling blanket, supplemental oxygen, pyrogens)
  3. intensive care admission (cardiac monitoring)

Hyperparathyroidism and Hypercalcemia

Types of Hyperparathyroidism

Primary Hyperparathyroidism

- **elevated serum PTH**
- **Benign Adenoma**: most common, single adenomatous gland; Rx: *see below*
- **Parathyroid Hyperplasia**: associated with MEN I & IIA (*see* Table 4–3), familial hypocalciuric hypercalcemia (autosomal dominant, increased renal calcium absorption), familial hyperparathyroidism
- **Carcinoma of the Parathyroid Gland**: rare tumor, suspect with a palpable, gray mass, vocal fold paralysis, or severe hypercalcemia; Rx: en bloc resection including thyroid lobectomy, monitor for recurrence with serial serum calcium levels
- must differentiate hyperparathyroidism from other causes of hypercalcemia (*see* Table 4–4)

TABLE 4–4. Causes of Hypercalcemia: CHIMPANZEES Method

Calcium: exogenous
Hyperparathyroidism
Immobility
Metastasis to bone
Paget's disease
Addison's disease
Neoplasms: typically solid tumors (prostate, lung, colon, breast cancers)
Zollinger-Ellison syndrome (hypergastrinemia)
Excess: vitamin A or D, thiazides, lithium, estrogens, milk-alkali syndrome
Endocrine disorders: familial hypocalciuric hypercalcemia, hyperthyroidism, pheochromocytomas
Sarcoidosis: also other granulomatous diseases (tuberculosis and berylliosis)

Secondary Hyperparathyroidism

- compensatory parathyroid hyperplasia secondary to malfunction of other organ system
- Causes: chronic renal disease, osteogenesis imperfecta, Paget's disease, multiple myeloma, bone metastasis, pituitary adenomas

Tertiary Hyperparathyroidism

- autonomous or irrepressible PTH production (parathyroid hyperplasia from secondary parathyroidism, persistent hyperfunction despite correction)
- may have normal or low calcium

Evaluation

History and Physical

- Contributing Factors: family history of MEN disorders, radiation exposure
- "**stones, bones, groans:"** nephrolithiasis, osteitis fibrosa cystica, cholelithiasis
- Renal: polyuria, nephrolithiasis, nephrocalcinosis
- Gastrointestinal: constipation, dyspepsia, pancreatitis
- Musculoskeletal: muscle weakness, bone and joint pain (osteitis fibrosa cystica, calcific tendonitis)
- Central Nervous System: slow mentation, fatigue, depression, poor memory, psychosis
- Cardiovascular: heartblock, hypertension

Diagnosis of Hyperparathyroidism

- Serum Electrolytes: ionized calcium (should be elevated at 3 different times), magnesium (usually low), chloride (usually elevated from PTH induced bicarburia), and phosphate (usually low)
- PTH levels: immunoradiometric assay
- Plain Films: brown tumors (osteitis fibrosis cystica), loss of lamina, resorption of terminal phalanges, soft-tissue calcification, chest x-ray (granulomatous diseases or metastasis), abdominal film (renal calculi)
- Others: alkaline phosphatase (suggests bone disease), BUN/creatinine (renal function), urine calcium (elevated in primary hyperparathyroidism, low levels with familial hypocalciuric hypercalcemia), TFT, ACE levels (sarcoidosis), serum prolactin and gastrin and urine catecholamines and metabolites (evaluate for MEN syndromes)
- think "CHIMPANZEES" for other causes of hypercalcemia (*see* Table 4–4)

Localization

- **Thallium-Technetium Subtraction**: thallium uptake by thyroid and parathyroid, technetium uptake by thyroid gland only, parathyroid glands identified by computer subtraction; up to 90% accurate (less accurate for multiple gland hyperplasia, associated thyroid disease, or small glands)
- **High-resolution Ultrasound**: can not locate mediastinal, retrotracheal, retroesophageal, or small nodes
- **Selective Venous Catheterization for PTH**: reserved after exploration has failed
- **CT/MRI**: poor resolution, MRI more useful in identification of glands

Surgical Management

- surgery is the only definitive cure, avoids long-term complications of nephrocalcinosis and bone demineralizations
- parathyroidectomy is indicated for symptomatic (bone pain, pathological fractures, ectopic calcifications, intractable itching, etc) or persistently elevated serum calcium
- Surgical Theory: must first remove pathologic gland (adenoma), hyperplasia versus adenoma can not be distinguished grossly, therefore, must identify one "normal" gland to evaluate hyperplasia, if other gland is also hyperplastic then assume parathyroid hyperplasia and perform a subtotal (3½ glands) or total parathyroidectomy with autotransplantation, if gland is normal may either assume adenoma and terminate case or further biopsy other contralateral glands *(controversial)*
- autotransplantation requires 20 mg into muscle bed (usually to forearm)
- pregnant women should undergo surgery during second trimester to avoid miscarriage

Complications

- **Persistent Hypercalcemia**: most commonly from missed adenoma (the most commonly missed location is the posterior mediastinum), also from supernumerary gland, second adenoma, failed recognition of parathyroid hyperplasia, incorrect diagnosis, and residual adenoma
- **Postoperative Hypocalcemia and Hypomagnesmia**: usually temporary until replacement of low bone calcium stores (increased risk with elevated alkaline phosphatase)
- **Nerve Injury and Hematomas**: *see* above

Medical Management of Hypercalcemia

- **Saline Diuresis**: restores extracellular fluid volume and promotes calcium excretion, loop diuretics can also be given (thiazides impair calcium excretion)

- **Biphosphonates**: inhibit bone resorption, calcium serum levels reduce over several days
- **Plicamycin**: inhibits bone resorption; toxic side effects include thrombocytopenia, hepatic dysfunction, and renal failure, therefore used only for malignant hypercalcemia
- **Calcitonin**: rapid onset (serum calcium falls within hours)
- **Glucocorticoids**: inhibit calcium intestinal absorption, may be effective for hypercalcemia secondary to malignancy
- **Gallium Nitrate**: inhibits bone resorption, used for parathyroid carcinoma
- **Hemodialysis**: indicated for life-threatening conditions

ESOPHAGEAL AND SWALLOWING DISORDERS

Swallowing Phases

Preparatory and Oral Phase

- voluntary phase
  1. solid mastication (lip closure)
  2. saliva mixes with food bolus; tongue and facial muscles prevent bolus from falling into lateral sulci
  3. food bolus molded by tongue and teeth (**ends Preparatory Phase**) and forced to the dorsum of the tongue
  4. anterior tongue and base of tongue elevate to contact palate, posterior pharyngeal wall, and floor of mouth; hyoid bone slowly elevates
  5. food bolus propelled into oropharynx (up to vallecula)

Pharyngeal Phase

- reflexive phase (posterior pharyngeal wall receptors, CN IX and CN X)
- transient time <1 sec in normal subjects
- includes fast anterior, superior motion of the hyoid bone
  1. **Nasopharynx (Velopharyngeous) Closure**
     - levator veli palatini muscle lifts the soft palate
     - palatopharyngeous (posterior pillar) muscle tightens and raises the pharynx and narrows the oropharyngeal inlet
     - superior pharyngeal constrictor (Passavant's pad) contracts to meet the soft palate and posterior pharyngeal walls
  2. **Base of Tongue Propels Bolus Past Vallecula**
     - base of tongue squeezes against posterior pharynx
     - glossectomy patients have difficulty with bolus propulsion
  3. **Laryngeal Elevation and Closure**
     - larynx elevates approximately 2 cm

- Three Sphincters for Laryngeal Closure
  1. **Laryngeal Inlet**: epiglottis, aryepiglottic folds, arytenoids
  2. **False Vocal Fold**
  3. **True Vocal Fold**: most effective
- **glottic closure reflex**: mediated principally by the superior laryngeal nerve, reflex loop involves nucleus ambiguus, results in closure of true vocal folds (excess stimulation causes **laryngospasm**)

4. **Pharynx Shortens**
   - pharynx shortens by approximately 2 cm
   - pharyngeal constrictors push bolus while epiglottis directs food to pyriform sinus → posterior pharyngeal wall → cricopharyngeal sphincter (pharyngeal weakness pools saliva and food bolus in pyriform sinus)
5. **Cricopharyngeous Sphincter (Upper Esophageal Sphincter) Opens**
   - relaxation of the muscle and anterior displacement of the hypolaryngeal complex opens the sphincter
   - food bolus enters the cervical esophagus

Esophageal Phase

- fluid movement is passive, solid movement is active
- *see* Esophageal Anatomy and Physiology *below*

Dysphagia and Aspiration

Introduction

- Causes of Dysphagia: obstruction, misdirection (retrograde [nasopharynx] and anterograde [aspiration]), fragmentation
- oropharyngeal secretions most common aspirated substance (gastric contents is the second most common)

Evaluation of Dysphagia and Aspiration

History and Physical Exam

- Character of Dysphagia: solid dysphagia (obstructive) vs. liquid dysphagia (neurological), progressive (tumor, scleroderma, achalasia), odynophagia (suggests acute process, foreign body, pharyngitis, laryngitis), regurgitation (nasal or gastric regurgitation, timing of regurgitation), type of regurgitated food (digested or undigested), aspiration (cough after ingestion, recurrent pneumonia, gagging, choking), difficulty with mastication or oral competence (drooling)
- Contributing Factors: history of GERD; history of recent caustic or foreign body ingestion or trauma; risk factors for malignancy (recent

weight loss, family history of cancer, hoarseness, odynophagia, otalgia, tobacco, smoking, alcohol abuse); history of neurologic, connective tissue, or autoimmune disorders; recent dietary changes
- Associated Symptoms: voice changes (laryngeal involvement), heartburn (reflux esophagitis), hypothyroidism (fatigue, hair loss, depression, weight gain), neurological changes (weakness, paresthesias, diplopia, vertigo, mental status changes)
- Physical Exam: full neurologic exam (cranial nerves and peripheral exam), indirect laryngeal exam or nasopharyngeal scope (pooling of secretion, vocal fold mobility, masses), complete head and neck exam with high suspicion for malignancy (palpation of base of tongue, indirect mirror exam for lesions, evaluate nasopharynx), thyroid palpation
- Think "KITTENS" for differential diagnosis (*see* Table 4–5)

Modified Barium Swallow (MBS)

- **MBS**: radiographic videofluoroscopic study that visualizes oral and pharyngeal phases of swallowing
- typically reviewed with a swallowing therapist
- utilizes varying bolus amounts and consistencies
- determines oral and pharyngeal motility, laryngotracheal elevation, laryngeal penetration or aspiration, and safety of oral feeding

Other Ancillary Tests

- **Esophagram** (**Barium Swallow**): evaluates esophageal phase of swallowing (motility), luminal integrity (large ulcers, intrinsic/extrinsic masses, strictures, webs), reflux
- **Manometry**: measures duration, amplitude, and velocity of peristaltic waves
- **Chest X-ray**: reveals pneumonitis, pneumonia, masses, or a displaced airway
- **Laryngoscopy and Esophagoscopy**: indicated if suspect malignancy, for uncertain etiology, to evaluate esophagus, to remove foreign bodies, and to biopsy a mass or lesion
- **Functional (Fiberoptic) Endoscopic Evaluation of Swallowing** (**FEES**): allows bedside evaluation of swallowing function, nasopharyngoscopy utilized to visualize swallowing phases, may assess for aspiration, penetration, and pharyngeal and laryngeal function
- **Dyed Food**: indirect evaluation of aspiration in the presence of a tracheotomy, oral feeds are dyed (methylene blue), tracheotomy is suctioned to evaluate for the presence of stained aspirates
- **GERD Evaluation**: (*see* GERD *below*)
- **Videostroboscopy**: evaluates vocal fold motion, pooled secretions, and anatomical defects (masses, glottal chinks, etc)

TABLE 4–5. Differential Diagnosis of Dysphagia and Aspiration: KITTENS Method

(K) Congenital	Infectious & Iatrogenic	Toxins & Trauma	Tumor (Neoplasia)	Endocrine	Neurologic	Systemic
Tracheoesophageal fistulas	Laryngitis	Caustic ingestion	CNS tumors	Hypothyroiditis	Altered mental status (alcohol, sedatives, head injury)	Vocal fold paralysis
Dysphagia lusoria	Pharyngitis	Foreign body ingestion	Esophageal tumors		Degenerative diseases (Parkinson's disease, multiple sclerosis)	Gastrointestinal disorders (Zenker's diverticulum, GERD, achalasia, esophageal diverticulum, cricopharyngeal spasm, Plummer-Vinson syndrome, etc)
Congenital esophageal webs	Esophagitis	Mallory-Weis syndrome	Extrinsic compression of esophagus		Motor neuron disease (Amyotrophic lateral sclerosis)	Myopathies (muscular dystrophy, metabolic myopathies, polymyositis)
Cleft palate	Chagas' disease				Stroke	Connective tissue disorders (progressive systemic sclerosis)
	Tracheotomy, endotracheal intubation				Encephalopathies	Globus hystericus
	Postsurgical head and neck resection				Guillain-Barré	
					Myasthenia gravis	
					Bulbar and pseudobulbar palsy	
					Dementia	

- **Manofluorography**: quantitative analysis of pressure generation at tongue, palate, larynx, and pharyngeal walls (labor intensive, utilized primarily for research)
- **CT/MRI**: may be considered to evaluate masses

Management

Medical Management

- if possible address underlying cause (eg, iron supplementation for Plummer-Vinson, pyridostigmine for myasthenia gravis, benztropine for Parkinson's disease, antibiotics for acute bacterial pharyngitis)
- utilize an alternative temporary route of nutrition (nasogastric tube feeds, parenteral nutrition)
- begin a reflux regimen (*see* GERD, *below*)
- aggressively address aspiration pneumonia (hold oral feeds, antibiotic regimen, and aggressive pulmonary toilet)
- **Botulinum Toxin Injections**: may be considered for cricopharyngeal spasms, inject toxin into cricopharyngeus muscle

Swallowing Rehabilitation

- change food consistencies (pureed diet easier to tolerate initially, liquids are more difficult to manage)
- posture techniques (chin tuck, head turn to the poorer functioning side), palatal prostheses, muscle strengthening exercises
- **Supraglottic Swallow**: patient voluntarily closes airway at vocal folds by holding breath before swallow, voluntary cough after swallow, follow with an additional swallow for residual bolus in pharynx or pyriform
- **Mendelsohn Maneuver**: voluntarily elevates and anteriorly displaces larynx to prolong upper esophageal sphincter opening

Surgical Management

- **Esophageal Dilation**: may be considered for achalasia (distal LES spasm), and pharyngeal or esophageal strictures, webs, postoperative scarring, and postradiation strictures
- **Cricopharyngeal Myotomy**: may be considered for cricopharyngeal spasms (incomplete UES relaxation) or abnormal muscular contraction during relaxation (*controversial*), theoretically relaxes pharyngoesophageal segment and results in anterior elevation of larynx; complete myotomy includes part of the lower inferior constrictor, cricopharyngeus muscle, and part of the upper cervical esophagus
- **Gastric or Jejunal Feeding Tube**: temporary or permanent enteric feeding
- **Vocal Fold Medialization**: for unilateral vocal fold paralysis, *see* pp. 116–117

- **Tracheotomy** (cuffed): indicated for severe pulmonary complications, prevents aspiration pneumonia by allowing easier pulmonary toilet and preventing gross aspiration (does not prevent microaspiration); however, increases risk of aspiration by inhibiting laryngeal elevation, interfering with ciliary motion, and preventing production of subglottic pressure for an adequate cough
- **Laryngeal Closure Techniques**: requires a permanent tracheostomy; glottic closure may be considered for bilateral vocal fold paralysis; epiglottoplexy allows voicing, improves aspiration, and is potentially reversible
- **Laryngeal Suspension**: indicated for severe aspiration from supraglottic and pharyngeal dysfunction (may be considered after supraglottic resection), suspends larynx anteriorly by positioning thyroid cartilage under mandible, may improve voicing and swallowing
- **Laryngeal Diversion (Lindeman procedure) or Separation**: indicated for severe aspiration, creates a permanent tracheostomy with proximal tracheal segment diverted back to the esophagus; a variation of Lindeman's method creates a blind pouch from the proximal trachea
- **Laryngectomy**: indicated for life-threatening complications, gold standard for definitive therapy
- **Bilateral Submandibular Excision and Parotid Duct Ligation**: voice sparing, lower morbidity than laryngotracheal separation, may be considered in neurologically impaired patients who aspirate saliva, minor salivary gland production of saliva prevents xerostomia

Esophageal Anatomy and Physiology

Anatomy

- Esophageal Layers
  1. **Mucosa**: epithelium, lamina propria, muscularis mucosa
  2. **Inner Circular Muscle**
  3. **Submucosa**
  4. **Outer Longitudinal Muscle**: no serosa layer, therefore minimal barrier to infection and tumor infiltration
- Three Physiologic Narrowings
  1. **Upper Esophageal Sphincter**: *see below*
  2. **Crossing of the Aortic Arch and Left Main Bronchus**: anatomical narrowing about 27 cm from incisor opening
  3. **Lower Esophageal Sphincter**: *see below*
- Embryology: epithelial proliferation obliterates the lumen, **recanalization** occurs by 8th week (abnormal recanalization results in congenital stenosis)

- Vasculature: segmented from inferior thyroid artery (upper 1/3), thoracic aorta (middle 1/3), and left gastric and inferior phrenic arteries (lower 1/3)
- Histology: stratified squamous epithelium except distal 1–3 cm (columnar epithelium)
- Innervation: mixed somatic innervation from CN IX and CN X (left vagus nerve passes anteriorly and the right vagus nerve passes posteriorly to form the esophageal plexus)
- **Auerbach's Plexus**: myenteric plexus, between muscle layers, parasympathetic
- **Meissner's Plexus**: submucosal plexus

Upper Esophageal Sphincter (UES)

- UES is composed of the **cricopharyngeus muscle** (approximately 16 cm from incisor opening)
- tonic contracture prevents air reflux, aspiration, and regurgitation
- relaxes during the pharyngeal phase

Lower Esophageal Sphincter (LES)

- normal tone: **10–40** mm Hg (achalasia: **>40**; scleroderma **<10**)
- normally positioned below diaphragm (hiatal hernias result in LES positioned above diaphragm)
- physiological sphincter (not a true anatomic sphincter), aided by the crura of the diaphragm (vagal innervation)
- Agents that Increase LES Pressure: proteins, acid, gastrin, vasopressin, α-adrenergics
- Agents that Decrease LES Pressure: secretin, nitrates, calcium channel blockers, glucagon, chocolate, fat, β-adrenergics, alcohol, mints, nicotine

Physiology

- minimal secretion and absorption
- upper 1/3 is composed of voluntary striated muscle (1 second of esophageal phase)
- lower 2/3 is composed of involuntary smooth muscle (3 seconds of esophageal phase)

Peristalsis Types

- **Primary Peristalsis**: initiated by food bolus, contracts proximally to distally
- **Secondary Peristalsis**: initiated by esophageal distention (residual food bolus) and reflux
- **Tertiary Contractions**: nonperistaltic, spontaneous contractions, may propel bolus in a retrograde direction to proximal esophagus

Gastroesophageal Reflux Disease (GERD)

Introduction

- Pathophysiology
  1. LES incompetence or transient LES relaxation
  2. delayed esophageal clearance
  3. delayed gastric emptying (may increase gastric volume)
- Risks: obesity, alcohol abuse, hiatal hernias, pregnancy, scleroderma, retained feeding tube
- SSx: postprandial heartburn, choking spells at night, regurgitation, hoarseness (worse in the morning), nausea, bitter taste in mouth, globus hystericus, cough, chronic throat clearing
- Complications: reflux esophagitis, Barrett's esophagus (gastric metaplasia of the distal esophagus), esophageal strictures, gastric and esophageal ulcerations, globus pharyngitis, chronic cough, aspiration pneumonia, laryngeal granulomas, laryngopharyngeal acid reflux, failure to thrive, sudden infant death syndrome (SIDS), possibly laryngeal carcinoma

Diagnosis

- **History**: may begin empiric reflux regimen based on history, consider ancillary testing after failed empirical management, atypical symptoms, recurrent symptoms, or complications (weight loss, dysphagia)
- **Nasopharyngoscopy**: reveals erythema and edema of the posterior commissure, arytenoids, superior surface of the vocal folds, and laryngeal surface of the epiglottis; diffuse supraglottic edema; laryngeal pachydermia (interarytenoid); granulomas of the vocal process
- **Barium Swallow**: good initial screening test for demonstrating esophageal anatomy, high false-negative rate for reflux
- **24 hour pH Monitor**: most sensitive, "gold standard," pH <**4.1** (5 cm above LES), 85% sensitivity
- **Esophagoscopy**: evaluates for esophagitis and esophageal strictures, allows histological confirmation of Barrett's esophagus or esophagitis
- **Gastroesophageal Scintiscan**: utilizes swallowed radiolabeled technetium, allows evaluation of gastric emptying and reflux

Management

1. **Behavior Management**
   - smoking cessation
   - elevate head of bed at night
   - avoid tight fitting clothing
   - avoid overeating, eating before sleep

- abstain from caffeine, fatty foods, alcohol, mints, chocolate, and other reflux inducing foods
- avoid aspirin, nitrates, and calcium channel blockers

2. **Medical Management**
   - **Liquid Antacids** (calcium carbonate, hydroxides of aluminum and magnesium, sodium bicarbonate): may be considered as first-line therapy, take after meals and before sleep, overuse may result in acid-base and other metabolic disturbances
   - **H_2-blockers** (cimetidine, famotidine, ranitidine, nizatidine): may be considered for uncomplicated GERD, blocks histamine interaction with its receptor; side effects include constipation, diarrhea, confusion, and elevated liver enzymes
   - **Proton Pump Inhibitors** (omeprazole, lansoprazole): highly effective, indicated as first-line agents for complicated GERD or failed first-line regimens, blocks the "proton pump" responsible for acid secretion
   - **Prokinetic Agents** (cisapride, metoclopramide): indicated for delayed gastric emptying, also increases lower esophageal sphincter pressure; side effects include tardive dyskinesia, drowsiness, depression, and confusion
   - **Sucralfate**: nonsystemic medication, protects exposed ulcerated mucosal surfaces
3. **Surgical Management**
   - indicated for failed medical regimen
   - fundoplication procedures increase tone of distal esophagus (LES)

Esophageal Disorders

Achalasia (Megaesophagus)

- Pathophysiology: degeneration of Auerbach's plexus resulting in **aperistalsis and increased LES pressure** (LES does **not** relax with bolus)
- SSx: progressive dysphagia, vomiting, malodorous breath, aspiration, weight loss
- Dx: esophagram (**bird beak** appearance, air-fluid levels, aperistalsis, failed LES relaxation), manometry (increased LES pressure), esophagoscopy to rule out masses (carcinoma) and esophagitis
- Complications: increased risk of esophageal carcinoma

Management

- "wash" meals down with fluids
- serial esophageal dilation (pneumatic dilators)
- **Heller's procedure**: esophagomyotomy with fundoplication
- may consider calcium channel blockers and nitrates to decrease LES pressure (*controversial*)

Progressive Systemic Sclerosis (Scleroderma)

- Pathophysiology: autoimmune disease causes small vessel vasculitis, widespread collagen deposition, and smooth muscle atrophy (associated with other connective tissue diseases)
- Dx: barium swallow (dilated, flaccid similar to achalasia, however, **patent LES**), manometry (normal UES pressure, loss of tone of LES)
- **CREST Syndrome**: variant, milder form, **C**alcinosis (cutaneous), **R**aynaud's phenomenon, **E**sophageal dysmobility, **S**clerodactyly, **T**elangiectasis
- Rx: reflux regimen, calcium channel blockers for Raynaud's phenomenon, consider corticosteroids and NSAIDs

Symptoms

- H&N SSx: dysphagia from esophageal dysmotility of the lower two thirds (smooth muscle), GERD, fixed face appearance (tight skin, thin lips, sclerotic skin)
- Cutaneous Involvement: initial edematous skin, later tight skin
- Visceral Involvement: pulmonary hypertension and fibrosis, pericarditis, microcytic anemia (hypertensive renal crisis)
- Other SSx: carpal tunnel syndrome, Raynaud's phenomenon, sclerodactaly, telangiectasis

Polymyositis (Dermatomyositis)

- Pathophysiology: idiopathic inflammatory myopathy of striated muscle
- **Dermatomyositis**: variation with associated rash, higher risk of malignancy
- associated with other connective tissue diseases, **malignancies**, hiatal hernias, reflux esophagitis, vasculitis
- SSx: proximal muscle (hip and shoulder) weakness and wasting, dysphagia, aspiration, dysmobility in upper 1/3 of the esophagus and pharyngeal weakness (**striated muscle**), nasal regurgitation, dysrhythmias
- Dx: EMG; muscle biopsy; increased serum creatine phosphokinase, liver enzymes, LDH
- Rx: antireflux regimen, corticosteroids, antimetabolites, immunosuppressives

Esophageal Diverticulum

- Pathophysiology: pouch created by herniation of mucosa through mucosal wall
- Upper Esophageal/Hypopharyngeal Areas of Weakness
  1. **Killian's Triangle**: inferior to posterior cricopharyngeal muscle, superior to cricothyroid muscles, below raphae of inferior constrictors

2. **Killian-Jamieson Space**: laterally, between cricopharyngeal and esophagus muscle
3. **Laimer-Haeckermann Space**: between cricopharyngeus superiorly and circular fibers inferiorly

Zenker's Diverticulum (Pharyngoesophageal Diverticulum)

- Complications: diverticulitis, fistula formation, perforation, bleeding
- Pathophysiology: pulsion diverticulum (created from elevated intraluminal pressure) creates a herniation typically at Killian's triangle (*see above*)
- **False Diverticulum**: diverticulum contains mucosa and submucosa only
- SSx: insidious dysphagia, spontaneous regurgitation of undigested food, malodorous breath, aspiration, may become obstructive
- Dx: esophagram and endoscopy (80–90% located on the left side)
- Rx: may observe or complete a cricopharyngeal myotomy for small defect and minimal symptoms; symptomatic defects and large diverticuli (>1.5 cm) require a transcervical diverticulectomy with cricopharyngeal myotomy (must complete a cricopharyngeal myotomy to prevent recurrence), diverticulopexy (theoretically reduces risk of fistula), or an endoscopic myotomy; may also consider botulinum toxin injection into cricopharyngeal muscle (*controversial*)

Traction Diverticulum

- Pathophysiology: "tugging" effect from inflammation (lymph nodes) and fibrotic adjacent tissue
- **True Diverticulum**: diverticulum contains all layers of esophageal wall (including muscularis)
- typically in middle third of esophagus
- Dx: esophagram and esophagoscopy (high risk of perforation)
- SSx: dysphagia, usually asymptomatic
- Rx: if symptomatic or complicated may undergo right thoracotomy with diverticulectomy

Epiphrenic Diverticulum

- Pathophysiology: pulsion diverticulum created above cardioesophageal junction
- associated with GERD, hiatal hernia, esophageal spasm, esophageal carcinoma, and other esophageal disease
- SSx: dysphagia, regurgitation, obstruction
- Dx: esophagram and esophagoscopy
- Rx: if symptomatic or complicated may consider diverticulectomy, must address underlying cause

Cricopharyngeal Dysfunction (Cricopharyngeal Spasm, Cricopharyngeal Dysphagia)

- Pathophysiology: abnormal coordination between pharynx and cricopharyngeus (UES)
- Types
  1. **Idiopathic**: may be secondary to subclinical reflux
  2. **Neurologic**: secondary to neurologic process
  3. **Reflux Induced**: most common
- SSx: localized dysphagia to the cervical esophagus, globus sensation, choking
- Dx: esophagram may be normal or reveal cricopharyngeal bar with functional obstruction, manometry more accurate
- Complications: diverticular formation, aspiration, pulmonary disease
- Rx: cricopharyngeal myotomy (open or endoscopic), **botulinum toxin injections**, reflux regimen

Esophagitis

- **Reflux Esophagitis**: most common, secondary to gastric acid reflux, causes mucosal erosion of distal esophagus
- **Candidal Esophagitis**: white plaques with erythematous base, associated with odynophagia, increased risk with immunocompromised patients and long-term antibiotics; Rx: systemic antifungals
- **Herpes Esophagitis**: multiple ulcerations, increased risk with immunocompromised patients
- **Drug Induced Esophagitis**: typically punctate ulcerations; common examples include tetracycline, potassium, quinidine, aspirin, and clindamycin; Rx: prevent by taking with sufficient fluid
- **Bullous Dermatoses**: pemphigoid, epidermolysis, and others
- **Radiation Esophagitis**; may be acute (during therapy) or chronic (scarring and stenosis may occur from 6–18 months after radiation therapy

Esophageal Rupture and Perforation

- Causes: iatrogenic instrumentation (most common cause), blunt and penetrating trauma, neoplasms, inflammation, increased abdominal pressure
- Variants
  1. **Mallory Weiss syndrome**: incomplete tear of esophageal mucosa and laceration of submucosal arteries from increased abdominal pressure (emesis in alcoholics), presents as an upper gastrointestinal bleed; Rx:

usually self limiting, may decompress (nasogastric tube), rarely requires endoscopic coagulation or open procedures

2. **Boerhaave Syndrome**: increased abdominal pressure results in spontaneous rupture of all 3 layers of the esophagus (usually distal, posterior wall), severe symptoms (bloodstained vomiting, chest pain, dyspnea, hypovolemic shock)

- SSx: chest pain, tachycardia, fever, respiratory distress, dysphagia, subcutaneous emphysema, **Hammer's sign** (crunching sound over heart from subcutaneous emphysema)
- Dx: clinical exam, chest x-ray (mediastinal widening, pneumothorax), esophagram (gastrografin)
- Complications: chemical mediastinitis (saliva, bile, gastric acid), septic shock
- Rx: early surgical repair and drainage (thoracotomy), may consider medical therapy (antibiotics and observation) for small perforation in select patients

Esophageal Foreign Bodies and Caustic Ingestion

(*see* pp. 481–484)

Diffuse Esophageal Spasm

- Pathophysiology: nonperistaltic contraction in esophageal smooth muscle
- SSx: odynophagia to solids and liquids, dysphagia, chest pain
- Dx: esophagram (corkscrew, normal LES relaxation), manometry
- Complications: diverticuli
- Rx: nitrates, calcium channel blockers, anticholinergics, may consider dilation and myotomy for severe symptoms

Dysphagia Lusoria (Bayford Syndrome)

- Pathophysiology: anomalous **right subclavian artery** from **descending** aorta (fourth branchial arch anomaly) has a retroesophageal course causing dysphagia from extrinsic compression of the esophagus
- associated with a **nonrecurrent** right recurrent nerve, aortic and subclavian aneurysms, and diverticuli
- SSx: intermittent dysphagia (usually presents at middle age with loss of elasticity of vessels), weight loss
- Dx: barium swallow, arteriogram, esophagoscopy (pulsating horizontal bar at obstruction site)
- Rx: ligate and reanastomose to right common carotid for severe symptoms

Eagle Syndrome (Styloid Process Syndrome)

- Pathophysiology: elongated styloid process or ossified stylohyoid ligament causes mechanical irritation of surrounding nerves
- SSx: odynophagia, unilateral tonsilar pain, pain behind mandibular angle, referred otalgia, palpation at site reproduces pain
- Dx: radiography
- Rx: excision from intraoral or external approach

Plummer-Vinson Syndrome

- Pathophysiology: unclear etiology, may be secondary to nutritional deficiency (iron)
- more common in young to middle-aged women
- Risks: northern hemisphere (Scandinavians)
- SSx: dysphagia (degeneration of esophageal muscle), **microcytic hypochromatic anemia** (iron deficiency), cervical (pharyngoesophageal) webs, chelitis (fissures at corners of lips), hypothyroidism, hiatal hernia, splenomegaly, achlorhydria
- Dx: clinical exam, iron levels, CBC, esophagram
- Complications: increased risk of upper esophageal and hypopharyngeal carcinoma
- Rx: iron supplements (improves most symptoms), esophageal dilation

Esophageal Webs and Rings

- **Web**: asymmetric, thin, membranous projection into the lumen, covered by esophageal epithelium (composed of mucosa and submucosa only)
- **Ring**: thicker, composed of mucosa, submucosa, and muscularis
- **Schatzki Rings**: web-like narrowing at **squamocollumnar junction** (junction of esophageal and gastric mucosa), only $^1/_3$ are symptomatic
- upper cervical webs may be associated with Plummer-Vinson syndrome
- SSx: usually asymptomatic, may cause dysphagia, weight loss
- Dx: esophagrams or esophagoscopy
- Rx: surgical intervention rare, dilation, endoscopic excision (laser)

Tracheoesophageal Fistulas

- Pathophysiology
  1. **Congenital**: failure of recannulation of the esophagus or developmental failure of the tracheoesophageal septum
  2. **Acquired**: secondary to tracheostomies, long-term intubation, tumor, inflammation, trauma results in communication between lumen of the esophagus and the trachea

- Types
  1. **Esophageal Atresia with Distal Tracheoesophageal Fistula**: most common form (80–90%)
  2. **Isolated Esophageal Atresia**: second most common form, associated with polyhydramnios
  3. **Isolated Tracheoesophageal Fistula** (**H-type**): may remain asymptomatic until later in life
  4. **Complete Esophageal Stenosis**: failure of recannulization
- associated with the **VATER complex** (**V**ertebral, **A**nal, **T**racheal-**E**sophageal, **R**adial limb or **R**enal defects)
- SSx: immediate gagging and cyanosis after birth, gas filled stomach; may present later in life with cough, recurrent aspiration pneumonia, gagging
- Dx: endoscopy, contrast radiography
- Rx: division and reconstruction of the trachea and esophagus

Other Systemic Diseases Effects on the Esophagus

- **Presbyesophagus**: reduced peristalsis and decreased LES pressure seen in the elderly, probably neuropathic
- **Chagas' Disease**: parasitic infection, destroys Auerbach's plexus, results in achalasia-like symptomatology
- **Diffuse Idiopathic Skeletal Hyperostosis** (**DISH**) (**Forestier's Disease**): paraspinous ligament calcification causes dysphagia from cervical osteophytes compression or periesophageal soft tissue inflammation; Dx: lateral neck film, esophagram; Rx: surgical reduction for severe symptoms

Esophageal Neoplasms

Benign Tumors and Cysts

- **less** common than malignant tumors
- may be intraluminal, intramural, or periesophageal
- SSx: dysphagia, pressure behind the sternum, bleeding, weight loss
- Dx: barium swallow (highly sensitive), endoscopy (biopsy), CT/MRI, chest x-ray (displacement of structures)
- Rx: typically requires endoscopic or open excision

Types

- **Leiomyoma**: most common benign tumor of the esophagus, intraluminal, arises from muscularis (usually distal 2/3); Rx: excision of symptomatic lesions

- **Polyps**: most common intraluminal lesion, most common in cervical esophagus; Rx: endoscopic removal
- **Others**: Myomas, Fibromas, Neurofibromas, Lipomas, Adenomas, Papillomas, Hemangiomas

Malignant Tumors

- Risks: elderly black males, tobacco and alcohol abuse, regional (China, Iran, Russia), caustic burns, radiation, petroleum, nitrates, esophageal webs, Plummer-Vinson disease, achalasia, pernicious anemia, nutritional deficiencies, esophagitis, tylosis (autosomal dominant disease characterized by thickened palms and soles), GERD
- SSx: initial painless dysphagia develops later into odynophagia, hemoptysis, cough, hoarseness, weight loss, emesis
- Dx: barium swallow, endoscopy (biopsy), CT/MRI (evaluate extent, invasion, displacement of structures, regional metastasis), endoscopic ultrasonography (may determine depth of invasion)

Types

- **Squamous-cell Carcinoma**: most common malignancy in the esophagus, middle third most common region
- **Adenocarcinoma**: associated with Barrett's esophagus
- **Others**: Adenoid Cystic Carcinoma, Mucoepidermoid Carcinoma, Small-cell Carcinoma, Sarcomas

Management

- absence of a serosa layer allows early transmural spread, overall a poor prognosis (10% 3-year survival)
- involvement of the prevertebral fascia, trachea, carotids or metastatic disease should be considered incurable
- high-grade Barrett's esophagus should undergo prophylactic excision
- Rx: surgical resection with adjunctive chemoradiation, may consider palliation chemotherapy and radiation
- Reconstruction Methods
  1. **Pectoralis/Deltopectorial Flaps**: may be tubed, bulky, poor speech, high fistula and stenosis rate
  2. **Colonic Interposition**: one-stage procedure, high infection rate
  3. **Gastric Pull-Up**: reliable, one-stage procedure, vagotomy and pyloroplasty required, single anastomosis (*see also* p. 248 for complications)
  4. **Jejunal Free Flap**: laryngeal preservation and replacement of cervical esophagus only (*see also* p. 394)

PHARYNGEAL AND ADENOTONSILLAR DISORDERS

Anatomy

Tonsils

- **Waldeyer's Ring**: circle of lymphoid tissue consisting of palatine (fauceal) tonsils, pharyngeal tonsils (adenoids), lingual tonsils, and **tubal tonsils of Gerlach** (near fossa of Rosenmüller)
- tonsils increase in size between 1–3 years old (after exposure to antigens), peak between 3–7 years old, involute after puberty
- Arterial Supply
  1. lingual artery → dorsal lingual branch (lower pole)
  2. facial artery → ascending palatine artery and tonsillar artery (main supply, lower pole)
  3. external carotid artery → ascending pharyngeal artery (upper pole)
  4. internal maxillary artery → greater palatine artery and descending palatine artery (upper pole)
- Venous Drainage: lingual and pharyngeal veins → internal jugular vein
- Lymphatics: **no afferent lymphatics**, drainage into superior deep cervical and jugular digastric lymph nodes
- Histology/Tonsillar Zones
  1. **Reticular Cell Epithelium**: squamous layer, contain antigen-presenting cells (M-cells) which transport antigen to the lymphoid germinal center
  2. **Extrafollicular Area**: contain T-cells
  3. **Lymphoid Follicle**: composed of the **Mantle Zone** (mature B-cells) and the **Germinal Center** (active B-cells)

Adenoids

- similar to palatine tonsils, present at birth, enlarge in childhood, and regress during puberty
- Arterial Supply: ascending pharyngeal artery from the external carotid artery, minor branches from internal maxillary artery (pharyngeal branch) and facial arteries (ascending palatine artery)
- Venous Drainage: pharyngeal veins → facial and internal jugular vein
- Histology: ciliated pseudostratified columnar, stratified squamous, and transitional layers

Pharyngitis

Acute Pharyngitis

- Pathogenesis: may be secondary to sinonasal disease, caustic injury, chronic allergy

- Pathogens: typically viral; most common types are adenovirus, rhinovirus, and enterovirus (less common are mumps, herpes simplex, and rubella); secondary bacterial infections are less common (streptococci, pneumococci, *H. influenza*)
- SSx: sore throat, odynophagia, otalgia (referred), malaise, fever, erythema, cervical adenopathy
- Dx: clinical exam, consider throat cultures, viral smears rarely indicated
- Rx: supportive care (bed rest, hydration, humidity, lozenges, anesthetic sprays [cetacaine or xylocaine], iodine glyceride solutions, antipyretics, decongestants), antibiotics for suspected bacterial infections

Other Causes of Infectious Pharyngitis

- **Syphilis** (*Treponema pallidum*): spirochete infection, may present as a form of secondary syphilis 2–3 months after primary infection, presents as dark-red papules on the tonsils or pharyngeal wall; Rx: penicillin, tetracycline, erythromycin
- **Pertussis** (*Bordetella pertussis*): gram-negative coccobacillus associated with paroxysmal coughing with loud inspiratory sound ("whooping" cough), effects tracheobronchial tree and pharyngeal and laryngeal mucosa, progresses in 3 stages (catarrhal, paroxysmal, convalescent); Rx: supportive care, typically self limiting, immunization (preventative)
- **Gonorrhea** (*Neisseria gonorrhea*): gram-negative diplococci transmitted by sexual contact, typically asymptomatic although may present as a sore throat, culture on chocolate agar stain for identification; Rx: penicillin, tetracycline, trimethroprim-sulfamethoxazole
- **Candidiasis**: *see* p. 174
- **Infectious Mononucleosis**: *see below*
- **Herpangina**: *see below*
- **Diphtheria**: *see* pp. 102–103
- **Scarlet Fever**: *see below*

Chronic Pharyngitis

- Pathogenesis: postnasal drip (chronic rhinosinusitis), irritants (dust, dry heat, chemicals, smoking, alcohol), reflux esophagitis, chronic mouth breathing (adenoid hypertrophy), voice abuse, allergy, granulomatous diseases and connective tissue disorders, malignancies
- SSx: constant throat clearing, dry throat, odynophagia, thickened and granular pharyngeal wall, pharyngeal crusting
- Dx: clinical history and exam, culture and biopsy if failed empiric therapies
- Rx: address underlying etiology, avoidance of contributing factors (smoking, dust, dry environments, etc), symptomatic treatment similar to acute pharyngitis

Adenotonsillar Pathology

Tonsillitis

- Pathophysiology: often a viral infection occurs prior to a bacterial superinfection
- Pathogens: most commonly Group A β-hemolytic *Streptococcus* (GABHS), *Moraxella*, and *H. influenzae*; other less common organisms include *Bacteroides, Staphylococci, E. coli* , diphtheria, syphilis, and viral etiologies (Epstein-Barr, Adenovirus, Influenza A and B)
- SSx: sore throat, fever, chills, odynophagia, trismus, cervical adenopathy, halitosis, erythema of tonsils and faucial pillars
- Phases of Tonsillitis: tonsillar erythema, exudative tonsillitis, follicular tonsillitis (yellow spots corresponding to lymphatic follicles), cryptic tonsillitis (chronic infection)
- Dx: primarily clinical history and physical, may consider throat cultures (may consider rapid Streptococcal Antigen test), 2 dilutional rise in antistreptolysin-O titer (carrier state may be detected with positive culture without a change in antistreptolysin-O titer)
- DDx: peritonsillar cellulitis/abscess, leukemia, or lymphoma
- Complications: acute airway compromise, peritonsillar abscess (*see below*), parapharyngeal or retropharyngeal space abscess, rheumatic fever (secondary to an immune response resulting in carditis, polyarteritis, chorea, erythema marginatum, subcutaneous nodules), glomerulonephritis (secondary to an immune response resulting in a nephritic syndrome), sepsis, scarlet fever (*see below*)
- Rx: antimicrobials against suspected pathogens for 10 days (penicillin antimicrobial of choice, may consider β-lactamase inhibitors, clindamycin for allergic patients), bed rest, hydration, analgesics, oral hygiene, tonsillectomy (*see below*)

Chronic Tonsillitis

- Pathophysiology: typically polymicrobial infection
- SSx: malaise, recurrent sore throat, **fibrotic** or **cryptic** tonsils, cervical lymphadenopathy, halitosis
- Dx: clinical history and physical exam
- Rx: long-term antibiotics against β-lactamase producing organisms and encapsulated organisms, tonsillectomy

Adenoiditis

- Pathogens: similar to tonsillitis
- SSx: sore throat, purulent rhinorrhea, halitosis, postnasal drip, nasal obstruction (snoring), fever

- Dx: clinical history and physical
- Rx: similar to acute tonsillitis, adenoidectomy (*see below*)

Chronic Adenoiditis

- Pathophysiology: typically a polymicrobial infection, may be related to GERD especially in children (difficult to distinguish from sinusitis)
- SSx: persistent nasal discharge, malodorous breath, nasal obstruction (snoring)
- Dx: clinical history and physical exam
- Rx: similar to chronic tonsillitis, adenoidectomy

Obstructive Adenoid and Tonsillar Hyperplasia

- Adenoid Hyperplasia SSx: nasal obstruction, hyponasal speech (pinching nose does not change speech, "M" words), snoring, chronic mouth breathing, rhinorrhea, "adenoid facies" (open mouthed, dark circles under eyes, flattened midface, high arched palate), eustachian tube obstruction (otitis media), sinusitis
- Tonsillar Hyperplasia SSx: snoring, hyponasal speech, sleep apnea
- Adenoid Grading: 0 (absent), 1+ (<25% obstruction), 2+ (25–50% obstruction), 3+ (50–75% obstruction), 4+ (>75% obstruction)
- Tonsillar Grading: 0 (unable to visualize tonsil), 1+ (tonsil within fossa), 2+ (<50% obstruction), 3+ (>50% obstruction), 4+ (tonsils touching, "kissing")
- severe presentations may result in complications of Obstructive Sleep Apnea such as failure to thrive or cor pulmonale (*see also* p. 166)
- Dx: clinical history and physical exam; visual inspection (nasopharyngoscopy); for unilateral tonsilar hyperplasia must rule out neoplasms (lymphoma, leukemia) or unusual infections (*M. tuberculosis*, atypical mycobacteria, actinomycosis, fungal); sleep study and lateral neck radiographs are typically not necessary
- Rx: for acute upper airway obstruction consider nasal trumpet (rarely requires intubation) and a short course of corticosteroids, may consider prolonged course of antibiotics (3–6 weeks) or nasal corticosteroid sprays for adenoid hyperplasia; tonsillectomy and adenoidectomy for definitive therapy (*see below*)

Peritonsillar Abscess and Peritonsillar Space Infection

- Pathophysiology: spread of infection outside tonsillar capsule into the peritonsillar space (typically begins at superior pole)
- Boundaries of Peritonsillar Space: palatal tonsil (medial border), superior constrictor muscles (lateral border), tonsilar pillars (superior, inferior, and posterior border)

- Risks: recurrent infections, allergy, dental caries
- SSx: unilateral otalgia, odynophagia, uvular deviation, pharyngotonsillar asymmetry, trismus, drooling
- Complications: airway distress, parapharyngeal or retropharyngeal abscess, aspiration pneumonia
- Rx: urgent incision and draining, may consider needle aspiration for minor cases, antibiotics, elective tonsillectomy after resolution of the infection (high risk of recurrence), "Quinsy tonsillectomy" (tonsillectomy at time of infection) may be considered for younger children or recurrent/unresponsive cases

Herpangina (Hand-Foot-Mouth Disease)

- Pathogen: **Coxsackie A virus**
- SSx: oral vesicles (anterior pillar, palate, and buccal mucosa), benign rapid course, high fevers, fatigue, anorexia
- Dx: clinical history and physical exam
- Rx: oral hygiene, observation (rapid self-limiting course), analgesics, hydration, bed rest

Scarlet Fever

- Pathophysiology: secondary effect from the endotoxin produced by Type A β-hemolytic *Streptococcus*
- SSx: tonsils and pharynx deep red, **strawberry tongue**, perioral skin erythema and desquamation, dysphagia, malaise, severe cervical lymphadenopathy
- Dx: clinical exam and history, cultures and sensitivity, **Dick Test** (involves injection of diluted erythrogenic toxin which elicits local erythema if positive)
- Rx: antibiotics (IV penicillin G) and oral hygiene

Infectious Mononucleosis

- Pathogen: Epstein-Barr virus (oral contact)
- SSx: high grade fever, lymphadenopathy (posterior triangle), general malaise, fibrinous exudative tonsillitis, associated hepatosplenomegaly, rhinopharyngitis
- Dx: clinical history, monospot test, Paul-Bunnel test (heterophil antibodies in serum), 80–90% mononuclear and 10% atypical lymphocytes on smear, viral culture
- Complications: acute airway obstruction, cranial nerve involvement, meningitis, autoimmune hemolytic anemia, splenic rupture

- Rx: secure airway for acute obstruction with a nasal trumpet or endotracheal intubation (corticosteroids for severe obstructing tonsillitis), hydration, analgesics, oral hygiene, antimicrobials for secondary infections (ampicillin may cause a severe rash), typically resolves after 2–6 weeks

Diphtheria (*see* pp. 102–103)

Tonsillectomy and Adenoidectomy

Tonsillectomy Indications and Contraindications

- significant decrease in incidence (>1 million cases/year in the 1950s)
- tonsillectomy/adenoidectomy is still the most common major surgical procedure in children in the United States

Absolute Indications

- tonsillar hyperplasia resulting in sleep disturbances or sleep apnea associated with cor pulmonale
- suspected malignancy (unilateral tonsilar hypertrophy)
- tonsillitis resulting in febrile convulsions (may require a Quincy tonsillectomy)
- persistent or recurrent tonsillar hemorrhage
- failure to thrive (not attributable to other causes)

Relative Indications

- recurrent acute tonsillitis (documented 7 infections in 1 year, 5 infections in 2 years, or 3 infections in 3 years or >2 weeks of missed school or work in 1 year)
- peritonsillar abscess
- chronic tonsillitis with persistent sore throat, halitosis, or cervical adenitis
- swallowing difficulties (not attributable to other causes)
- tonsillolithiasis
- orofacial or dental disorders (results in a narrow upper airway)
- *Streptococcus* carrier unresponsive to medical management
- recurrent or chronic otitis media

Contraindications

- leukemias, hemophilias, agranulocytosis, uncontrolled systemic disease (diabetes, tuberculosis)
- Relative Contraindications: cleft palate, acute infection

Adenoidectomy Indications and Contraindications

Absolute Indications

- adenoid hyperplasia resulting in sleep disturbances or sleep apnea associated with cor pulmonale
- nasal obstruction associated with orofacial abnormalities
- failure to thrive (not attributable to other causes)

Relative Indications

- recurrent acute adenoiditis (documented 7 infections in 1 year, 5 infections in 2 years, or 3 infections in 3 years or >2 weeks of missed school or work in 1 year)
- chronic adenoiditis with persistent sore throat, halitosis, or cervical adenitis
- swallowing difficulties (not attributable to other causes)
- recurrent or chronic otitis media or sinusitis

Contraindications

- palatal clefting
- velopharyngeal insufficiency (hypernasal speech, nasal regurgitation)

Complications

- **Immediate and Delayed Hemorrhage**: 0.5–1% incidence, intraoperative hemorrhage may be caused by an aberrant carotid artery, retained adenoid tissue, or tears in the posterior pharyngeal wall injury, delayed hemorrhage may occur 7–10 days postoperatively when the eschar sloughs off; Rx: intraoperative pressure packing, hemostatic agents (eg, thrombin, cellulose, microfibrillar collagen), suction cautery, sutures, posterior nasopharyngeal pack (for recalcitrant intraoperative bleeding), external carotid artery ligation (life-threatening bleeding); delayed postoperative bleeding may be addressed in the treatment room in the cooperative patient with topical hemostatic agents, electrical or chemical cautery, if unsuccessful should be controlled in the operating room
- **Postoperative Airway Compromise**: rare, may occur from dislodged clots, dislodged adenotonsillar tissue, postoperative oropharyngeal edema, or a retropharyngeal hematoma; Rx: manage airway (reintubation, surgical airway) corticosteroids, address retropharyngeal hematoma
- **Dehydration**: typically secondary to pain; Rx: analgesics, parenteral hydration
- **Pulmonary Edema**: caused by the sudden relief of airway obstruction from long-standing adenotonsillar hypertrophy resulting in a sudden drop of intrathoracic pressure, increased pulmonary blood volume, and increased hydrostatic pressure, may occur immediately or after a few hours; Rx: positive end-expiratory pressure ventilation, mild diuresis

- **Atlantoaxial (C1–C2) Subluxation**: rare incidence, increased risk with Down syndrome patients
- **Velopharyngeal Insufficiency**: results from an incompetent velopharyngeal inlet, increased risk with the presence of a submucosal cleft, history of nasal regurgitation, or preoperative hypernasality; Rx: speech therapy (typically resolves), pharyngeal flap
- **Nasopharyngeal Stenosis**: rare complication from scarring; Rx: difficult to treat (high rate of recurrence), may consider pharyngeal flaps or stents
- **Eustachian Tube Dysfunction**: from scarring and stenosis of the eustachian tube orifice; Rx: pressure equalization tubes, prevent by avoiding curettage near torus tubaris
- **Aspiration Pneumonia**: rare, occurs from aspiration of blood clots of adenotonsillar issue

SLEEP APNEA AND SNORING

Introduction

Physiology

Types of Sleep Apnea

- **Central Sleep Apnea**: apneas caused by decreased respiratory motor drive, more common in infants and the elderly
- **Obstructive Sleep Apnea (OSA)**: apneas caused by anatomical narrowing of the upper airway
- **Mixed**: sleep apnea caused by both central and anatomical components
- **Pickwickian**: OSA secondary to obesity, obstruction results from redundancy in neck, soft palate, and base of tongue

Stages of Sleep

- **Slow Wave Sleep**: stage of sleep in which brain waves are slowed, deep and restful sleep, decrease in vascular tone, respiratory rate, and basal metabolic rate
- **Rapid Eye Movement (REM) Sleep**: stage of sleep in which the brain is quite active, not restful, associated with active dreaming ("remembered"), increased metabolic and physical activity, occurs more frequently with longer sleep periods, quicker onset with sleep deprivation and other sleep disturbances (eg, narcolepsy)

Pathophysiology of OSA

1. during REM or deep sleep, obstruction occurs resulting in decreased arterial oxygen pressure (PaO_2) and increased arterial carbon dioxide pressure ($PaCO_2$)

2. nocturnal desaturation arouses patient and causes increased pulmonary and systemic arterial pressure and stimulates erythropoiesis (polycythemia)
3. sleep deprivation leads to daytime hypersomnolence

Causes of Sleep Apnea

- oropharyngeal and hypopharyngeal laxity
- nasal obstruction
- tonsil and adenoid hypertrophy
- macroglossia, retrodisplaced tongue, lingual tonsil hypertrophy
- micrognathic or retrognathic mandible
- midface abnormalities

Complications

- nocturnal death
- cor pulmonale, cardiac heart failure, and hypertension
- auto and industrial accidents
- growth delay or failure to thrive in children
- poor work performance

Evaluation of Sleep Apnea

History and Physical

- Symptoms: snoring, apnea (cessation of breathing >10 seconds), morning headaches, chronic mouth breathing, restlessness, daytime hypersomnolence, poor work performance, enuresis
- Contributing Factors: obesity, drug history (sedating medications, alcohol), medical profile (right cardiac failure, thyroid and pulmonary disorders)
- Physical Exam: weight and overall body habitus, craniofacial abnormalities (mandibular and maxillary deformities), nasal obstruction (deviated septum, hypertrophic turbinates, nasal polyposis), allergic rhinitis, oropharyngeal redundancy, tonsil and adenoid hypertrophy, elevated blood pressure
- DDx: narcolepsy (episodes of sudden attacks of sleep), nocturnal myoclonus (periodic sleep movement), REM disorders, sleep deprivation)

Sleep Study (Polysomnography)

- sensitive and specific, expensive
- Components: EEG, EKG, EOG (electrooculargram), pulse oximeter, respiration rate, nasal and oral air flow
- **Apnea**: cessation of respirations >10 seconds

- **Hypopnea**: cessation of respirations for 5–10 seconds
- **Apnea Index**: number of apneic episodes per hour (>5–10 episodes/hour suggests sleep apnea)
- **Oxyhemoglobulin Desaturations**: <85% saturations highly significant; <60% severe obstruction
- **Respiratory Disturbance Index (RDI)**: total number of apneas and hyponeas per hour of sleep, normal (<5 apneas/night), mild (10–30 apneas/night), moderate (30–50 apneas/night with SaO_2 <85%), severe (>50 apneas/night with SaO_2 <60%)
- In general, the longest duration of apnea and the lowest O_2 desaturation are more important than mean values

Other Diagnostic Tests

- **Multiple Sleep Latency Test (MSLT)**: series of 4–5 daytime naps 2 hours apart, excessive daytime sleepiness (sleep apnea or narcolepsy) results in a short onset of sleep (< 5minutes) or short-onset REM sleep (narcolepsy)
- **Home Sleep Study**: less expensive, less specific
- **Home Audio/Video Monitoring**: observe sleeping children
- **Muller's Maneuver**: nasopharyngoscope is placed in hypopharynx, examine site of velopharyngeal collapse during inhalation with pinched nose and closed mouth (reverse Valsalva); collapse that occurs maximally at the tonsillar fauca and soft palate correlates with better outcome after a uvulopalatopharyngoplasty

Ancillary Tests

ECG, chest x-ray (cor pulmonale), ABG (CO_2 retention, hypoxia), CBC (polycythemia), lateral neck films (evaluate adenoid bed, airway patency), thyroid function tests (hypothyroidism), cephalometric studies

Management

Nonsurgical Management

- Behavior Modifications: weight loss, cessation of alcohol and sedating medications, lateral decubitus sleeping (securing an object to the back at night)
- Medical Management: address rhinitis, sinusitis, or hypothyroidism if present
- **Oxygen Supplementation**: considered in mild sleep apnea
- **Continuous Positive Airway Pressure (CPAP)**: highly effective if tolerated, 30% noncompliance
- **Mechanical Prostheses**: tongue retaining devices, mandibular positioning devices, nasal airways

- **Protriptyline**: tricyclic antidepressant (nonsedating), decreases REM sleep, controversial effectiveness

Surgical Management

- **Tracheotomy**: most effective management, "**gold standard,**" procedure of choice for serious OSA with complications (morbid obesity, pulmonary or cardiac sequelae)
- **Nasal Obstruction Management**: septoplasty, turbinate reduction, polypectomy
- **Tonsillectomy and Adenoidectomy**: most common surgical management in children; confirmation with polysomnography not necessary with classic history in children (snoring, apneic episodes, restlessness); may also consider lingual tonsillectomy, linguoplasty, or midline laser glossectomy for prominent, obstructing lingual tonsils
- **Uvulopalatopharyngoplasty (UPPP)**: removes the palatine tonsils, posterior edge of soft palate, and uvula; 50% improvement of OSA and 96% improvement of snoring; complications include velopharyngeal insufficiency (VPI), nasopharyngeal stenosis (increased risk with concurrent adenoidectomies), postobstructive pulmonary edema
- **Laser Assisted Uvuloplasty** (LAUP) or **Bovie Assisted Uvuloplasty (BAUP)**: amputates uvula and 1 cm trench in soft palate
- **Somnoplasty**: utilizes low-power, low-temperature radiofrequency energy directed into the uvula and soft palate to reduce volume of the tissue, may be completed in the office, recently approved by U.S. Food and Drug Administration for management of snoring

Mandibular and Midface Advancement Techniques

- **Total Mandibular Advancement**: indicated for retrognathic mandibles, requires bilateral osteotomies and plating
- **Anterior Mandibular Advancement**: requires a myotomy of the infrahyoid muscles, resuspension of the hyoid bone more superiorly and anteriorly, and advancement of the genial tubercle
- **Le Fort I Osteotomies**: indicated for midfacial defects, requires bilateral maxillary osteotomies

BENIGN ORAL CAVITY LESIONS

Anatomy and Physiology

Tongue and Palate Anatomy

Surface Anatomy

- **Fungiform Papillae**: small red structures found at tip and edge of anterior 2/3 of tongue

- **Foliate Papillae**: located at the posterolateral tongue base
- **Circumvallate Papillae**: raised circular structures, arranged in V-shape at junction of anterior $^{2}/_{3}$ and posterior $^{1}/_{3}$ of tongue
- **Filiform Papillae**: distributed throughout tongue, bumpy appearance, does not participate in taste
- **Sulcus Terminalis**: groove between anterior and posterior tongue
- **Foramen Cecum**: central point of the sulcus terminalis (origin of thyroid gland)

Muscular Anatomy

- **Tongue Extrinsics**: genioglossus, palatoglossus, styloglossus, hyoglossus muscles (innervated by hypoglossal nerve)
- **Tongue Intrinsics**: superior and inferior longitudinal, vertical, transverse layers (innervated by hypoglossal nerve)
- **Palatal Muscles**: musculus uvulae, levator veli palatini, tensor veli palatini, palatoglossal (anterior pillar), palatopharyngeus (posterior pillar), innervated by glossopharyngeal and vagus nerves except the tensor veli palatini which is innervated by CN V_3

Taste

- Sensations of Taste: salty, sweet, sour, and bitter (topographic mapping of taste does not exist, all sensations are perceived by each taste receptor)
- taste buds are found in fungiform, foliate, and circumvallate papillae (not found in filiform papillae) of the tongue; taste buds are also located on the hard palate, anterior pillar, tonsil, and posterior pharyngeal wall
- saliva is required for taste

Afferent Taste Innervation

- Anterior Two-Thirds of the Tongue: taste receptors (fungiform and foliate papillae)—chorda tympani nerve → **geniculate ganglia** (taste fiber cell bodies) → nervus intermedius (**CN VII**) → nucleus solitarius
- Posterior One-Third of the Tongue: taste receptors (foliate and circumvallate papillae; posterior oropharynx, valleculae, and base of tongue)—**CN IX** → inferior petrosal ganglion → nucleus solitarius
- laryngeal surface of epiglottis—**CN X** → nucleus solitarius

Evaluation of Disorders of Taste

Determine Classification of Taste Disorder

- **Ageusia**: no taste sensation (eg, congenital aplasia, toxins, cranial nerve injury)

- **Hypogeusia**: decreased sensitivity to taste (eg, radiation effect)
- **Hypergeusia**: increased sensitivity to taste (eg, glossopharyngeal neuralgia)
- **Dysgeusia/Parageusia**: foul or abnormal perception of taste (eg, infectious stomatitis)

History and Physical

- History and Associated Symptoms: medications (*see below*); recent toxin exposure; other neurologic symptoms (paresthesia, stroke, weakness, anosmia); history of diabetes, endocrine disorders, connective tissue disease, or depression
- Medications that Cause Taste Disturbances: antibacterial mouthwash, anticholenergics, aspirin, antiparkinson drugs, acetazolamide, lithium, penicillamine, others
- Physical Exam: complete head and neck exam including oral and oralpharyngeal exam, otologic exam (chorda tympani), cranial nerve assessment, and other neurologic evaluation
- Think "KITTENS" for differential diagnosis (*see* Table 4–6)

Evaluation of Oral and Oropharyngeal Lesions

Introduction

- **5–20%** of leukoplakia is malignant or premalignant
- Erythroplakia has higher risk of malignancy (approximately **25%**)

History and Physical Exam

- History of Lesion: onset, duration, and progression of lesion; painful
- Contributing Factors: trauma (gum biting, poor fitting dentures) or caustic ingestion; risk factors for malignancy (weight loss, smoker, alcohol abuse, family history, etc); history of connective tissue diseases, autoimmune disorders, immunodeficiency, diabetes, radiation therapy, other malignancies
- Associated Symptoms: taste disturbances, persistent sore throat (>3 weeks), odynophagia, dysphagia, halitosis, hoarseness, trismus, fever, malaise, persistent otalgia (referred pain with normal otologic exam)
- Physical Evaluation of Lesion: describe lesion (eg, macular, papular, ulcerative, vesicular), color (leukoplakia, erythroplakia, *see* Table 4–7), adherency, induration, tenderness
- Type of Leukoplakia
  1. **Keratotic**: adherent, insidious development, protracted course, nonerosive surface (**higher risk of carcinoma**)

TABLE 4–6. Differential Diagnosis of Taste Disturbances: KITTENS Method

(**K**) **C**ongenital	**I**nfectious & **I**diopathic	**T**oxins & **T**rauma	**T**umor (Neoplasia)	**E**ndocrine	**N**eurologic	**S**ystemic
Aplasia of taste buds	Otitis media (direct damage to chorda tympani) Oral stomatitis (Thrush)	Medications Facial nerve or glossopharyngeal injury (tonsillectomy, otological surgery) Toxins	Neoplasms of the floor of mouth, infratemporal fossa, cerebellopontine angle Glomus jugulare	Addison's disease Diabetes Pregnancy Hypothyroidism	Epilepsy Cerebral cortical disease Bell's palsy Herpes zoster oticus	Mucosal atrophy secondary to Sjögren's Radiation effects Depression Malnutrition

TABLE 4–7. Differential Diagnosis of Benign Pigmented Oral Lesions

Color	Causes
Generalized pale mucosa	Anemia, Thalassemia
Black/brown discolorization	Bismuth and arsenic intoxication
Blue-gray gingival margin (**Burton's Line**)	Lead intoxication
Generalized redness	Polycythemia vera, hepatic insufficiency
Perioral melanotic macules	Puetz-Jeghers syndrome (GI hamertomatous polyps)
Small yellow spots	Fordyce's disease (sebaceous gland histology)
Black-hair tongue	Elongated (hyperplastic) filiform papillae
Telangiectasia	Osler-Weber-Rendu syndrome
Diffuse hyperpigmentation of mucosa	Addison's disease

2. **Nonkeratotic**: nonadherent, acute onset, erosive and ulcerative features (**higher risk of acute infections**)

- Oral and Oropharyngeal Exam: visually inspect of all areas of oral and oral pharyngeal cavity (indirect mirror exam) for masses and lesions (ulceration, leukoplakia, erythroplakia); palpate floor of mouth and base of tongue; assess mobility of tongue and any involvement of mandible; inspect dentition (quality of teeth, occlusion); surface and consistency of the tongue; palpate salivary glands and Stenson's duct
- Other Physical Exam: complete H&N exam (signs of underlying malignancy, cervical adenopathy), associated cutaneous lesions

Diagnostic Testing

Biopsy

- **all chronic leukoplakia or ulcerative lesions that fail to heal after 1–2 weeks should undergo excisional biopsy** (must keep a high suspicion for malignancy)
- biopsy specimen should include a clear margin
- consider direct and indirect immunofluorescence staining

Other Ancillary Tests

- Culture and Sensitivity: consider culture of oral/oropharyngeal mucosa or lesion if suspect infection (eg, fever, tender cervical adenopathy); must evaluate for aerobic, anaerobic, and fungal organisms
- CT /MRI: indicated if suspect tumor, evaluates extent of tumor size and involvement of adjacent structures; aids in staging and determining nodal status
- Laboratory Tests: complete blood count with differential, autoimmune and connective tissue profiles (eg, ANA, SS-A, SS-B, ESR, Rh factor, LE cell), ACE levels

Infectious Stomatitis

Herpetic Gingivostomatitis

- Pathogenesis: activation of dormant Herpes Simplex Virus Type I (HSV-II associated with genital lesions, although may be found in oral lesions)
- Types
  1. **Primary**: most common in children, associated with fever, malaise, cervical lymphadenopathy
  2. **Secondary**: recurrence of dormant virus in the trigeminal ganglion (migrates along axonal sheath); triggered by stress, trauma, or immunosuppression

- SSx: small painful vesicles that ulcerate leaving an erythematous base with a gray cover, heals without scar, odynophagia, fever, malaise, cervical adenopathy, usually resolves in 1–2 weeks
- Dx: history and clinical exam, viral culture in early stages, monoclonal antibodies and DNA hybridization
- Rx: oral acyclovir for primary infections or for prophylaxis (immunocompromised patient), topical acyclovir for active lesions

Acute Necrotizing Ulcerative Gingivitis—Trench Mouth—Vincent's Gingivitis

- Pathophysiology: mucosa infected by multiple synergistic pathogens including spirochetes (***Borrelia vincentii***), fusiform rods, and anaerobic bacteria
- Risks: malnutrition, degenerative diseases, immunocompromised, debilitating diseases
- SSx: ulcerative ("punched out") craters of the interdental papilla, gray pseudomembranous cover, malaise, fever, cervical adenopathy, halitosis
- Dx: culture, clinical exam and history
- Rx: oral hygiene, antibiotics

Oral Candidiasis (Thrush)

- Pathophysiology: opportunistic infection, typically *Candida albicans* (*Aspergillus* may also be cultured)
- Risks: long-term antibiotics, infants, elderly, immunosuppression (corticosteroids, poorly controlled diabetes), poor nutrition status, radiation and chemotherapy
- SSx: friable, white (cheesy) plaques that scrape off leaving an erythematous base, pseudomembranous erythematous plaques, odynophagia, taste disturbances
- Dx: clinical exam and history, culture (fungal stain reveals 90 degree, pseudohyphae yeast)
- Rx: topical antifungals (eg, nystatin swish and swallow), systemic antifungals for severe forms, oral hygiene

Actinomycosis

- Pathophysiology: *Actinomyces israelii*, present within normal teeth, invade tissue through an extraction site or a damaged tooth
- Risks: oral mucosal trauma, poor oral hygiene, dental infections, immunocompromised
- SSx: head and neck mass, may have a purplish discolorization of overlying skin

- Dx: culture (requires 1–2 weeks for growth)
- Histopathology: branching anaerobic gram negative bacteria, **sulfur granules**
- Rx: surgical debridement and long-term antibiotics (penicillin, tetracycline, erythromycin)

Syphilis (*see* pp. 208–209)

Noninfectious Stomatitis

Erythema Multiforme

- Pathophysiology: antigen-antibody complex deposit in small vessels of the dermis and submucosa, may occur spontaneously (50%) or in association with a hypersensitivity reaction (medication or infection)
- SSx: oral or cutaneous target lesion with concentric erythematous rings, explosive presentation, self limiting, fever, regional adenopathy
- Dx: positive direct and indirect immunofluorescence (nonspecific presentation)
- Rx: supportive, corticosteroids

Stevens-Johnson Syndrome

- severe life-threatening form of Erythema Multiforme
- SSx: widespread lesions (mouth, eyes, genitalia, respiratory tract), photophobia, blindness, fever
- Rx: supportive care (hydration, analgesics, antipyretics), airway management, high-dose corticosteroids

Pemphigus Vulgaris

- Pathophysiology: autoantibodies against desmosome-tonofilament complexes (intracellular bridges) results in **acantholysis** (loss of cellular cohesiveness)
- Risks: Ashkenazic Jews, Mediterranean region, other connective tissue disorders
- SSx: painful blisters in oral or pharyngeal mucosa, desquamative gingivitis, **Nikolsky's sign** (rubbing or trauma of uninvolved mucosa produces an ulcer)
- Histopathology: intraepithelial cell splitting (**suprabasilar**), attached rows of basal cells to lamina propria (row of tombstones), **Tzank cells** (free squamous cells, more spherical from loss of intracellular attachment)
- Dx: direct immunofluorescence of **intracellular substance** (intraepithelial blistering), **positive** serum antibodies (indirect immunofluorescence)
- Rx: oral corticosteroids, may consider immunosupressive agents

Cicatricial Pemphigoid (Ocular Pemphigus, Benign Mucous Membrane Pemphigoid, Mucosal Pemphigoid)

- Pathophysiology: autoantibodies result in subepidermal blistering
- SSx: subepithelial bullae or desquamative gingivitis primarily involving attached gingiva, **Nikolsky's sign** (*see* Pemphigus Vulgaris), **ocular lesions** (50–70% incidence; conjunctivitis, blindness, symblepharon, entropion), presents in 4th–5th decade
- Dx: direct immunofluorescence in **basement membrane** (**subepithelial clefting**), **negative** indirect immunofluorescence (too localized)
- Rx: topical, intralesional, and oral corticosteroids, may consider immunosupressives and antimalarials

Bullous Pemphigoid

- Pathophysiology: autoantibodies results in subepidermal blistering
- similar to Cicatrical Pemphigoid, however Bullous Pemphigoid has cutaneous lesions (rare oral lesions), self limiting, affects both genders, and is positive for serum indirect immunofluorescence
- SSx: subepithelial bullae of flexor surfaces, groin, and abdomen; self limiting; **rare oral lesions**, seen in 7th–8th decade
- Dx: direct immunofluorescence of **basement membrane** (subepithelial clefting), **positive** serum indirect immunofluorescence (70%)
- Rx: oral corticosteroids, may consider immunosuppressive agents

Lichen Planus

- Pathophysiology: autoimmune disease in which the basal layer is destroyed by activated lymphocytes, may be familial, may be induced by medication (eg, penicillamine, methyldopa, phenothiazide, antimalarials)
- Types
  1. **Reticular**: lacy white lines (**Wickham's striae**), most commonly occurs on the buccal cheek mucosa (may also be found on the palate, lips, and tongue)
  2. **Plaque**: appears like leukoplakia, may manifest on the lips or palate
  3. **Atrophic**: atrophy in the center of the papule
  4. **Erosive and Bullous**: painful, ulcerative lesions, common on the buccal mucosa and dorsum of the tongue
  5. **Ulcerative**: may involve buccal mucosa (more common of the feet and toes), painful
  6. **Annular**: more common on the lips (may also be found on the penis), ringed edges composed of small papules
- SSx: recurrent papules of various features (depending on type) with a predilection for the flexor surfaces and trunk; however, 60–70% may present on the lips, oral mucosa, or eyelids; oral lesions tend to be chronic

- Dx: clinical exam, biopsy
- **Kobner Isomorphic Phenomenon**: lesions may be provoked by physical trauma (eg, itching, scratching)
- Histopathology: vacuolar alteration of the basal cell layer resulting in **Civatte bodies** (degenerative eosinophilic ovoid keratinocytes), "saw tooth" pattern of epidermal hyperplasia, lymphocytic infiltration of lamina propria
- Complications: 1–4% risk of malignant transformation (higher risk with ulcerative lesions)

Management

- no cure, treat painful, erythematous, and erosive lesions
- Identify Reversible Contributing Factors: medications, dental restoration, improve oral hygiene (frequent teeth cleaning), avoid tobacco, alcohol, and smoking abuse
- Medical Therapy: may consider oral or topical corticosteroids and retinoids; may also consider cryotherapy, UV light, and laser surgery

Aphthous Ulcers

- most common oral ulcer
- Pathophysiology: idiopathic, may be immunologic, infectious, hormonal, stress induced, traumatic, or nutritional
- Types
  1. **Minor**: most common, burning and tingling before ulcer formation, <1.0 cm in diameter, painful
  2. **Major**: more painful and larger (1–3 cm in diameter), multiple (1–10), risk of scar formation
  3. **Herpetiform**: numerous small ulcers 1–3 mm in diameter, lasts >1 month, risk of scar formation
- **Sutton's Disease**: recurrent aphthous ulcers (major type)
- SSx: painful, white ulcerations on the keratinized gingiva surrounded by a border of erythematous mucosa, may be multiple
- Dx: clinical exam and history
- Rx: observation (self-limiting), consider anti-inflammatory agents, antibiotics, antivirals, oral and topical corticosteroids, cauterization (silver nitrate), *Lactobacillus* capsules

Behçet's Disease

- Pathophysiology: idiopathic vasculitis
- SSx
  1. recurrent painful aphthous ulcers of the upper respiratory tract and genitalia

2. ocular inflammation (uveitis, iritis, papilledema, blindness)
3. cutaneous vasculitis
4. progressive SNHL, tinnitus, vertigo
5. systemic involvement: CNS, large vessel vasculitis

- Dx: clinical symptomatology
- Rx: no proven cure, may consider immunosuppressives, corticosteroids, or gamma globulin

Other Oral Lesions

Amyloidosis

- Pathophysiology: abnormal deposition of fibrillar protein and polysaccharide complexes
- Types
  1. **Primary Systemic**: (>50%) cardiac, tongue, and GI involvement
  2. **Secondary Systemic**: (<10%) associated with other chronic diseases
  3. **Localized**: <10%
  4. **Multiple Myeloma Associated**: 25%
  5. **Hereditary-Familial Amyloidosis**: 1%
- more common in African-Americans or Puerto Ricans, 3rd–4th decade (rare before 15 years old)
- H&N SSx: (10–15% present in the H&N) macroglossia (tongue most common site in H&N, 50%), anterior subglottic mass, orbital deposition
- Dx: biopsy and stain with Congo red (**apple-green birefringent** under polarized light)
- Rx: conservative surgical excision for symptomatic lesions

Primary Leukoplakia

- Pathophysiology: many potential etiologies including chronic irritation (dentures, teeth), smoking, and infection
- SSx: white patch that cannot be removed by rubbing (keratotic)
- Dx: excisional biopsy (must rule out malignancy)
- Histopathology: hyperkeratosis, acanthosis, atypia
- Complications: 10–30% malignant potential
- Rx: excision (usually at time of biopsy)

Hairy Leukoplakia

- Pathophysiology: benign mucosal hyperplasia associated with Epstein-Barr Virus
- associated with HIV patients
- SSx: painless, lateral tongue lesions

- Dx: clinical history and physical exam, biopsy
- Histopathology: hyperkeratosis, acanthosis, atypia
- Rx: observation, may consider high-dose acyclovir

Squamous Papillomas

- most common benign tumor of oral cavity and pharynx
- Pathophysiology: benign lesion associated with the human papilloma virus
- associated with tonsillar carcinoma
- SSx: well-demarcated papillary lesion, painless
- Dx: biopsy
- Rx: excision

Salivary Gland Cysts Mucocele (*see* p. 73)

ODONTOGENIC, JAW, AND BONE PATHOLOGY

Evaluation of the Jaw Mass

History and Physical Exam

History

- Character of Jaw Mass: onset, duration, and progression of growth, presence of pain (pain is typically associated with infected or malignant lesions)
- Contributing Factors: complete dental history including recent dental work and caries, history of other skin lesions (Basal Cell Nevus Syndrome), recent trauma, history of other congenital defects (cleft palate or lip)
- Associated Symptoms: fever, weight loss, malaise, temporomandibular joint pain

Physical Exam

- Character of Jaw Mass: size, distribution, tenderness, consistency, solitary versus multiple lesions
- Dental Exam: complete dental exam including percussion, presence of impacted teeth (follicular or dentigerous cysts) and nonviable teeth, malalignment (may result from divergent roots from compressive effects of underlying mass)

Management

Radiographs

- **Intraoral Radiographs**: indicated for screening dental disease; provide excellent detail; available in dental offices; inadequate evaluation of ramus, condyle, and inferior aspect of the mandible
- **Panorex**: assesses entire mandible in a single view, inadequate visualization of symphyseal region
- **Mandibular Series**: multiple extraoral views, difficult to delineate small lesions
- **CT/MRI**: not cost effective for screening; indicated for large, distorting lesions and preoperative planning for malignancy

Biopsy

- **Fine-Needle Aspiration**: allows acquisition of cells with minimal morbidity, may aspirate cysts for culture and sensitivity (vascular lesions may be aspirated to avoid hemorrhage from open biopsies)
- **Excisional Biopsy**: indicated for small lesions with low suspicion for malignancy by radiographs
- **Incisional Biopsy**: indicated for larger, potentially malignant lesions to obtain diagnosis prior to definitive resection

Jaw Cysts

Odontogenic Cysts

Periapical Cyst (Radicular Cysts)

- most common odontogenic cyst
- Pathophysiology: **nonviable tooth** (dental decay) results in osteitis of periapical bone from dental canal, epithelial cell rests of Malassez proliferate in the periodontal membrane causing cyst formation
- typically located on anterior maxilla and posterior mandible
- **Lateral Periodontal Cyst**: less common variant of radicular cyst which is found on the lateral aspect of the tooth
- SSx: typically asymptomatic, may illicit pain with percussion or heat
- Radiographic Findings: radiolucency at root apex of tooth with associated dental caries or pulp injury
- Histopathology: stratified squamous epithelium lining
- Complications: local cellulitis; fascial plane infections; abscess formation; fistula to oral cavity, skin, or nasal cavity; septicemia; jugular vein thrombosis; osteomyelitis; orbital extension
- Rx: antibiotics, drainage of abscess, and endodontic therapy (root canal to remove tooth)

Follicular or Dentigerous Cyst

- Pathophysiology: disruption of late odontogenesis (associated with an **impacted tooth**)
- SSx: often diagnosed late with jaw deformity, associated with an impacted tooth (especially the mandibular and maxillary third molar)
- Radiographic Findings: radiolucency at crown of an unerupted tooth, may displace teeth
- Histopathology: outer thin connective tissue wall with a thin stratified squamous epithelium inner layer
- Complications: risk of malignant transformation in cyst wall (unicystic ameloblastoma, epidermoid carcinoma)
- Rx: enucleation and curettage

Primordial Cyst

- Pathophysiology: degeneration of odontogenesis results in cyst development where a tooth would normally develop
- SSx: typically asymptomatic
- Radiographic Findings: ovoid, well demarcated lesions, may be multiloculated
- Complications: may become secondarily infected, 50% are considered odontogenic keratocysts
- Rx: enucleation and curettage, must consider odontogenic keratocysts if cyst recurs

Odontogenic Keratocyst (OKC)

- Pathophysiology: similar to primordial cysts
- may be associated with a crown of unerupted tooth (follicular cyst) or tooth root (periapical cyst)
- most commonly found in the mandibular third molar and ramus
- SSx: usually asymptomatic
- Dx: based on histology, FNA (white keratin-containing aspirate)
- Radiographic Findings: similar to primordial cysts
- Histopathology: (determines diagnosis) thin stratified squamous epithelium, columnar or cuboidal basal cell layer, parakeratin or orthokeratin surface layer, thin connective tissue layer, lumen may be filled with keratin
- Complications: may become secondarily infected, **10–60% recurrence**
- Rx: enucleation and aggressive curettage (rotary burr), careful follow-up for recurrence, consider resection with 1 cm margins for recurrent OKCs

Basal Cell Nevus Syndrome

- suspect with multiple OKCs

- autosomal dominant trait
- Other SSx: multiple basal cell CA, bifid ribs, hypertelorism, mandibular prognathism, calcification of falx cerebri (85%), palmar pitting (65%), depressed midface, frontal and parietal bossing

Eruption Cyst

- Pathophysiology: developmental cyst within the soft tissue
- SSx: bluish cyst overlying the alveolar ridge, typically resolves
- Radiographic Findings: translucent, dome shaped cyst
- Rx: excise if symptomatic

Nonodontogenic Cysts

- **Midpalatal Cyst of Infants**: arise from epithelium trapped between embryologic palatal shelves ("fissural"), midline palatal mass; Rx: enucleation and curettage
- **Nasolabial Cyst**: arise within the labial vestibule, presents as a swelling of the upper lip or nasal floor; Rx: excision
- **Nasopalatine Duct Cyst** (**Incisive Canal Cyst**): derived from embryological remnant of the nasopalatine duct, located between the maxillary central incisors (heart-shaped lucency, >10 mm); Rx: enucleation and curettage for symptomatic lesions
- **Aneurysmal Bone Cyst**: more common in the teenager with a history of trauma to the mandible, painful; Rx: rapid enucleation to avoid hemorrhage
- **Idiopathic Bone Cavity**: not a true cyst, may be secondary to a traumatic intramedullary hemorrhage with degeneration of the clot resulting in an air-filled bony space; Rx: biopsy to rule out other lesions

Odontogenic Neoplasms

Epithelial Neoplasms

Ameloblastoma

- Pathophysiology: benign neoplasm of uncertain origin, locally invasive (thought of as the oral counterpart to basal cell carcinoma)
- peak occurrence between 3rd and 4th decade
- 80% located in the mandible
- rare malignant transformation (*see below*)
- Types
  1. **Central**: arise in bone (intraosseous)
  2. **Plexiform Unicystic**: more aggressive central variant, occurs in the lining of **follicular cysts** or impacted teeth

3. **Peripheral**: arise in soft tissue around alveolar bone, much less aggressive, may be treated with local excision

- SSx: slow growing, painless, intrabony mandibular swelling, may resorb tooth roots
- Radiographic Findings: radiolucent multiloculations ("soap bubbles" or "honeycombed")
- Histopathology: various histological patterns, most common is the follicular pattern with islands of epithelium lined with columnar cells, central mass of loosely arranged cells (stellate reticulum), collagenous stroma
- Rx: wide excision with 1 cm bony margin and immediate reconstruction for central ameloblastomas (3–5 cm margin for plexiform unicystic types)

Malignant Ameloblastoma

- Pathophysiology: rare malignant degeneration from ameloblastoma resulting in metastasis
- SSx: regional or distant metastasis with aggressive local invasion
- Radiographic Findings: radiolucency with poorly defined margins
- Dx: biopsy of metastasis reveals similar histology to primary ameloblastoma with malignant features (atypical mitotic figures, invasive, pleomorphism)
- Rx: aggressive local resection, neck dissections for positive lymphadenopathy, consider postoperative radiation and/or chemotherapy

Pindborg Tumor (Calcifying Epithelial Odontogenic Tumor)

- benign infiltrating lesion
- median age of 40 years old
- associated with impacted molars
- SSx: slow growing, painless, intrabony mandibular swelling (similar to ameloblastomas)
- Radiographic Findings: unilocular radiolucency with calcifications
- Histopathology: sheets or islands of epithelial cells with eosinophilic cytoplasm, may contain amyloid with concentric calcifications or psammoma-like bodies (Liesegang rings)
- Rx: similar to ameloblastomas

Ameloblastic Fibroma

- Pathophysiology: true mixed tumor with epithelial and mesenchymal elements
- occurs in younger patients (5–20 years old)
- SSx: similar to ameloblastomas
- Radiographic Findings: similar to ameloblastomas

- Histopathology: islands of columnar or cuboidal epithelium with various arrangements (islands, cords, strands), central papilla-like connective tissue
- Rx: enucleation with bony curettage (does not require 1 cm margin typically required for an ameloblastoma)

Ameloblastic Odontoma

- Pathophysiology: hamartomatous lesion derived from epithelial (enamel secreting) and mesenchymal (dentin-secreting) elements
- associated with dentigerous cysts
- SSx: typically asymptomatic, may prevent tooth eruption
- Radiographic Findings: radiopaque mass surrounded by a thin radiolucency (may be associated with an unerupted tooth)
- Histopathology: presence of dentin and enamel (similar to a dental follicle), "ghost" cells
- Rx: simple enucleation (low risk of recurrence)

Adenomatoid Odontogenic Tumor

- Pathophysiology: hamartomatous growth (not neoplastic)
- more common in young women in first two decades
- associated with impacted teeth (cuspids)
- SSx: maxillary mass, typically anterior to the molars
- Histopathology: spheres of cuboidal and spindle cells, amyloid and dystrophic calcifications
- Rx: simple enucleation (low risk of recurrence since often degenerates)

Calcifying Odontogenic Cyst (Gorlin's Cyst)

- Pathophysiology: derived from proliferating epithelium, necrotic degeneration forms cysts
- SSx: mandibular asymptomatic swelling
- Radiographic Findings: well circumscribed radiolucency with central calcifications
- Histopathology: cysts with solid forms, lined by eosinophilic cells and "ghost" cells, may cause giant-cell reaction
- Rx: enucleation and curettage (en bloc resection for rare neoplastic variants)

Mesenchymal Neoplasms

Cementomas

- class of benign tumors that secrete cementum

- Types
  1. **Periapical Cemental Dysplasia**: more common in black females, multiple lesions, develops through varying stages (osteolytic, cementoblastic, and maturation)
  2. **Cementoblastoma**: true benign tumor of cementoblasts of the tooth root, usually from first mandibular tooth
  3. **Cementifying Fibroma**: similar to ossifying fibroma (*see below*)
- SSx: asymptomatic mandibular bony mass
- Rx: observation for periapical cemental dysplasia, curettage with extraction for cementoblastomas

Odontogenic Fibroma and Myxoma

- Pathophysiology: arise from periodontal ligament, dental papilla, or dental follicle
- SSx: slow-growing, asymptomatic mandibular mass
- Radiographic Findings: smooth-bordered radiolucency, may occur around the crown of an unerupted tooth
- Histopathology: fibromas contain an abundance of collagen producing fibroblasts, myxomas have fewer cells with a mucoid intracellular matrix
- Rx: fibromas require simple enucleation, myxomas are more difficult to excise and have a 25% rate of recurrence

Nonodontogenic Masses

Fibro-osseus Jaw Lesions

Osseous Dysplasia Cementoma

- Pathophysiology: **reactive disorder**
- more common in young blacks
- Types
  1. **Florid Osseous Dysplasia**
  2. **Traumatic Simple Bone Cyst**
- SSx: alveolar bone mass, typically asymptomatic
- Dx: biopsy
- Rx: observation, excision if symptomatic

Ossifying Fibroma

- Pathophysiology: benign, nonodontogenic tumor of the mandible or maxilla
- more common in females
- SSx: painless, slow-growing mandibular mass, may displace teeth
- Radiographic Findings: well demarcated, cannonball-like, homogeneous opacity

- Histopathology: abundant collagen with varying calcification
- Rx: enucleation with curettage, may consider en bloc resection for aggressive or recurrent lesions

Fibrous Dysplasia

- Pathophysiology: **developmental**, hamartomatous lesion of maxilla or mandible, **medullary bone replaced by fibro-osseous tissue**
- more common in middle-aged women
- Types
  1. **Monostotic**: one bone involved
  2. **Polyostotic**: more than one bone involved (**Albright Syndrome**: polyostotic fibrous dysplasia, precocious puberty, abnormal skin pigmented lesions)
  3. **Juvenile**: rapidly destructive, aggressive, destroys teeth, refractory to treatment
- SSx: slow growing, painless, well circumscribed, marble-like mass, destroys bone, unilateral facial deformity (mass does **not** cross midline, developmental plates)
- Dx: biopsy, panorex and plain films (ground glass, "Chinese writing," irregular, multiloculated, **diffuse margins**, eggshell thin cortex from destruction of cortical bone), CT/MRI
- Rx: excision and curettage, cosmetic shaping

Other Nonodontogenic Lesions

Torus Mandible and Palatinus

- Pathophysiology: autosomal dominant, enlargement of bone of the lingual surface of the mandible or the hard palate
- SSx: typically asymptomatic
- Dx: clinical exam
- Rx: observation, excision if symptomatic

Cherubism

- Pathophysiology: autosomal dominant, results in symmetric enlargement of mandible body and ramus (rarely involves maxilla)
- SSx: painless, round "cherub" face, regresses at puberty
- Radiographic Findings: multiple, multiloculated, well defined radiolucencies, thin cortex
- Rx: observation, may consider surgical option after puberty for persistence

Paget's Disease

- Pathophysiology: increased bone resorption and formation, involves the lumbosacral regions and skull

- Phases
  1. **Lytic**: replacement of bone with vascularized stroma, destructive phase
  2. **Mixed**: increased osteoclastic and osteoblastic activity, formation of Paget bone
  3. **Sclerotic**: decreased osteoclastic activity, increased hard, dense bone formation
- more common in patients >40 years old (fibrous dysplasia affects younger patients)
- SSx: enlarged skull, dorsal kyphosis, bowed legs, nonunion fractures, hearing loss, neurologic sequelae from compressive effects
- Dx: biopsy, panorex and plain films (polyostotic, mosaic pattern of bone), CT/MRI; bone scan, hypercalcemia, elevated alkaline phosphatase and urine hydroxyproline from increased bone turnover
- Radiographic Findings: multiple, multiloculated, well defined radiolucencies (lytic lesions), thin cortex, sclerotic lesions in later phases
- Complications: hypercalcemia, polycythemia, cardiac failure, malignant transformation (osteosarcoma, giant cell tumor), neurologic compression, normal pressure hydrocephalus, pathologic fractures
- Rx: no treatment require if asymptomatic and localized, medical management for symptomatic or complicated disease includes calcitonin, cytotoxic agents, etidronates, and analgesics

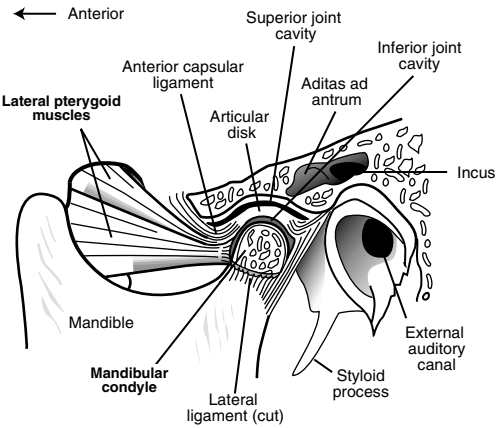

Figure 4–2. Components of the temporomandibular joint.

Temporomandibular Joint Disorders

Temporomandibular Joint (TMJ) Anatomy (Fig. 4–2)

- Vascular Supply: superficial temporal artery
- Sensory Innervations: auriculotemporal, masseteric, and deep posterior temporal nerves (CNV_3)
- **Diarthrodial Joint**: true synovial joint (freely mobile) with two modes of movement (gliding and hinging)
- **Glenoid Fossa**: socket within the temporal bone, lined with fibrocartilages
- **Articular Eminence**: anterior aspect of the glenoid fossa which articulates to the **condyle**

Fibrocartilagenous Articular Disk (Meniscus)

- the articular disk is interposed between the articulating components (separates the superior and inferior synovial compartments)
- attaches to the condyloid process by the **lateral and medial ligaments** and by the inferior lamina of the **meniscotemporomandibular frenum**
- Disc Bands: anteriorly attaches to the lateral pterygoid muscle (2 mm disc thickness), intermediate region (thinnest zone, 1 mm disc thickness), posteriorly (thickest, 3 mm disc thickness)
- Functions: stability, allows rotational and translatory motions, absorbs shock forces

Evaluation

History and Physical

- must differentiate myogenic versus articular and osteogenic pathology
- History: jaw pain (worse with movement or chewing), may manifest as otalgia, headaches, toothache, facial pain, vertigo, trismus, **nocturnal bruxism** (jaw clenching during sleep, associated with muscle strain), recent trauma
- TMJ Exam: evaluate occlusion and intraincisor opening (normal 40–50 mm); palpate TMJ and jaw muscles; examine for tenderness, warmth, crepitus, and joint clicking
- H&N Exam: otologic exam (exclude primary ear disease), nasopharynx, oropharynx, nasal, sinus, and laryngeal exam (evaluate for possible sources of referred pain), parotid palpation (parotitis)

Initial Management

- early management is key to preventing progression of joint degeneration
- Behavior Modifications: soft diet (may require liquid diet, smaller bites), avoid gum chewing

- Physical Therapy: repositioning, repetitive jaw stretching, active and passive mobilization
- Medical Management: analgesics and NSAIDs (narcotics should be avoided), muscle relaxants, warm compresses, ultrasound, may consider corticosteroid injections
- Dental Management: consult for malocclusion, carious teeth, poorly fitting dentures, possible orthognathic surgery
- Prosthetic Devices: occlusal splints

Imaging

- indicated for failed conservative management
- **MRI**: indicated if suspect disc displacement or dysmorphology
- **CT**: indicated if suspect degenerative joint disease, bony deformities, or ankylosis
- **Arthrotomogram**: requires injection of contrast into joint space, can detect disc displacement

Surgical Management *(controversial)*

- indicated for confirmed TMJ intracapsular derangements
- **Arthroscopy and Arthrotomy**: exploration of TMJ, removes debris, lyses adhesions
- **Discplasty, Discectomy**: repositioning, plication, removal, or reduction of articular disc, indicated for fractured or malaligned discs
- **Total Joint Prosthesis**
- Complications: 10–20% failure rate, joint degeneration, malocclusion, osseous ankylosis, foreign-body reaction (implants)

Articular and Osteogenic Pathology

Congenital and Developmental Diseases

- **Agenesis**: typically unilateral, associated with otologic abnormalities
- **Maxillomandibular Hypoplasia or Hyperplasia**: facial asymmetry, malocclusion (cross-bite), hypoplasia may be acquired (radiation effects, infection, trauma)

Arthritis

- **Degenerative Arthritis**: occurs in the elderly, primarily a mechanical process, minimal inflammatory changes, destroys articular tissue; radiograph may reveal erosion of condyle; Rx: conservative management (*as above*), rare surgical indications
- **Infectious Arthritis**: rare, unilateral lesion; Rx: antibiotics, restrict movement, consider aspiration or incision and drainage for suppurative infections

- **Rheumatoid Arthritis**: usually bilateral lesions, radiograph reveals bone destruction; Rx: medical management (eg, anti-inflammatory agents, hydroxychloroquine, gold), jaw exercises, consider surgery for ankylosis
- **Traumatic Arthritis**: trauma-related edema, intraarticular hemorrhage, and disc injury; Rx: NSAIDs, heat compress, restrict jaw motion

TMJ Disc Displacement

- Pathophysiology: joint dislocation secondary to disruption of the ligamentous attachments
- more common in women
- Causes: hyperextension of jaw secondary to intubation, forceful teeth extraction (especially third molars), or retraction from tonsillectomy; chronic clenching, facial trauma
- Types
  1. **Reducing**: joint "clicks" back into alignment with jaw opening
  2. **Nonreducing**: joint "locks" with jaw opening
- most commonly displaced **anteriorly** (due to the vector force of the lateral pterygoid muscles)
- SSx: clicking or locking during jaw motion, pain
- Dx: clinical exam, MRI
- Rx
  1. observation for painless (reducing) clicking
  2. painful clicking may require conservative management (*see above*)
  3. persistent painful clicking or nonreducing displacement require reduction under sedation

Other Causes

- **Mandible and Maxillary Fractures**: *see* pp. 449–460
- **Dislocations**: results in an open bite deformity; Rx: manual reduction for single acute instances, chronic dislocation may require sclerosing injections to stimulate scarring and contracting or a capsulorrhaphy
- **Neoplasms**: rare benign and malignant tumors of bone and cartilage; Rx: surgical management
- **Ankylosis**: most often from trauma or rheumatoid arthritis, typically unilateral, fibrous and bony types; Rx: surgical management (*see above*), physical therapy

Myogenic Pathology

- the TMJ is susceptible to any muscular pathology
- Pathophysiology: typically psychophysiologic, stress results in muscle hyperactivity, clenching, or grinding

- SSx: poorly localized and diffuse pain or ache, tenderness on palpation of muscle of mastication or tendinous attachments, limited jaw opening secondary to pain, tension-like headache
- Dx: clinical evaluation, may consider radiographs to rule out articular and osteogenic pathology
- Complications: may lead to degenerative arthritis, internal derangements, contracture or malocclusion
- Rx: NSAIDs, analgesics, soft diet, warm compress, restrict jaw movement, muscle relaxants, oral splints (nocturnal bruxism), antidepressants, anxiolytics, stress management

NECK MASSES

Anatomy of the Neck

Anterior Cervical Triangles

- Boundaries: midline of the neck, posterior border of the sternocleidomastoid muscle, and inferior border of the mandible
- **Submandibular Triangle**: bordered by the inferior border of the mandible, and by anterior and posterior digastric muscles
- **Submental Triangle**: bordered by the anterior belly of the digastric, hyoid bone, and midline of the neck
- **Carotid Triangle** (**Superior Carotid Triangle**): bordered by the omohyoid muscle, posterior belly of the digastric muscle, and the posterior border of the sternocleidomastoid muscle
- **Muscular Triangle** (**Inferior Carotid Triangle**): bordered by the omohyoid muscle, midline of the neck, posterior border of the sternocleidomastoid muscle

Posterior Cervical Triangles

- Boundaries: clavicle, posterior border of the sternocleidomastoid muscle, anterior border of the trapezius muscle
- **Occipital Triangle**: bordered by the posterior border of the sternocleidomastoid muscle, anterior border of the trapezius muscle, and the omohyoid muscle
- **Subclavian Triangle**: bordered by the posterior border of the sternocleidomastoid muscle, the clavicle, and the omohyoid muscle
- Contents of the Posterior Triangle: scalene, posterior cervical, and supraclavicular nodes; pleural apices; phrenic nerve, brachial plexus (trunks); subclavian artery and vein; anterior scalenes; thyrocervical trunk branches (dorsal scapular, transverse cervical, supraclavicular, and inferior thyroid arteries)

Lymphatic System

- Deep Jugular Chain: superior, middle, and inferior groups; jugulodigastric node is located at the junction of the posterior belly of the digastric muscle and the deep jugular chain; receive drainage from the **parotid**, **retropharyngeal**, **spinal accessory** (drain the upper retropharyngeal and parapharyngeal nodes), **superficial cervical** (drain the parotid, retroauricular, and occipital nodes), **paratracheal**, and **submandibular nodes**
- Jugular Trunk: drains the deep jugular drain at the root of the neck, drains into the internal jugular or subclavian vein (right) and the thoracic duct (left)

Evaluation of the Neck Mass

History

- Character of Neck Mass: onset, duration, and progression of growth, pain
- Contributing Factors: recent upper respiratory infection, sinus infection, otitis media, or other head and neck infection; exposure to pets and other animals; recent travel; exposure to tuberculosis; risk of malignancy (previous excision of skin or scalp lesions, family history of cancer, smoking and alcohol abuse, radiation therapy, other malignancies); recent trauma; immunodeficiency (risk of HIV, corticosteroids, uncontrolled diabetes); age (often infectious in children, higher risk of malignancy in adults)
- Associated Symptoms: fever, postnasal drip, rhinorrhea, sore throat, otalgia, night sweats, weight loss, malaise, dysphagia, hoarseness
- Think "KITTENS" for differential diagnosis (*see* Table 4–8)

Physical Exam

- Character of Neck Mass: size (normal hyperplastic nodes rarely exceed 2 cm), distribution, mobility, tenderness and fluctuance (infectious), consistency (firm, elastic, soft, compressible), solitary mass versus general cervical adenopathy, lesions and character of the overlying skin (eg, erythematous, blanching, vascular signs, fistulas, induration, radiodermatitis, necrotic)
- Physical Exam: thorough head and neck exam for primary malignancies (attention to nasopharynx, oral cavity, base of tongue, tonsilar fossa, nasal cavity, external ear canal, scalp, thyroid, and salivary glands); palpate other lymphatic sites (eg, inguinal, axillary, supraclavicular); palpate thyroid gland, liver, and spleen (lymphoma, mononucleosis); auscultation for vascular abnormalities

TABLE 4–8. Differential Diagnosis of the Neck Mass: KITTENS Method

(K) Congenital	Infectious & Iatrogenic	Toxins & Trauma	Endocrine	Neoplasms	Systemic
Branchial cleft cysts	Bacterial or viral lymphadenitis	Hematoma	Thymic cyst	Metastatic or regional malignancy	Granulomatous diseases
Cystic hygromas	Tuberculosis		Thyroid hyperplasia	Thyroid neoplasia	Laryngoceles
Teratomas and dermoid cysts	Cat scratch disease		Aberrant thyroid tissue	Lymphoma	Plunging ranula
Thyroglossal duct cyst	Syphilis		Parathyroid cyst	Hemangiomas	Kawasaki disease
External laryngoceles	Atypical mycobacteria			Salivary gland tumors	
	Persistent generalized lymphadenopathy			Vascular tumors	
	Mononucleosis			Neurogenic tumors	
	Sebaceous cyst			Lipomas	
	Deep inflammation or abscess				

Ancillary Tests

- **CT/MRI of Neck**: provides greater differentiation of abscess, neoplasms, vascular lesions, hematomas, or congenital abnormalities
- **Laboratory Evaluation**: complete blood count with differential, monospot, Purified Protein Derivative (PPD), HIV testing, cat-scratch antigen titers, toxoplasmosis titers, Epstein-Barr virus serology tests
- **Ultrasound**: identifies cystic masses, when combined with Doppler further defines vascular lesions
- **Fine-needle Aspirate (FNA)**: provides fluid for culture and sensitivity, indicated for nonresolving masses suspicious for malignancy (firm, large [>2 cm], nontender, asymmetric neck masses) without a known primary
- **Open Biopsy**: indicated for persistent idiopathic adenopathy or high suspicion of malignancy (if FNA is negative or nondeterminate and complete work-up does not reveal a primary site), prepare for possible completion of neck dissection if frozen section is positive
- **Angiography**: demonstrates primary vascular diseases
- **Panendoscopy**: direct laryngoscopy, esophagoscopy, and bronchoscopy may be considered to evaluate for a primary site for malignancy

Congenital Neck Masses

Branchial Cleft Anomalies

- Pathophysiology: developmental alterations of the branchial apparatus results in cysts (no opening), sinuses (single opening to skin or digestive tract), or fistulas (opening to skin and digestive tract)
- SSx: neck mass in anterior neck (**anterior** to SCM, deep to platysma); may have an associated subcutaneous palpable cord; fistulas and sinuses may express mucoid discharge; secondary infections cause periodic fluctuation of size, tenderness, and purulent drainage
- Dx: CT with contrast (may consider injecting contrast into fistula), laryngoscopy to visualize internal opening
- Histopathology: lined by squamous epithelium

First Branchial Cleft Cyst

- SSx: usually presents as a preauricular cyst (may also be infra- or postauricular), opening within external auditory canal
- Types
  1. **Type I**: ectodermal elements only; duplicated external auditory canal; typically begin periauricularly, pass lateral (superior) to facial nerve, parallel the external auditory canal, and end as a blind sac near the mesotympanum
  2. **Type 2**: more common; ectodermal and mesodermal elements; duplicated membranous external auditory canal and pinna; presents

near the angle of the mandible, passes lateral or medial to facial nerve, may end near or into the external auditory canal

- Rx: full excision after resolution of infection (risk of **facial nerve injury**), may need superficial parotidectomy, avoid incision and drainage

Second Branchial Cleft Cyst

- SSx: cyst along anterior border of the SCM (most common branchial cleft cyst)
- Fistula Pathway: external opening at lower anterior neck → along carotid sheath → **between** external and internal carotid arteries → **over** (lateral to) hypoglossal and glossopharyngeal nerves → internal opening at **middle constrictors** or in **tonsillar fossa**
- the course of the second branchial cleft cyst runs deep to second arch derivatives and superficial to third arch derivatives
- Rx: full excision after resolution of infection, avoid incision and drainage

Third Branchial Cleft Cyst

- SSx: cyst in lower anterior neck (less common)
- Fistula Pathway: external opening at lower anterior neck → **over** (superficial to) vagus nerve and common carotid artery → **over** hypoglossal nerve → **inferior** to glossopharyngeal nerve → pierces thyrohyoid membrane → internal opening at **upper pyriform sinus**
- Rx: full excision after resolution of infection, avoid incision and drainage

Thyroglossal Duct Cyst

- Pathophysiology: failure of complete obliteration of thyroglossal duct (created from tract of the thyroid descent from the foramen cecum down to midline neck)
- SSx: midline neck mass with cystic and solid components, elevates with tongue protrusion (attached to hyoid bone), typically inferior to hyoid bone and superior to thyroid gland, may have fibrous cord, dysphagia, globus sensation
- Histopathology: lined with respiratory and squamous epithelium
- Complications: rare malignant potential, secondary infection
- Rx: **Sistrunk procedure** (excision of cyst and tract with cuff of tongue base and mid-portion of hyoid bone, 3% recurrence)

Cystic Hygromas (Lymphangiomas)

- Pathophysiology: abnormal development or obstruction of jugular lymphatics

- spontaneous remission is not common
- SSx: soft, painless, multiloculated, compressible neck mass (most common located in posterior triangle); presents at birth; transilluminates; may cause stridor, dysphagia, torticollis, cyanosis, or parotid swelling
- Dx: CT or MRI of neck
- Complications: infection, compressive effects may cause respiratory compromise or dysphagia, facial deformity
- Rx: excision without violating important structures, difficult to excise since lesions do not follow typical anatomical planes, high rate of recurrence

Thymic Cysts

- Pathophysiology: remnant of third pharyngeal pouch between angle of mandible to midline neck
- SSx: midline neck mass, lower neck
- Dx: biopsy, serum calcium (associated parathyroid disorders, DiGeorge's syndrome), CT/MRI
- Rx: excision (may require thoracic surgery)

Dermoid Cysts and Teratomas

- Pathophysiology: derived from pleuripotent embryonal crest cells
- Types
  1. **Teratomas**: composed of all 3 embryologic layers
  2. **Dermoid Cyst**: ectodermal and mesodermal elements only, most common type
  3. **Teratomoma**: differentiated to organ structure (usually fatal)
  4. **Epignathi**: differentiated to body parts (usually fatal)
- SSx: soft midline neck mass, may be associated with tufts of hair
- Dx: biopsy
- Complications: rare malignant potential
- Rx: excision

Congenital Torticollis

- Pathophysiology: intrauterine or birth trauma causing muscle injury, hematoma, and resultant fibrosis (typically of the sternocleidomastoid muscle)
- SSx: head and neck are held to the diseased side, chin toward the healthy side, firm thickened mass confined to sternocleidomastoid muscle (may be tender)
- Dx: clinical history and exam
- Rx: physical therapy, observation

Infectious Neck Masses

Bacterial Cervical Adenitis

- Pathogens: most commonly from group A streptococci and *S. aureus*
- SSx: tender, mobile cervical mass, associated with constitutional symptoms (malaise, fever), initially may present with a single discrete node before developing generalized adenopathy
- Dx: clinical exam, aspiration for culture and sensitivity
- Complications: septic shock
- Rx: incision and drainage, antibiotic regimen

Cat-Scratch Disease

- Pathogen: cat-scratch bacillus
- SSx: cutaneous lesions at primary site, tender cervical adenopathy (later becomes painless), mild fever and malaise, pustulous lesions tend to ulcerate (risk of fistula formation)
- Dx: culture (Warthin-Starry stain), cat-scratch antigen test, history of cat exposure
- Histopathology: intracellular, gram-negative bacillus, **Warthin-Starry stain**
- Rx: observation with supportive care (self-limiting), avoid incision and drainage to prevent sinus formation (may consider aspiration)

Atypical Mycobacteria

- Pathogens: *Mycobacterium avium, M. scrofulaceum, M. intracellulare*
- typically less virulent than *M. tuberculosis;* however, less responsive to antituberculosis medications
- may colonize respiratory tract
- Risks: children, immunocompromised hosts, history of foreign travel
- SSx: unilateral cervical adenopathy (adhesive to overlying skin), corneal ulceration (most common head and neck manifestation)
- Dx: acid-fast stain and culture, culture requires 2–4 weeks for growth, tuberculin skin testing is often negative
- Rx: complete excision (avoid incision and drainage), antibiotics (may consider rifampin to reduce the bulk of the infected node prior to excision)

Other Specific Lymphadenitis

- **Tuberculosis**: typically presents as a postprimary disease, pulmonary tuberculosis must be examined, *see also* pp. 209–210
- **Syphilis**: may present in primary or secondary disease; *see also* pp. 208–209 for management

- **Toxoplasmosis**: presents as an influenza-like illness in adults; congenital form presents with hydrocephalus, chorioretinitis, and intracerebral calcifications; diagnosed with serum titers and lymph node biopsy

Nonspecific Lymphadenitis

- Pathophysiology: reactive adenitis typically secondary to a nasopharyngeal infection, may occur from any infection of the head and neck
- primary infection may have resolved with persistent, enlarged cervical lymph nodes
- SSx: painful swelling of the cervical lymph nodes, induration, fluctuating size or tenderness
- Dx: clinical history and exam, must undergo a careful exam for the primary infection, open biopsy if suspect malignancy
- Rx: address primary infection; if no primary infection site may consider a trial of broad-spectrum antibiotics versus close observation

Other Neck Masses

Kawasaki Disease (Mucocutaneous Lymph Node Syndrome)

- Pathophysiology: vasculitis of multiple organ systems
- SSx: fever, cervical lymphadenopathy, conjunctivitis, red and dry blistering lips, desquamating rash, cervical lymphadenopathy, "strawberry tongue" (prominent papillae)
- Dx: clinical history and exam
- Complications: associated with coronary aneurysms (acute myocardial infarction), vasculitis
- Rx: supportive therapy, high-dose gamma globulins and aspirin for complications (coronary aneurysms)

Plunging Ranula (*see* p. 73)

External Laryngoceles (*see* pp. 112–113)

Cervical Adenopathy in the HIV Patient

Introduction

- idiopathic follicular hyperplasia is the most common cause of cervical adenopathy in the HIV-infected patient

- higher risk for lymphoma, mycobacterium, carcinoma, tuberculosis
- open biopsy should be reserved for highly suspicious lesions (failed antibiotic trial, enlarging, mediastinal adenopathy, suspicious FNA, >2 cm nodes, asymmetric lesions)

Persistent Generalized Lymphadenopathy (PGL)

- Pathophysiology: idiopathic lymphadenopathy, may be a direct effect of the HIV
- cervical adenopathy is the third most common lymphatic site (axillary and inguinal more common)
- SSx: typically asymptomatic adenopathy
- Dx: based on clinical history and exam, neoplastic and infectious causes must be ruled out, must have adenopathy of 2 or more sites (extra-inguinal) for greater than 3 months in the HIV patient
- Histopathology Patterns: follicular hyperplasia, follicular involution (small follicles), and lymphoid depleted (no follicles)
- Rx: observation

NECK PLANES, SPACES, AND INFECTION

Cervical Fascial Planes
(Fig. 4–3)

Superficial Cervical Fascia

- condensed sheath of connective tissue under skin from the zygomatic process to the thorax and axilla
- Contents: platysma and facial muscles of expression

Deep Cervical Fascia

Superficial Layer of the Deep Cervical Fascia (Investing Fascia)

- forms stylomandibular ligament
- Contents: trapezius, SCM, and masseter muscles; submandibular and parotid glands

Middle Layer of the Deep Cervical Fascia (Visceral Fascia)

- forms midline raphe, pteryogomandibular raphe, and pretracheal fascia
- Muscular Division Contents: strap muscles
- Visceral Division Contents: pharynx, larynx, trachea, esophagus, thyroid, constrictor and buccinator muscles (buccopharyngeal fascia)

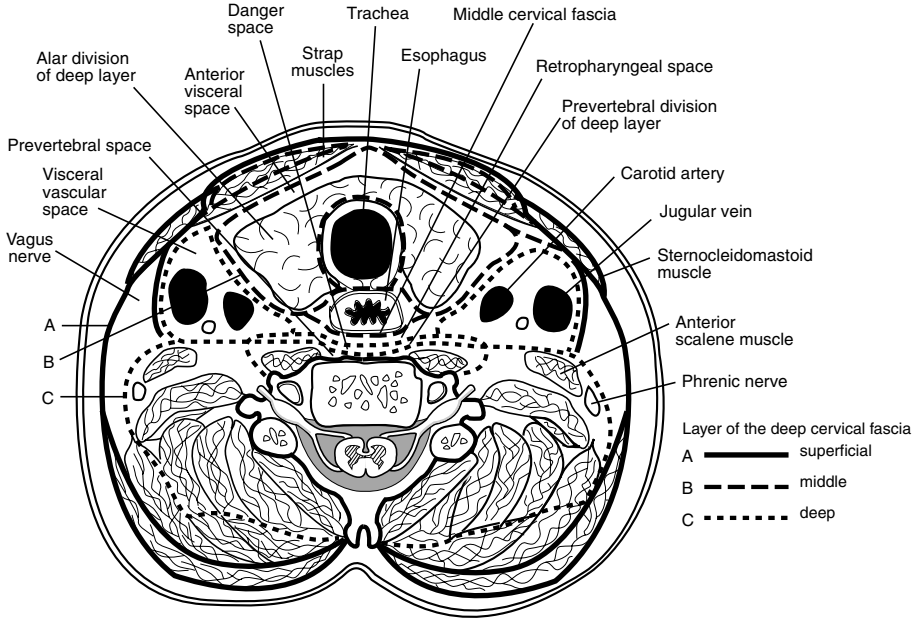

Figure 4–3. Cross-section anatomy of the neck demonstrating the cervical fascial planes. Adapted with permission from Rosse C., Gaddum-Rosse P. *Hollinshead's Textbook of Anatomy,* 5th ed, p. 710. Philadelphia, PA: Lippincott Williams & Wilkins; 1997.

Deep Layer of the Deep Cervical Fascia (Prevertebral)

- divided into prevertebral and alar layers
- Contents: paraspinus muscles and cervical vertebrae

Carotid Sheath

- made up of all layers of deep cervical fascia
- attaches to jugular and carotid foramina at the base of skull
- Contents: carotid artery, internal jugular vein, and vagus nerve

Neck Space Infections

Organisms and Management

- Sources: odontogenic (most common), tonsils (most common pediatric source), trauma, upper respiratory infections, salivary glands, intravenous drug injections
- Pathogens: usually wide array of organisms including aerobic and anaerobic, must also consider *Actinomycosis* which may cross fascial planes (*see,* pp. 174–175)
- Dx: lateral and AP neck radiographs may be consider for screening (examine for soft tissue swelling in the posterior pharyngeal region, abnormal if space >50% the width of the vertebral body), CT/MRI of neck with contrast media, chest x-ray
- Rx: secure airway (tracheotomy), aggressive antibiotic therapy, local drainage (culture and sensitivity), extraction of tooth (if source), debridement of osteomyelitic bone

Necrotizing Fascitis of the Head and Neck

- Pathophysiology: aggressive polymicrobial infection of the subcutaneous tissue and fascia typically from trauma or dental infections
- Pathogens: *Staphylococci,* hemolytic *Streptococci,* and gram-negative rods (myonecrosis occurs from superinfections from anaerobes and other gram negatives)
- SSx: rapid, severe necrosis of the skin, fascia, and soft tissues, may form crepitus, sepsis
- Dx: CT reveals subcutaneous emphysema, necrotic soft tissue
- Complications: rapid spread to mediastinum and abdomen, systemic infection (sepsis), death
- Rx: aggressive debridement, fasciotomy, broad-spectrum parenteral antibiotics, consider hyperbaric oxygen

Parapharyngeal Space (Lateral Pharyngeal Space, Pharyngomaxillary Space)

- Boundaries: cone-shaped with base at base of skull and apex at lesser cornu of hyoid bone, bounded by pharynx, parotid, mandible, and pterygoid muscles
- Compartments
  1. **Prestyloid**: contains internal maxillary artery; fat; inferior alveolar, lingual, and auriculotemporal nerves
  2. **Poststyloid**: neurovascular bundle (carotid artery, internal jugular vein, sympathetic chain, CN IX, X, and XI)
- Source of Infections: odontogenic, tonsils, pharynx, nasopharynx, and parotid gland
- SSx: pronounced trismus, fever, muffled voice, intraoral bulge, dysphagia
- Complications: aspiration, airway compromise, cranial nerve palsies, septic thrombophlebitis, carotid blowout, endocarditis, extension into adjacent spaces
- Rx: external drainage, aggressive antibiotics, airway management

Peritonsillar Space (*see* pp. 161–162)

Pterygopalatine Fossa (Pteryogomaxillary Space)

- Boundaries: pyramidal-shaped space below the apex of the orbit
- Contents: maxillary division of the trigeminal nerve, vidian nerve, sphenopalatine nerve, lesser and greater palatine nerves, sphenopalatine ganglion, and internal maxillary artery
- Source of Infections: maxillary teeth, osteomyelitis
- SSx: gingival edema, facial cellulitis, trismus, ocular manifestations, extension into infratemporal fossa
- Rx: drainage procedure from a Caldwell-Luc or alveobuccal sulcus approach, antibiotic regimen

Masticator Space

- Compartments
  1. **Masseteric**: space between masseter muscle and ramus of the mandible
  2. **Pterygoid**: space between pterygoid muscles and ramus of the mandible
  3. **Superficial Temporal**: space between superficial temporal fascia and temporalis muscle
  4. **Deep Temporal**: space between temporal fascia and temporal bone

- Contents: muscles of mastication, internal maxillary artery, mandibular nerve
- Source of Infections: molars (third molar most common)
- SSx: edema over posterior ramus, trismus
- Complications: osteomyelitis of the mandible, extension into neck spaces
- Rx: incision and drainage procedure, antibiotics

Submandibular and Sublingual Space

- Compartments
  1. **Sublingual**: between floor of mouth and mylohoid muscle; contains sublingual gland, CN XII, Wharton's duct; communicates with opposite space and submental space
  2. **Submandibular**: between body of mandible and mylohoid, hyoglossus, and styloglossus muscles; contains submandibular gland, lingual nerve, and facial artery; communicates with sublingual and pharyngeal spaces
- Source of Infections: odontogenic, submandibular gland, paranasal, pharynx
- SSx: salivary gland tenderness, odynophagia
- Complications: Ludwig's angina (*see below*)
- Rx: external or internal drainage, aggressive antibiotics, airway management

Ludwig's Angina

- Pathophysiology: bilateral cellulitis of submandibular and sublingual spaces
- SSx: "wooden" floor of mouth, neck swelling and induration, drooling, respiratory distress, swollen tongue, dysphagia, trismus, may rapidly progress to airway compromise
- Complications: rapid respiratory compromise, sepsis
- Rx: early local tracheotomy, external incision (usually straw-colored weeping but no true abscess fluid), aggressive antibiotic therapy

Parotid Space

- formed by the splitting of the superficial layer of the deep cervical fascia
- Contents: parotid gland, facial nerve, posterior facial vein, lymphatics, external carotid artery
- Source of Infections: parotid
- SSx: tenderness over parotid region
- Complications: extension into parapharyngeal space
- Rx: treat for parotiditis (hydration, sialogogues, oral hygiene, antibiotics), may require aspiration or incisional drainage

Buccal Space

- Boundaries: buccinator muscle, cheek, pterygomandibular raphe, zygomatic arch, and inferior mandible
- Source of Infections: odontogenic
- SSx: buccal swelling that may extend up to eyelid (preseptal) and orbicularis oris
- Complications: cavernous sinus thrombosis (from angular vessels), intracranial infections, extension into other spaces and orbit
- Rx: urgent draining (external approach), aggressive antibiotic regimen

Carotid Sheath Space

- Boundaries and Contents: extends from base of skull to thoracic inlet; contains carotid artery, internal jugular vein, and vagus nerve
- Source of Infections: extension from adjacent fascial planes
- SSx: torticollis (toward uninvolved side)
- Complications: shock, carotid blow-out, endocarditis, cavernous sinus thrombosis
- Rx: surgical draining, aggressive antibiotic regimen

Visceral Space

- Boundaries: between pharyngeal constrictors muscles and alar fascia, extends from skull base to mediastinum
- Contents: pharynx, esophagus, larynx, trachea, thyroid
- Source of Infections: typically from perforations of the anterior esophageal wall (instrumentation, trauma)
- SSx: dysphagia, hoarseness, emphysema, respiratory compromise, subcutaneous emphysema
- Complications: mediastinitis, sepsis, pneumonia, laryngeal edema
- Rx: nothing by mouth, external drainage, aggressive antibiotic regimen, possible tracheotomy

Retropharyngeal Space (Retrovisceral Space)

- infection more common in children
- Boundaries: between pharyngeal constrictors muscles and alar fascia, extends from skull base to **mediastinum**
- Contents: retropharyngeal lymphatics that drain nose, nasopharynx, and paranasal sinuses, adenoids, sinus, nasopharynx
- SSx: odynophagia, "hot potato" voice, drooling, stiff neck, stridor, spiking fevers
- Source of Infections: tonsils, paranasal sinus, and nasopharynx

- Complications: mediastinitis (50% mortality), respiratory distress, ruptured abscess (aspiration pneumonia), spread into danger and prevertebral space
- Rx: urgent draining (external versus internal approach), aggressive antibiotic regimen

Danger Space

- Boundaries: between alar and prevertebral fascia, extends from skull base to **diaphragm**
- Contents: loose areolar tissue (spreads fast)
- SSx: same as retropharyngeal space infections
- Complications: same although may spread into abdominal cavity
- Rx: same as retropharyngeal space infections

Prevertebral Space

- Boundaries: deep to prevertebral fascia, extends from skull base to **coccyx**
- Contents: longus colli muscle
- SSx: same as retropharyngeal and danger space infections
- **Pott's abscess**: tuberculous osteomyelitis
- Complications: same as retropharyngeal and danger space infections, osteomyelitis of the spine
- Rx: same as retropharyngeal and danger space infections

HEAD AND NECK MANIFESTATIONS OF SYSTEMIC DISEASES

Noninfectious Granulomatous Diseases

Histocytosis X (Reticuloendotheliosis)

- Pathophysiology: granulomatous disease of unknown etiology, manifests as a proliferation of histiocytes
- Histopathology: sheets of polygonal histiocytes, Birbeck granules ("zipper" pattern)

Eosinophilic Granuloma

- localized form
- presents in children and young adults
- excellent prognosis
- SSx: **monostotic or polyostotic osteolytic bone lesions** (predilection for temporal and frontal bones, ribs, and long bones), proptosis (sphenoid involvement), acute mastoiditis, middle ear granulation tissue, tympanic membrane perforations, facial paralysis

- Rx: surgical excision, radiation therapy reserved for recurrence, inoperable sites, and high risk patients

Hand-Schüller-Christian Disease

- chronic disseminated form
- presents in children and young adults (rare in the elderly)
- 30% mortality (higher risk with heart, lung, brain involvement)
- SSx: **polyostotic osteolytic lesions** (skull), exophthalmos, diabetes insipidus (from erosion from sphenoid into sella turcica), facial paralysis, external auditory canal polypoid lesions
- Rx: radiation therapy, chemotherapy, corticosteroids, Medical Oncologists consult

Letterer-Siwe Disease

- acute disseminated form
- uniformly fatal
- presents in children <3 years old
- SSx: fever, proptosis, splenomegaly, hepatomegally, exfoliative dermatitis, thrombocytopenia
- Rx: radiation therapy and chemotherapy, Medical Oncologists consult

Midline Destructive Syndromes

Angiocentric Immunoproliferative Lesions (Angiocentric T-cell Lymphoma, Polymorphic Reticulosis, Lymphomatoid Granulomatosis)

- Pathophysiology: originates from T-cells, form of **extranodal Non-Hodgkin's Lymphoma**, subtypes based on the β-chain of the T-cell receptor, forms perivascular infiltrates (angiocentric, **not a true vasculitis**)
- SSx
  1. pansinusitis and nasal obstruction (most common initial finding)
  2. often with midface (paranasal, oral, orbital), pulmonary, renal, GI, or CNS involvement
  3. cutaneous involvement (maculopapular rash, ulcerative cutaneous lesions)
- Stages
  1. **Prodromal**: clear rhinorrhea, nasal obstruction
  2. **Ulcerative** (**Active**): purulent rhinorrhea, ulceration and septal perforations (epistaxis), destruction of osteocartilaginous tissue
  3. **Terminal**: malaise, fever, sloughed tissues, death
- Dx: biopsy, CT/MRI of paranasal sinuses
- Histopathology: sheets of **atypical** polymorphonuclear cells, no granuloma, no palisading histiocytes

- Rx: radiation therapy for local disease, cyclophosphamide and prednisone for multiregional disease

Idiopathic Midline Destructive Disease

- Pathophysiology: idiopathic midline destructive disease, localized to the head and neck, more aggressive
- SSx: ulcerative lesions of the nose or sinus, pansinusitis, nasal obstruction
- Dx: biopsy, CT/MRI of paranasal sinuses
- Histopathology: sheets of **typical** polymorphonuclear cells, no granuloma, no vasculitis (pseudovasculitis)
- Rx: radiation therapy for local disease, cyclophosphamide and prednisone for multiregional disease

Wegener's Granulomatosis

True vasculitis (*see below*), most common midline granuloma syndrome

Sarcoidosis

- Pathophysiology: idiopathic systemic granulomatous disease of unknown etiology, mononuclear cells accumulate in affected organs followed by formation of granulomas, may lead to irreversible fibrosis of tissue
- more common in African-American women
- Histopathology: **noncaseating granulomas**, accumulation of T-cells, mononuclear phagocytes, derangement of normal tissue architecture
- Complications: progressive interstitial lung disease, blindness (progression of uveitis), airway obstruction (rare)
- Rx: corticosteroids for significant exacerbations, do not treat asymptomatic lesions, may require surgical excision of obstructing laryngeal lesions

Symptoms

- 40% asymptomatic (incidental chest x-ray finding)
- pulmonary involvement (88%, cough, hilar adenopathy, dyspnea)
- cervical adenopathy (25–50%, most common H&N presentation)
- Salivary Glands: parotid mass, uveopartoid fever or Heerfordt's disease (*see,* p. 70)
- Laryngeal: supraglottic submucosal mass (epiglottis most common), TVF paralysis
- Other H&N: uveitis, nasal mass, orbital mass, nasal perforations
- Other Systemic: cutaneous lesions (erythema nodosum, rashes), "Darrier Rousey" nodule (subcutaneous lesion), hepatic and renal involvement, splenomegaly, cardiac (arrhythmias), bone lesions, neuropathies, fever, weight loss

Evaluation

- Dx: biopsy of lung or affected organ
- chest x-ray, anergy skin tests, ECG
- CBC, serum protein electrophoresis (hypergammaglobulinemia), LFT, electrolytes (hypercalcemia), ESR, ACE level

Pyogenic Granuloma (Lobular Capillary Hemangioma)

- Risks: young males, postpubescent females, pregnancy (hormonally related)
- SSx: painless, friable, ulcerated, or polypoid lesion on lips (40%), nasal cavity (30%, epistaxis), tongue (20%), or oral mucosa (15%), difficult to distinguish from hemangiomas
- Dx: biopsy
- Histopathology: circumscribed capillaries arranged in lobules
- Rx: surgical excision

Necrotizing Sialometaplasia

- Pathophysiology: infarct of mucosal salivary gland tissue resulting in a self-healing inflammatory process
- Risks: smokers
- SSx: deep ulcerated lesions in salivary tissue (most commonly between the junction of the hard and soft palate), usually asymptomatic (painless)
- Dx: biopsy (**may be mistaken for squamous cell carcinoma in the oral cavity**)
- Histopathology: lobular necrosis, pseudoepitheliomatous hyperplasia, no architectural destruction
- Rx: oral hygiene and observation (resolves in a month)

Infectious Granulomatous Diseases

Syphilis

- Pathophysiology: chronic infection caused from the spirochete *Treponema pallidum*, usually as a sexually transmitted disease or from maternal transmission
- Dx: nonspecific screening RPR or VDRL (Venereal Disease Research Laboratories), specific FTA-ABS (Fluorescent Treponemal Antibody-Absorption Test), dark field microscopy, Warthin-Starry tissue staining
- Histopathology: mononuclear infiltrate, obliterative arteritis, hydrops, gummas and osteolytic lesions in optic capsule

- Rx: penicillin, ampicillin, tetracycline, erythromycin, steroids for otologic involvement

Early Stages

- **Primary**: chancre at inoculation site (penis, cervix, anal canal), lymphadenopathy
- **Secondary**: "The Great Imitator," highly contagious, general malaise and fever, arthralgia, hepatosplenomegally, genital condyloma lata, nephrotic syndrome, "mucous patches"

Late Stages

- **Latent**: (1/3) asymptomatic phase (may have return to mucocutaneous lesions)
- **Tertiary**: (1/3) noninfectious stage, may occur years after initial infection, slowly progressive, neurosyphilis, aortic involvement, gummas
- **Remission**: (1/3) resolution

H&N Symptoms

- Lymphadenopathy: generalized cervical adenopathy
- Laryngeal: laryngitis with mild edema of the larynx, vocal fold paralysis, dysphagia
- Oral Cavity: chancre, granulomatous infiltration of tongue, palate
- Otologic: abrupt profound SNHL, Ménière's symptoms, interstitial keratitis, TM perforation, gummas of the temporal bone
- Nasal: saddle nose deformity, rhinitis, osseous and cartilaginous destruction, septal perforation
- Other: patchy alopecia, cutaneous lesions, cranial nerve involvement

Congenital Syphilis

- often fatal
- Early SSx: rhinitis, cutaneous lesions (bullae, papulosquamous lesions, condyloma lata), hepatosplenomegally, jaundice, lymphadenopathy
- Late SSx: labyrinthitis, Clutton's joints (bilateral knee effusion), neurosyphilis, "mulberry molars," frontal bossing, mental retardation, saber chins, saddle nose deformity (from epiphysitis)
- Hutchinson's Triad: abnormal central incisors (Hutchinson's teeth), interstitial keratitis, deafness

Tuberculosis

- Pathophysiology: inhalation of acid-fast bacilli, *Mycobacterium tuberculosis*, into lungs

- Risks: immunocompromised (50% of HIV population), health care workers, immigrants, elderly, poor
- Dx: skin test (positive if any reaction in HIV patients, >5 mm in household contacts, >10 mm in health care workers, >15 mm in low risk population), sputum culture, chest x-ray, biopsy
- Rx
  1. isoniazid (INH) and rifampin for active disease (for drug resistance consider adding ethambutol, pyrazinamide)
  2. prophylaxis INH for immunosuppressed patients with exposure or positive PPD, history of TB (without active disease) and without proper previous treatment, positive PPD with inactive disease, or household contacts of patients with active disease

Symptoms

- **Primary**: asymptomatic; lower lobe lesion, **Ghon complex** (calcified lung lesion and draining lymph node)
- **Secondary**: reinfection; weight loss, fever, night sweats, nonproductive cough; endogenous, caseating granulomatous tubercles, apical segments lung involvement
- H&N manifestations usually secondary from pulmonary source
- Cervical Lymphadenopathy: bilateral, anterior and posterior, firm, nontender, most common H&N manifestation
- Larynx: granulation and ulcerative tissue in **posterior glottis** (posterior interarytenoids, laryngeal surface of epiglottis, vocal folds)
- Otologic: painless, odorless, watery otorrhea, multiple TM perforations
- Salivary Glands: diffuse glandular involvement
- Oral Cavity: ulcerative lesions
- Ocular: conjunctivitis, uveitis

Fungal Granulomatous Disease

- **Aspergillosis**: allergic, noninvasive, or invasive forms (*see* Rhinology and Paranasal Sinus: Sinusitis, p. 38)
- **Rhinosporidiosis**: fungal sporangium, paranasal involvement (*see* Rhinology and Paranasal Sinus: Nonallergenic Rhinitis, pp. 34–35)
- **Phycomycosis** (**Mucor/Rhizopus**): *see* Rhinology and Paranasal Sinus: Sinusitis, p. 38

Histoplasmosis

- Pathophysiology: airborne transmission (endemic to Missouri and Ohio River Valley, found in bird feces) causes chronic pulmonary infection, may disseminate

- H&N SSx: (disseminated form) painful, ulcerative, granulomatous lesions (heaped edges) of pharynx, larynx, epiglottis, oral cavity (tongue, lip, oral mucosa)
- Histopathology: intracellular organisms
- Dx: culture on Sabouraud's medium, skin test, complement fixation test, latex agglutination
- Rx: amphotericin B

Blastomycosis

- Pathophysiology: airborne transmission (endemic to Central America and the Midwest) causes chronic pulmonary infection, may disseminate
- H&N SSx: (disseminated form) oropharyngeal and verrucous laryngeal lesions
- Histopathology: pseudoepitheliomatous hyperplasia, intraepithelial microabscess, single bifringent broad based bud ("figure 8" formation)
- Dx: culture on Sabouraud's medium, skin test
- Rx: amphotericin B

Coccidiomycosis

- Desert and Latin America, "Valley Fever"
- H&N SSx: nodules or erosions of epiglottis, mucous membranes, thyroid, trachea, and salivary glands
- Histopathology: "sac with bugs"
- Dx: skin test, complement fixation
- Rx: amphotericin B

Cryptococcosis

- Risks: immunocompromised, pigeon carriers
- H&N SSx: meningitis (hearing loss), membranous nasopharyngitis
- Dx: culture (capsule seen with India ink stain), fluorescent antibody staining
- Rx: amphotericin B

Other Bacterial Granulomatous Disease

- **Cat-Scratch Disease**: intracellular, pleomorphic, gram-negative bacilli, cervical and general adenopathy, cutaneous lesions (*see* p. 197)
- **Brucellosis**: aerobic, gram-negative bacilli, acquired from products from butter, pigs, cattle, goats; presents with flu-like symptoms; diagnose with serum titers; Rx: tetracycline
- **Rhinoscleroma**: *Klebsiella rhinoscleromatis,* affects paranasal sinus (*see* p. 34)

- **Leprosy** (**Hansen's Disease**): *Mycobacterium leprae*, cutaneous lesions, may present with ulcerative lesions in the larynx and oral mucosa, nasal collapse, anesthetic plaques, lymphadenopathy; Rx: chronic dapsone (Diaminodiphenylsulfone), quinolones, consider steroids
- **Atypical Myobacteria**: presents most commonly with corneal ulceration and cervical adenopathy (*see* p. 197)
- **Actinomycosis**: branching anaerobic or microaerophilic gram-negative bacteria, may present as a mass anywhere in the H&N (*see* pp. 174–175)
- **Nocardiosis**: subtype of *Actinomycosis,* soil saprophyte, primary lung disease with hematogenous spread, Rx: sulfa antibiotics may require I&D

Connective Tissue Diseases

Lupus Erythematosis

- Pathophysiology: idiopathic autoimmune vasculitis, causes damage by deposition of antibodies and immune complexes
- more common in African-Americans and women
- Risks: pregnancy, genetic disposition (HLA DR2 & DR3)
- Rx: no cure, control inflammation with oral and topical steroids, NSAIDs, salicylates, may consider immunosuppressives and antimalarials, avoid sun exposure

Types

1. **Discoid** (DLE): women (3rd and 4th decade), oral lesions (20–25%) and cutaneous lesions (elevated erythematous plaques, hypopigmented edges, alopecia, leaves a scar), no visceral involvement, least aggressive form, ANA(−), LE cell test(−)
2. **Subacute Cutaneous** (SCLE): mild systemic form, oral and cutaneous lesions (papulosquamous lesions, no scar), ANA (equivocal), SSA and SSB(+/−)
3. **Systemic** (SLE): occasional oral lesions, butterfly rash, multiple visceral organs involvement, ANA(+) (98%), LE cell test(+), SSA(+) and SSB(+)

Diagnosis

- Nonspecific Markers: ESR, ANA, SS-A (anti-Ro) & SS-B (anti-La)
- Specific Markers: anti-DS DNA and Sm Ag
- Direct Immunofluorescence: IgG, C3 positive in subepithelial layer
- LE Cell Phenomenon: serum from patient is added to buffy coat of normal blood, reaction is a rosette of neutrophils surrounding a pale nuclear lymphocytic mass
- ECG, UA, CBC

Symptoms

- H&N SSx: mucosal oral ulcerations with an erythematous halo, cutaneous lesions (*as above*), ulcerated nasal septum (nasal perforation), hoarseness from thickened vocal cords and cricoarytenoid joint arthritis, acute enlargement of parotid, cranial neuropathies, SNHL (autoimmune inner ear disease), nonspecific lymphadenopathy
- Other SSx: malar rash, fever, malaise, weight loss, nonerosive polyarthritis, pleurositis, proliferative glomerulonephritis, anemia, Raynaud's phenomenon, diffuse interstitial pulmonary fibrosis, pericarditis, endocarditis

Rheumatoid Arthritis

- Pathophysiology: idiopathic autoimmune disease causing inflammation of synovial joints
- Dx: clinical history, rheumatoid factor (titer >1:64), chest x-ray, ESR, complement levels, synovial fluid analysis
- Rx: ASA, NSAIDs, corticosteroids, antimetabolites

Symptoms

- H&N SSx: TMJ (50%), hoarseness (cricoarytenoid joint alkylosis), ossicular joints (CHL)
- symmetrical polyarthritis arthritis (MCP and PIP joints common, morning stiffness), joint erosion, cartilaginous destruction, ulnar deviation
- rheumatoid nodules (subcutaneous nodules), visceral nodules
- rheumatoid vasculitis
- sicca syndrome

Mixed Connective Tissue Disease

- Pathophysiology: syndrome of unknown etiology that causes a combination of features of SLE, RA, polymyositis, and scleroderma
- SSx: lupus erythematous cutaneous and oral manifestations, esophageal dysmotility, polyarthralgia, Raynaud's phenomenon, visceral disease (pulmonary, renal, CNS)
- Dx and Rx: similar to SLE and scleroderma, hypogammaglobulinemia, high titer of ribonucleoprotein antibody

Other Connective Tissue Disorders

- **Scleroderma (Progressive Systemic Sclerosis)** (*see* p. 151)
- **Polymyositis and Dermatomyositis** (*see* p. 151)
- **Sjögren's Disease** (*see* pp. 71–72)
- **Relapsing Polychondritis** (*see* p. 324)

Vasculitis

Wegener's Granulomatosis

- Pathophysiology: idiopathic granulomatous vasculitis of upper and lower respiratory tract with glomerulonephritis (may be autoimmune)
- Types: limited (no renal involvement), systemic (pulmonary and renal involvement)
- Histopathology: **necrotizing granulomas** (with multinucleated giant cells) with **vasculitis** of upper and lower respiratory tract
- Rx: corticosteroids for initial control followed by cyclophosphamide, azathioprine, or methotrexate; may consider prophylaxis trimethroprim-sulfamethoxazole; nasal hypertonic saline irrigations and nasal debridement for local therapy

Systemic Symptoms

- Pulmonary: **universal**, hemoptysis, cough, dyspnea
- Nonspecific Focal Glomerulonephritis: hematuria, urine blood casts (renal involvement accounts for most deaths)
- Other: systemic fever, nausea, malaise, night sweats, arthralgia, myalgia

Head and Neck Symptoms

- Paranasal: recurrent and chronic sinusitis (60–80%, most common H&N symptom), nasal deformity, septal perforation, nasal obstruction, epistaxis
- Laryngeal: ulcerative lesions (25%), subglottic stenosis (<10%)
- Otologic: CHL, serous chronic otitis media (20–25%)
- Ocular: uveitis, keratitis (40%)
- Oral: gingival hyperplasia, gingivitis

Evaluation

- **cANCA** (**antineutrophil cytoplasmic autoantibodies**): suggests active disease, (cANCA + neutrophil Ab = **86%** specific)
- Pulmonary or Nasal Biopsy: pulmonary highest yield, nasal biopsy often obscured with acute inflammatory cells
- Renal Biopsy: glomerulonephritis
- CBC, ESR, SPEP, Bun/Creatinine, VDRL, U/A, chest x-ray, autoimmune panel, smooth muscle and neutrophil cytoplasm antibodies, sinus films

Giant Cell (Temporal) Arteritis

- most common vasculitides
- Pathophysiology: focal granulomatous inflammation of medium and small arteries
- Risks: elderly

- SSx: headache (most common initial symptom), tenderness over scalp, jaw and tongue claudication
- Dx: elevated ESR, biopsy temporal artery (up to 40% false negative rate, if negative biopsy contralateral side)
- Complications: intracranial involvement resulting in blindness (1/3 of untreated patients), cranial nerve defects, psychosis, or vertebrobasilar insufficiency
- Rx: long-term corticosteroids

Behçet's Syndrome (*see* Oral Cavity Lesions, pp. 177–178)

CHAPTER

5

Head and Neck Cancer

R. Pasha, George H. Yoo, and John R. Jacobs

continued

NOTE: salivary gland, temporal bone, thyroid, parathyroid, esophageal, and odontogenic tumors are reviewed in other chapters

INTRODUCTION TO HEAD AND NECK CANCER

Evaluation of the Head and Neck Cancer Patient

History

- Otological: persistent otalgia (referred pain with normal otologic exam), hearing loss, aural fullness, pulsatile tinnitus
- Nasal/Paranasal/Nasopharynx: recurrent epistaxis, unilateral nasal obstruction, persistent rhinorrhea or sinusitis
- Oral/Pharyngeal: persistent sore throat (>3 weeks), odynophagia, dysphagia, trismus, presence of nonhealing ulcers, halitosis, numbness in the lower teeth
- Laryngeal: persistent hoarseness and throat pain (>3 weeks), difficulty breathing
- Neck: character and duration of neck masses
- Neurological: diplopia, cranial nerve palsies, mental status changes
- Risk Factors: pack-year history of smoking, alcohol use, tobacco abuse, sun exposure, previous cancers, family history of cancer, radiation exposure, exposure to wood dust or heavy metals
- Constitutional Symptoms: extent of weight loss, bone pain, hemoptysis, malaise, anorexia
- Other History: co-morbid medical conditions including psychological profile, cardiac history, pulmonary disease, diabetes, hepatic dysfunction, previous stroke, and renal failure; previous skin lesion removal

Physical Exam

- Otological: assess for middle ear fluid, presence of external auditory canal masses and lesions
- Nasal/Paranasal/Nasopharynx: rhinoscopy and endoscopic exam for masses including the nasopharynx (fossa of Rosenmüller)
- Oral/Pharyngeal: visually inspect all areas of oral and oral pharyngeal cavity (indirect mirror exam) for masses and lesions (ulceration, leukoplakia, erythroplakia); palpate floor of mouth and base of tongue; assess mobility of tongue and any involvement of mandible; evaluate airway, inspect dentition (quality of teeth, occlusion)
- Laryngeal: indirect mirror exam or fiberoptic laryngoscopy, examine all areas of larynx (vallecula, pyriform sinus, postcricoid region, supraglottis, vocal folds) and vocal fold motion

- Neck: palpate nodes (mobility, size, level), palpate salivary and thyroid glands
- Ocular/Neurological: assess visual acuity, extraocular motility, and presence of proptosis, complete cranial nerve exam
- Skin: thoroughly examine skin including scalp, presence of jaundice
- General Physical Exam: complete physical exam to evaluate co-morbidities and overall nutritional status

Initial Evaluation

- typically an accurate history and physical exam, endoscopy with biopsy, chest x-ray, and CT of neck with contrast are all that is essential for an initial work-up for most head and neck cancers; possible additional studies are listed below

Imaging and Ancillary Studies

- Fine Needle Biopsy: indicated for suspicious neck nodes if no primary identified
- Biopsy: biopsy may be performed in the office for most oral, oropharyngeal, nasal, and skin tumors; difficult biopsy sites may be taken during endoscopy
- Chest Radiographs: screening for pulmonary metastasis or primary pulmonary cancer
- CT of Primary Site and Neck: obtained with contrast, evaluates extent of primary tumor size and involvement of adjacent structures (carotid artery, base of skull, floor of neck, bone); aids in staging and determining nodal status (radiographic criteria for nodal malignancy is lesions >1 cm and presence of central necrosis, CT up-stages 10–20% nodal disease); CT not indicated for the T_1N_0 glottic cancer
- MRI of Head and Neck: may be considered for nasopharyngeal, infratemporal fossa, temporal bone, parotid, parapharyngeal, skull base, or intracranial involvement
- CT of Chest: obtained with contrast, indicated for suspicious findings on chest radiographs
- CT of Abdomen: indicated if suspect abdominal metastasis or primary (elevated hepatic transaminases, abdominal masses), lymphoma evaluation
- Panorex: indicated to evaluate dentition or tumor involvement, preoperative films for a mandibulotomy
- Modified Barium Swallow with Esophagram: evaluates aspiration and swallow, indicated for suspicion of esophageal carcinoma or lesions
- Videostroboscopy: provides documentation of laryngeal tumors and allows for patient education

- Nuclear Medicine Studies: may be considered in evaluating thyroid, parathyroid, mandible invasion, and metastatic bone disease, usually not indicated as a screening tool for head and neck cancer

Lab Work

- Preoperative Evaluation: coagulation studies, complete blood count, electrolytes, blood urea nitrogen and serum creatinine, electrocardiogram, liver function tests (albumin, transaminases, alkaline phosphatase)
- Pulmonary Function Tests: preoperative evaluation for pulmonary disease, preoperative assessment for consideration of partial laryngectomy

Tracheotomy, Examination Under Anesthesia, Endoscopy, Feeding Tube

- Tracheotomy: low threshold for surgical airway management in anticipation of radiation effects, postoperative effect, and tumor growth
- Examination Under Anesthesia: reevaluate nodal disease (bimanual exam), palpate primary, base of tongue, and floor of mouth
- Direct Laryngoscopy With Possible Biopsy: evaluate oral cavity, soft palate, hard palate, tonsillar fossa, base of tongue, and larynx (vallecula, pyriform sinus, postcricoid region, supraglottis, and vocal folds) for lesions
- Esophagoscopy: evaluate esophagus for abnormalities and second primaries
- Bronchoscopy: evaluate tracheobronchial tree for abnormalities and second primaries
- Feeding Tube: permanent (gastrostomy tube) or temporary (nasogastric tube), allows enteral feedings with lower risk of aspiration, may place with anticipation of radiation effects, postoperative effect, and tumor growth
- Triple Endoscopy: direct laryngoscopy, esophagoscopy, and bronchoscopy may be considered as routine screening for second primaries and other abnormalities (controversial for negative chest x-ray and no signs or symptoms of esophageal or tracheobronchial involvement)

Immunohistochemistry

- immunohistochemistry utilizes antigen-antibodies reactions that bind to specific cellular components that aid in the histological diagnosis

- most markers are not tumor-specific and not utilized on a routine basis (due to expense)
- useful for paranasal malignancy and poorly differentiated cancers (small- and large-cell tumors)
- squamous cell carcinoma typically is not difficult to distinguish, possible false positives include necrotizing sialometaplasia and mucoepidermoid carcinoma

Common Immunohistochemical Markers

- **Lymphoma**: common leukocyte antigens, T-cell and B-cell markers
- **Carcinoma**: cytokeratin
- **Melanoma**: S-100 (also found in neural and cartilaginous tumors), HMB-45 (diagnostic)
- **Neuroendocrine**: chromogranin, neuronspecific endolase (NSE)
- **Sarcomas**: vimentin, desmin (smooth and skeletal muscle), myoglobin (skeletal muscle, rhabdomyosarcoma)

Determine Classification of Neoplasms

- based on histological specimen and evaluation of size and spread of primary tumor (tumor mapping)
- Grading: categorizes the histological type of cancer according to the degree of differentiation (well differentiated [$\mathbf{G_1}$], moderately well differentiated [$\mathbf{G_2}$], poorly differentiated [$\mathbf{G_3}$], undifferentiated [$\mathbf{G_4}$]); not significant to prognosis
- TNM: categorizes size and spread of cancer (primary tumor [$\mathbf{T_{1–4}}$], regional lymph nodes [$\mathbf{N_{0–3}}$], and presence of distant metastasis [$\mathbf{M_0}$–$\mathbf{M_1}$])
- Staging: grouped TNM classification used for statistical analysis (prognosis, treatment effectiveness, etc), *see* Table 5–1

Initial Management and Prognostic Evaluation

Management

Consultations

- Medical and Radiation Oncologist: assess potential for the role of radiation and chemotherapy
- Speech and Swallow Therapists: may be considered postoperatively to assess swallowing and speech
- Dental: evaluate dentition and possibility of teeth extraction (radiation therapy)

TABLE 5–1. General Staging for Head and Neck Cancer (Except Salivary and Thyroid Glands)

Stage	TNM Classifications
I	$T_1N_0M_0$
II	$T_2N_0M_0$
III	T_1 or $T_2N_1M_0$
	$T_3N_{0-1}M_0$
IV	any T4
	any N_2 or N_3
	any M_1

- Reconstructive Surgery: may be considered to evaluate complex reconstructive issues including free-flap transfer
- Neurosurgery: assist in skull base and cranial procedures
- Ophthalmology: assist in surgical management of the eye
- Prosthodontist: preoperative consultation for oral-maxillary, orbital, and other head and neck prostheses
- Dermatology: consideration of Mohs micrographic surgery for dermal lesions
- Vascular Surgery: consultation for possible carotid resection or bypass
- Medicine: preoperative evaluation (preoperative clearance), management of coexisting medical conditions
- Dietitian: may be considered preoperatively for malnourished patients to assess nutrition status and recommend methods to obtain a positive nitrogen balance

Treatment Concepts

- Single-modality Therapy: treatment of early staged disease with primary surgery or radiation therapy alone
- Multimodality Therapy: combined therapeutic approaches for advanced disease
- Surgical Therapy: advantages include quicker extirpation of the tumor and provision of specimen for margin analysis; disadvantages include potential risk of anesthesia and functional disability; a tumor is considered **unresectable** if it involves the base of skull, nasopharynx, prevertebral fascia, floor of neck, mediastinum, or

subdermal lymphatics (carotid artery involvement is a relative contraindication)
- Radiation Therapy: (*see below*) generally indicated as primary therapy for early glottic, hypopharyngeal, and nasopharyngeal cancers and as adjuvant therapy for advanced head and neck cancer; advantages include easier access for poorly exposed tumors and generally less functional disability; disadvantages include lengthy course treatment, less responsiveness to larger or deeper tumors, inability for second course radiation therapy for recurrence within previously irradiated fields, difficult to detect recurrent cancer, salvage surgery for radiation failure is associated with higher morbidity (conservation surgery may not be possible for recurrence)
- Chemotherapy: (*see below*) generally indicated for palliation for disseminated disease, recurrent or unresectable cancer, laryngeal preservation, esophageal and nasopharyngeal cancers, chemoprevention (retinoids), and clinical protocols
- Clinical Trial Designs: **Phase I** trials define the maximum tolerated dose of the treatment regimen; **Phase II** trials test the efficacy of the treatment of regimen on one or more tumor sites and toxicity; **Phase III** trials are randomized, prospective trials that evaluate the new treatment effect compared to the standard treatment

Post-treatment Follow-up

- follow-up every 1 month for first year post-therapy, every 2 months after 2 years post-therapy, every 3 months after 3 years post-therapy, every 6 months after 4 years post-therapy, every 1 year after 5 years post-therapy
- encourage continued smoking and alcohol cessation
- yearly chest radiographs (may consider CT of primary site and chest for high recurrence risk patients)
- yearly liver enzymes and thyroid function tests (indicated for patients who receive radiation therapy to the neck)

Prognostic Evaluation

- overall 5-year survival for head and neck cancer is **<40%** (50–60% of mortality from head and neck cancer is from failed loco-regional control, 20–30% from metastatic disease, and 10–20% from second primaries)
- premorbid conditions account for the mortality of 30–35% early-staged head and neck cancer patients and 10–15% of late-staged head and neck cancer patients

- Tumor Staging (Presence of Regional and Distant Metastasis): poorer prognoses are suggested with invasiveness and metastasis within each site; presence of regional nodal disease is the strongest predictor of prognosis for most head and neck cancer (may decrease survival as much as 50%)
- Tumor Volume and Thickness: as determined by CT/MRI scan, poorer prognosis suggested with increased tumor thickness and tumor volume
- Tumor Location: presence of abundant lymphatic drainage at the primary site increases risk of regional disease
- Character of Nodal Disease: presence of nodal extracapsular spread reduces overall patient survival, presence of metastasis to lower nodal levels (level IV and V) correlates with poorer prognosis, skipped nodes (eg, nodal involvement of level II and IV without involvement of level III) and increased number of positive nodes indicate decreased survival, lymphocytic predominant immunomorphology of the lymph node has a better prognosis (denotes a host immunological response) than lymphocyte-depleted patterns
- Vascular and Perineural Invasion: histologic predictors of cancer spreading beyond the margins of resection, indicate aggressive cancer behavior suggesting decreased survival
- Cytomorphometric Parameters: (*controversial*) DNA content (ploidy) may predict prognosis (aneuploid DNA content associated with worse prognosis than diploid DNA content)
- Grade of Differentiation: not an important determinant in prognosis for most head and neck cancers

CHEMOTHERAPY AND RADIATION THERAPY

Chemotherapy

Indications for Chemotherapy

- **Nasopharyngeal Cancer**: Phase III Intergroup Study 0099 showed increased survival benefit with chemoradiotherapy (cisplatin/5-FU) over radiotherapy alone for stage III and stage IV nasopharyngeal cancer (76% versus 46% 3-year survival, $P<0.001$) (*J Clin Oncol.* 1998; 16:1310–1317)
- **Unresectable Head and Neck Cancer**: Phase III Multicenter Study showed increased survival benefit with concomitant chemotherapy (cisplatin/5-FU) and radiation therapy over radiation therapy alone

for advanced unresectable head and neck cancer (48% versus 24% 3-year survival, $P<0.0003$) (*J Clin Oncol.* 1998; 16:1318–1324)
- **Laryngeal Organ Preservation**: Veterans Affairs Laryngeal Study Group showed that induction chemotherapy (cisplatin/5-FU) followed by radiation therapy for partial and complete responders results in preserving a functional larynx without compromising survival (68% 2-year survival for both groups, $P=0.98$) (*N Engl J Med.* 1991; 324:1685–1690)
- **Pyriform Sinus Cancer Organ Preservation**: Phase III European Organization for Research and Treatment of Cancer (EORTC) showed no significant change in survival with induction chemotherapy (cisplatin/5-FU) followed by adjuvant radiation therapy (preserves a functional larynx) over surgical management for advanced hypopharyngeal cancer (30–35% 5-year survival), approximately half of the patients who were alive at 3 years in the induction chemotherapy arm retained their larynx (*J Natl Cancer Inst.* 1996; 88:890–899)
- **Recurrent and Distant Metastatic Disease**: original traditional role in head and neck cancer, essentially provides palliation for recurrent unresectable disease or incurable cancer with distant metastatic disease
- **Investigative Protocols**: numerous multi-institutional studies are currently investigating the role of chemotherapy in head and neck cancer

Chemotherapy Strategies

Neoadjuvant (Induction)

- Definition: sequential chemotherapy before local treatment modality (surgery or radiation)
- Advantages: initial chemotherapy may allow better drug penetration prior to impaired vascularity secondary to radiation effects, initial chemotherapy may also reduce tumor bulk or "cure" patient of disease (organ preservation)
- Disadvantages: postchemotherapy patients are often more debilitated which may increase complications of surgical intervention or radiotherapy, margins difficult to access
- Phase III Trial by the Southwest Oncology Group and a multi-institutional study by The Head and Neck Contracts Program (HNCP) failed to demonstrate improved survival in advanced resectable head and neck cancer with induction chemotherapy prior to standard therapy (*Laryngoscope.* 1988; 98:1205–1211; *Cancer.* 1987; 60:301–311)

- The HNCP and other phase III trials have shown a tendency for decreased rate of distant metastasis (without improved overall survival)

Concominant (Radiation Therapy and Chemotherapy)

- Definition: concurrent chemotherapy and radiation (chemotherapy may be given continuously or interrupted [split course])
- Advantages: simultaneous therapies may synergistically maximize therapeutic effect (chemotherapy sensitizes cells to radiation and simultaneously kills micrometastatic disease, *see below*)
- Disadvantages: results in increased side effects from concurrent therapies (mucositis, infection, malnutrition)
- Phase III trial by a single-institution study demonstrated improved loco-regional control (41% versus 23%, P=0.08) and overall 5-year survival (36% versus 13%, P<0.01) with concominant postoperative radiotherapy and cisplatin infusion for stage III or IV squamous cell carcinoma of the head and neck versus radiation therapy alone (*Int J Radiation Oncology Bio Phys.* 1996; 36:999–1004)
- Phase III multi-institutional study by the French "Groupe d'Oncologie Radiothérapie Tête et Cou" (GORTEC) demonstrated improvement in 3-year disease-free survival (42% versus 20%, P=0.04) and overall 3-year survival (51% versus 31%, P=0.02) with the use of concomitant chemoradiation for advanced-staged oropharyngeal cancer versus radation therapy alone (*J Natl Cancer Inst.* 1999; 91:2081–2086)
- Phase III multicenter prospective randomized trial demonstrated improved 3-year loco-regional control (36% versus 17%, P<0.004) and overall 3-year survival (48% versus 24%, P<0.0003) with the use of concomitant chemotherapy for unresectable advanced head and neck cancer versus radiation therapy alone (*J Clin Oncol.* 1998; 16:1318–1324)
- Phase III Intergroup Trial investigating concominant chemoradiation for unresectable head and neck cancer is currently under study

Mechanisms of Increased Effectiveness of Concominant Radiation and Chemotherapies

- chemotherapy inhibits the repair of cells that may otherwise recover from radiation
- concominant radiation and chemotherapies result in synchronization of cell cycles increasing the effectiveness of both therapies
- resistant cells of one mode of therapy may be susceptible to the other mode of therapy
- shrinking the tumor with radiation improves drug delivery for increased effectiveness of chemotherapy

- combined therapy may initiate cells from G_0 phase to active phases rendering the cells more susceptible to therapy
- concominant modalities damage cells in different ways and increase chances of cells' death

Adjunctive

- Definition: chemotherapy after primary treatment modality to control microscopic disease
- Indications: locally advanced disease, multiple positive regional neck nodes, nodal extracapsular spread, positive margins, recurrence
- Advantages: may improve survival
- Disadvantages: exposes patient to side effects of chemotherapeutic agents
- The Intergroup Study 0034 showed no significant increased survival advantage with adjuvant chemotherapy (cisplatin/5-FU) followed by postoperative radiation therapy versus postoperative therapy alone for advanced resectable head and neck cancer (*Int J Radiation Oncology Biol Phys.* 1992; 23:705–713)
- The Head and Neck Contracts Program failed to demonstrate improved survival in advanced resectable head and neck cancer with induction chemotherapy, standard treatment, followed by maintenance therapy (*Cancer.* 1987; 60:301–311)
- Phase III Intergroup Study of adjuvant radiochemotherapy for resectable high risk head and neck cancer (positive margins, 3 or more positive nodes, extracapsular spread) is currently under investigative trials

Chemoprevention

- **Retinoids** (analogues of vitamin A) have been shown to reduce malignant potential of premalignant oral lesions (54% reversal of dysplasia with retinoids versus 10% for controls, P=0.01) (*N Engl J Med.* 1986; 315:1501–1505) and prevent the development of second primaries in a single-institution phase III trials (4% developed second primaries with retinoids versus 24% in controls, P=0.005) (*N Engl J Med.* 1990; 323:795–801); the effect, however, is not sustained once treatment is stopped
- Mechanism of Action: regulate cellular differentiation and proliferation in epithelial tissue (via retinoic acid receptors) and modulate neoplastic cell growth differentiation and apoptosis
- Common Side Effects: mucous membrane and skin dryness, conjunctivitis, transitory elevated transaminases, hyperlipidemia, myalgia, teratogenicity, skeletal effects

- A Phase III Intergroup Trial is currently investigating the efficacy of moderate-dose 13-cis retinoic acid in preventing second primaries in head and neck cancer

Common Chemotherapy Agents and Combinations in Head and Neck Cancer

Cisplatin

- Mechanism of Action: heavy metal that acts as an alkylating agent that covalently binds DNA and RNA
- Common Side Effects: nausea, nephrotoxicity, peripheral neuropathy, ototoxicity, electrolyte disturbances, anorexia
- Indications: best single-agent against squamous cell carcinoma of the head and neck in recurrent disease; common combination agent for neoadjuvant, adjuvant, and concomitant chemotherapy of the head and neck; radiation sensitizer

Carboplatin

- Mechanism of Action: similar to cisplatin (less reactive)
- Common Side Effects: better tolerated than cisplatin (less nephrotoxicity, nausea, neurotoxicity, and ototoxicity)
- Indications: not been fully investigated in head and neck cancer, often used in combination with taxol

5-Fluorouracil (5-FU)

- Mechanism of Action: antimetabolite that binds to thymidilate synthetase blocking the conversion of uridine to thymidine preventing DNA synthesis in S-phase
- Common Side Effects: anorexia and nausea, mucositis, diarrhea, alopecia, myelosuppression, cardiac toxicity
- Indications: similar to cisplatin (cisplatin and 5-FU is the most studied combination chemotherapy regimen in head and neck cancer)

Methotrexate

- Mechanism of Action: antimetabolite that binds to dihydrofolate reductase preventing DNA synthesis in S-phase
- Common Side Effects: bone marrow suppression, gastrointestinal disturbances, mucositis, alopecia, dermatitis, nephrotoxicity, teratogenicity, interstitial pneumonitis
- Indications: "standard" palliative therapy for recurrent or metastatic disease

- **Leucovorin** (**Tetrahydrofolic Acid**): utilized as a "rescue" agent, competitively overcomes increases intracellular pools of dUMP (also used with 5-Fluorouracil)

Taxanes (Paclitaxel and Docetaxel)

- Mechanism of Action: prevent normal microtubular reorganization
- Common Side Effects: neutropenia, alopecia, mucositis
- Indications: currently being investigated for recurrent disease and as a potential radiation sensitizer

Radiation Therapy

Mechanism of Injury

- **Rad** (**Radiation Absorbed Dose**): amount of energy deposited by ionizing radiation per gram of tissue (1 Gy = 100 rads)
- radiation cell "kill" is expressed as a logarithmic cell survival curve (radiation does not kill a certain number of cells but a percentage of cells)
- cells are considered "killed" when they lose clonogenic survival
- **Direct Mechanism of Radiation Injury**: direct damage of radiation with critical elements in a cell (eg, DNA, cell membranes)
- **Indirect Mechanism of Radiation Injury**: secondary damage from direct radiation effects on other cell moieties; primary mechanism of cell death (eg, DNA injury from production of free radicals)

Determinants of Sensitivity of Radiation Therapy

- larger tumors have a more hypoxic center, therefore are less sensitive to radiation because less free radicals are generated (1 cm tumor = 10^9 cells; 3 cm tumor = 10^{10})
- oxygenated cells are more susceptible to radiation than hypoxic cells (exophytic tumors are typically well vascularized and therefore more susceptible to radiation injury, ulcerative and infiltrative tumors are less vascularized and therefore more resistant to radiation)
- cell death occurs with proliferation (usually 4–5 times before lysis), therefore rapidly growing tumors are more susceptible to injury
- cells tend to be more radiosensitive in mitosis and late G_1 and early S-phases

Fractionation and the "4 Rs" of Radiation Biology

- **Conventional Fractionation**: radiotherapy given in smaller interval dosing rather than given all at once
- Reassortment: fractionation allows cells to proceed in their cycle to more radiosensitive stages in their cell cycle

- Reoxygenation: fractionation allows for reoxygenation of previously more hypoxic cells (more susceptible)
- Repopulation: prolonged waiting between fractions results in regrowth of tumor cells from sublethal damage
- Repair: normal tissue tends to have better repair than tumor cells, therefore recovery more quickly from sublethal damage
- fractionation results in total less biological injury due to the greater opportunity for repair, therefore requires a higher total dose than single or hypofractionated dosing
- fractionation in general is less toxic
- hypofractionization (less fractions) are used for tumors with good reparation of sublethal injury (malignant melanoma)

Methods of Fractionation

- **Conventional Fractionation**: typically uses 1 treatment per day (generally 1.8–2.2 Gy) for 5 days a week
- **Pure Hyperfractionation**: increases rate of treatments (eg, 2-times a day dosing), smaller dose per fraction, same duration of therapy, higher total dose than conventional fractionation; decreases late side effects
- **Accelerated (Standard) Fractionation**: decreases duration of therapy, increases rate, higher dose per fraction, with same decreased total dose as conventional fractionation; increases acute side effects, decreases late side effects, increases tumor cell kill (prevents tumor cell proliferation)
- **Accelerated Hyperfractionation**: increases rate of treatments, decreases duration of therapy, increases total dose
- **Concominant Boost Accelerated Fractionation**: changes rate to twice a day dosing in the last 2 weeks of therapy
- Phase III EORTC 22851 Trial showed that accelerated fractionation improved loco-regional control in comparison to conventional fractionation (59% versus 46% 5-year loco-regional control, P=0.02), but no survival benefit (*Radiother Oncol.* 1997; 44:111–121)
- Phase III Trials are currently investigating the effectiveness of hyperfractionation and accelerated fractionation in survival of head and neck cancer

Radiation Strategies

- **Shrinking Fields**: selective radiation dosing to varying region depending on primary size and shape (eg, 7,000 cGy to primary site as primary therapy, 5,000–5,500 cGy to N_0 neck or adjuvant treatment)

- **Brachytherapy**: delivery of radiation to malignant tissue by placement of permanent radioisotopes intraoperatively via a temporarily placed radioactive source within tumor bulk
- **Three-Dimensional Multiple Treatment Beam Therapy**: utilizes CT and MRI imaging, multiple treatment fields arranged to maximize radiation dose to target area yet achieve maximum normal tissue sparing
- **Concominant Radiation Therapy and Chemotherapy**: (*see above*)
- **Hyperfractionation**: (*see above*)
- **Neutron Beam Therapy**: "heavy" neutron particle that result in greater direct mechanisms of injury, less dependence on cell cycling and proliferation; however, less repair of sublethal damage by normal tissue

Preoperative Radiation Therapy

- Advantages: may reduce tumor bulk which may cause inoperable lesions to become operable, reduce surgical excision area, or cure (organ preservation); requires smaller portals than postoperative radiation therapy; microscopic tumor is more sensitive than postoperative residual tumor (microscopic disease may be in scar tissue, therefore more hypoxic)
- Disadvantages: increased difficulty in operating in irradiated tissue, increased postoperative complications (wound infections, carotid blowout)

Postoperative Radiation Therapy

- Advantages: allows for adequate postoperative healing prior to beginning adjuvant therapy, accurate assessment of pathologic staging and factors for recurrence
- Disadvantages: requires higher dose of radiation due to hypoxia in operated tissue, microscopic disease may be within scar tissue resulting in decreased radiosensitivity, requires larger fields than preoperative radiation therapy
- Phase III Radiation Therapy Oncology Group (RTOG) Trial did not demonstrate a significant overall survival difference between preoperative and postoperative radiation therapy for advanced head and neck carcinoma, although loco-regional control was significantly better with postoperative radiation therapy (*Int J Radiation Oncology Biol Phys.* 1991;20:21–28)

Side Effects

- **Cutaneous Reactions**: may result in dryness, erythema, hyperpigmentation, desquamation, telangiectasias, or subcutaneous

fibrosis; Rx: skin moisturizers, mild skin cleaning, oral diphenhydramine (pruritis), corticosteroid creams
- **Mucositis**: presents as tender, erythematous, and swollen mucous membranes; increased risk of coexisting infections with *Candida*, herpes simplex virus, and other bacteria; may result in sepsis for severe cases; Rx: aggressive oral hygiene (oral irrigations), adjust dental appliances, smoking and alcohol cessation, cool food, nutritional supplements, oral and topical anesthetics (viscous lidocaine, diphenhydramine, aluminum hydroxide-magnesium hydroxide-simethicone mixture)
- **Alopecia**: temporary hair loss may occur in radiation field, may be permanent at higher doses
- **Xerostomia/Dental Caries**: salivary acinar cells are extremely sensitive to radiation therapy causing irreversible xerostomia, increased risk of dental caries, change in taste, and tenacious oral secretions; Rx: fluids with meals, artificial saliva, pilocarpine, fluoride treatment, aggressive oral hygiene (prevent with preoperative dental evaluation)
- **Osteoradionecrosis of the Mandible/Cartilage Radionecrosis of the Larynx**: hypocellularity, hypovascularity, and ischemia of tissue (not infectious); Rx: initially should treat conservatively with antibiotics, analgesics, meticulous oral hygiene, and soft diet; debridement may be required; may also consider hyperbaric oxygen therapy
- **Radiation Induced Cancer**: increased risk of thyroid, salivary gland cancer, leukemia, sarcomas (may have lag period of 20 years)
- **Otologic Sequelae**: increased incidence of otitis externa, sterile otitis media, and auricular chondritis

CANCER OF THE NECK

Introduction

- most malignancies in the neck are from metastasis (approximately 85%), primary neck cancers in the neck are less common (approximately 15%), usually from salivary tumors, thyroid cancers, or lymphoma
- bilateral regional metastasis is common with primaries of the base of tongue, supraglottis, ventral tongue, and soft palate
- Mechanisms of Regional Metastasis: malignant tumors extend to surrounding tissue, tumor cells invade blood vessels and/or lymphatics (via hydrolytic enzymes), microemboli of tumor cells become trapped in the lymph node resulting in seeding for further proliferation (most cells that enter the blood vessels are rapidly destroyed)

Evaluation and Management of the Neck Mass

Initial Evaluation

- History and Physical Exam: complete head and neck history and physical exam (*as above*), attention to nasopharynx, oral cavity, base of tongue, tonsilar fossa, nasal cavity, external ear canal, scalp, thyroid, and salivary glands (also consider breast, rectal, and pelvic exams)
- Primary Cancer Identified: biopsy primary site, treatment based on site of primary
- FNA Biopsy: indicated for any suspicious nodes without a known primary
- FNA Reveals Benign Lymph Tissue or is Indeterminant: may consider a CT scan of neck to re-evaluate for criteria of malignancy in the neck mass, may repeat the FNA, or may observe if low suspicion
- FNA Reveals Lymphoid Cells: perform an excisional open biopsy (fresh specimen), if frozen section suggests a lymphoma may evaluate patient per protocol for lymphomas (*see below*), if frozen section is nonlymphoma then consider complete neck dissection if appropriate
- FNA Suggests Adenocarcinoma: for high-level nodal disease consider neck dissection with submandibulectomy and possible parotidectomy (depending on preoperative imaging); for low-level nodal disease consider excisional biopsy (may also consider thyroid scan)
- FNA Suggests Other Primary Cancer: sarcomas, thyroid cancer, salivary gland malignancy; treatment based on type of cancer

Evaluation and Management for FNA-proven Carcinoma of a Neck Mass with an Unknown Primary

- complete imaging and lab workup including chest radiograph, CT or MRI of head and neck
- complete endoscopy with biopsy of suspicious lesions; may consider blind biopsies of nasopharynx, base of tongue, tonsils, and pyriform sinuses; may also consider ipsilateral tonsillectomy
- may consider bone scan, CT of chest and abdomen, mammography, GI imaging (barium swallow), and thyroid scans
- Primary Cancer Identified: treatment based on site of primary
- Primary Cancer Unknown: treatment of neck disease as described below, close monthly follow-up with low threshold for biopsy

Staging

Nodal Levels

I submental and submandibular triangles
II upper third (superior to the hyoid bone or carotid bifurcation to the base of skull); contain the upper jugular lymph nodes
III middle third (superior to the omohyoid muscle to the carotid bifurcation); contain the middle jugular lymph nodes
IV lower third (superior to the clavicle to the omohyoid muscle); contain the lower jugular lymph nodes
V posterior triangle
VI anterior neck, between carotid sheaths

Staging (*based on the American Joint Commission on Cancer Staging*, 1997)

N_0 no regional lymph nodes
N_1 metastasis to single ipsilateral lymph node, <3 cm
N_{2a} metastasis to single ipsilateral lymph node, 3–6 cm
N_{2b} metastasis to multiple ipsilateral lymph nodes, <6 cm
N_{2c} metastasis to bilateral or contralateral lymph nodes, <6 cm
N_3 metastasis to any lymph node >6 cm

Management of the Neck in Head and Neck Cancer

Primary Cancers

- management for lymphoma, thyroid malignancy, salivary gland tumors is discussed in other sections
- sarcomas and branchial cleft cyst carcinomas are quite rare and typically require wide resection with neck dissection

Regional Nodal Metastasis

- Early Staged Neck Disease (N_1): resection of the primary site with a neck dissection (preferably en bloc if possible) with consideration of adjuvant radiation therapy (to the primary site and neck) for extracapsular spread or advanced primary disease versus primary radiation therapy of **6,500 cGy** (especially if the primary site is being irradiated) with planned or salvage neck dissection
- Late Staged Neck Disease (N_2–N_3): multimodality therapy with a neck dissection and postoperative radiation therapy

Clinically Negative Neck (N_0)

- controversial management between observation, elective treatment with neck dissection, or radiation therapy
- Observation: may be considered if risk of occult metastasis is **<15–25%**
- Elective Neck Dissection: generally indicated if risk of regional metastasis **>15–25%** (eg, supraglottis, base of tongue, tonsil, oral tongue, and advanced staged cancer), typically a modified neck dissection of selected nodal groups provides the least morbidity with adequate excision, specimen provides histology to evaluate for positive nodes and extracapsular spread (adjuvant therapy)
- Radiation Therapy: may also be considered to eradicate occult neck disease if risk of regional metastasis is >15–25%, indicated especially if primary site is being irradiated

The Fixed Neck (unresectable)

- fixed nodes suggest adherence to the vertebrae, branchial plexus, major vessels, mastoid process, or other structures that are nonmobile
- unresectability is typically considered for branchial plexus, floor of neck, or vertebral involvement (carotid artery involvement is considered a relative contraindication)
- multimodality therapy is offered as an initial radiation and chemotherapy in hopes of "freeing up" the tumor to allow for resection

Bilateral Positive Nodes (N_{2c})

- therapy typically consists of excision of primary site with bilateral neck dissections with adjuvant radiation therapy
- a modified neck dissection (preserving the internal jugular vein and spinal accessory nerve) should be attempted first on the least involved side prior to operating on the more involved side; if unable to preserve the internal jugular vein should consider staging the necks 3–4 weeks apart to allow for dilation of the intracranial venous vasculature to avoid increased intracranial pressure

Unknown Primary

- Early Staged Neck Disease (N_1): may manage with a neck dissection with adjuvant radiation therapy to the neck, Waldeyer's ring, nasopharynx versus radiation therapy alone
- Late Staged Neck Disease (N_2–N_3): multimodality therapy with neck dissection and adjuvant radiation therapy to the neck, Waldeyer's ring, and nasopharynx (no role for adjuvant chemotherapy)

Neck Dissections

Classifications of Neck Dissections

Radical Neck Dissection

- **Radical**: comprehensive (removes all level nodes)
- Indication: clinically positive nodes with primary cancer that has a high risk of occult nodes; advance nodal disease; presence of large matted nodes or posterior nodes; involvement of sternocleidomastoid muscle, internal jugular vein, or spinal accessory nerve; recurrence or radiation failure
- Technique: removes submandibular gland, tail of parotid, sternocleidomastoid muscle, internal jugular vein, spinal accessory nerve, and cervical nodes
- Advantages: technically easier, lower risk of residual disease
- Disadvantages: neck deformity (removal of sternocleidomastoid muscle), shoulder drop (removal of spinal accessory nerve), risk of facial edema (removal of internal jugular vein), hyposthesia of the neck and periauricular region

Modified Radical Neck Dissection

- comprehensive (removes all level nodes)
- **Type I**: spares spinal accessory nerve
- **Type II**: spares internal jugular vein and spinal accessory nerve
- **Type III** (**Functional, Bocca**): spares sternocleidomastoid muscle, internal jugular vein, and spinal accessory nerve
- Indication: clinically positive nodes with primary cancer with a lower risk of occult nodes or the N_0 neck; no involvement of sternocleidomastoid muscle, internal jugular vein, or spinal accessory nerve
- Advantages: lower morbidity from preservation of sternocleidomastoid muscle, spinal accessory nerve, or internal jugular vein
- Disadvantages: technically more difficult, higher risk of residual disease

Selective Neck Dissection

- does not remove all nodal levels (removes only nodal disease at high risk)
- **Supraomohyoid** (**Anterolateral**) **Neck Dissection**: removes nodal levels I–III (expanded supraomohyoid removes level IV); indicated for larger oral cancers with a N_0 or select N_1 (mobile) neck

- **Lateral Neck Dissection**: removes nodal levels II–IV; indicated for select supraglottic, oropharyngeal, hypopharyngeal cancers, typically bilateral
- **Posterior Lateral Neck Dissection**: removes nodal levels II–V (also retroauricular and suboccipital nodes); indicated for select posterior scalp cancers

Complications

- **Wound Infections and Wound Breakdown**: incidence higher in irradiated tissue; soilage of wound by saliva, tracheal aspirates, or gastric secretions; tight wound closure; immunocompromised and malnourished states; and presence of a foreign body, hematoma, or seroma; Rx: aggressive antibiotic regimen, monitor for fistula, control diabetes, maximize nourishment, meticulous wound care (debridement, wet to dry dressings), evaluate potential for carotid blow-out (*see below*)
- **Flap Necrosis**: poorly planned incision that compromises vascular supply may result in tissue loss, wound infection, fistulas, or vessel exposure; Rx: avoid with properly designed neck incisions (eg, curvilinear, MacFee, modified Schobinger incision; trifurcation should begin at right angles from the main incision), manage wound infection and breakdown as above
- **Shoulder Syndrome**: injury or sacrifice of spinal accessory nerve; Rx: physical therapy, may also consider early cable grafting or orthopedic reconstruction
- **Injury to the Vagus Nerve**: injury results in varying presentations depending on level and branch of injury; superior laryngeal nerve injury typically may present with only subtle voice changes, recurrent laryngeal nerve injury may result in hoarseness, aspiration, or airway compromise; Rx: may require tracheotomy for airway management (especially for bilateral vocal fold paralysis), management of aspiration and vocal fold paralysis (*see* pp. 114–119)
- **Injury to the Marginal Branch of the Facial Nerve**: at risk during elevation of the cervical flap, should be identified and protected, bilateral injury may result in oral incompetence; Rx: if discovered interoperatively immediate neurorrhaphy should be performed, may consider facial reanimation procedures (*see* pp. 369–371)
- **Hematoma/Seroma**: prevented with meticulous hemostasis and placement of suction drains; Rx: consider reopening for major hematomas with removal of clot and controlling bleeding; for minor

hematomas may wait 7–10 days to allow for liquification then aspirate; may also consider pressure dressings and prophylactic antibiotics; seromas may also be aspirated

- **Chylous Fistula**: typically left sided from injury to thoracic duct, milky drainage, appears within first few days, incidence from 1–2%; Rx: initial conservative management (pressure drainage, head elevation, restrict fats and medium chained triglycerides, manage electrolyte abnormalities), consider tetracycline sclerosing therapy versus surgical re-exploration for failed conservative therapy or if output >600 cc/day
- **Cerebral and Facial Edema**: higher risk with internal jugular vein ligation (especially bilateral internal jugular vein ligation), may present with Syndrome of Inappropriate Antidiuretic Hormone Secretion (SIADH) or mental status changes; Rx: cerebral edema must be addressed urgently; Neurosurgery consultation; consider corticosteroids, lumbar drainage, hyperventilation, hyperosmolar agents (mannitol), and diuretics
- **Blindness**: caused by optic nerve infarction (uncertain etiology, may be from hypotension and increased intracranial pressure), rare
- **Postoperative Dyspnea**: possible etiologies include pneumothorax, phrenic nerve injury, intrinsic lung disease (atelactasis), and congestive heart failure
- **Carotid Blow-out**: typically from infected wound site and wound breakdown, high mortality rate (approximately 10%); Rx: airway management, compression, large bore catheters with fluid replacement, immediate blood replacement, intraoperative ligation

ORAL CANCER

Introduction

Introduction

- 30% of all head and neck cancers (most common head and neck site)
- oral cavity has the highest rate of second primaries (10–40%)
- >90% of occult metastatic disease in oral cancer involves nodal groups I–III (the supraomohyoid dissection is oncologically sound especially in the N_0 neck)
- SSx: nonhealing ulcers, denture difficulties, dysphagia, odynophagia, trismus, halitosis, numbness in the lower teeth (suggests mandible involvement of the inferior alveolar nerve)

- Risk Factors: smoking, alcohol, and tobacco abuse; radiation and ultraviolet radiation exposure (lip cancer), Plummer-Vinson syndrome (*see* p. 155), human papilloma virus, poor oral hygiene, oral leukoplakia (5–20% malignant potential) and erythroplakia (approximately 25% malignant potential *see* pp. 170–173)

Anatomy

- **Oral Cavity**: extends from lips to junction of hard and soft palate and circumvallate papillae
- other than the mandibular periosteum there is no finite fascial plane to inhibit tumor's extension in the oral cavity

Subsites and Regional Lymph Node Potential

- **Lips**: most common location of oral cancer, **90%** on lower lip, **90%** 5-year survival if <2 cm, **90%** squamous cell carcinoma (**Rules of 90's**), basal cell carcinoma is more common on upper lip; 2–15% regional metastasis (for all stages); lower lip has **bilateral and ipsilateral** lymphatic drainage into level I–III nodal groups and upper lip has **ipsilateral** lymphatic drainage into level I–III nodal groups (no contralateral drainage due to embryological fusion plates); overall 5-year survival for all stages for squamous cell carcinoma is 70–90% for the lower lip and 40–60% for the upper lip; poorer prognosis is associated with upper lip and commissure involvement
- **Buccal Mucosa**: common site near mandibular third molar (site of chewing tobacco), most common site for verrucous cancer, more common in India; 50% regional metastasis (for all stages), occult neck metastasis is approximately 10%
- **Alveolar Ridge**: more common in edentulous and molar areas of the mandible, must differentiate from invasive maxillary cancer, high rate of bony involvement; 50–65% overall 5-year survival
- **Retromolar Trigone**: triangle-shaped region with the base at the last mandibular molar and the apex at the maxillary tuberosity; typically presents in an advanced stage, bony invasion common, 50% regional metastasis (for all stages); approximately 25–55% 5-year survival for all stages (due to advance initial staging and poor salvage potential)
- **Hard Palate**: incisive foramen allows tumor extension into anterior nose, palatine foramen allows tumor extension to pterygopalatine fossa; less aggressive (10–25% occult regional metastasis), minor salivary gland tumors are common

- **Oral Tongue**: movable portion, anterior to circumvallate papillae; second most common site of oral cancer; 25–66% regional metastasis (for all stages); 60–80% 5-year survival for early disease (T_1–T_2)
- **Floor of Mouth**: dependent site for alcohol and chewing tobacco, 30% present with regional metastasis, overall 5-year survival is 30%–65%

Staging and Pathological Classification

Staging (*based on the AJCC Staging*, 1997)

- T_1: primary tumor <2 cm
- T_2: primary tumor 2–4 cm
- T_3: primary tumor >4 cm
- T_4: primary tumor invades adjacent structures (eg, through cortical bone, skin, through floor of mouth)

Pathology

- **Squamous Cell Carcinoma** (**SSC**): >90% of oral cancers
- **Verrucous Carcinoma**: variant of SSC, broad based, warty growth, most common site is the **buccal mucosa**, lateral growth, rare metastasis and deep invasion
- **Basal Cell Carcinoma**: more common on the upper lip
- Other Types: Lymphoma, Kaposi's Sarcoma, Salivary Gland Malignancies, Melanoma
- NOTE: **Necrotizing Sialometaplasia** and **Granular Cell Tumors** may be mistaken for squamous cell carcinoma in the oral cavity due to similar histology (pseudoepitheliomatous hyperplasia)

Management

Early Oral Cancer (T_1–T_2)

- Single-Modality Therapy: excision of primary tumor with primary reconstruction (*see below*), may consider primary radiation (external beam versus brachytherapy)
- N_0 Neck: elective ipsilateral or bilateral (midline or oral tongue cancer) selective neck dissection (supraomohyoid) versus external beam therapy (early stage hard palate or lower lip do not require elective neck dissections because of lower rate of occult metastasis); if surgical specimen is positive for tumor may consider observation,

completion of a comprehensive neck dissection, or radiation therapy to neck
- N_{1-3} Neck: radical neck dissection for clinical nodes; parotid nodes require a superficial parotidectomy

Advanced Oral Cancer (T_3–T_4)

- Single-Modality Therapy: excision of primary tumor with primary reconstruction (*see below*) versus primary radiation for nonoperable candidates (external beam versus brachytherapy)
- N_0 Neck: elective ipsilateral or bilateral (midline or oral tongue cancer) selective neck dissection (supraomohyoid) versus external beam therapy; if surgical specimen is positive for tumor may consider observation, completion of a comprehensive neck dissection, or radiation therapy to neck
- N_{1-3} Neck: radical neck dissection for clinical nodes; parotid nodes require a superficial parotidectomy
- Adjuvant Therapy: postoperative radiation therapy may be considered for positive margins; multiple positive neck nodes or extracapsular extension; perineural or intravascular invasion; or bone, cartilage, or soft tissue invasion
- chemotherapy indicated for palliation or may be considered for adjuvant treatment for advanced disease

Lip Cancer

- Single-Modality Therapy: excision of primary tumor (may consider Mohs micrographic excision) with primary reconstruction versus primary radiation therapy (5,000–7,000 cGy) for small tumors or nonoperable candidates (must also consider functional and cosmetic outcomes)
- Adjuvant Therapy: postoperative radiation therapy may be considered for advanced stages (T_{3-4}, N_{2-3}), close or positive margins, multiple positive neck nodes, perineural or intravascular invasion, or extracapsular extension
- N_0 Neck: elective ipsilateral or bilateral (for lower lip midline disease) selective neck dissection (supraomohyoid) versus external beam therapy for advanced diseases (T_3–T_4); if surgical specimen is positive for tumor may consider observation, completion of a comprehensive neck dissection, or radiation therapy to neck
- N_{1-3} Neck: radical neck dissection for clinical nodes; parotid nodes require a superficial parotidectomy
- chemotherapy may be considered for palliation or adjuvant treatment for advanced disease

Surgical Management of Oral Cancer

- Approach: anterior and small tumors (<2 cm) may be approached intraorally, larger and more posterior tumors require a transmandibular or transcervical approach
- Mandible: invasion of the mandible (as diagnosed by CT or multiplanar reformation CT) requires a segmental mandibulectomy, direct abutment of tumor against periosteum requires a mandible-sparing procedure (marginal or rim mandibulectomy)
- Reconstruction: primary closure and split-thickness skin grafts provides the best speech and swallowing outcomes, large defects require regional or distant flaps for bulk and adequate closure

OROPHARYNGEAL CANCER

Introduction

Introduction

- SSx: sore throat, odynophagia, dysphagia, voice quality changes, neck mass, referred otalgia, globus sensation, trismus, dysarthria, decreased tongue mobility, base of tongue mass (on palpation)
- Risk Factors: smoking, alcohol, and tobacco abuse; Epstein-Barr virus (lymphoepitheliomas)

Anatomy

- **Oropharynx**: anterior boundary from junction of hard and soft palate (above) and circumvallate papillae (below), superior boundary at level of hard palate, inferior boundary at level of pharyngoepiglottic folds

Subsites and Regional Lymph Node Potential

- **Soft Palate**: rare, since more visible than other sites typically found at early stages, 20–45% regional metastasis, 70% 5-year survival
- **Base of Tongue**: more aggressive tumor than oral tongue, high rate of cervical metastasis (>60%, 20% bilateral metastasis), poorer prognosis (approximately 65% 5-year survival for all stages)
- **Tonsil/Lateral Pharyngeal Wall**: most common site of oropharynx, may present as an exophytic mass or an ulcerative lesion, aggressive usually presents with regional neck disease (65–75%), higher instances of lymphomas and lymphoepithelomas (*see below*)
- **Posterior Pharyngeal Wall**: less common, aggressive although less metastatic potential than base of tongue

Classification and Management

Staging (*based on the AJCC Staging*, 1997)

- T_1: primary tumor <2 cm
- T_2: primary tumor 2–4 cm
- T_3: primary tumor >4 cm
- T_4: primary tumor invades adjacent structures (eg, mandible, hard palate, deep muscles of tongue, larynx, pterygoid muscles)

Pathology

- **Squamous Cell Carcinoma** (**SSC**): >95% of oropharyngeal cancer
- **Lymphoepitheliomas**: subgroup of poorly differentiated carcinoma, may present in the tonsil, exophytic, radiosensitive
- **Lymphomas**: (*see* Lymphomas) 10–15% of base of tongue and tonsillar cancers
- Other Types: Sarcomas, Salivary Gland Malignancies, metastatic disease

Management of Oropharyngeal Cancer

- Single-Modality Therapy: indicated for T_1 or T_2 oropharyngeal tumors (except base of tongue involvement); excision of primary tumor with primary reconstruction (*see below*) versus primary external beam radiation or brachytherapy
- Multimodality Therapy: indicated for T_3 or T_4 oropharyngeal tumors (or base of tongue involvement); excision of primary tumor with primary reconstruction (*see below*) and postoperative radiation therapy versus primary radiation with salvage surgery
- N_0 Neck: elective bilateral neck dissection versus external beam therapy
- N_{1-3} Neck: radical neck dissection for clinically evident nodes
- Adjuvant Therapy: postoperative radiation therapy may be considered for advanced or aggressive disease (tongue base cancer); close or positive margins; multiple positive neck nodes or extracapsular extension; perineural or intravascular invasion; or bone, cartilage, or soft tissue invasion
- chemotherapy induction protocols may be considered for select cases (surgical salvage for nonresponders and adjuvant radiation therapy for responders)

Surgical Management of Oropharyngeal Cancer

- typically requires an initial tracheotomy

Approaches

- **Transoral**: may be used for limited tumors (eg, posterior pharyngeal wall, anterior pillar, uvula, soft palate), no external scar, poor exposure
- **Transcervical/Visor Flap**: may be considered for large tumors of the base of tongue or tonsil, access oropharynx from a transoral incision of the floor of the mouth, preserves mandibular integrity, poor exposure, chin numbness
- **Mandibulectomy**: indicated for larger lesions, mandible extension, or multiple sites (composite resection); may be approached laterally or medially with a lip-splitting incision (**mandibular swing**); provides excellent exposure, easier soft tissue closure, risk of malocclusion and plate extrusion
- **Mandibulotomy**: spares mandible, may be approached laterally or midline with a lip-splitting incision, osteotomy is performed in a stepwise fashion to create a favorable repair followed by rigid fixation, provides excellent exposure, less risk of malocclusion
- **Lateral Pharyngotomy**: may be considered for small base of tongue or posterior pharyngeal wall tumors; enters pharynx between hypoglossal and superior laryngeal nerves; limited exposure, spares mandible, avoids lip-splitting incision
- **Transhyoid Pharyngotomy**: may be considered for small base of tongue or posterior pharyngeal wall tumors without significant superior or tonsillar extension; enters pharynx above or through hyoid bone; spares mandible, avoids lip-splitting incision, vallecula must be free of tumor, poor exposure superiorly

Reconstruction

- **Primary Closure**: simplest and best functional outcome (speech and swallowing), ideal method of reconstruction if a tension-free closure can be obtained without resulting in significant stenosis
- **Split-thickness Skin Grafts**: allows resurfacing with good functional outcomes, does not provide tissue bulk
- **Pedicled Regional Flaps**: provides soft tissue bulk with compromise of speech and swallowing function (*see* Facial Plastics and Reconstruction, for complete discussion)
- **Free Tissue Transfers**: provides soft tissue bulk with compromise of speech and swallowing function, **Fibular Free Flap** (or **Iliac Crest**)

methods of choice for mandibular reconstruction of >5 cm of bone loss (*see* Facial Plastics and Reconstruction for complete discussion)

HYPOPHARYNGEAL CANCER

Introduction

Introduction

- usually advanced when diagnosed (poor prognosis)
- SSx: airway obstruction, progressive dysphagia, odynophagia, neck mass, referred otalgia, globus sensation, sore throat, hoarseness, weight loss, laryngeal lesions (ulcerated, exophytic), vocal fold paralysis
- Risk Factors: smoking, alcohol, tobacco abuse, Plummer-Vinson syndrome (*see* p. 155)

Anatomy

- **Hypopharynx**: level of hyoid bone to esophageal introitus, lies behind and around the larynx

Subsites and Regional Lymph Node Potential

- **Pyriform Sinus**: most common site for hypopharyngeal cancer (65–75%); may extend into the subglottis, cricoarytenoid joint of muscle (vocal fold fixation), thyroid cartilage, or postcricoid region; 75% regional metastasis; apical primaries are associated with a worse prognosis
- **Posterior Pharyngeal Wall**: 20–25% of hypopharyngeal tumors; may extend into the oropharynx, postcricoid region, or prevertebral fascia (late in disease); 60% regional metastasis
- **Postcricoid Region**: rare (<5%), associated with Plummer-Vinson Syndrome, may extend into the cricoid cartilage, cricoarytenoid muscle, or cervical esophagus; 40% regional metastasis

Classification and Management

Staging (*based on the AJCC Staging*, 1997)

- T_1: primary tumor limited to one subsite and <2 cm
- T_2: primary tumor involves more than one subsite or >2 cm
- T_3: primary tumor >4 cm or vocal fold fixation

- **T_4**: primary tumor invades adjacent structures (eg, thyroid cartilage, carotid artery, soft tissues of the neck, thyroid, esophagus)
- NOTE: previous AJCC guidelines did not account tumor size for staging criteria (1 subsite, T_1; more than 1 subsite, T_2; more than 1 subsite or fixation of the hemilarynx T_3; invasion into adjacent structures, T_4)

Pathology

- **Squamous Cell Carcinoma (SSC)**: poorly differentiated (most common)
- Other Types: adenomas, salivary gland malignancies, metastatic disease

Management

- tumors of the hypopharynx may extend **submucosally** resulting in "**skip lesions**"
- most present in advanced state with clinical cervical nodes (40–75%)

Early Hypopharyngeal Cancer (T_1 or T_2)

- Single-Modality Therapy: primary radiation with surgical salvage (includes irradiating both sides of the neck), surgical excision with postoperative radiation may also be considered for limited select cases (*see below*)
- Neck: elective radiation therapy to bilateral necks in conjunction with treatment of the primary, elective ipsilateral neck dissection with surgical management of primaries (bilateral neck dissections for tumors that cross midline)

Advanced Hypopharyngeal Cancer (T_3 or T_4)

- Multimodality Therapy: organ preservation approach with induction chemotherapy (cisplatin/5-FU) followed by adjuvant radiation therapy (Phase III EORTC Trial, *see* p. 226); surgical salvage (total laryngectomy) for poor responders
- Neck: elective radiation therapy to bilateral necks in conjunction with treatment of the primary

Surgical Management

- **Partial Laryngopharyngectomy**: laryngeal conservation procedure that may be considered for early T_1–T_2 pyriform sinus cancer if tumor involves primarily the medial wall and **>1.5 cm** clear from the apex

- **Combined Suprahyoid and Lateral Pharyngectomy**: may be considered for early T_1–T_2 posterior cricopharyngeal wall cancer; includes removal of lateral third of the thyroid cartilage
- **Total Laryngectomy with Partial Pharyngectomy**: typically required for most postcricoid primaries, advanced hypopharyngeal disease, recurrence, and radiation failures
- **Esophagectomy**: indicated for cervical esophageal involvement
- **Wookey Procedure**: turned-in cervical skin flaps
- Reconstruction: primary closure or skin grafts are rarely appropriate for tension free closure, typically require regional pedicled flap (pectoralis major or trapezius myocutaneous flap, gastric pull-up) or microvascular free flap (*see* Facial Plastics and Reconstruction for complete discussion)

Complications of Gastric Pull-up Reconstruction

- **Anastomosis Breakdown and Leak**: salivary leak results in wound infection, fistula formation, sepsis, mediastinitis, and death; 10–37% incidence of anastomotic disruption; Rx: prevented with tension-free closure, avoidance of infection, maintenance of vascular supply; may require external diversion or re-exploration and repair (increased risk of mortality)
- **Abdominal Complications**: not common, various complications including perforation of the pyloroplasty, peritonitis, wound dehiscence, and splenic injury
- **Cardiopulmonary Complications**: may result from fluid shifts, hemorrhage, pulmonary atelectasis, respiratory failure, pneumothorax
- **Hypocalcemia**: secondary to decreased calcium absorption from decreased gastric acidity and decreased intestinal transient time from the truncal vagotomy and from disruption of the vascular supply to the parathyroid glands; Rx: calcium supplementation
- **Tracheobronchial Injury**: most commonly from injury to the posterior membranous tracheal wall during dissection; Rx: requires primary closure of defect or tissue coverage of defect (eg, adjacent muscle or pedicled flap)

LARYNGEAL CANCER

Introduction

Introduction

- 1–5% of all malignancies
- second most common site for head and neck malignancy

- SSx: hoarseness, aspiration, dysphagia, odynophagia, sore throat, hemoptysis, airway obstruction (stridor), referred otalgia, weight loss, globus sensation
- Risk Factors: smoking and alcohol use; radiation exposure; history of juvenile papillomatosis (HPV), Plummer-Vinson syndrome; exposure to metal, plastics, paint, wood dust, and asbestos

Barriers and Spaces of the Larynx (*see* Figure 5–1)

- **Quadrangular Membrane**: fibroelastic membrane, supports supraglottis, extends from epiglottis to arytenoid and corniculate cartilage
- **Conus Elasticus**: fibroelastic membrane, supports vocal fold, extends from cricoid cartilage to merge with vocal ligament (resists spread of glottic and subglottic cancers)
- **Pre-epiglottic Space**: midline fibrofatty-filled space bounded by the hyoid bone, thyrohyoid membrane, hyoepiglottic ligament, thyroepiglottic ligament, and epiglottis; tumor may enter from anterior commissure or supraglottic extension; continuous with paraglottic space
- **Paraglottic Space**: fibrofatty-filled space outside of conus elasticus and quadrangular membrane; allows transglottic extension
- **Reinke's Space**: superficial lamina propria of true vocal fold, lack of lymphatics and blood vessels permits rapid tumor extension

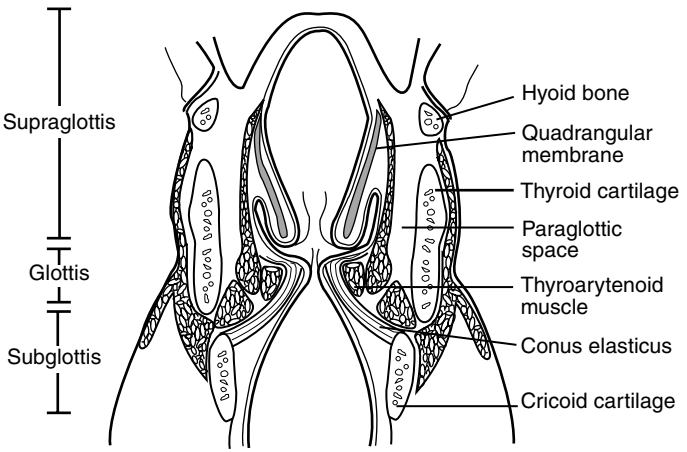

Figure 5–1. Coronal section of the larynx demonstrating the barriers and divisions of the larynx

- **Broyles' Tendon**: insertion of vocalis tendon to thyroid cartilage; allows tumor spread into thyroid cartilage (no perichondrium found at insertion site)

Classification by Site and Staging

Supraglottis

- approximately 30–40% of laryngeal cancer
- SSx: **sore throat, hemoptysis**, aspiration, dysphagia, odynophagia, airway obstruction (stridor), referred otalgia, weight loss, globus sensation
- Inferior Border: apex of the ventricle
- Histology: ciliated pseudostratified columnar epithelium
- Subsites: suprahyoid epiglottis, infrahyoid epiglottis (most common supraglottic site for cancer), aryepiglottic fold, arytenoids, false cords
- supraglottis is derived from the third and fourth branchial arches (glottis and subglottis develop from the sixth branchial arch), an embryologic fusion plate forms between supraglottis and glottis that functions as a tumor barrier and creates separate vascular supplies
- tumor invasion typically occurs superiorly toward base of tongue or pre-epiglottic space
- overall 25–75% risk of regional metastasis, primarily to nodal levels II, III, and IV

Staging (*based on the AJCC Staging*, 1997)

- T_1: primary tumor limited to one subsite
- T_2: primary tumor involves mucosa of adjacent subsite or outside region of supraglottis (glottis, base of tongue, medial wall of pyriform sinus)
- T_3: vocal cord fixation or primary tumor involves postcricoid, pre-epiglottic space, or deep base of tongue
- T_4: invades thyroid cartilage, soft tissues of neck, thyroid, or esophagus

Glottis

- approximately 50–75% of laryngeal cancer
- SSx: **hoarseness**, aspiration, dysphagia, odynophagia, airway obstruction, weight loss, sore throat
- Inferior Border: 1 centimeter below apex of ventricle
- Histology: stratified squamous epithelium
- typically diagnosed at early stages due to early symptomatology (hoarseness) and the barriers of tumor spread (vocal ligament, thyroglottic ligament, and conus elasticus)

- anterior commissure which does not have an inner perichondrium allows for thyroid cartilage invasion
- limited regional metastasis, primary drainage to nodal levels II, III, and IV
- vocal fold fixation suggests involvement of thyroarytenoid, lateral or posterior cricoarytenoid, and interarytenoid muscles; extension into cricoarytenoid joint; or perineural invasion

Staging (*based on the AJCC Staging*, 1997)

- T_1: primary tumor limited to vocal folds (may involve anterior and posterior commissures; one vocal fold involvement [T_{1a}], both vocal fold involvement [T_{1b}]); <5% regional metastasis
- T_2: primary tumor involves subglottis or supraglottis, impaired vocal fold mobility; 5–10% regional metastasis
- T_3: vocal fold fixation; 10–20% regional metastasis
- T_4: invades thyroid cartilage or beyond larynx; 25–40% regional metastasis

Subglottis

- **rare** primary site
- SSx: **airway obstruction** (**biphasic stridor**), hoarseness, dysphagia, odynophagia, hemoptysis, weight loss, sore throat
- Inferior Border: level of cricoid cartilage
- Histology: ciliated pseudostratified columnar epithelium
- often silent, poorly differentiated, usually extends into cricoid cartilage
- poor prognosis
- <20% regional metastasis

Staging (*based on the AJCC Staging*, 1997)

- T_1: primary tumor limited to subglottis
- T_2: primary tumor involves vocal folds with normal mobility
- T_3: primary tumor limited to larynx with fixed cords
- T_4: primary tumor invades thyroid cartilage or beyond larynx

Pathology

Squamous Carcinoma

- >95% of laryngeal cancer
- **Basaloid Squamous**: more aggressive high-grade variant

Verrucous Carcinoma (Akerman's Tumor)

- slow-growing, locally destructive (**rare metastasis**)
- excellent prognosis
- glottis most common site in the larynx
- Gross Lesion: rough, exophytic (warty), fungating, gray-white color
- Histopathology: benign-appearing (nonmitotic, no infiltration), well-differentiated squamous epithelium with papillary projections, extensive hyperkeratosis, basement membrane intact, "**pushing**" margins
- Rx: single-modality radiation versus surgery (controversy surrounds possible malignant transformation of verrucous carcinoma with radiation therapy)

Adenocarcinoma

- 1% of laryngeal cancer
- primarily supraglottic and subglottic regions (follow distribution of the laryngeal mucous glands)
- more aggressive than squamous cell carcinoma
- present submucosal and nonulcerative

Others

- **Adenoid Cystic Carcinoma**: perineural spread, indolent course, may present with distant metastasis years after primary therapy
- **Spindle Cell Carcinoma**: poorly differentiated variant of squamous cell carcinoma with malignant spindle cell stromal component
- **Neuroendocrine Tumors**: paragangliomas, carcinoid tumors, small (oat) cell carcinoma
- **Mucoepidermoid Carcinomas**: low, intermediate, and high grades
- **Sarcomas**: fibrosarcoma, chondrosarcoma, malignant fibrous histocytoma, rhabdosarcoma
- **Metastatic**: rare; primary sites from kidney, prostate, breast, stomach, and lung

Premalignant Glottic Lesions

Introduction

Histological Classification

- **Hyperplasia and Hyperkeratosis**: an increase in the number of cells and keratin production, not a significant risk factor for malignant degeneration
- **Mild Dysplasia**: abnormal dyskaryotic squamous cells with mild atypical cytological alteration involving cell size, shape, color, and organization, not a significant risk factor for malignant degeneration

- **Moderate Dysplasia**: atypical cytological alteration involving cell size, shape, color, and organization, risk factor for malignant degeneration
- **Severe Dysplasia** (**Carcinoma In Situ**): cells demonstrate cytological features of malignancy without invasion beyond the basement membrane (risk of developing invasive carcinoma varies from 15–25%)
- **Microinvasive Carcinoma**: malignant cells involving entire thickness of mucosa with discrete foci that invade beyond the basement membrane
- **Invasive Carcinoma**: malignant cells that frankly invade through the basement membrane

Biopsy Techniques

- **Incisional Biopsy**: more random, risk of "missing" tumor, less morbidity
- **Excisional Biopsy**: may be therapeutic for early glottic carcinoma (more cost effective)
- **Vocal Fold Stripping**: excisional biopsy technique that removes the vocal fold cover, risk of normal tissue stripping, imprecise, impairs mucosal wave
- **Microflap Excision**: excisional biopsy technique that dissects superficial lamina propria; spares vocal ligament; better preservation of the mucosal wave; indicated for superficial and midmembraneous lesions

Management of Severe Dysplasia (Carcinoma In Situ) and Microinvasive Carcinoma

- smoking and alcohol cessation
- may treat initially with radiation therapy versus surgical excision (vocal fold stripping or microflap excision, *see above*)
- may consider radiation for recurrence after initial excision
- close follow-up every 2–3 months for 5 years with low threshold to repeat biopsy
- may consider excisional biopsy every 3 months until 2 consecutive negative results for microinvasive carcinoma

Management of Laryngeal Carcinoma

Early Supraglottic Carcinoma (T_1N_0–T_2N_0)

- Single-Modality Therapy: primary radiation (external beam) to primary site (surgical salvage for failure) versus supraglottic laryngectomy

- N_0 Neck: elective bilateral selective neck dissection versus elective radiation therapy to neck regardless of presence of clinically evident nodes; if surgical specimen is positive for tumor may consider observation, completion of a comprehensive neck dissection, or radiation therapy to neck
- N_{1-3} Neck: radical neck dissection for clinical nodes
- Adjuvant Therapy: postoperative radiation therapy may be considered for positive or close margins; multiple positive neck nodes or extracapsular extension; perineural or intravascular invasion; bone, cartilage or soft tissue invasion; or if an emergent tracheotomy was required (increased risk of tumor seeding)

Advanced Supraglottic Cancer (T_3–T_4)

- Multimodality Therapy: total laryngectomy (extended supraglottic laryngectomy for select T_3 and T_4) and postoperative radiation therapy versus chemotherapy and radiation therapy for organ preservation or for nonoperable candidates
- N_0 Neck: elective ipsilateral selective neck dissection (lateral) regardless of presence of clinically evident nodes
- N_{1-3} Neck: radical neck dissection for clinical nodes

Early Glottic Carcinoma (T_1–T_2)

- Single-Modality Therapy: limited-field primary external beam radiation therapy or surgical management (cordectomy, endoscopic procedures, or partial laryngectomies) for failed radiation therapy or patient requesting quick definitive therapy (away from radiation centers)
- Neck: elective neck dissections are **not** indicated for early glottic cancer, however, for rare clinical nodes may address with radiation or modified neck dissection

Advanced Glottic Cancer (T_3–T_4)

- Multimodality Therapy: total laryngectomy (may consider conservation laryngectomy) with postoperative radiation versus organ preservation trial (radiation and chemotherapy) with salvage total laryngectomy for poor responders (Veterans Affairs Laryngeal Study Group, *see* Chemotherapy above) or primary radiation therapy with surgical salvage
- Neck: elective ipsilateral radical neck dissection (may consider selective neck dissection) regardless of presence of clinical nodes
- Phase III trials comparing induction chemotherapy versus radiation therapy alone versus concomitant chemotherapy (organ preservation) is currently being investigated

Subglottic Cancer

- no large investigative series exist due to the rarity of subglottic cancer as a primary tumor site
- Single or Multimodality Therapy: extended total laryngectomy with postoperative radiation and/or chemotherapy (for advanced disease) versus primary radiation or chemotherapy and radiation therapy to primary and neck for nonoperable candidates (organ preservation)
- Neck: ipsilateral radical neck dissection for clinical nodal disease

Surgical Techniques

General Contraindications for Partial Laryngectomies

- Fixed cords (except supracricoid laryngectomies)
- Cartilage invasion
- Subglottic extension
- Significant oropharyngeal extension
- Interarytenoid involvement
- Tumor spread into neck

Cordectomy via Laryngofissure or Transoral (Laser)

- Indications: T_1 glottic cancer limited to middle third of the vocal fold; no extension of tumor to vocal process or anterior commissure; no invasion into subglottis, ventricle, or false cord
- Technique: external approach, divides laryngeal cartilage at midline, enter glottis at anterior commissure to remove involved vocal fold up to the vocal process of the arytenoid; transoral approaches may utilize laser (CO_2)
- Advantages: external approach provides better access, transoral approach avoids external scar and tracheotomy
- Disadvantages: external approach requires initial tracheotomy, morbidity associated with external approach

Supraglottic Laryngectomy (Horizontal Hemilaryngectomy)

- Rationale for Procedure: embryological boundary between false and true vocal folds results in independent lymphatic drainage, supraglottic cancer tends to have pushing borders rather than infiltrating borders
- Indications: T_1 or T_2 (limited T_3) supraglottic tumors; tumor does not involve vocal fold, ventricle, thyroid cartilage, arytenoid, interarytenoid region, pyriform, or base of tongue; good pulmonary function tests (forced expiratory volume [FEV_1] >50–60%); patient must also give consent for possible total laryngectomy

- Technique: removes epiglottis, aryepiglottic folds, false vocal folds, preepiglottic space, portion of the hyoid bone and thyroid cartilage (spares true vocal folds and arytenoids)
- Advantages: good voice quality, potential for decannulation, adequate swallow
- Disadvantages: requires initial tracheotomy, extensive postoperative rehabilitation for swallowing (especially after postoperative radiation)
- **Extended Supraglottic Laryngectomy**: may extend to include excision of base of tongue, hypopharynx, or 1 arytenoid
- **Radiation Salvage**: surgery possible for select small original primaries in which the recurrent cancer correlates with the original primary lesion
- **Endoscopic Laser Supraglottic Laryngectomy**: may be considered for T_1 or T_2 (limited T_3) supraglottic tumors that may be accessed endoscopically (eg, suprahyoid epiglottis, aryepiglottic folds, vestibular folds, pharyngoepiglottic folds), tracheotomies are not routinely required, typically provides improved postoperative swallowing function when compared with the transcervical technique (preserves superior laryngeal nerve, tongue base, hyoid bone, and suprahyoid muscles)

Vertical Partial Laryngectomy (Hemilaryngectomy)

- Indications: select T_1–T_2 glottic carcinomas; tumor does not extend beyond ⅓ of opposite cord; <10 mm of anterior subglottic extension, or <5 mm of posterior subglottic extension; no posterior commissure, cricoarytenoid joint, aryepiglottic fold, posterior surface of the arytenoid, or paraglottic space involvement, good pulmonary function tests (forced expiratory volume [FEV_1] >50–60%), patient must also give consent for possible total laryngectomy
- Technique: removes one vocal fold from anterior commissure to vocal process (½ of the opposite vocal fold may be removed), ipsilateral false cord, ventricle, paraglottic space, and overlying thyroid cartilage (3 mm posterior strip of cartilage preserved)
- Advantages: allows decannulation, functional glottic voice
- Disadvantages: risk of aspiration, requires initial tracheotomy
- **Radiation Salvage**: surgery possible for select small original primaries in which the recurrent cancer correlates with the original primary lesion
- **Extended Hemilaryngectomy**: select T_3 lesions or arytenoid involvement; removes one vocal fold, arytenoid, and overlying thyroid cartilage (3 mm posterior strip of cartilage preserved)

Supracricoid Laryngectomy

- Rationale for Procedure: goal is to achieve decannulation and good swallowing and voice function with comparable local control and survival rates versus a total laryngectomy

- Indications: select $T_{2–4}$ glottic and supraglottic cancers that may involve the pre-epiglottic space, paraglottic space, ventricle, limited thyroid cartilage, or epiglottis; good pulmonary function tests (forced expiratory volume [FEV_1] >50–60%); patient must also give consent for possible total laryngectomy
- Contraindications: arytenoid fixation; infraglottic extent of tumor reaching upper border of cricoid cartilage; major pre-epiglottic involvement; invasion of the cricoid cartilage, perichondrium of thyroid cartilage, hyoid bone, posterior arytenoid mucosa; extralaryngeal involvement; or poor pulmonary function
- Technique: remove entire thyroid cartilage bilateral true and false vocal folds, one arytenoid (may spare both arytenoids if not involved), and paraglottic space; spares cricoid cartilage, hyoid bone, and at least one arytenoid cartilage (for speech and swallowing); may reconstruct with **cricohyoidopexy** (CHP) or **cricohyoidoepiglottopexy** (CHEP) if epiglottis is spared
- Advantages: allows decannulation, functional glottic voice
- Disadvantages: risk of aspiration, requires initial tracheotomy, dysphonia
- **Radiation Salvage**: surgery possible for select small original primaries in which the recurrent cancer correlates with the original primary lesion

Total Laryngectomy

- Indications: standard therapy for any laryngeal cancer that precludes conservative management (advanced disease, recurrence, radiation failure, patients with poor pulmonary function)
- Technique: removes entire larynx (true and false vocal folds, cricoid and thyroid cartilage, both arytenoids, epiglottis, pre-epiglottic and paraglottic spaces, and hyoid bone), creates complete separation between pharynx and trachea
- Advantages: no risk of aspiration, fair voice quality (with tracheoesophageal speech)
- Disadvantages: requires a permanent stoma
- **Near-Total Laryngectomy (3/4 Laryngectomy)**: creates a communication between trachea and pharynx for phonation, must keep one arytenoid to prevent aspiration through shunt

Postoperative Complications

- **Fistulas**: increased instance with radiation therapy; Rx: hold oral feeding (must however maximize nutritional status by parenteral or tube feeding for wound healing), medialize fistula if possible involvement of carotid artery (to prevent carotid blow-out),

antibiotic regimen (culture and sensitivity of wound drainage), daily loose wet to dry packing of fistula site to stimulate granulation tissue, local debridement of necrotic tissue, consider flap closure for recalcitrant drainage

- **Tracheotomy Complications**: pneumothorax, hemorrhage, subcutaneous emphysema (*see also* p. 97)
- **Speech Alteration**: variety of postoperative causes granulation tissue, scar formation, glottic incompetence, vocal fold paralysis; Rx: postoperatively all patients should follow up with speech therapist for videostroboscopy and voice analysis to address phonatory defects
- **Persistent Aspiration, Bronchopneumonia, and Deglutition Problems**: increased incidence with the elderly (poor pulmonary function), conservation laryngectomies, excision of arytenoid or base of tongue, and vagal nerve injury; Rx: postoperatively patients must follow-up with a swallowing therapist to evaluate for aspiration and swallowing disorders; therapy varies depending on etiology from swallowing rehabilitation to laryngeal reconstruction surgery (*see also* pp. 143–147)
- **Delayed Decannulation**: typically secondary to laryngeal edema and laryngeal stenosis (*see below*); Rx: must examine endoscopically for recurrence or persistent disease; may then consider steroid injections, endoscopic excision (CO_2 laser), or open thyrotomy with mucosal flap and resurfacing
- **Esophageal or Pharyngeal Stenosis**: typically secondary to scarring; Rx: must examine endoscopically to examine for recurrence; may then consider serial dilation, endoscopic excision (CO_2 laser), or pharyngoesophageal reconstruction with regional or free flap transfer
- **Perichondritis and Chondritis**: increased risk with inadequate coverage of bare cartilage; Rx: antibiotic regimen; may consider local debridement, resection of infected cartilage with reconstruction, or hyperbaric oxygen therapy
- **Stomal Stenosis**: prevented with bivona prosthesis; Rx: must evaluate for stomal recurrence, may require surgical dilation
- (*see also* Complications of Neck Dissections, pp. 238–239)

Postoperative Voice Management

Artificial Larynx (Electrolarynx)

- Mechanism: sound introduced into neck tissue and emanated from the mouth or oral type that directs sound directly to the oral cavity
- Advantages: easy to learn, inexpensive, loud

- Disadvantages: requires a power source, produces a mechanical voice, oral tubes may be required

Esophageal Speech

- Mechanism: esophageal speech is produced by vibration of pharyngoesophageal mucosa; requires trapping air in mouth, injecting air into the esophagus, and expulsion of air to create voice
- Advantages: more natural voice, hands free, no appliance required
- Disadvantages: difficult learning (30% success rate), quiet speech

Tracheoesophageal Puncture

- Mechanism: one-way valve allows air to enter esophagus to allow esophageal speech which is produced by vibration of the pharyngoesophageal mucosa
- Advantages: more natural voice quality, less conspicuous
- Disadvantages: may not work (most common failure from pharyngeal constrictor spasm), requires hands, risk of aspiration and esophageal prolapse

NOTE: postlaryngectomy patients may always elect writing as their form of communication without voicing

NASOPHARYNGEAL CANCER

Introduction

Introduction

- SSx: **neck mass** (most common initial symptom, 70%), serous otitis media from eustachian tube obstruction (second most common presentation, 50%), nasal obstruction, cranial nerve palsies (abducens nerve most common cranial nerve palsy), recurrent epistaxis, trismus, headache
- Risk Factors: regional distribution (Southern China, Northern Africa, Southeast Asia, Alaska, Greenland), Epstein-Barr Virus (EBV), genetic predisposition

Epstein-Barr Virus Association

- **Early Intracellular Antigen (EA)**: early intracellular antigen
- **Viral Capsule Antigen (VCA)**: late antigen, most specific immunological finding in nasopharyngeal cancer

- low titers of the IgA EA and IgA VCA and Antibody Dependent Cell-mediated Cytotoxicity (ADCC) for other EBV antigens predict poorer prognosis
- elevated IgA VCA and EA may be used for screening for nasopharyngeal carcinoma in high risk regions

Anatomy

- **Nasopharynx**: anteriorly bordered by the choanae, superiorly bordered by the base of skull (body of the sphenoid), inferiorly bordered by the plane of the soft palate, laterally bordered by eustachian tube orifices and superior constrictor muscles
- **Fossa of Rosenmüller**: slit-like region medial to the medial crura of the eustachian tube orifice, most common site of tumor in the nasopharynx
- **Passavant's Ridge**: interdigitating superior constrictor muscles that form a band at the posterior pharyngeal wall during swallowing that abuts the soft palate
- Subsites: posterior nasopharyngeal wall, lateral nasopharyngeal wall, soft palate
- Histology: lymphoid tissue and epithelium (pseudostratified ciliated columnar cells and stratified squamous cells)

Classification and Management

Staging (*based on the AJCC Staging*, 1997)

- T_1: primary tumor confined to nasopharynx
- T_2: primary tumor extension into nasal fossa or oropharynx (without parapharyngeal extension [T_{2a}], with parapharyngeal extension [T_{2b}])
- T_3: invasion of bony structures or paranasal sinuses
- T_4: invasion into intracranium, cranial nerves, infratemporal fossa, hypopharynx, or orbit
- NOTE: previous AJCC early staging was based on subsite involvement (1 subsite, T_1; more than 1 subsite, T_2; invasion of nasal cavity/oropharynx, T_3; invasion into skull or cranial nerves, T_4)

World Health Organization (WHO) Classification

- **WHO Type I**: **Keratinizing Squamous Cell Carcinoma**, squamous differentiation, **not** associated with EBV, worse prognosis, less sensitive to radiation

- **WHO II**: **Nonkeratinizing Squamous Cell Carcinoma**, does not demonstrate definite squamous differentiation, associated with EBV, better prognosis, sensitive to radiation
- **WHO III**: **Undifferentiated** (includes lymphoepitheliomas, anaplastic, and clear cell variants): indistinct cell margins, may have lymphocytic stroma (**lymphoepitheliomas**), associated with EBV, better prognosis, sensitive to radiation
- Other Types: Lymphoma, Adenocarcinoma, Plasma Cell Myelomas, Cylindromas, Adenocystic Carcinoma, Melanoma, Carcinosarcoma, Unclassified Spindling Malignant Neoplasm

Management

- Stage I and II: not a surgical disease, radiotherapy to primary site and bilateral necks (regardless of nodal status)
- Stage III and IV: concurrent chemoradiotherapy (cisplatin/5-FU) followed by adjuvant chemotherapy (Phase III Intergroup Study 0099, *see* p. 227)
- limited surgical role (neck dissections may be considered for postradiation salvage surgery)

NASAL AND PARANASAL CANCER

Introduction

Introduction

- Risk Factors: toxin exposure (nickel, chromium, mustard gas, hydrocarbons, and radium); woodworkers (adenocarcinoma); chronic infection; previous radiation
- SSx: unilateral persistent sinusitis, nasal obstruction, or epistaxis; nasal lesions (exophytic, ulcerative, septal perforation); orbital symptoms (proptosis, diplopia, vision loss); anosmia and taste disturbances; palatal fistula; cranial nerve palsies, headache, facial paresthesias, and pain
- Prognosis: maxillary tumors in general have 5-year survival rates between 20–50%, tumors posterior or superior to **Ohngren's line** (plane from medial canthus to angle of jaw) suggest poorer prognosis due to higher instances of skull base invasion and perineural spread
- maxillary sinus is the most common site for nasal and paranasal cancer (nasal cavity second most common)

Paranasal Subsites

- **Paranasal Sinuses**: (*see* pp. 2–8 for anatomy) maxillary sinus most common site involved (ethmoid sinus is the second most common),

typically presents in an advanced stage (90% invade more than one wall), sphenoid sinus has a higher instance of neurological sequelae (cranial nerve palsies, orbital symptoms, headache), frontal sinus rare site of primary cancer

- **Nasal Cavity**: (*see* pp. 5–8 for anatomy) <10% regional metastatic potential; 60% 5-year survival; worse prognosis for larger tumors, extension outside the nasal cavity, bony involvement, and regional metastasis
- **Anterior Cranial Fossa**: bordered posteriorly by the lesser wing of the sphenoid and optic chiasm and extends to the frontal bone; the floor contains the orbital plates of the frontal bone, fovea ethmoidalis, and cribriform plate
- **Pterygopalatine Fossa**: pyramidal-shaped space below the apex of the orbit and between the posterior wall of the maxillary sinus and the pterygoid plates; contains the foramen rotundum (CN V_2), vidian nerve, sphenopalatine nerve, lesser and greater palatine nerves, sphenopalatine ganglion, and internal maxillary artery; invasion of this space carries a poor prognosis
- **Infratemporal Fossa**: bounded anteriorly by the maxilla, posteriorly by the glenoid fossa and mandible, and medially by the lateral pterygoid plates; roof contains the foramen ovale (CN V_3) and spinosum (middle meningeal artery and vein), also contains the pterygoid muscles
- **Orbital Cavity**: invasion into bony orbit or orbital apex requires orbit exenteration

Pathology

Epithelial Malignancy

- **Squamous Cell Carcinoma**: most common malignancy in sinonasal tissue, nasosinusoidal lesions are more aggressive and have a higher rate of metastasis, majority are moderately differentiated keratinizing type
- **Adenocarcinoma**: increased instances with wood and leather workers, presents more commonly in the ethmoid sinuses
- **Adenocystic Carcinoma**: salivary gland tumor (more common in the minor salivary glands), insidious growth (distant metastasis may present years later), perineural extension may present with pain
- **Mucoepidermoid Carcinoma**: rare, salivary gland tumor
- **Melanoma**: usually metastatic, more common in nasal cavity (anterior nasal septum), high risk of local recurrence

- **Olfactory Neuroblastoma** (**Esthesioneuroblastoma**): rare tumor, arises from olfactory epithelium, bimodal frequency (presents in teenagers and the elderly), presents as a pink or brown friable nasal mass, excellent prognosis if confined to nasal cavity; Rx: surgical excision with postoperative radiation treatment of choice
- **Sinonasal Undifferentiated Carcinoma** (**SNUC**): characterized by extensive tissue destruction, rapidly progressing tumor, extremely poor prognosis
- **Small Cell Carcinoma**: extremely poor prognosis; Rx: nonsurgical management, chemotherapy and radiation therapy
- NOTE: pathological differentiation may require immunohistochemical analysis (*see* pp. 221–222)

Nonepithelial Malignancy

- **Rhabdomyosarcoma**: more aggressive than other head and neck sites; embryological, alveolar, and pleomorphic (more common in adults) subclasses; management in children is typically radiation and chemotherapy, adults typically require initial wide surgical excision with postoperative radiation
- **Hemangiopericytoma**: rare tumor, highly vascular, arises from **pericytes of Zimmermann** (located outside of endothelial cells, capable of changing the caliber of the capillaries), presents as a lobulated mass and epistaxis, may require preoperative embolization prior to excision
- **Neurogenic Sarcoma**: rare, aggressive, associated with neurofibromatosis
- **Leiomyosarcoma**: smooth muscle tumor, poor prognosis
- **Fibrosarcoma**: tumors of fibroblasts, associated with trauma and radiation
- **Angiosarcomas**: slow growing but locally aggressive with indistinct margins
- **Osteogenic Sarcomas and Chondrosarcomas**: aggressive, poor prognosis (10–20% 5-year survival), arise from the facial skeleton, more common in the mandible than in the maxilla
- **Lymphomas**: typically non-Hodgkin's (*see* pp. 274–276)
- **Metastatic Tumors**: renal cell carcinoma most common

Management

Treatment Concepts

- Early Stages: single-modality primarily with surgical resection for early maxillary or nasal cavity tumors except for small cell carcinoma,

lymphoma, and rhabdomyosarcoma (except for debulking, postoperative radiation therapy may be considered for close or positive margins or bony invasion) versus primary radiation therapy (radiation therapy is limited by orbital and intracranial complications, eg, blindness, keratitis, brain necrosis)
- Advanced Stages: multimodality therapy with either primary radiation therapy with surgical salvage or primary surgical excision with postoperative radiation therapy, inoperable patients for whom resection would result in severe cosmetic and functional morbidity (eg, sphenoid tumors) should undergo primary radiation therapy with surgical salvage
- Unresectable/Nonoperable Candidates: may consider primary radiation, chemotherapy, or combination radiochemotherapy
- N_0 Neck: elective neck dissection generally not indicated (may be considered if tumor involves the nasopharynx or soft palate) otherwise treat neck for clinical nodal disease only (<10% occult metastasis)
- N_{1-3} Neck: radical neck dissection for clinical nodes; parotid nodes require a superficial parotidectomy
- some nonsquamous tumors (eg, rhabdomyosarcomas, small cell carcinomas) may benefit from chemotherapy

Surgical Management

- surgery may be considered for cranial base extension (infratemporal fossa), absolute contraindications for surgical resection include involvement of significant brain parenchyma, cavernous sinus, bilateral orbits, optic chiasm, or carotid artery (relative contraindication)
- orbit exenteration is indicated when there is periorbital bony erosion or tumor is within the orbital apex (via direct bony erosion, perineural or perivascular spread, or infraorbital fissure or nasolacrimal duct extension)
- craniotomy (craniofacial resection) is indicated if tumor is involved superior to cribriform plate or roof of the ethmoid sinuses

Transfacial Approaches

- **Endoscopic Excision**: low morbidity, no external scar, limited to benign small lesions
- **Lateral Rhinotomy**: incision begins at medial aspect of the eyebrow, follows along the nasal lateral wall, and around alar crease to the philtrum and a sublabial incision; may extend incision to include the

lip for exposure of hard palate; standard approach for medial maxillectomies, provides excellent visualization of maxillary wall, ethmoid sinus, sphenoid sinus, medial orbital wall, and nasal cavity
- **Midfacial Degloving**: requires a transfixation, bilateral intercartilaginous and gingivobuccal incisions; limited to lesions of inferior and medial maxillary walls (limited superior and posterior visualization); avoids external incisions and allows bilateral exposure
- **Facial Translocation**: indicated for wide exposure of the middle cranial base, infratemporal fossa, pterygopalatine fossa, and nasopharynx, requires extensive facial flaps, more limited degree of facial nerve transection (requires only transection of the frontal branch)
- **Transpalatal Approach**: indicated for tumors involving the floor of nose or inferior portion of the maxilla, may be used in conjunction with a lateral rhinotomy
- **Weber-Fergusson Approach**: consists of a lateral rhinotomy incision with a lip-splitting incision to connect with the sublabial incision (and possibly a subciliary or transconjuctival incision), allows additional exposure of maxilla for a total maxillectomy
- **Infratemporal Fossa Approach**: access obtained via a preauricular or postauricular incision with extension in a hemicoronal and cervical fashion, may gain additional exposure by retracing the temporomandibular joint
- **Combined Craniofacial Approach**: combines the above facial incisions with a bicoronal incision to allow exposure for a craniotomy

Cranial Approaches

- **Wide Frontal Craniotomy**: standard approach
- **Narrow Frontal Craniotomy**: limited approach
- **Subfrontal Craniotomy**: remove frontal bar complex, allows option to avoid a transfacial approach

Surgical Resection Types

- **Medial Maxillectomy**: removes lateral nasal wall and medial maxilla, may also include a complete sphenoethmoidectomy, standard resection for inverting papillomas (*see* pp. 18–19)
- **Inferior Maxillectomy**: resects the inferior portion of the maxillary sinus inferior to the infraorbital nerve, indicated for tumors involving the maxillary alveolar process or limited hard palate lesions

- **Total Maxillectomy**: en bloc removal of entire maxilla, indicated for tumors involving the maxillary antrum
- **Radical Maxillectomy**: total maxillectomy with orbital exenteration
- **Craniofacial Resection**: en bloc removal of the anterior cranial base including the cribiform plate and ethmoid sinuses, may require dural excision

Reconstruction

- **Fascial, Temporalis Muscle, or Pericranial/Galeal Flaps**: provides additional support for watertight closure of skull base and dural defects allowing for adequate cranionasal separation
- **Regional or Microvascular Free Flaps**: provides soft tissue bulk (*see* Facial Plastics and Reconstruction for a complete discussion)
- **Prosthetic Obturator**: indicated for inferior maxillary or palatal defects, provides oronasal separation, a surgical obturator is placed initially to allow better speech and swallowing function and is later replaced with a permanent device

Surgical Complications

- **Intracranial Infections** (**Meningitis, Cerebritis**): most significant complication, may occur with a CSF leak, life threatening; Rx: *see* pp. 47–48
- **CSF Leak**: prevented by avoiding dissection above cribiform plate, watertight closure of the dura, consider lumbar drainage for high-risk procedures, avoid postoperative straining; Rx: *see* p. 330
- **Cerebrovascular Occlusion**: may be secondary to excessive brain retraction
- **Orbital Complications**: extraocular muscle entrapment, optic nerve compression, blindness; Rx: *see* pp. 45–47
- **Frontal Lobe Syndrome**: frontal lobe dysfunction results in changes in patient's affect, may be permanent
- **Osteitis**: may be secondary to a wound infection or skin flap necrosis
- **Epiphora**: typically from lacrimal duct injury; Rx: consider dacryocystorhinostomy if no resolution
- **Hemorrhage**: increased risk with vascular tumors, may consider preoperative embolization of select tumors; Rx: electrocautery, nasal or surgical cavity packing
- **Tension Pneumocephalus**: occurs from trapped air that enters into the cranial cavity from the sinonasal tract, high risk of brain herniation, may be caused by excessive lumbar CSF drainage; Rx: emergent aspiration, re-exploration to define leak

- **Cranial Nerve Injuries**: combined injuries to cranial nerves III, IV, and VI occur in **cavernous sinus syndrome** which causes total ophthalmoplegia of the affected globe; *see also* pp. 114–119 *for management of vagal injuries; see also* p. 368 *for facial nerve injuries*; other cranial nerve injury at risk are cranial nerves II and V

CUTANEOUS MALIGNANCIES

Basal Cell Carcinoma

Introduction

- approximately 60% of cutaneous neoplasms
- indolent course, extends peripherally without vertical invasion
- rare metastasis
- cutaneous lesions located in the embryonic fusion plates of the face (nasolabial folds, floor of nose, columella, preauricular regions, inner and outer canthus of the eye) are more aggressive and have a higher risk of recurrence, therefore require close follow-up
- 90% cure rate in the head and neck regions
- Risks: ultraviolet light exposure (UV-B), fair skin, blue-green eyes, sunburns as a child, regions of previous burns (**Marjolin's ulcer**) and scars, radiodermatitis, arsenic exposure, immunosuppression, xeroderma pigmentosum (autosomal recessive disorder that causes a defect in DNA repair), previous basal cell carcinoma
- Dx: excisional biopsy of suspicious lesions

Histopathology

- principal cells resemble basal cells (small, dark-blue cells with minimal cytoplasm) of the epidermis
- **Undifferentiated** (**Solid Type**): proliferation of basaloid cells that extend into the papillary dermis, peripheral columnar cells arranged in palisades
- **Keratotic**: differentiated to hair-like structures
- **Cystic**: differentiated to sebaceous gland-like structures
- **Adenoid**: differentiated to tubular structures, lace-like histological pattern

Types

- **Nodular** (**Noduloulcerative**): **most common**; pearly, telangiectatic papule, central ulceration, "rolled" appearance at the base

- **Superficial**: typically found in the trunk and extremities (rare in the head and neck); scaly, waxy, indurated, irregular shapes (similar to eczema)
- **Morphea** (**Sclerosing** or **Fibrosing**): common on the face; flat or depressed, indurated, yellow, indistinct borders (similar appearance to a scar); resembles the cutaneous form of scleroderma; **aggressive**, higher rate of recurrence, worst prognosis
- **Pigmented**: similar to nodular type, however more pigmented, resembles a melanoma or benign nevus
- **Fibroepitheliomas**: raised, firm, pedunculated or sessile, red with smooth skin surface

Management

- avoid excess sun exposure (sunblock)
- careful follow-up for recurrence and second primaries every 4–6 months
- **Excisional Curettage with Electrodesiccation**: most common treatment modality, ideal for small (<2 cm) solid-type, contraindicated for morphea lesions
- **Cryosurgery**: intense cold causes tissue necrosis, requires freeze-thaw-freeze technique for deeper destruction of tissue, requires a 5 mm margin, may be considered for small (<1 cm) lesions
- **Scalpel Excision**: should have 4 mm margin with primary reconstruction (consider intraoperative frozen sections)
- **Radiation Therapy**: may be considered where cosmetic outcome is important (eyelid, nose, lip) or nonoperable candidates, advanced stages may be followed by surgical salvage

Mohs Micrographic Surgery (Chemosurgery)

- technique utilizing intraoperative microscopic examination of all resection margins for complete excision (approximately 96% success)
- Indications: recurrence, morphea type, high risk of recurrence regions ("H-zone"), cosmetic regions in which reconstruction is difficult (eyelids, lips, nose, ears, nasolabial fold)
- Technique: excise lesion while maintaining proper orientation in order to create a tumor map of the specimen, section specimen and fix to slides, examine microscopically for adequate margins, re-excise regions involved with tumor, repeat until histologic sections are free of tumor

Nevoid Basal Cell Carcinoma Syndrome (Gorlin's)

- Pathophysiology: autosomal dominant disorder
- SSx: multiple basal cell carcinomas appear at early age, **odontogenic keratocyst** (*see* pp. 181–182), rib abnormalities (bifid ribs), scoliosis, mental retardation, frontal bossing
- Rx: excise malignant lesions, close follow-up every 3–6 months

Squamous Cell Carcinoma

Introduction

- approximately 30% of cutaneous neoplasms
- 1–4% metastatic potential
- tendency for vertical growth
- more aggressive lesions and higher recurrence rates arise from lesions from previous scars and wounds, lesions located on embryonic fusion plates (nasolabial folds, floor of nose, columella, preauricular regions, inner and outer canthus of the eye), lesions arising de novo (non-sun-exposed skin), and lesions deep (>6 mm) and large in size (>2 cm)
- Risks: similar to basal cell carcinoma; additional risk factors include **actinic keratosis** (warty, sandpaper-like, scaly lesions on sun-exposed skin), previous squamous cell carcinoma lesions, and infection by the human papilloma virus
- SSx: erythematous, hyperkeratotic, opaque nodule, ulcerative, granular base, bleeds easy

Histopathology Variants

- **Solar Keratosis**: atypical squamous cells extend past the dermal-epidermal junction into the papillary dermis, may not exhibit keratin pearls
- **Spindle Cell**: occurs more commonly at site of a scar, trauma, or burn; elongated nuclei arranged in a swirling pattern, more aggressive form
- **Adenoid**: pseudoglandular arrangement, acantholytic dyskeratotic cells, commonly located periauricularly in the elderly, more aggressive form
- **Verrucous**: warty appearance, less aggressive (low-grade malignancy)
- **Bowen's Disease**: erythematous patch or plaque, may have scales, noninvasive (carcinoma in situ)

Management

- avoid excess sun exposure (sunblock)

- careful follow-up for recurrence and second primaries initially every 1 month for 6 months then every 4–6 months
- **Surgical Excision**: should have 4–6 mm margins for early disease and 1–2 cm margins for advanced disease
- **Radiation Therapy**: may be considered where cosmetic outcome is important (eyelid, nose, lip) or nonoperable candidates, advanced stages may be followed by surgical salvage
- **Mohs Micrographic Surgery** (**Chemosurgery**): similar indications as basal cell carcinoma
- **Neck Dissection and Superficial Parotidectomy**: indicated for clinically positive nodes only
- **Postoperative Radiation Therapy**: may be considered for close or positive margins; multiple positive neck nodes or extracapsular extension; perineural or intravascular invasion; recurrent lesions; or bone or cartilage invasion

Melanoma of the Head and Neck

Introduction

- **10–25%** of melanomas present in head and neck (most common site in the head and neck is on the skin of the cheek and the occipital scalp)
- Risks: ultraviolet light exposure (UV-B), sunburns in childhood, fair skin (blue-green eyes), immunosuppression, large congenital nevi (>20 cm), sporadic or inherited **dysplastic nevi** (**Familial Dysplastic Nevus Syndrome**, autosomal dominant), genetic disposition, previous melanomas
- ABCD's of Melanoma: **A**symmetric (irregular shaped), **B**order irregularity, **C**olor variation (color changes within the lesion), **D**iameter (>1 cm, increasing size), also bleeding and ulceration
- Dx: excisional biopsy of suspicious lesions (shave biopsy, curettage, and laser excision is contraindicated if melanoma is suspected); **HMB-45** tumor marker specific for melanoma (*see* Immunohistochemistry *above*)
- Mucosal Melanomas: 6–10% of head and neck melanomas, most common mucosal location is on the hard palate, lower rate of metastasis

Differential Diagnosis of Neuroendocrine Malignancies

- Pigmented: melanoma
- Nonpigmented Small Cell: lymphoma, small cell carcinoma, melanoma, neuroendocrine carcinoma, esthesioblastoma

- Nonpigmented Large Cell: lymphoma, melanoma, neuroendocrine carcinoma
- Nonpigmented Mixed Cell: lymphoma, undifferentiated carcinoma, sarcoma, melanoma
- Nonpigmented Spindle Cell: sarcoma, undifferentiated carcinoma, melanoma

Types and Histopathology

- **Superficial Spreading Melanoma**: most common, 70% from pre-existing **junctional nevi**, radial phase predominates (lateral growth), ulceration suggests vertical phase growth, neoplastic melanocytes form aggregates and invades all levels of the dermis
- **Nodular Melanoma**: very aggressive (rapid vertical phase), may present on non-sun-exposed areas (de novo), **worst prognosis**, atypical melanocytes invade into the reticular dermis
- **Lentigo Maligna**: irregularly hyperpigmented macule, sun-exposed skin, more common in the elderly, common on the head and neck, confined to epidermis, spreads laterally (extended radial phase), **best prognosis**, atypical melanocytes tend to remain at the dermal-epidermal junction
- **Acral Lentiginous Melanoma**: presents on soles of feet and hand, common among Blacks, histologically presents with acanthosis and lentiginous proliferation with lateral growth

Level Classification, Staging, and Prognosis

Clark's Levels

I primary tumor involves epidermis only
II primary tumor invades through basal cell layer
III primary tumor fills papillary dermis
IV primary tumor involves reticular dermis
V primary tumor involves subcutaneous tissue

Breslow's Levels

I <0.76 mm
II 0.76–1.49 mm
III 1.50–3.99 mm
IV ≥4.0 mm

Pathologic Staging (*based on the AJCC Staging,* 1992)

- **pT_0**: atypical melanocyte hyperplasia, Clark Level I

- **pT_1**: invasion of papillary dermis (Level II) or <0.76 mm thick (superficial)
- **pT_2**: invasion of papillary-reticular dermis (Level II–III) or 0.76–1.5 mm thick (intermediate)
- **pT_3**: invasion of reticular dermis (Level IV) or 1.51–3.99 mm thick (intermediate)
- **pT_4**: invasion of subcutaneous tissue (Level V) or ≥4.00 mm thick (deep)

Prognosis and Metastasis Potential

- overall 5–20% recurrence rate
- *see* Table 5–2
- poorer prognosis also associated with the presence of ulceration, scalp lesions, microscopic satellites, and nodal disease
- distant metastasis has a grave prognosis

Management

- avoid excess sun exposure (sunblock)
- careful follow-up for recurrence and second primaries every 4–6 months

Melanomas <0.76 mm Thick (Superficial)

- Single-Modality Therapy: excision with **1 cm** margin down to fascia
- N_0 Neck: elective neck dissection not indicated for superficial lesions

Melanomas 0.76–3.99 mm Thick (Intermediate)

- Single-Modality Therapy: excision with **2 cm** margin down to fascia
- Adjuvant Therapy: may consider interferon α-2b (*see* next page)
- N_0 Neck: may consider sentinel lymph node biopsy (*see below*), if positive should complete a neck dissection (posterior lateral neck

Table 5–2. Melanoma 5-Year Survival and Metastasis Potential Based on Pathological Staging

Stage	5-Year Survival	Nodal Metastasis	Distant Metastasis
pT_1	>95%	2–3%	
pT_2	80–94%	20–25%	8%
pT_3	40–84%	57%	15%
pT_4	10–30%	62%	72%

dissections may be considered for scalp lesions); Intergroup Melanoma Surgical Program revealed an overall 5-year survival benefit with elective neck dissections for patients <60 years old with 1–2 mm melanomas, especially for tumors without ulcerations (*Ann Surg.* 1996; 224:255–266)
- $\underline{N_{1-3}\ \text{Neck}}$: neck dissection (may require a superficial parotidectomy), may consider chemotherapy

Melanomas ≥4.0 mm Thick (Deep)

- Single-Modality Therapy: excision with **3 cm** margin down to fascia
- Adjuvant Therapy: may consider interferon α-2b (*see below*)
- $\underline{N_0\ \text{Neck}}$: elective neck dissections not indicated
- $\underline{N_{1-3}\ \text{Neck}}$: neck dissection (may require a superficial parotidectomy), may consider chemotherapy

Distant Metastasis

- dismal prognosis
- may consider chemotherapy

Interferon α-2b (IFNα-2b)

- Eastern Cooperative Oncology Group [ECOG] Trial 1684 showed increased relapse-free interval (1 year for controls versus 1.7 years for interferon treatment, P=0.0023) and increased survival (2.8 years for controls versus 3.8 years for interferon therapy, P=0.0237) with the use of IFNα-2b for deep primary (pT_4) or regionally metastatic melanoma (*J Clin Oncol.* 1996; 14:7–17)
- confirmatory trials (ECOG 1692) are currently investigating the role of high-dose versus low-dose interferon (abstract presented for the April 1999 meeting of the American Society of Clinical Oncology)
- typical duration of therapy is 1 year
- adverse effects include cardiac and hepatic toxicity, constitutional symptoms (fever, flu-like symptoms), and myelosuppression

Lymphatic Mapping/Sentinel Lymph Node Biopsy

- **Sentinel Node**: first echelon node that drains primary tumor
- **Lymphoscintigraphy and Lymphatic Mapping**: localizes the sentinel node and lymphatic drainage pattern
- Technique: preoperatively undergo a lymphoscintigraphy to identify region of the draining basin and sentinel node, interoperatively inject lymphozarin dye intradermally to visualize sentinel node during dissection, confirm sentinel node by utilizing a radioactive probe (β-

probe), biopsy sentinel node and send for frozen section (4–8% false-positive rate)
- if node is **positive** for melanoma should undergo a neck dissection and consideration for interferon therapy
- if node is **negative** for melanoma no neck dissection required, may undergo neck dissection and consideration for interferon therapy if clinical nodes develop
- frozen section positive for tumor 15–20% of time

OTHER HEAD AND NECK MALIGNANCIES

Lymphoma

Introduction

- Pathophysiology: lymphoproliferative disorder
- Risks: irradiation, Epstein-Barr virus (Burkitt's lymphoma), human immunodeficiency virus (HIV), immunosuppression, organic toxins (phenols, benzenes), human T-cell lymphotrophic virus (HTLV-1, T-cell malignancies), immunological diseases (rheumatoid arthritis, celiac disease)
- SSx: nodal masses (may be painful), non-Hodgkin's lymphoma may present with extranodal masses as initial presentation (gastrointestinal tract most common extranodal site, also tonsils, paranasal cavity, base of tongue, salivary glands, thyroid, and orbit), fever, night sweats, weight loss, symptoms from mass effect from extranodal site (throat pain, nasal obstruction, exophthalmos, hoarseness)

Evaluation

- Complete Physical Exam and History: including palpation of other nodal sites (supraclavicular, axillary, epitrochlear, inguinal, femoral) and spleen; inquire about fever, weight loss, and night sweats (B-symptoms)
- **Fine Needle Aspirate**: may determine immunohistochemical subtype (B-cell or T-cell), DNA character (ploidy, phase), and RNA content (cannot differentiate follicular versus diffuse)
- **Open Biopsy**: usually required for definitive diagnosis prior to treatment, fresh sample required for immunochemistry
- Labs: complete blood count, blood urea nitrogen, lactate dehydrogenase levels, β_2-microglobulin, liver transaminases
- Staging: bone scan (gallium); CT/MRI of neck, chest, abdomen and pelvis; bone marrow biopsy

- Staging Laparotomy: may be considered for aggressive Hodgkin's lymphoma (not NHL); includes inspection of abdominal cavity, liver biopsy, nodal biopsy, and oophoropexy
- may also consider serum protein electrophoresis, upper gastrointestinal series

Hodgkin's Lymphoma

- bimodal incidence curve (teenagers and middle-aged adults)
- spreads via **contiguous** lymph nodes
- **Reed-Sternberg Cells** (**R-S cells**): binucleated, malignant giant cells, eosinophilic inclusions
- Rx: early stages may consider single-modality radiation therapy; advanced disease requires multimodality therapy with radiation and chemotherapy, **MOP**(**P**) (mechlorethamine, oncovin (vincristin), prednisone, procarbazine)/ **ABVD** (adriamycin, belomycin, vinblastine, DTIC); no surgical intervention except to obtain biopsy specimen
- Prognosis: prognosis dependent on staging, overall >75% 5-year survival; better prognosis for lymphocytic predominance types; worse prognosis with presence of Reed-Sternberg cells, class B symptoms, and higher staging

Ann Arbor Staging

I involvement of a single lymph node region or single extralymphatic site
II involvement of two or more lymph node regions on the same side of the diaphragm
III involvement of lymph node regions on both sides of the diaphragm
IV involvement of more than one extralymphatic organ
Symptom Classes: **A class** (asymptomatic), **B class** (weight loss, fever, night sweats)

Rye Modification of the Lukes and Butler Classification

- **Lymphocytic Predominance**: predominantly lymphocytic cells with few R-S cells, typically presents at an early stage, better prognosis
- **Nodular Sclerosis**: more common in women, typically presents at an early stage
- **Mixed Cellularity**: most common type
- **Lymphocytic Depleted**: rare, few lymphocytes with many R-S cells, usually advanced, poor prognosis

Non-Hodgkin's Lymphoma (NHL)

- most are **B-cells** or **null cells** (except lymphoblastic lymphomas)
- Rx: typically treated with **CHOP** (cytoxan, doxorubicin, vincristine, prednisone) chemotherapy followed by radiation to involved sites, low-grade B-cell lymphomas and MALT syndrome may be observed
- Prognosis: grading important; better prognosis associated with smaller and nodular types; poor prognosis associated with larger or diffuse types, higher grading, and central nervous system or bone marrow involvement
- **Mucosa-Associated Lymphoid Tissue** (**MALT**): low-grade, B-cell tumors that may occur in the gastrointestinal tract, conjunctiva, lungs, and thyroid, associated with *Helicobacter pylori*, may respond to antibiotics and anti-ulcer medications

Rappaport Classification and Working Formulation

Low Grade: typically B-cell lymphomas, better survival

- **Small Lymphocytic**: may be found in the orbit, elderly, consistent with chronic lymphocytic leukemia
- **Follicular** (**Small Cleaved Cells**): common, indolent course
- **Follicular** (**Mixed Cells**): small and large cell components

Intermediate Grade: most common type in the head and neck, potentially curable with chemotherapy

- **Follicular** (**Large Cell**)
- **Diffuse** (**Small Cleaved Cell**)
- **Diffuse** (**Mixed Cells**): small and large cell components
- **Diffuse** (**Large**): may involve sanctuary sites (CNS, testes), rapidly fatal, most common NHL type in the head and neck

High Grade: usually presents in the head and neck as part of a more extensive disease, extremely aggressive, requires immediate chemotherapy

- **Large Cell**
- **Lymphoblastic**: usually of T-cell origin
- **Burkitt's** (**Small Noncleaved Cell**): maxilla or mandible involvement (EBV associated, common in Africa), abdominal organ involvement (HIV associated)

Pediatric Malignancies

Introduction

- 5–10% of malignant tumors in children arise from the head and neck

- epidermoid cancers are relatively rare in the pediatric population in comparison to adults
- the neck is the most common presenting anatomical location

Pathology

- **Lymphoma**: (*see above*), most common pediatric malignancy of the head and neck (excluding retinoblastoma and neoplasms of the central nervous system), both Hodgkin's and Non-Hodgkin's are more common in boys, Burkitt's lymphoma almost exclusively affects children
- **Sarcomas**: rhabdomyosarcomas (*see below*); Ewing's sarcoma, fibrosarcomas, chondrosarcomas, and others are less common
- **Thyroid Carcinoma**: papillary carcinoma most common, medullary carcinoma, radiation exposure is the most significant risk factor
- **Nasopharyngeal Carcinoma**: typically undifferentiated type
- **Neuroblastomas**: arise from neural crest cells, typically originate from sites below the diaphragm and may metastasize to skull and facial bones
- **Salivary Gland Malignancies**: mucoepidermoid carcinoma most common
- **Malignant Teratoma**

Rhabdomyosarcomas

- most common soft tissue malignancy in children
- most common sites of origin in the head and neck in children are the pharynx and orbit
- 20% regional nodal involvement (although rare for orbital tumors)
- rare distant metastasis
- **Parameningeal Sites**: (middle-ear-mastoid, sinonasal, nasopharyngeal, and infratemporal fossa) may spread into meninges, less favorable prognosis
- Types
  1. **Embryonal:** poorly differentiated, **botryoid type**, more common in infants and young children, consists of spindle shaped, round, and "tadpole" shaped cells
  2. **Alveolar:** more common in adolescence, composed of round cells with reticulin-staining trabeculae
  3. **Pleomorphic:** more common in adults, well differentiated, consists of spindle and "strap" cells
- SSx: commonly misdiagnosed early due to vague symptomatology; rapidly progressive proptosis, vision loss, unilateral otitis media, nasal obstruction, headache, cranial nerve palsies
- Dx: CT/MRI, fine-needle aspiration, open biopsy

- Rx: chemotherapy is essential for management (either as a primary regimen, concominant with radiation therapy, or less commonly as postoperative adjuvant therapy), surgical debulking may be required without causing morbidity (the **Intergroup Rhabdomyosarcoma Studies** [**IRS**] which investigate multi-modality therapy have had several on-going trial groups which has led to a tripling of the 2-year survival rate in the last 20 years)

Parapharyngeal Space Tumors

Introduction

- 80% of parapharyngeal space tumors are benign
- SSx: displaced medial wall of the oropharynx or tonsil (if >3 cm), mass at angle of the mandible, otologic symptoms (mass effect on eustachian tube), cranial nerve palsies (CN IX–XII), dysphagia, obstructive sleep apnea, Horner's syndrome (mass effect on sympathetic chain), trismus, pain
- Dx: CT/MRI with contrast, carotid angiography, fine-needle aspiration, may consider open biopsy (intraoral route is contraindicated to avoid contamination)

Anatomy of the Parapharyngeal Space (*see* Figure 5–2)

- potential space shaped like an inverted pyramid that lies between the angle of the mandible and the tonsil
- attaches to sphenoid bone, foramen lecerum, petrous bone
- Superior Boundary: skull base
- Inferior Boundary: junction of the posterior belly of the digastric muscle and the greater cornu of the hyoid bone
- Medial Boundary: tensor veli palatini, levator veli palatini, and superior constrictor muscles and tonsillar fossa
- Lateral Boundary: ramus of the mandible, lateral and medial pterygoid muscles
- Posterior Boundary: prevertebral fascia
- Anterior Boundary: pterygomandibular raphe and medial pterygoid fascia

Compartments (divided by fascia between the styloid process and tensor veli palatini)

- **Prestyloid**: internal maxillary artery, inferior alveolar and lingual nerve (CN V_3), deep lobe of the parotid, fat; majority of tumors are **salivary gland tumors**

- **Poststyloid**: carotid sheath (internal carotid artery, jugular vein), cranial nerves IX–XII, sympathetic chain, lymph nodes; majority of tumors are **neurogenic and vascular**

Tumors of the Parapharyngeal Space

- **Salivary Gland Tumors**: most common primary tumor of the parapharyngeal space, pleomorphic adenoma (most common salivary gland tumor in parapharyngeal space, dumbbell shaped)
- **Neurogenic Tumors**: second most common primary tumor of the parapharyngeal space, schwannomas (most common neurogenic tumor in the parapharyngeal space), paragangliomas, neurofibromas, neuromas
- Other Primary Tumors: liposarcomas, chondrosarcomas, lymphomas, meningiomas
- Secondary Lesions: extension of oropharyngeal, parotid, submandibular, intercranial, or base of skull tumors; metastatic disease from nasopharynx, tongue base, oropharynx, and salivary glands

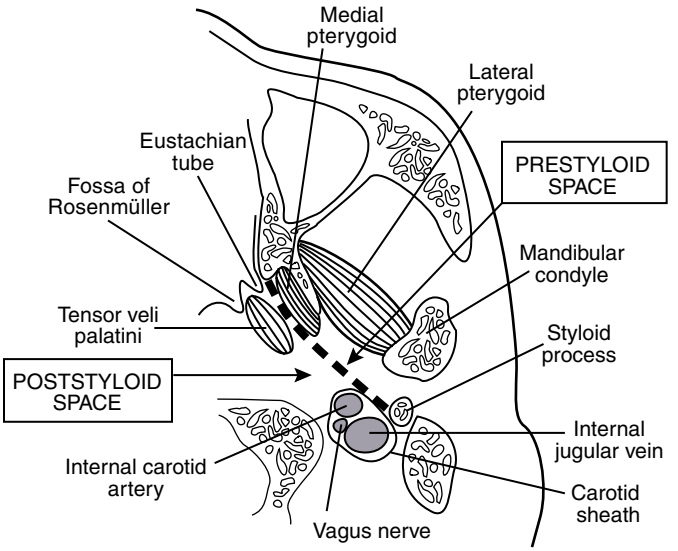

Figure 5–2. Cross-section at the level of the nasopharynx demonstrating the anatomy of the parapharyngeal space.

- Other Masses: lymphadenopathy, branchial cleft cyst, cystic hygromas, lipomas

Surgical Approaches

- **Transcervical Approach**: access via transverse incision at the level of the hyoid bone, allows direct access to the poststyloid compartment, excellent postsurgical cosmesis, mandibular osteotomy may be used to improve exposure
- **Transcervical Submandibular Approach**: transcervical approach with dissection of the submandibular triangle to gain access to the prestyloid compartment, mandibular osteotomy may be used to improve exposure
- **Parotidectomy-Submandibular Approach**: tumor is excised from the parotidectomy and exposure from the submandibular space, indicated for deep-lobe parotid tumors
- **Lip Splitting with Mandibulotomy**: indicated for pharyngeal malignancies that extend into the parapharyngeal space as part of a composite resection
- **Infratemporal Fossa Approach or Craniofacial Approaches**: indicated for tumors involving the base of skull

CHAPTER

Otology and Neurotology

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continued

ANATOMY, EMBRYOLOGY, AND PHYSIOLOGY OF HEARING AND BALANCE

External Ear and Temporal Bone Anatomy and Embryology

Auricle

- Embryology: around the sixth week of gestation the external ear arises from the proliferation of mesenchymal cells from the first and second branchial clefts
  1. **First Branchial Arch**: derives **Hillock of His 1–3** (1. tragus; 2. helical crus; 3. helix)
  2. **Second Branchial Arch**: derives **Hillock of His 4–6** (4. antihelix crus; 5. antihelix; 6. lobule and antitragus)
- the auricle is constructed from a framework of **elastic cartilage** and perichondrium with tightly adherent skin anteriorly and more loosely adherent skin posteriorly
- attachment of the auricle to the skull is by ligaments (anterior, posterior, and superior ligaments), muscles (anterior, posterior, and superior auricular muscles), skin, and the cartilage of the external auditory canal
- **Darwin's Tubercle**: small protuberance that may arise from the posterior-superior helix

External Auditory Canal (EAC)

- Embryology: **first pharyngeal groove** (cleft) forms the EAC; in the embryo the EAC fills with epithelial cells that recannulize by apoptosis in a medial to lateral direction around the seventh month (failure of recannulization results in aural atresia)
- **Cartilaginous EAC**: lateral ⅓, fibrocartilage, contains apopilosebaceous units (cerumen glands, hair follicles, sebaceous glands)
- **Osseous EAC**: medial ⅔, periosteum, is tightly adherent to the skin, contains no subcutaneous tissue
- Boundaries: infratemporal fossa, bony wall of the mastoid cavity, parotid gland and temporomandibular joint, tympanic membrane, epitympanum
- Sensory Contributions to the Auricle and EAC: **CN V_3** (EAC, TM, middle ear), **CN VII** (posterior concha, EAC), **CN IX** (Jacobson's nerve, with Arnold's nerve forms tympanic plexus on the

promontory), **CN X** (Arnold's nerve, concha, and antihelix), **great auricular nerve** (cervical plexus)
- **Fissures of Santorini**: lymphatic channels that connect the **lateral** cartilaginous EAC to the parotid and glenoid fossa region, allows for extension of infection and malignant tumors
- **Foramen of Huschke**: embryological remnant that forms a defect that connects the **medial** EAC to the parotid and glenoid fossa region, allows for extension of infection and malignant tumors outside the temporal bone

Tympanic Membrane (TM, Fig. 6–1)

- Embryology and the Formation of the Three Layers
  1. **First Branchial Pouch** (**Endoderm**): forms the eustachian tube, middle ear space, and the **inner mucosal layer** of the tympanic membrane
  2. **Mesoderm**: forms the **middle fibrous layer** of the tympanic membrane
  3. **First Pharyngeal Groove** (**Ectoderm**): forms the EAC and the **outer epidermal layer** of the tympanic membrane
- **Pars Flaccida**: superior portion of the TM, located in the notch of Rivinus, less stiff, sparse disorganized fibrous layer
- **Pars Tensa**: larger, stiff, vibrating surface of the TM, organized fibrous layer (**Schrapnell's membrane**)
- **Annulus Fibrosus**: thickened circumference of the pars tensa forming a fibrous outer ring for attachment to the temporal bone

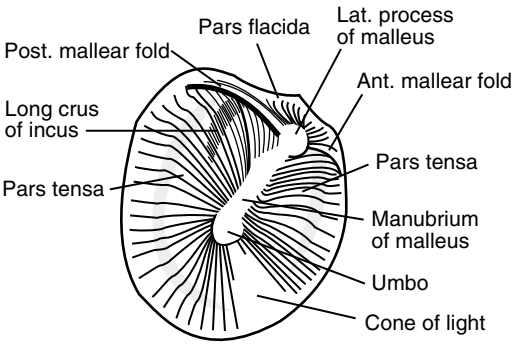

Figure 6–1. Anatomy of the tympanic membrane.

- **Notch of Rivinus**: notch of the squamose portion of the temporal bone located superiorly, within lies the pars flaccida

Surface Anatomy and Mastoid Cavity of the Temporal Bone

Lateral Surface

- **Temporal line:** bony ridge for the attachment of the temporal muscle fascia
- **Spine of Henle**: eminence located near the posterosuperior wall of the EAC to which the soft tissue of the EAC
- **Tympanomastoid suture line**: located posteriorly in the EAC, reliable landmark for the exit of the facial nerve
- **Tympanosquamous suture line**: embryonic fusion plane located posteriorly and inferiorly in the EAC, divides tympanic and mastoid portions of the temporal bone
- **Macewen's triangle**: bordered by the EAC, inferior temporal line, and the spine of Henle, aids in identifying the antrum

Superior Surface (Floor of Middle Cranial Fossa)

- **Arcuate eminence**: landmark of the superior semicircular canal
- **Foramen lacerum**: contains the internal carotid artery
- **Facial hiatus**: contains the greater (superficial) petrosal nerve
- **Tympanic canaliculus**: contains the lesser (superficial) petrosal nerve

Posterior Surface

- **Vestibular aqueduct**: contains endolymphatic sac and duct (*see also* p. 287)
- **Subarcuate canal**: contains the vestibular, facial, and acoustic nerve
- **Sigmoid sulcus**: located in medial aspect of the mastoid process
- **Cochlear aqueduct**: contains periotic duct (*see also* p. 287)
- **Internal auditory meatus (porus acusticus):** opening of the IAC that contains the facial, acoustic, vestibular, and intermediate nerves and the internal auditory artery

Mastoid Cavity

- **Tegmen tympani**: thin plate of bone that separates the middle fossa dura from the temporal bone
- **Sinodural angle**: region between the sigmoid sinus and the tegmen of the middle fossa
- **Korner's septum**: bony plate that separates the squamous and petrous air cells
- **Mastoid antrum**: first air cell that communicates the middle ear with the mastoid air cells

- **Facial recess**: triangular space bounded by the short process of the incus (fossa incudis) superiorly, facial nerve medially, and chorda tympani laterally; potential route to the mesotympanum
- **Donaldson's line**: imaginary line from the lateral semicircular canal bisecting perpendicularly the posterior semicircular canal, identifies the superior aspect of the endolymphatic duct

Middle Ear Anatomy and Embryology

Ossicles

- Embryology
  1. **First Branchial Arch** (**Meckel's cartilage**): derives the malleus head and neck, anterior malleal ligament, incus body and short process
  2. **Second Branchial Arch** (**Reichert's cartilage**): derives the manubrium of the malleus, long process and lenticular process of the incus, stapes (except vestibular part of footplate)
- ossicles reach their adult size at birth
- **Malleus**: manubrium, lateral and anterior process, neck and head; **tensor tympani muscle** (CN V) attaches to the malleus neck by tendon from the **cochleariform process**
- **Incus**: body, short and long process (crus), lenticular process; articulates with the malleus and the stapes via diarthrodial joints
- **Stapes**: head, anterior and posterior crus, base (footplate); **stapedius muscle** (CN VII) attaches to the stapes from the **pyramidal eminence**
- **Footplate**: attaches to the bony margins of the oval window by the **annular ligament** forming a joint (**syndesmosis**)

Eustachian Tube (ET)

- Embryology: derived from the **ventral first branchial pouch**
- consists of a posterior osseous portion (⅓, remains open to the middle ear) and an anterior membranous-cartilaginous portion (⅔, mobile portion) ends in the nasopharynx (**torus tubaris**)
- in the adult the ET is 45° from horizontal; in the infant the ET is shorter and is 10° from horizontal (more susceptible to regurgitation with feeding)
- Associated Muscles: **tensor veli palatini** (V_3, predominant dilator, the medial bundle of the tensor veli palatini forms the **dilator tubae**), **levator veli palatini** (pharyngeal plexus), **salpingopharyngeus** (pharyngeal plexus), **tensor tympani** (V_3) muscles
- Functions: ventilation (equalization of pressure between nasopharynx and middle ear), clearance (drainage), and protection (from sound and secretions)

Other Important Landmarks

- **Promontory**: basilar turn of the cochlea, floor of the middle ear
- **Jacobson's Nerve**: tympanic branch of the glossopharyngeal nerve located on the promontory
- **Arnold's nerve:** auricular branch of the vagus nerve, traverses the tympomastoid fissure to innervte the posterior aspect of the EAC
- **Ponticulus**: ridge of bone below the oval window
- **Subiculum**: ridge of bone between the round window niche and the pyramidal eminence
- **Sinus Tympani**: located in the mesotympanum between the ponticulus and subiculum

Inner Ear Anatomy and Embryology

Bony Labyrinth

- Embryology: mesenchyme which encapsulates the otocyst forms the cartilaginous **otic capsule**; ossification of the otic capsule occurs around the sixteenth week
- Components: vestibule, semicircular canals, and cochlea
- derived from the labyrinthine capsule from periosteal and enchondral ossification (susceptible to bony diseases such as Paget's disease, osteodystrophies, or otosclerosis)
- **Cochlear Aqueduct**: contains the **Periotic Duct**, continuous with the subarachnoid space (ends in posterior cranial fossa), inner ear opening is at the base of the scala tympani
- **Vestibular Aqueduct**: contains the **Endolymphatic Duct** and the veins of the vestibular aqueduct, runs from the vestibule to the posterior surface of the petrous pyramid in the posterior cranial fossa (opening is the **operculum**)

Membranous Labyrinth

- Embryology: around embryonic day 22–23 ectodermal thickening of the side of the head forms the **otic placode** which deepens to form the **optic pit** which later forms the **otocyst**; around embryonic week 25 the otocyst forms the membranous labyrinth
- **Membranous Labyrinth**: enclosed within the bony labyrinth
- **Endolymphatic Duct**: connects the endolymphatic compartment to the endolymphatic sac
- **Periotic Duct**: within the cochlear aquaduct, connects the scala tympani to the posterior cranial fossa
- **Ductus Reuniens**: narrowest segment between cochlea and saccule

Labyrinthine Fluids

- **Perilymph**: fluid within the osseous labyrinth, similar in composition to **extracellular fluid** and **cerebrospinal fluid** ($[Na^+] > [K^+]$), formed from the filtrate of blood and diffusion of cerebrospinal fluid
- **Endolymph**: fluid within the membranous labyrinth, similar in composition to intracellular fluid ($[K^+] > [Na^+]$), formed from the filtrate of perilymph

Semicircular Canals (SSC)

- senses rotational (angular) acceleration
- on each side the 3 SSCs are orthogonal (90° of each other)
- **Ampulla**: pear-shaped expansion located at one end of each SSC near the vestibular opening
- **Cupula**: gelatinous layer located within each ampulla which extends to the roof of the ampulla sealing the SSC
- **Crista Ampullaris**: group of hair cells at the base of the cupula
- **Horizontal** (**Lateral**) **SSC**: 30° from horizontal; ampullopetal (toward vestibule) flow of endolymph **increases** vestibular neuron firing rate
- **Vertical** (**Superior and Posterior**) **SSC**: both canals share a nonampullated common crus, ampullofugal (away from vestibule) flow of endolymph **decreases** vestibular neuron firing rate
- **Scarpa's Ganglion**: vestibular nerve cell bodies located in the distal vestibular nerve

Physiology

- **rotational** acceleration causes inertial deflection of the cupula which deflects the hair cells on the crista ampullaris causing depolarization
- **Type I** (flask-shaped) and **Type II** (cylindrical-shaped) hair cells contain 1 **kinocilia** and about 60 stereocilia, deflection of the stereocilia toward the kinocilia results in increased vestibular neuronal firing rate
- in the **horizontal** SSC, the kinocilia are located on the utricular side, therefore displacement of the stereocilia toward the vestibule (ampullopetal) causes increased vestibular neuronal firing rate and displacement away from the vestibule (ampullofugal) causes decreased vestibular neuronal firing rate
- in the **vertical** SSCs (posterior and superior), the kinocilia is located on the semicircular canal side, therefore the reverse occurs, displacement of the stereocilia away from the vestibule (ampullofugal) causes increased vestibular neuronal firing rate and displacement toward the vestibule (ampullopetal) causes decreased vestibular neuronal firing rate
- each canal exhibits a resting basal discharge rate

Otolith Organs

- **Utricle and Saccule**: vestibular end-organ cells that sense linear acceleration and changes in gravity
- **Static Maculae**: consists of **supporting cells** and **hair cells** whose cilia are within an overlying gelatinous mass on the surface of which lie the **otolithic membrane** and **otoliths** (calcium carbonate)
- **Striola**: line of orientation of hair cells in the utricle and saccule around which the polarity of the sensory cells changes 180°

Physiology

- **linear** acceleration causes inertial displacement of the otoliths which displace the cilia resulting in hair cell stimulation
- gravity accelerates the otoliths resulting in a sensation of up or down
- the sensory cells in the otolithic organs act similarly to the SSC in regard to the kinocilia and the cilia, however the utricle and saccule's sensory hair cells are arranged in a specific pattern (striola, *see above*)

Cochlea (Fig. 6–2)

- **Cochlea**: spiral-shaped acoustic end-organ centered on the central **modiolus** (2½ turns)
- **Scala Vestibuli**: contains perilymph, begins in vestibule
- **Scala Tympani**: contains perilymph, begins at the round window
- **Helicotrema**: opening between the scala tympani and the scala vestibuli at the cochlear apex
- **Scala Media**: contains endolymph and organ of Corti (*see below*)
- **Reissner's Membrane**: 2-cell layer membrane between the scala media and scala vestibuli
- **Basilar Membrane**: between the scala media and scala tympani, stiffer and skinnier at base, supports the organ of Corti
- **Stria Vascularis**: forms lateral wall of the scala media, sodium-potassium ATPase keeps membrane potential at −80 mv, contains highly metabolically active cells (dense mitochondria, Golgi apparatus, and endoplasmic reticulum), supports cochlear function
- **Spiral Ganglion**: ganglion of the cochlear nerve, located within the central modiolus in lateral end of cochlear nerve
- **Osseous Spiral Lamina**: bony plate forms the cochlear framework

Organ of Corti

- supporting cells (**Deiter's, Hensen's, Claudius' cells**) provide nutrients and support

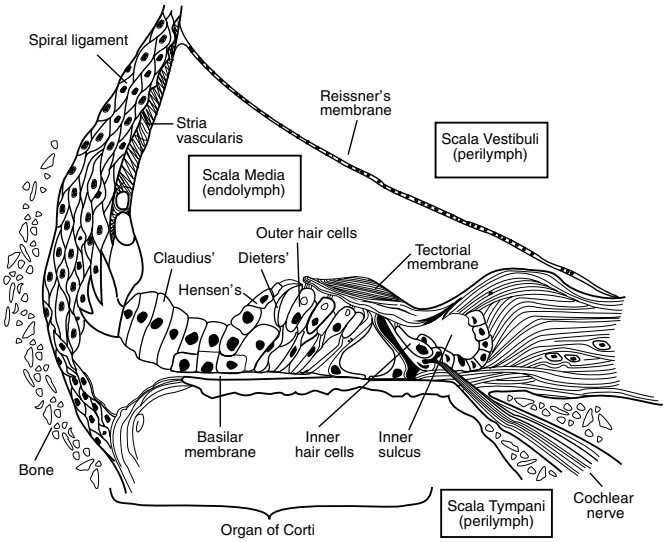

Figure 6–2. Cross-sectional anatomy of the cochlea. Reprinted with permission from Bailey BJ. *Head and Neck Surgery—Otolaryngology* [ill by Tony Pazos]. Copyright 1993, Lippincott, Williams & Wilkins.

- **Tectorial Membrane**: fibrogelatinous structure arises from the bony spiral lamina, the tips of the stereocilia of the outer hair cells are partially embedded into the tectorial membrane which causes shearing forces with vibration of the basilar membrane resulting in stimulation of the hair cells

Inner Hair Cells

- may act as the principal transducer of motion from the basilar membrane to a nerve impulse
- **single** row, fewer than outer hair cells
- **rounded**, flask-like shape with nucleus in the center, low intracellular glycogen
- few cilia in a curved shape
- loose connection to tectorial membrane
- completely surrounded by supporting cells
- Afferent Innervation: **Type I** (**radial, bipolar, myelinated**), form **95%** of fibers of the cochlear nerve, each inner hair cell is

innervated by 10–20 neurons (low hair to ganglion ratio) → cochlear nucleus

Outer Hair Cells

- may act as a motor to amplify motion from the basilar membrane (**cochlear amplifier**)
- source of **otoacoustic emissions**
- **3 rows**, more numerous than inner hair cells
- **cylindrical**-shaped with nucleus at base and organelles aligning the cell membrane, high intracellular glycogen
- many stereocilia in a "w" or "v" shape
- tight connection to tectorial membrane
- supported only at base
- Afferent Innervation: **Type II** (**spiral, pseudomonopolar, unmyelinated**), form **5%** of fibers of the cochlear nerve, 10 outer hair cells are innervated by one neuron (high hair to ganglion ratio) → cochlear nucleus
- Efferent Innervation: begins from the auditory cortex down to the level of the cochlear nuclei, additional contributions from the superior olive join and terminate predominantly on the outer hair cells

Physiology of Hearing

Conductive Pathway

- sound energy from the air is conducted to the inner ear via the tympanic membrane and the ossicles which provide hydraulic (area) advantage and a lever effect
- **Area Advantage**: area of the vibrating portion of TM is 55 mm^2; the area of the stapes foot plate 3.2 mm^2; advantage ratio = **17:1**
- **Lever Effect**: lever action of the ossicles affords an advantage ratio of **1.3:1**
- **Transformer Ratio**: area advantage + lever advantage = **22:1** or approximately **25–30 dB gain**
- TM protects the round window by allowing a **phase difference** between sound conducting to the oval window and sound directly reaching the round window; if sound waves would strike the oval and round windows simultaneously the resultant forces would suppress the cochlear fluid wave
- **Place Theory** (**Tonotopic Organization of the Cochlea**): pressure at the oval window produces a "traveling wave" along the basilar membrane from the base to the apex of the cochlea with a maximal amplitude of higher frequencies at the base and lower frequencies at the apex of the cochlea thus creating a tonotopical arrangement

Resonance Frequencies: natural frequencies that vibrate a mass with the least amount of force

- **Concha**: 4000–6000 Hz
- **EAC**: 2000–3000 Hz (primary frequencies of speech)
- **TM**: 800–1600 Hz
- **Ossicles**: 500–2000 Hz
- **Middle Ear**: 800 Hz

Auditory Nerve Pathway

1. stimulation of hair cells from vibration of the basilar membrane stimulates the bipolar neurons of the spiral ganglion that form the cochlear division of the eighth nerve
2. the cochlear nerve unites with the vestibular division as it courses through the internal auditory canal, the cochlear neurons then synapse with the cochlear nuclei (anterior ventral, posterior ventral, and dorsal)
3. **bilateral** innervation from the cochlear nucleus (via the acoustic striae) synapse on the superior olivar nuclei
4. superior olivary nuclei → lateral lemniscus → inferior colliculus → thalamus (medial geniculate body) → auditory cortex at Sylvian Fissure of Temporal Lobe (Brodmann's area 41)

*NOTE: the mnemonic **E COLI** outlines the neural pathway (**E**ighth nerve, **C**ochlear nucleus, superior **O**livary nuclei, **L**ateral lemniscus, **I**nferior colliculus)*

Physiology of Balance

The 5 Receptors of Balance (per Labyrinth)

- vestibular receptors respond to acceleration (linear and angular changes) only, not velocity
- **Otolithic Organs (2)**: linear accelerometers, oriented vertically in the erect position; unlike the crista of the SSCs the maculae of the otolithic organ have an extremely complex array of stereocilia; *see also* p. 289
- **Semicircular Canals (3)**: rotational accelerators (sense turning motion), the cristae of the SSC are arranged in an orthogonal pattern that creates functional pairs (right and left horizontal SSCs, right posterior and left superior SSCs, right superior and left posterior SSCs) in that increased output from one canal results in decreased output from its paired canal thus creating a "push-pull" mechanism that enhances the sensitivity of the system; *see also* p. 288

Nystagmus and Reflexes

- **Vestibulo-Ocular Reflex** (**VOR**): head rotation in a plane results in an opposite eye rotation to allow the eye to maintain fixation on an object (ie, motion of 10° to the left produces eye movement 10° to the right)
- **Vestibulospinal Reflex**: the otolithic organs modulate the antigravitational muscles in a similar way as the vestibulo-ocular reflex modulates the eye to allow for postural control (ie, simultaneous contraction of the extensor muscles with contralateral flexor muscles to maintain posture); position and motion of muscles and joints may also modify vestibulospinal reflex via receptors found in intervertebral joint receptors (upper cervical)
- **Nystagmus**: rapid involuntary movement of the eyes; the direction of nystagmus is determined by the **fast** phase; the **slow** phase is the **physiological** component (the reticular formation corrects the pathological slow phase component with a fast phase response); classically a **right** peripheral disorder results in **left** beating nystagmus

Vestibular Central Nervous System Processing and Compensation

- unlike the auditory nerve, the vestibular neural pathways have a balance of bilateral continuous neural traffic; unilateral interruption of any vestibular end organ results in perceptual dizziness
- imperfect correlation of receptors (vestibular, visual, or somatosensory) produces a "sensory conflict" resulting in perceptual dizziness
- habituation occurs with repeated or sustained receptor mismatch resulting in a decreased vestibular response via processing in the brain stem and cerebellum (**central compensation**)

AUDIOLOGY AND HEARING AMPLIFICATION DEVICES

Audiograms

Measurements of Intensity (Decibels)

- **Decibel** (**dB**): unit of measurement of sound pressure or intensity based on a logarithmic ratio

Derivation of the Decibel

- **Bell** (B) = log ($I_{output}/I_{reference}$); I = unit of intensity level
- dB = 10 log ($I_{output}/I_{reference}$)

- $I = P^2$; P = unit of sound pressure level
- **dB** = 10 log ($P_{output}^2/P_{reference}^2$) = **20 log** ($\mathbf{P_{output}/P_{reference}}$)

Reference Levels of the Decibel

- **Hearing Level** (**HL**): threshold dB based on normative hearing data as a reference (ie, 0 dB HL is the least intensity for the average normal ear to perceive a specific frequency)
- **Sensation Level** (**SL**): level in dB above an individuals threshold (eg, if an individual threshold is 20 dB HL then 50 dB SL = 70 dB HL)

Basic Audiogram

- **Pure-Tone Audiometry**: utilizes pure tones (single-frequency sound) at 250, 500, 1000, 2000, 4000, and 8000 Hz at selected intensities; complete series consists of air and bone conduction thresholds (bone conduction thresholds are not measured at 8000 Hz)
- **Air Conduction Threshold**: lowest level dB HL (hearing thresholds) at which the subject perceives 50% of pure tones introduced via earphones or speakers (sound field), represents conduction from the auricle to the cochlea
- **Bone Conduction Threshold**: lowest level dB HL (hearing thresholds) at which the subject perceives 50% of pure tones introduced via a bone oscillator apparatus, represents conduction from the bones of the skull to the inner ear (bypassing the TM and ossicles)
- **Pure Tone Average** (**PTA**): average thresholds for the speech frequencies (500, 1000, and 2000 Hz); typically should be within 10 dB of the speech reception threshold
- **Air-Bone Gap** (**ABG**): decibel difference between bone and air conduction thresholds
- **Speech Audiometry**: utilizes spoken voice as a sound stimulus at selected intensities
- **Speech Reception Threshold** (**SRT**): lowest dB HL patient can repeat a **spondee** (2-syllable words with balanced accents) 50% of the time
- **Speech Discrimination Score** (**SDS**): percentage of phonetically balanced monosyllabic words (**phonemes**) repeated correctly after being presented at 20–40 dB SL above the SRT

Hearing Threshold Levels

- <20 dB: normal
- 21–40 dB: mild hearing loss
- 41–60 dB: moderate hearing loss

- 61–80 dB: severe hearing loss
- >80 dB: profound hearing loss

Masking

- **Masking**: noise introduced with air conduction into the non-test ear to prevent crossover (perceived sound from an acoustic signal introduced to the opposite ear)
- air conduction crossover occurs if the intensity level of pure tones of the test ear is **40 dB or greater** than the bone conduction threshold of pure tones of the non-test ear
- bone conduction crossover occurs at **0 dB**
- bilateral ABG of 50 dB cannot be masked (**masking dilemma**)

Common Audiogram Patterns

- **Conductive Hearing Loss** (**CHL**): normal bone conduction thresholds with abnormal air conduction thresholds (presence of an ABG), maximal conductive hearing loss is 60 dB (ossicular chain discontinuity with an intact TM)
- **Sensorineural Hearing Loss** (**SNHL**): abnormal air conduction thresholds, no ABG
- **Mixed Hearing Loss**: hearing loss from conductive and sensorineural components, presence of an ABG with abnormal bone conduction thresholds
- **Low Frequency SNHL:** characteristic of endolymphatic hydrops
- **High Frequency SNHL:** characteristic of presbycusis
- **Bilateral Down-sloping High Frequency SNHL:** characteristic of presbycusis
- **Carhart's Notch**: falsely depressed bone conduction peaking at 2000 Hz suggestive of otosclerosis, secondary to a reduction of the inertial component of bone conduction
- **4 KHz Notch**: high frequency SNHL at 4000–5000 Hz that occurs with noise-induced hearing loss
- **Cookie Bite** (**U-Shape**): associated with hereditary hearing impairment
- **Recruitment**: increasing signal intensity produces an out-of-proportion perception of loudness, suggests **cochlear hearing loss** (eg, endolymphatic hydrops), recruitment causes discomfort within a shorter range of the SRT (normally 95 dB above SRT causes discomfort)
- **Rollover**: a paradoxical decrease in discrimination ability with increasing signal intensity, suggestive of a **retrocochlear disorder** (eg, Acoustic Neuroma, posterior fossa lesions)

- **Tone Decay and Fatigue**: a decrease in auditory perception with a sustained signal stimulus, suggestive of a **retrocochlear disorder**

Acoustic Impedance

- **Acoustic Impedance**: opposition to the flow of energy in a sound wave
- acoustic impedance measurements provide information about the middle ear (tympanometry) and inner ear, acoustic nerve, and brainstem function (acoustic reflex testing)

Tympanometry

- indirect tests of middle ear function by the transmission/reflection of sound energy
- **Tympanogram**: plots compliance changes of the TM versus air pressure in the external auditory canal
- **Peak**: point on the tympanogram that represents the point of maximum compliance, in which pressure of the external ear canal equals the pressure of the middle ear space

Types

A: normal peak between −150 and +50 daPa
A_S: "shallow" peak (reduced compliance), TM stiff, suggests otosclerosis, or tympanosclerosis
A_D: "deep" peak (hypercompliant), TM flaccid, suggests ossicular discontinuity or a monomeric TM
B: flat, no peak, nonmobile TM, suggests effusion, perforation, or an open pressure equalization tube
C: peak shifted to a more negative pressure (<−150), retracted TM, suggests eustachian tube dysfunction

Acoustic Reflex Testing

- **Acoustic Reflex**: reflexive contraction of the stapedial muscle in response to high intensity sound (usually at 500, 1000, and 2000 Hz)
- measured by introducing acoustic signals at varying frequencies (typically elicited at 70–100 dB SL) in one ear and measuring the TM motility (stapedial response) in the probe ear
- Reflex Arch: acoustic signal produces an action potential via CN VIII → ipsilateral cochlear nucleus → trapezoid body → **bilateral** superior olive → facial nuclei → CN VII → stapedial contraction (ipsilateral contraction stronger than contralateral contraction)

- CHL: may not be able to **stimulate** response initially if >40 dB CHL in receiving ear or may not be able to **receive** signal because ossicles cannot transmit stapes signal to TM in probe ear
- SNHL: may not be able to **stimulate** response initially if >60 dB SNHL
- **Acoustic Reflex Decay Test**: measures maintenance of the stapedial reflex for a sustained signal at 10 dB SL for 10 seconds, inability to maintain reflex suggests retrocochlear lesions
- **Absence Reflexes**: minimal CHL (5–10 dB), SNHL (>60 dB), mild CN VIII impairment (0–40 dB), brain stem lesion, or CN VII dysfunction proximal to the stapedial branch

Otoacoustic Emissions (OAE)

- **Otoacoustic Emissions**: objective sounds in the external auditory canal emitted from **outer hair cells** spontaneously or more commonly following acoustic stimulation, presence of OAE suggests a normal organ of Corti and middle ear system
- cochlear disorders >30 dB HL and middle ear disease may not have OAE
- retrocochlear lesions typically have normal OAE (as long as audiometric thresholds are within 30 dB HL))

Types

- **Spontaneous OAE**: does not require external stimulus, not present in all normal ears
- **Transiently Evoked OAE** (**Cochlear Echo**): evoked by broad-band tones, indicated for hearing screening, present in most normal ears
- **Stimulus Frequency OAE**: evoked with a particular frequency (low-tone), present in most normal ears
- **Distortion Product OAE**: elicited by simultaneous application of 2 pure tone frequencies, indicated for hearing screening (sensitive to cochlear disorders), present in most normal ears and ears with a mild or moderate SNHL

Pseudohypacusis

- **Pseudohypacusis**: subjective loss of hearing by a patient with the absence of organic pathology
- subjectively may be suspected when patient hesitates, shifts pure tone thresholds, or expresses confusion
- average PTA of 500, 1000, and 2000 Hz should be within 10 dB of SRT

- acoustic reflexes should be absent if tests suggests significant hearing loss
- **Stenger's Test**: the principle is that 2 tones of the same frequency presented to each ear cannot be heard simultaneously if one is louder; subject is given 2 simultaneous tones with matched frequency but the alleged poorer ear receives a tone at a greater intensity; a genuinely hearing impaired subject **or** normal subject should hear the tone with the better hearing ear; pseudohypacusis is suggested if the subject reports hearing nothing (denying any sound in the poorer hearing ear)
- **Lee's Speech Delay Test**: patient is played back his or her own speech at a delay that would cause the subject to stutter
- **Lombard Test**: background noise is gradually introduced below the subject's recorded response threshold as the subject is asked to read aloud; pseudohypacusis is suspected if the volume of the reader's voice increases as the masking noise is presented

Electrical Response Audiometry

Electrocochleography (ECoG)

- **Electrocochleography**: recording of evoked neuroelectrical events from the auditory nerve and cochlea in response to auditory clicks via air conduction
- Technique: probe is placed either via a transtympanic placement of a needle electrode on promontory (most accurate) or a tympanic probe membrane may be placed on the TM near the umbo, a reference electrode is placed on the vertex or pinna, wide-band clicks or tone pips are introduced and electrical potentials are recorded
- Indications: direct measurement of cochlear and auditory nerve function, may exhibit marked negative summation potential and a larger SP amplitude/AP amplitude ratio; intraoperative monitoring with certain diseases (eg, endolymphatic hydrops, perilymphatic fistula, *see also* p. 356)

Cochlear Potentials

- **Endolymphatic Potential** (**EP**): determined by the stria vascularis maintaining electrolyte balance in the scala media producing a DC potential (−80 to −100 mv)
- **Cochlear Microphonic Potential** (**CM**): alternating current generated predominantly from outer hair cells, normal CM suggests normal inner ear function

- **Summation Potential** (**SP**): direct current generated predominantly from the stria vascularis or hair cells, cochlea response to sound reflects changes in endolymphatic pressures
- **Whole-Nerve** (**Compound**) **Action Potential** (**CAP**): represents the summation of many individual nerve fibers, "all or none response," $\mathbf{N_1}$ (the first component of the CAP) is identical to the ABR Wave I

Auditory Brainstem Response (ABR)

- **Auditory Brainstem Response**: recording of the activity of the eighth nerve and central nervous system's response to an auditory signal, occurs during the first 10–20 ms after the stimulus
- Technique: active and reference electrodes are placed on the vertex, high forehead, mastoid, or earlobe; clicks, filtered clicks, or tone bursts are introduced to the ear between 5 and 50 times per second; surface electrodes utilize **signal averaging** to record specific ABR waves
- Indications: asymmetric hearing loss, unilateral vestibular weakness, or unilateral tinnitus suggestive of retrocochlear pathology; high risk children or with failed auditory screening (OAE); suspected malingerers; intraoperative monitoring
- unaffected by sedation; affected by phenytoin, lidocaine, diazepam, and halothane
- waveform ABR may not be measurable with SNHL of ±60 dB

ABR Peaks: each peak may correlate to an anatomical location (E COLI)

- I–II: **E**ighth nerve (distal and proximal segments)
- III: **C**ochlear nuclei
- IV **O**live (superior)
- V **L**ateral lemniscus (largest wave)
- VI–VII: **I**nferior Colliculus
- only waves I, III, and V are observed at birth, I–V is prolonged at birth

Retrocochlear and Acoustic Tumors

- abnormal **interaural wave V latency difference** (normal difference <0.2 msec), may be relative to size of tumor (reliable)
- abnormal **interaural wave I–V and III–V latency difference** (most reliable in the presence of a CHL)
- prolonged **interwave** latency between I–V or I–III
- increased **absolute** latency measurements (eg, 5.7 msec for wave V), may not be as accurate since latency may be affected by CHL

Sound Amplification Devices

Introduction

- Components: receiver, filter, amplifier, microphone, battery, volume control
- **Gain**: ratio of the output to the input
- **Saturation Sound Pressure Level** (**SSPL**): input level that produces the maximum output level
- **Frequency Response**: gain across a range of frequencies
- **Compression**: allows the dynamic range of the output to be less than the input in a nonlinear fashion
- **Venting**: a hole within a hearing aid placed in the EAC that reduces low frequency amplification by increasing resonant frequency
- in general, binaural aids are preferred for binaural hearing loss to provide better discrimination, loudness, and sound localization

Types

- **Digital Hearing Aids**: process information in digital form, provide differential amplification of frequency groups to adjust for specific frequency losses and produce less background and circuit noise, more expensive
- **Contralateral Routing of Signal** (**CROS**): may be considered for unilateral profound hearing loss, microphone placed on poorer hearing ear and sound presented to better ear for sound localization; (**Bi-CROS** hearing aids also amplify sound to the better hearing ear)
- **Bone-Conduction Hearing Aids**: indicated for severe conductive loss when air conduction aids cannot be used
- **Compression Hearing Aids**: do not maintain a linear relationship from input level to output level, indicated for patients who have limited dynamic range or recruitment
- **Programmable Hearing Aids**: hearing aid is programmable to more than one listening situation (eg, variable background noise)

Body Hearing Aids

- microphone placed on torso
- Advantage: allows high level output with low feedback, easy to manage
- Disadvantage: conspicuous, clothes and body may rub against microphone, poor localization of sound

Behind-the-Ear (BTE)

- microphone and amplifier placed behind the ear

- Advantage: allows adequate gain from mild to profound hearing loss, allows for open ear molds (large vents)
- Disadvantage: conspicuous

In-the-Ear (ITE)

- placed in concha
- Advantage: less conspicuous, allows gains for moderate to severe hearing loss
- Disadvantage: limited venting, more difficult to adjust settings

In-the Canal (ITC)

- hearing aid placed entirely in ear canal
- Advantage: inconspicuous
- Disadvantage: limited gain because of feedback (limited to mild to moderately-severe hearing loss), difficult to adjust settings, limited venting

Contained-In-Canal (CIC)

- placed deep into ear canal
- Advantage: most inconspicuous, may be used from mild to severe hearing loss (does not require as much gain since placed close to TM), lower instance of feedback (since requires less gain), reduced noise, less distortion
- Disadvantage: difficult to adjust settings, difficult to insert and remove, limited venting

Cochlear Implants

- cochlear implants convert sound into an electric signal and stimulate the cochlear nerve directly
- Components: microphone (placed above ear), processor (placed behind the ear or worn in the pocket), receiver-stimulator, and implantable electrode (typically placed **intracochlearly** in the scala tympani via the round window)
- Contraindications: infected ear, retrocochlear disease, Michel's deformity (Mondini deformity and other deformed cochlea are not contraindicated), inability to participate in an extensive rehabilitation program (noncompliance)
- Complications: device failure (eg, electrode malfunction), infection (eg, wound infection, meningitis), skin flap breakdown, device migration or extrusion, facial paresis or paralysis, perilymphatic gusher (increased risk with Mondini's deformity)

Indications

- Adults: **postlingual**, bilateral, severe to profound cochlear hearing loss with minimal benefit from hearing aids, ability to particpate in rehabilitation
- Children: >2 years old, bilateral, severe to profound cochlear hearing loss with minimal benefit from hearing aids, ability to participate in rehabilitation, may be **pre- or postlingual**

Types

- **Single-Channel**: utilizes 1 electrode pair, provides temporal and intensity information, largely been replaced by multi-channel systems
- **Multi-Channel**: provides improved sense of pitch by delivering signals to different locations in the cochlea (tonotopic organization), results in better speech recognition, typically up to 24 pairs of electrodes may be used

Results

- better results are associated with shorter duration of deafness, postlingual deafness, and increased implant usage
- cochlear implants have been shown to reduce tinnitus, improve warble tone thresholds in children, and improve overall communication ability (including speech recognition)
- implantees typically have a reduced dynamic range (difference between threshold and discomfort levels)

HEARING LOSS AND TINNITUS

Evaluation for Hearing Loss

History

- Character of Hearing Loss: onset and duration, constant versus intermittent, progression, unilateral versus bilateral, high or low tone loss, decreased speech intelligibility
- Contributing Factors: recent infection (fevers, upper respiratory infection); loud noise exposure; recent trauma (barotrauma, straining, weight lifting, head injury); exacerbating factors of tinnitus (sleep, exercise, caffeine, alcohol, stress); previous otological history (surgery, infections); toxin exposure and medications (*see* Table 6–1); history of autoimmune disease, hypertension, diabetes, vascular disorders, temporomandibular joint disease, neurologic disease (stroke), depression, and otologic disorders; family history of deafness

TABLE 6–1. Common Ototoxic Medications

Category	Representative Examples and Comments
Antibiotics	aminoglycosides exert their ototoxic effects by damaging vestibular and cochlear hair cells, streptomyocin and gentamicin affect vestibular function more than auditory function (hearing loss occurs initially with supra-audiological thresholds, >8 kHz), vancomyocin causes synergistic ototoxic effects with aminoglycosides
Chemotherapy Agents	cisplatin may cause auditory nerve and vestibular toxicity
Diuretics	furosemide and ethacrynic acid are the most common diuretic ototoxic agents, often potentiate other ototoxic medications
NSAIDs and Salicylates	aspirin is the most common medication to cause tinnitus (reversible with discontinued use)

- Associated Symptoms: aural fullness, fevers, vertigo, tinnitus (*see below*), otalgia, otorrhea, weight loss, other neurologic complaints
- Think "KITTENS" for differential diagnosis (*see* Table 6–2)

Physical Exam and Hearing Tests

- Otoscopy/Microscopy: inspection of EAC (cerumen impaction, lesions, masses) and tympanic membrane (color, thickness, presence of fluid, myringosclerosis, perforations, lesions), Valsalva test (test patency of eustachian tube by having subject perform Valsalva maneuver and inspect TM for mobility)
- Pneumatic Otoscopy: test mobility of TM with positive and negative pressure, **fistula test** (positive pressure causes nystagmus which reverses with negative pressure, may be seen in perilymph fistulas and syphilitic labyrinthitis)
- Inspection and Palpation: inspect outer ear for lesions, malformations, auricular pits, scars, edema, swelling, mastoid tenderness, tragal tenderness
- Complete Head and Neck Exam: cervical lymphadenopathy, neurologic and vestibular exam, bruits

Tuning Fork Tests

- **Rinne Test**: typically uses a 512 Hz (C^1 fork) and 1024 Hz tuning forks to compare air conduction (AC) and bone conduction (BC);

TABLE 6–2. Common Differential Diagnosis of Hearing Loss: KITTENS Method

(**K**) Congenital	**I**nfectious and Idiopathic	**T**oxins and Trauma	**T**umor (Neoplasms)	**E**ndocrine	**N**eurologic	**S**ystemic/ PSychological
Hereditary progressive SNHL	Labyrinthitis	Ototoxic medication	Temporal Bone neoplasms	Hypothyroiditis	Multiple sclerosis	Ménière's disease
Congenital auricular malformations	Otitis media	Noise induced hearing loss	Cholesteatomas	Hyperlipidemia	Meningitis	Paget's disease
Congenital hearing loss	Luetic hearing loss	Head trauma			Stroke	Otosclerosis
	Presbycusis	Lead and mercury toxicity			Central auditory processing disorder	Presbycusis
		Barotrauma				Cerumen impaction
						Pseudohypacusis
						Autoimmune disorders

strike tuning fork then place within 1 cm of the entrance to the ear canal (AC) and then immediately place on the mastoid (BC); when the tuning fork was better heard by AC then the test is referred to as **positive** (normal ears or most SNHL); if BC is perceived louder than AC the test is referred to as **negative** (CHL >15–30 dB HL or severe to profound SNHL with cross-over)

- **Weber Test**: typically utilizes a 512 Hz (C^1 fork) and 1024 Hz tuning forks; strike tuning fork and place in center of forehead, vertex, or upper teeth; perceived sound should normally be heard centrally (or in "both ears"); unilateral SNHL should lateralize to better hearing ear, unilateral CHL should lateralize to diseased ear

Audiometric Tests

- essential to identify auditory function, CHL versus SNHL, cochlear versus neural dysfunction, central auditory dysfunction, and pseudohypacusis (*see* pp. 293–299)

Ancillary Tests

Imaging

- **Radiographs**: special views of the temporal bone (Schuller's, Stenvere's, Towne's views) have largely been replaced by computed tomography (CT) and magnetic resonance imaging (MRI)
- **CT of Temporal Bones**: indicated to evaluate the complications of suppurative ear disease, neoplasms, cholesteatoma, mastoiditis, temporal bone fracture, or a congenital disorder
- **MRI of Brain and Brainstem with Gadolinium**: indicated if suspect cerebellopontine angle tumors, meningiomas, demyelinating lesions (multiple sclerosis), or petrous apex lesions (cholesterol granulomas)

Ancillary Laboratory Studies (may be considered for specific circumstances)

- **Complete Blood Count**: results may suggest active inflammation or leukemic process
- **Treponemal Studies**: Lyme titers and VDRL/FTA-ABS
- **Lipid Profile**: high risk of artherosclerotic disease (associated with hearing loss)
- **Immunological Profiles**: *see below*
- **Glucose**: screen for diabetes (associated with hearing loss)
- **Coagulation Profile**: coagulopathies are associated with hearing disorders

Presbycusis

- **Presbycusis**: progressive deterioration of hearing associated with aging
- most common cause of adult auditory deficiency (approximately 50% of the elderly have hearing impairment)
- Pathophysiology: uncertain; possible etiologies include vascular disorders causing atrophy of the stria vascularis; the aging process resulting in degeneration of hair cells, loss of ganglion cells, or degeneration of nerve fibers; stiffening of the basilar membrane; or chronic insults such as noise
- SSx: progressive, symmetric SNHL initially of frequencies >2000 Hz (sloping configuration)
- Dx: clinical history with high-frequency SNHL without other sources of hearing loss
- Rx: sound amplification if required

Sudden Sensorineural Hearing Loss (SSHL)

Introduction

- **Sudden Sensorineural Hearing Loss**: loss of significant hearing (>35 dB in at least 3 adjacent frequencies) that occurs over less than 3 days, typically unilateral
- ⅓ return to normal hearing, ⅓ result in a profound hearing loss
- worse prognosis if associated with vertigo, total deafness, advanced age, down-sloping audiogram (high-frequency loss), or associated vascular risk factors
- better prognosis if minimal hearing loss and low-frequency loss
- Causes: typically idiopathic, may be autoimmune, viral, or vascular in etiology
- Evaluation: similar to hearing loss evaluation (*as above*); however, must keep a high suspicion for reversible causes of SSHL (eg, perilymphatic fistulas, ossicular trauma); initial investigations should include an immediate audiogram and immittance testing
- Rx: in the absence of a known cause, medical treatment is empirical and controversial; high-dose oral corticosteroids are the most commonly employed therapy, may consider antivirals (less common agents such as vasodilators and anticoagulants to improve cochlear blood flow or reverse vasospasm are controversial)

Autoimmune Hearing Loss

- may be associated with systemic immune diseases (rheumatoid arthritis, lupus erythematosus, etc), allergy, and vasculitides
- most common between ages 20–50 years old

- Pathophysiology: may arise from host's defense from infection causing autoimmunity, cross reactivity from distant antigens, or circulating immune complexes affecting circulation in the stria vascularis
- SSx: rapidly progressive or fluctuating SNHL (bilateral), normal otoscopic exam, tinnitus
- Dx: rapidly progressive SNHL with serological evidence of immune dysfunction
- Serological Evaluation for Autoimmunity in Hearing Loss: CBC, erythrocyte sedimentation rate (ESR), antinuclear antibody (ANA), rheumatoid factor (RF), C1q binding test, Raji cell assay, cryoglobulins, complement profiles, lymphocyte transformation test, 68 kD proteins, collagen II ABS
- Rx: high-dose oral corticosteroid trial with audiogram follow-up to assess response, if an immunological diagnosis is highly suspected (positive serology) cytotoxic medications for nonresponsive cases

Noise-Induced Hearing Loss

- according to OSHA regulations exposure equivalent above 85 dBA time-weighted average for 8 hours requires hearing protection programs (approximate equivalent exposure duration: 95 dBA for 4 hours, 100 dBA for 4 hours, or 130 dBA for less than 2 minutes)
- **Temporary Threshold Shift (TTS)**: loud noise exposure can cause a temporary SNHL that typically resolves within 24 hours
- **Permanent Threshold Shift (PTS)**: permanent hearing loss from noise exposure that does not improve after the acoustic trauma event
- rifle gunfire poses a risk to the opposite ear of the shooting arm, the same side ear is protected by the head shadow effect; pistol gunfire poses a risk to the same side of the shooting arm
- Pathophysiology: outer hair cells are susceptible to damage to chronic loud noise exposure; a single intense exposure (eg, explosion or gunfire) may mechanically damage the organ of Corti or rupture cochlear membranes (resulting in mixing perilymph and endolymph-injuring cells)
- SSx: SNHL (typically bilateral), high-pitched tinnitus
- Dx: history and audiogram findings (usually initially effects 4–5 kHz frequencies)
- Rx: hearing protection programs, amplification if indicated, and yearly audiograms until hearing is stable

Central Auditory Processing Disorder

- **Central Auditory Processing Disorder (CAPD)**: central receptive language disorder of adults and children from difficulty in decoding and storing auditory information

- higher risk with children with learning disabilities, family history, Attention Deficit Disorder, other neurological disorders (many children have normal hearing and intelligence)
- SSx: perceptual hearing loss (especially with background noise), delayed communication abilities (eg, speech problems, delayed response, use of gestures instead of speech), echolalia (repeating back words without comprehension), behavior problems
- Dx: normal audiogram, tested by audiologists (series of auditory processing tests), and child psychologists
- Rx: preferential seating, reduction of background noise (consider FM devices in school), speech therapy

Hereditary and Congenital Hearing Loss

- **Congenital Hearing Loss**: hearing loss that is present at birth, may be inherited or acquired (eg, infection, birth trauma)
- **Hereditary Hearing Loss**: hearing loss that presents after birth in which the cause is present in the genetic code; may be associated with a variety of syndromes (eg, Alport Syndrome, Waardenburg Syndrome)
- *see* pp. 339–347 for complete evaluation and management

Tinnitus

Introduction

- **Subjective Tinnitus**: perception of sound in the absence of any acoustic, electrical, or external stimulation, more common than objective tinnitus, typically associated with a high frequency hearing loss (pitch of tinnitus may correlate with the frequency of hearing loss, most common is 3000–5000 Hz)
- **Objective Tinnitus**: perception of sound caused by an internal body sound or vibration, may be exacerbated with a conductive hearing loss
- Pathophysiology: the pathophysiology of subjective tinnitus is largely unknown; however, current tinnitus models center around subcortical auditory pathways rather than cochlear dysfunction

Work-up for Tinnitus

- similar work-up as hearing loss (*see* pp. 302–305) with special attention to retrotympanic masses, audible bruits, history of trauma, medications, presences of TMJ, and psychological factors
- Character of Tinnitus: unilateral or bilateral, high-pitched (ringing, hissing) or low-pitched (roaring, buzzing), pulsatile, clicking, progression and frequency, loudness, pure or multiple tones, level of discomfort (difficulty with sleeping)

- consider appropriate evaluation for retrocochlear lesions if suspected (acoustic neuromas may present with tinnitus)
- **Subjective Tinnitus**: *see* Table 6–3 for Differential Diagnosis

Causes of Objective Tinnitus

Vascular Causes/Pulsatile Tinnitus

- **Benign Intracranial Hypertension Syndrome**: most common cause of pulsatile tinnitus (especially in overweight females, 20–30 years old), increased intracranial pressure without focal neurological signs, caused by systolic pulsations of CSF, diagnosis is by lumbar puncture to measure CSF pressure, may be associated with a mild SNHL; Rx: weight loss and diuretics (may consider subarachnoid-peritoneal shunts in select cases

TABLE 6–3. Differential Diagnosis of Subjective Tinnitus

Hearing Loss and Otologic Disorders

- **Hearing Loss (Presbycusis, Autoimmune Hearing Loss)**
- **Retrocochlear Lesions (Acoustic Neuromas)**
- **Ménière's Disease**

Medications: (*see* Table 6–1)

- **Aspirin/NSAIDs**
- **Hypertensive Agents**
- **Aminoglycosides**
- numerous medications have been reported to have the potential to cause tinnitus

Trauma

- **Head Injuries (Whiplash)**
- **Loud Noise Exposure**
- **Barotrauma**

Systemic Diseases

- **Hypertension**
- **Depression and Anxiety**
- **Neurological Disease (Multiple Sclerosis, Brainstem Stroke)**

- **Arteriovenous Malformations** (**AVM**): causes pulsatile tinnitus which is synchronous with the heart beat, may be secondary to trauma; Rx: surgical excision or embolization
- **Arterial Bruits and Venous Hums**: "whooshing" sound synchronous with the heart beat, may be caused by turbulent blood flow in nearby vessels (stenosis) or by aberrant vessels
- **Hypertension**: may present with pulsatile tinnitus
- **Vascular Tumors**: most common are glomus tympanicum and glomus jugulare (*see* p. 319)

Mechanical Causes

- **Patulous Eustachian Tube**: noise occurring synchronous with nasal respiration from an overly patent eustachian tube, may occur in postradiation patients or after significant weight loss; Rx: mucosal irritants, eustachian diathermy probe (often requires a pressure equalization tube), may consider Teflon paste injection into the torus tubaris (less effective)
- **Palatal Myoclonus**: rapid clicking sound caused by the contraction of the palatal muscles; may be evaluated by nasopharyngoscopy in an awake patient; Rx: consider muscle relaxants
- **Tensor Tympani Syndrome**: spasm or myoclonus of the tensor tympani muscle resulting in a fluttering, low frequency tinnitus; Rx: reassurance, rarely requires section of the tensor tympani muscle
- **Spontaneous Otoacoustic Emissions**: rare cause of objective tinnitus

Management of Subjective Tinnitus

Conservative Management

- evaluate medications for possible causes or contributing factors (eg, aspirin and NSAIDs may cause worsening of subjective tinnitus)
- broad band masking at night (eg, placing the radio between stations)
- relaxation techniques (biofeedback)
- consider referral to a tinnitus support group

Hearing Aids

- indicated for tinnitus associated with hearing loss
- simplest method of "direct masking"
- reduces tinnitus by amplifying ambient sound to mask the tinnitus

Maskers

- maskers utilize a band of white noise centered around the tinnitus frequency (pitch matched)

- indicated for intractable tinnitus in patients with normal or near normal hearing
- **Pitch and Loudness Matches**: determine frequency and intensity of tinnitus, difficult to obtain and often inaccurate
- **Minimal Masking Level**: amount of sound (pure tone, narrow band, or broad band of noise) required to cover the tinnitus (requirements of >10–15 dB are associated with a poor success rate)
- **Residual Inhibition**: length of time of decreased or absent tinnitus after exposure of 1 minute of masking, typically lasts seconds or minutes

Medical Management

- may consider Benzodiazepines, Tricyclic Antidepressants, and Carbamezapine
- medical management may result in a >50% improvement of tinnitus

INFECTIONS OF THE EAR AND TEMPORAL BONE

Infections of the External Ear

Acute Otitis Externa ("Swimmer's Ear")

- **Acute Otitis Externa**: bacterial infection involving the skin of the external auditory canal
- Pathophysiology
  1. aggressive washing of cerumen or retention of water ("Swimmer's Ear") results in a more alkalotic EAC and decreased production of antibacterial agents (eg, lysozyme) which are permissive for bacterial overgrowth and penetration into the apopilosebaceous unit
  2. microtrauma (eg, cotton swabs, fingernails, or hairpins) directly injure EAC soft tissue
- Pathogens: *Pseudomonas aeruginosa* (most common, opportunistic infection), *Staphylococcus*, gram-negative bacilli
- Risks: immunocompromised patients (diabetics, HIV), swimmers
- SSx: pain with posterosuperior manipulation of auricle, tragal tenderness, otalgia, and pruritus; edematous and erythematous EAC (may have exudative or purulent discharge); may have a conductive hearing loss
- Dx: clinical history and physical exam (perforation of the TM may suggest an underlying chronic otitis media)

Management

- thorough and frequent ear canal debriclement with fine suction to allow opening of apilosebaceous unit

- aggressively manage diabetes
- evaluate for signs and symptoms of malignant external otitis (granulation tissue in the canal, cranial nerve involvement)
- **Otic Drops**: acidification, drying, antibiotic, or antibiotic/corticosteroid combination drops for 7–10 days (consider placing an otowick to aid drops to reach medial EAC if canal is too edematous to visualize TM)
- **Oral Pain Medication**: may require narcotics
- **Oral Antibiotics**: typically not required unless signs of cellulitis, concurrent otitis media, persistent or severe symptoms, or systemic illness
- consider culturing the EAC for recalcitrant infection

Chronic Otitis Externa and Eczematous Otitis Externa

- **Chronic Otitis Externa**: thickening of the EAC from a persistent low-grade infection or inflammation
- **Eczematous Otitis Externa**: broad term that describes the dermatological conditions affecting the EAC (eg, atopic dermatitis, contact dermatitis, psoriasis, lupus erythematosus)
- **risk of secondary infection** by bacteria or fungal agents
- SSx: dry and flaky erythematous EAC, pruritis, mild pain
- Dx: clinical history and physical examination
- Rx: clean and debride EAC, corticosteroid cream/lotion, consider dermatology consult for Eczematous Otitis Externa, surgical intervention (canalplasty with STSG) may be considered in recalcitrant disease

Osteomyelitis of the Skull Base (Necrotizing or Malignant External Otitis)

- Pathophysiology: extension of infection from the EAC into the temporal bone or skull base resulting in progressive **osteomyelitis**
- Pathogens: *Pseudomonas aeruginosa* (most common)
- Risks: debilitated and immunocompromised patients (diabetics, HIV) radiation exposure
- SSx: granulation tissue in EAC typically at the bony-cartilagenous junction, persistent otalgia and otorrhea, cranial nerve involvement
- Dx: clinical history and physical exam, CT of temporal bones, Technetium bone scan (evaluates osteoblastic activity, excellent for localization) and Gallium bone scan (evaluates inflammation, follow course with gallium scans), may consider biopsy with culture of EAC, elevated CSR

- Complications: cranial neuropathy, sinus thrombosis, septicemia, meningitis, intracranial infections, death

Management

- prolonged parenteral anti-*Pseudomonus* antibiotics
- acidic, antibiotic, or antibiotic/corticosteroid combination otic drops
- aggressive diabetic control
- meticulous cleaning and debridement
- follow course with periodic gallium bone scans
- hyperbaric oxygen may be considered for recalcitrant cases
- rarely surgical debridement of involved tissue may be required for failed medical management

Otomycosis

- Common Pathogens: *Aspergillus* (most common) and *Candida*
- Risks: immunocompromised patients (diabetics, HIV), poor hygiene, increased ear moisture, long-term topical antibiotics
- often occurs as a superinfection with external otitis
- SSx: moist, "tissue-paper" sheets of keratin within the EAC, dotted gray membrane (after removal of membrane reveals an erythematous canal), pruritic, musty exudate, *Aspergillus* produces distinct small black conidiophores on top of fluffy white filamentous hyphae
- Dx: clinical physical exam, may see fungal hyphae under microscope, culture
- Rx: meticulous and frequent debridement of ear canal; acidification, drying, or fungicidal otic drops; debridement; for refractory cases consider 2% aqueous gentian violet or methyl-cresyl acetate

Perichondritis

- Pathophysiology: infection of the auricular cartilage possibly from extension from inadequately treated auricular cellulitis, exposed cartilage, trauma, external otitis, or infected endaural incisions
- Pathogens: *Pseudomonas aeruginosa* (most common), *S. aureus, Streptococcus*
- risk of cauliflower ear from cartilage erosion if not treated aggressively
- SSx: tender, erythematous, warm, edematous auricle, occasional systemic symptoms (fevers, cervical adenopathy)
- Dx: clinical history and physical exam (also condider Relapsing Periochrondritis, *see* p. 324)
- Rx: aggressive systemic antibiotics regimen (anti-*Pseudomonus*)

Infections of the Middle Ear

Acute Otitis Media (Acute Suppurative Otitis Media)

- **Acute Otitis Media** (**Acute Suppurative Otitis Media**): acute infection (<3 weeks) causing inflammation of the middle ear space
- second most common disease in children (upper respiratory infection is the most common)
- Pathophysiology: eustachian tube dysfunction results in negative middle ear pressures leading to transudative fluid collection in the middle ear space and subsequent infection
- Common Pathogens: ***S. pneumoniae*** (most common), *H. influenza*, *Moraxella (Branhamella) catarrhalis;* gram negative bacilli and Group B *Streptococcus* may be found in infants; viral pathogens are often present alone (sterile otitis media) or concurrently with bacterial pathogens
- Risks: factors that may contribute to eustachian tube dysfunction (craniofacial or skull base abnormalities, recurrent upper respiratory infections, nasal allergy), attendance at a child care facility, bottle feeding or supine baby feeds (reflux into ear), smoking or presence of second-hand smoke, history of immunological disorders (especially IgA and IgG subclass 2 and 3 deficiencies) or ciliary dysfunction (Kartagener syndrome), gastroesophageal reflux, prolonged nasotracheal intubation or nasogastric tubes, adenoid hypertrophy
- SSx: otalgia (irritability and tugging of ears in children), aural fullness, hearing loss, tinnitus, fever, hyperemic or thickened TM, fluid in middle ear space (nonmobile or bulging TM, air-fluid levels, bubbles, yellow hue)
- Dx: clinical and physical exam, audiogram (CHL <30 dB) and tympanometry

Management of Acute Otitis Media

Medical Therapy

- Antimicrobial Therapy: first-line antimicrobials should include gram positive and gram negative coverage, may consider β-lactamase resistant agents due to increasing β-lactamase activity in certain regions; treat for at least 7–10 days (typically resolves infection within 72 hours); if no resolution may consider broader spectrum coverage or β-lactamase resistant agents
- Adjunctive Therapy: antipyretics, analgesics, oral or nasal decongestants (relieve associated nasal congestion, no proven benefit in the treatment of acute otitis media), myringotomy may be considered for severe otalgia or toxic patients

- Prophylaxis: indicated for recurrent acute otitus media, full course of antibiotics for 10 days followed by reduced bedtime dose for 5–6 weeks (eg, Amoxicillin 250 mg tid × 10 days then 250 mg qH5 × 6 weeks)

Management of Recurrent Acute Otitis Media

Myringotomy with Pressure Equalization Tubes (PET)

- Indications: recurrent otitis media, persistent effusion (>3 months), poor response to antibiotic regimen, immunocompromised patient, presence of a cleft palate, impending complication (hearing loss), severe retraction pocket, barotitis media, autophony from eustachian tube dysfunction
- PET Complications: persistent otorrhea (10%), early extrusion, persistent perforation, tympanosclerosis (fibrosis of the TM), granuloma formation, cholesteatoma, hearing loss (similar complication may occur with non-surgically-treated recurrent otitis media)

Additional Contributing Factors

- **Adenoiditis/Adenoid Hypertrophy**: adenoids may harbor infection or obstruct eustachian tube especially in children; may consider adenoidectomy in children >4 years old with recurrent infections or hypertrophied adenoids
- **Immunodeficiency Profile and Ciliary Disorders**
- **Reflux Evaluation**: associated with recurrent otitis media in the pediatric population

Secretory (Serous) Otitis Media or Otitus Media With Effusion

- **Secretory** (**Serous**) **Otitis Media**: persistence of fluid in the middle ear space without evidence of infection
- Pathophysiology: may be from persistent fluid from suppurative acute otitis media (≈**10%** of acute otitis media cases have persistent fluid 3 months after resolution of the infection) or secondary to eustachian tube dysfunction
- SSx: serous fluid in middle ear space (nonmobile TM, air-fluid levels), aural fullness, hearing loss, tinnitus
- Dx: clinical and physical exam, audiogram (CHL <30 dB) and tympanometry
- Rx: observation (frequent autoinflation), management of nasal congestion (eg, decongestants, nasal corticosteroids), may consider

myringotomy with pressure equalization tube, and/or adenoidectomy after failed conservative measures, antimicrobials (similar to acute otitis media) reduce the risk of secondary infection
- NOTE: any adult with unilateral, persistent middle ear effusion should undergo inspection of the nasopharynx for nasopharyngeal tumors with biopsy of suspicious lesions

Chronic Otitis Media (With Otorrhea)

- **Chronic Otitis Media**: persistent (>6 weeks) or recurrent drainage from infection of the middle ear and/or mastoid in the presence of a TM perforation (or ventilation tube)
- Pathophysiology: chronically inflamed or infected middle ear space or mastoid secondary to poor aeration (chronic eustachian tube dysfunction) or the presence of a cholesteatoma
- Pathogens: mixed infections; gram-negative bacilli (*Pseudomonas, Klebsiella, Proteus, E. coli,*), *Staphylococcus*, and anaerobes
- Risks: abnormal eustachian tube function (cleft palate, Down syndrome), immune deficiency, ciliary dysfunction (Kartagener syndrome), gastroesophageal reflux
- SSx: otorrhea (mucopurulent, odorous), TM perforation, inflamed middle ear mucosa, conductive hearing loss
- **Tuberculous Otitis Media**: uncommon; presents as insidious, painless, odorless, scant otorrhea, lymphadenopathy, multiple minute TM perforations with pale granulation tissue; usually from pulmonary source
- **Wegener's Granulomatosis**: uncommon; may present with serous otorrhea or a SNHL secondary to cochlear vasculitis
- Dx: clinical and physical exam, audiogram (CHL), use of CT is controversial, however CT should be obtained if suspect labyrinthe fistula, presence of facial weakness, or possible intracranial complications; relative indications for CT include previous surgery (revisions), only hearing ear, and inability to visualize TM

Medical Management of Chronic Otitis Media

- Aural Hygiene: cleaning of discharge and debris from EAC, water protection (ear plugs during swimming and bathing)
- Otic Drops: antimicrobial or antimicrobial /corticosteroid combination drops
- Systemic Antimicrobial Therapy: consider 3–4 weeks of oral anti-microbials (broad spectrum including anti-Pseuodomonal coverage), may consider parenteral antibiotics for persistent drainage
- Address Eustachian Tube Dysfunction/Sinonasal Disease: topical nasal coricosteroids, decongestants

Mastoidectomy

- Indications: persistent chronic otitis media despite appropriate medical therapy, coalescent mastoiditis, subperiosteal abscess, or cholesteatoma
- Goals: create a "safe" ear by eradicating infection and removal of cholesteatoma (first priority), preservation of hearing and vestibular function is second in priority
- **Intact Canal Wall Mastoidectomy**: preserves the posterior and superior wall of the EAC, procedure of choice to avoid mastoid bowl and provide a water safe ear, indicated for limited disease and a reliable patient for follow-up (to check for recurrent disease); increased risk of undetected recurrence or residual cholesteatoma (may require a "second look" procedure), other advantages include faster healing rate
- **Modified Radical Mastoidectomy** (**Canal Wall Down Technique**): posterior EAC is removed creating a "mastoid cavity or bowl"; indicated for advanced disease (eroded canal wall, large cholesteatoma), noncompliant patients, only hearing ear; provides good exposure and easier recognition of recurrent disease; disadvantages include required cleaning of mastoid cavity, difficulty with hearing aid fitting, and increased risk of infection with water exposure
- **Radical Mastoidectomy**: totally exteriorizes the middle ear, attic, and mastoid cavity and obliterates eustachian tube; rarely indicated today

Reconstruction of the Tympanic Membrane and Middle Ear

Reconstruction of the Tympanic Membrane/Tympanoplasty

- **Simple Closure** (**Paper Patch**) **Technique**: may be considered for small perforations, requires "rimming" the perforation to stimulate regrowth with placement of a cotton disc, onionskin paper, cigarette paper, silastic film, or collagen film to act as a scaffolding for cellular migration
- **Tympanoplasty**: repair of tympanic membrane typically utilizing a tissue graft such as temporalis fascia (most common), perichondrium, periosteum, cartilage, vein, areolar tissue, fat, and dura
- **Medial** (**Underlay**) **Technique**: graft placed under the annulus and remnant TM and either over or under the malleus, adequate for most TM perforations (especially smaller posterior perforations), technically easier, shorter operating time, fewer complications
- **Lateral** (**Overlay**) **Technique**: graft placed lateral to the annulus, indicated for larger perforations (especially anterior perforations), and problem perforations; longer operating time; risk of lateralizing graft and anterior blunting of the graft; requires a

longer healing process; risk of entering the glenoid fossa, greater postoperative CHL

Ossicular Chain Reconstruction (OCR)

- ossicular reconstruction re-establishes the sound conduction mechanism from the TM to the inner ear fluids
- ossicular disruption most commonly occurs at the incudostapedial joint secondary to necrosis of the lenticular process (most susceptible site of avascular necrosis)
- in addition to discontinuity of the ossicles, ossicular fixation must also be assessed
- Incus Replacement Techniques
  1. **Transposed or Sculptured Incus Autograft**: incus is removed, resculptured, and placed between the malleus and the stapes suprastructure
  2. **Homograft**: well tolerated, provides excellent sound conduction, may be presculpted, requires storage, risk of disease transfer
  3. **Synthetic Incus Strut**: constructed from a variety of materials (eg, hydroxyapatite, titanium, porous polyethylene) to recreate the connection from the malleus to the stapes suprastructure
  4. **Partial Ossicular (Replacement) Prosthesis (POP or PORP)**: replaces malleus and incus, connect the TM to the stapes capitulum, constructed from a variety of material (eg, hydroxyapatite, titanium, porous polyethylene with a cartilagenous cap)
- Incus-Stapes Replacement Techniques
  1. **Transposed or Sculptured Incus Autograft**: incus is removed, resculptured, and placed between the malleus and the stapes footplate
  2. **Homograft**: similar to incus homografts
  3. **Synthetic Incus-Stapes Strut**: similar to incus strut, re-creates connection from malleus to stapes footplate
  4. **Total Ossicular (Replacement) Prosthesis (TOP or TORP)**: replaces malleus incus, and stapes suprastructure, connect the TM to the stapes footplate, material similar to PORPs

Complications of Otitis Media

Intracranial Complications

- High Risk Pathogens: **Type III *Pneumococcus*** (intracranial predilection), ***H. influenzae* type B** (higher risk of meningitis), and presence of coexisting anaerobes

- Routes of Spread into the Intracranium: direct extension from bone erosion, lymphatic or hematogenous spread, invasion through normal anatomic structures (labyrinth), spread through iatrogenic or traumatic defects, extension through **Hyrtle's Fissure** (embryologic remnant that connects hypotympanum to the subarachnoid space)
- Dx: CT of brain with contrast or MRI of brain with gadolinium, lumbar punctures (LPs) may be required if meningitis is suspected (LPs are contraindicated with increased intracranial pressure to avoid herniation)

Epidural (Extradural) Abscess

- pus collection between skull and dura
- most commonly from direct extension via bone erosion
- SSx: may be asymptomatic, headaches, low-grade fevers, malaise
- Rx: high-dose parenteral antibiotics and surgical drainage (debride diseased bone until normal dura is exposed)

Subdural Abscess

- pus collection between dura and arachnoid membrane
- SSx: rapid neurological deterioration (may develop quickly to seizures, delirium, hemiplegia, aphasia, or coma)
- Dx: CT/MRI reveals crescent shaped enhancement (less resistance to spread) that does not cross midline
- Rx: high-dose parenteral antibiotics and neurosurgical consultation for drainage, surgical exploration with mastoidectomy and exploration of middle ear once patient is stable

Brain Abscess

- most common sites are the temporal lobe or cerebellum
- SSx and Stages
  1. **Encephalitis**: (initial invasion) fevers, headache, nuchal rigidity
  2. **Latency**: (organization of abscess, liquification necrosis) minimal symptoms, may last weeks
  3. **Expanding abscess**: intracranial hypertension, seizures, localizing signs (nominal aphasia, quadrantic homonymous hemianopia, and motor paralysis for a temporal lobe abscess; nystagmus and gait disturbances for cerebellar lesions)
  4. **Termination**: rupture of abscess, death
- Rx: high-dose parenteral antibiotics, neurosurgical consultation for drainage, surgical exploration with mastoidectomy and exploration of middle ear once patient is stable

Meningitis

- most common intracranial complication of otitis media (especially in children < 5 years old)
- increased risk with Mondini's aplasia (from dilated cochlear aqueduct)
- Pathogens: ***H. influenzae*** **(type B)**, *Pneumococcus,* hemolytic *Streptococcus*
- SSx: headache, lethargy, nuchal rigidity, irritability, fever, **Kernig's sign** (with hip in flexion, pain is elicited with leg extension), **Brudzinski's sign** (flexion at neck causes a reflexive flexion of the legs), seizures, photophobia
- Dx: lumbar puncture (after checking for increased intracranial pressure with CT or assessing for papilledema) for cells and culture (elevated CSF pressure, decreased CSF glucose, presence of inflammatory cells and bacteria, increase in protein content), may consider MRI
- Rx: high dose parenteral antibiotics, wide myringotomy or tympanectomy with culture of middle ear fluid, possible mastoidectomy and surgical exploration (may cause ossification of the labyrinth or cochlea), audiogram when stable (10–20% risk of postmeningitic partial or total, unilateral or bilateral SNHL)

Lateral Sinus Thrombophlebitis

- inflammation with subsequent thrombus formation of the sigmoid and/or transverse sinus
- Pathophysiology: mural thrombus forms in vessel wall (thrombophlebitis) → propagates distally and begins to seed
- SSx: "picket fence" spiking fevers, headache, papilledema, **Griesinger's sign** (pain over mastoid from occlusion of the mastoid emmisary vein)
- Dx: typically determined with imaging (CT may reveal enhancement within the sinus, MRI reveals increased signal intensity in both T_1 and T_2 weighted images, MRA may reveal total/partial occlusion), **Tobey-Ayer or Queckenstedt's test** (normally external compression of jugular vein results in a rapid increase in CSF pressure of 50–100 mm Hg, compression on the side of lateral sinus thrombosis results in a slow rise or no rise in CSF pressure secondary to obstruction), may consider blood cultures or LP for cells and culture
- Rx: parenteral antibiotics and possible surgical exploration via a mastoidectomy, may require ligation of internal jugular vein recalcitrant disease, anticoagulants (*controversial*)

Otitic Hydrocephalus

- (*misnomer*: does not cause dilation of the ventricles) raised intracranial hypertension associated with otitis media
- Pathophysiology: lateral sinus mural thrombus formation prevents CSF absorption which results in intracranial hypertension (*theoretical*)
- SSx: chronic course (weeks), papilledema, diplopia, nausea, headache, lethargy, abducens palsy
- Rx: address thrombophlebitis (*as above*), lower intracranial pressure (consider corticosteroids, mannitol, serial lumbar punctures), consider surgical exploration with debridement once patient is stable
- Complications: risk of blindness secondary to optic neuropathy

Acute/Subacute Mastoiditis

- Pathophysiology: suppurative infection of the middle ear spreads to the mastoid resulting in an osteitis of the mastoid air cells, continued infection may develop into a purulent infection and result in breakdown of bony septa and coalescence of air cells (**acute coalescent mastoiditis**)
- SSx: tenderness over the mastoid, associated suppurative otitis media, edema over the mastoid (indicates involvement of the cortex), fever, adenopathy
- Dx: CT of temporal bone (coalescence or lack of septations of the mastoid with the presence of fluid or soft tissue)
- Rx: parenteral antibiotics; possible wide myringotomy, tympanectomy, or pressure equalization tubes; consider mastoidectomy for recalcitrant disease

Labyrinthine Fistula

- Pathophysiology: chronic suppurative otitis media from a cholesteatoma erodes the bone of the labyrinth (most commonly at the arch of the horizontal canal)
- SSx: may be asymptomatic with progression of the cholesteatoma, dizziness, hearing loss (SNHL)
- Dx: CT may reveal bony erosion of the labyrinth, **Fistula Test** (nystagmus induced with pneumatoscopy, high false negative rate)
- Rx: surgical exploration via a mastoidectomy with exteriorization of the cholesteatoma (matrix should be left intact over the lateral SSC), graft site (fascia) if fistula is exposed, parenteral antibiotics if infected

Subperiosteal Abscess

- Pathophysiology: mastoiditis results in spread of infection to involve the outer mastoid cortex elevating the periosteum
- SSx: edema, erythema, and tenderness over site of abscess; associated suppurative otitus media; fever
- **Postauricular abscess**: most common site, spread through emissary veins or through bone, may present with a pinna protruding outward from mass effect
- **Bezold's abscess**: spread through a perforation in the mastoid cortex, tracts into sternocleidomastoid muscle, presents as a mass in the **posterior triangle of the neck**
- Dx: CT of temporal bone
- Rx: parenteral antibiotics, mastoidectomy with drainage of the abscess

Petrous Apicitis

- Pathophysiology: extension of infection into the air cell tracts around the labyrinthine capsule
- SSx: **Gradenigo triad** (otorrhea, retroorbital pain, diplopia [abducens palsy occurs from involvement of **Dorello's canal**]), fever
- Dx: CT of temporal bones
- Rx: parenteral antibiotics, consider mastoidectomy and petrous apicectomy

Facial Nerve Paralysis

- Pathophysiology: infection from suppurative otitis media that invades a dehiscent facial nerve in the middle ear space or the fallopian canal resulting in facial nerve paralysis
- Rx: surgical exploration for chronic otitis media, wide myringotomy or tympanectomy and antibiotics for acute otitis media
- *see also* p. 367

Infections of the Inner Ear

Labyrinthitis

Suppurative Labyrinthitis

- bacterial invasion into labyrinth, associated with permanent hearing loss and vestibular dysfunction, can cause meningitis
- Pathophysiology: bacterial invasion from the middle ear space through the round window membrane or oval window niche or bone erosion with spread though the otic capsule (may occur secondarily from meningitis from invasion through the cochlear aqueduct)

- SSx: sudden vertigo and permanent hearing loss, typically in the presence of a middle ear infection, tinnitus
- Dx: clinical history and exam, audiogram (SNHL)
- Rx: high-dose parenteral antibiotics, vestibular suppressants, surgical management of middle ear infection (if indicated)

Serous ("Toxic") Labyrinthitis

- toxic or virally determined serous inflammation of the labyrinth resulting in hearing loss and vertigo
- Pathophysiology: presumed diffusion of toxic inflammatory products into the labyrinth, typically from the middle ear
- SSx: dizziness, hearing loss, tinnitus
- Dx: clinical history and exam, audiogram (SNHL)
- Rx: vestibular suppressants, management of underlying infectious process
- **Luetic Labyrinthitis**: *Treponema pallidum* infection that may present in primary, secondary, or tertiary syphilis; may present with a **Tullio's sign** (vertigo and nystagmus elicited with loud noise)

Vestibular Neuronitis

- Pathophysiology: presumed viral infection of the vestibular nerve
- SSx: vertigo lasting several hours to days, not associated with hearing loss, may be associated with a prodromal upper respiratory infection, full recovery may require weeks or months
- months to years after resolution may develop BPPV
- Dx: clinical history and physical exam, unilateral vestibular weakness (decreased caloric response) on ENG, no associated hearing loss on audiogram
- Rx: consider hospitalization with parenteral hydration, high-dose corticosteroids, antivertiginous and antiemetics (long-term antiverginous medications tend to decrease vestibular habituation), vestibular rehabilitation training

NONINFECTIOUS DISORDERS OF THE EAR AND TEMPORAL BONE

Diseases of the External Ear

Cerumen Impaction and Foreign Bodies

- SSx: conductive hearing loss (if >95% of canal is occluded), tinnitus, aural fullness
- Dx: otoscopic inspection

- Rx: manual removal, consider low pressure irrigation with warm water, cerumen softening agents for severely impacted cerumen

Keratosis Obliterans (Canal Cholesteatoma)

- **Keratosis Obliterans**: external auditory canal cholesteatoma caused by blockage of the EAC permitting accumulation of epithelial debris, bone remodeling from pressure of the keratin plug may cause bony enlargement
- **Invasive Keratitis**: local accumulation of epithelial debris that occurs on the floor of the EAC
- SSx: keratin debris in EAC, bony expansion of canal wall, CHL, may become secondarily infected, may erode into middle ear or attic
- Dx: clinical history and physical examination
- Rx: frequent and aggressive debridement of EAC, cleansing with ototopical (softening) drops

Relapsing Perichondritis

- Pathophysiology: unknown etiology, inflammation of elastic cartilaginous tissue with high concentration of glycosaminoglycans
- SSx: episodic and progressive symptoms
  1. auricular chondritis, cochlear and vestibular injury (vertigo, hearing loss)
  2. respiratory chondritis (laryngeal collapse)
  3. nasal chondritis (saddle nose deformity)
  4. polyarthritis (nonerosive, migratory)
  5. cardiac valve insufficiency
- Histopathology: perichondrial inflammation, fibrous tissue replacement
- Dx: clinical history and physical exam, elevated nonspecific immunological markers (eg, ESR, IgG), elevated antibodies to type II and type IV collagen (must differentiate from rheumatoid arthritis, gout, and other connective tissue diseases)
- Rx: NSAIDs, corticosteroids for severe attacks

Exostoses

- Pathophysiology: unknown etiology (associated with **cold water swimming**) resulting in a benign bony outgrowth that arises from the tympanic bone in the EAC
- SSx: single or **multiple**, unilateral or **bilateral** smooth sessile protrusions of the medial bony canal, results in narrowing of the canal with progressive growth

- Dx: clinical history and physical exam
- Rx: excise if symptomatic

Osteoma

- Pathophysiology: true neoplastic benign outgrowth in the bony canal
- SSx: **single**, **unilateral** protrusion at the **bony cartilage junction**, asymptomatic unless becomes obstructive
- Dx: clinical history and physical exam
- Rx: excise if symptomatic

Temporal Bone Neoplasms and Lesions

Cholesteatoma

- (*misnomer*: not a true neoplasm) squamous epithelium in the middle ear, epitympanic, and mastoid cavities, accumulation results in bone and soft tissue erosion and recurrent infections
- **Primarily Acquired**: cholesteatoma that extends from the invagination of a retraction pocket, associated with negative ear pressure (eustachian tube dysfunction); pars flacida pockets are the most common and initially invade the epitympanum via **Prussak's space** (bounded by lateral malleal fold [roof], lateral process of the malleus [floor], neck of the malleus [medial], and Shrapnell's membrane [lateral]), pars tensa retraction pockets more directly invades the middle ear
- **Secondarily Acquired**: cholesteatoma that forms secondarily from ingrowth of keratinizing epithelium into the middle ear space through a pre-existing TM perforation (previous otitis media) or secondary to trapping of canal skin from trauma
- **Congenital**: cholesteatoma that occurs from an embryonic epithelial tissue within the temporal bone (commonly associated with the tensor tympani muscle), presents as a white mass generally in the anterosuperior middle ear with an intact TM
- Pathophysiology: squamous epithelium formed from trapped epithelium from a retraction pocket, TM perforation, or temporal bone trauma (acquired cholesteatoma); congenital rest cells (congenital cholesteatomas); proteolytic enzymes and infection lead to osteolytic bone destruction; presence of cholesteatoma may harbor infection resulting in a chronically draining ear
- SSx: white "pearly" mass within the middle ear or mastoid space, nonresolving chronic suppurative otitis media, progressive

conductive hearing loss ("silent cholesteatoma" may occur when a large cholesteatoma acts as a sound conductor)
- Dx: visualization of squamous epithelium by otomicroscopic evaluation, CT of temporal bone may be considered in select cases (see p. 316, abnormal soft tissue mass within middle ear, epitympanum, or mastoid cavity with associated bony erosion, most commonly of the scutum), audiometry (CHL)
- Histopathology: compact sac of keratinizing squamous epithelium with a central core of keratin debris
- Complications: destruction of the ossicular chain, chronic otitis media labyrinthine fistula, intracranial complications, facial nerve paralysis
- Rx: surgical removal or exteriorization with possible primary or secondary ossicular chain reconstruction or tympanoplasty, address chronic otitis media (*see* pp. 316–318)

Glomus Tumors (Paragangliomas, Chemodectomas)

- **Glomus Tumor**: slow growing, benign tumor although locally destructive (expansive, bone eroding) of the chemoreceptive cells (paraganglion cells, neural crest in origin) distributed along parasympathetic nerves in the base of skull, neck, and chest
- most common benign tumor of the temporal bone of adults (rare in pediatrics)
- **10%** multiple
- **<5%** malignant degeneration
- **1–3%** associated with a paraneoplastic syndrome (paroxysmal hypertension, headache, and palpitations) from secretion of vasoactive catecholamines and other neuropeptides
- Associated: other MEN syndromes, genetic disposition (**10% family history**)
- Types
  1. **Carotid Body**: arise from carotid body, most common type, typically does not involve the temporal bone
  2. **Glomus Tympanicum**: arise from the promontory along the course of Jacobson's nerve, tympanic branch of CN IX, confined to the middle ear space
  3. **Glomus Jugulare**: arise from the region of the jugular foramen
  4. **Glomus Vagale**: arise from paraganglia around the vagus nerve at the base of skull
- SSx: pulsatile tinnitus (most common symptom), hearing loss, cranial nerve palsies, aural discharge, otalgia, dizziness (invasion of the labyrinth), reddish middle ear mass, **Brown's sign** (blanching of the TM with positive pressure)

- Dx: high-resolution CT of temporal bones with constrast bolus; may also consider MRI/MRA ("salt and pepper" lesion on T2, weighted images, may miss small tumors), bilateral four-vessel angiography (assesses collateral circulation and allows for preoperative embolization), audiometry (baseline hearing), and 24-hour urine catecholamine screen (vanillylmandelic acid, metanephrine, normetanephrine)
- Histopathology: nests of nonchromaffin staining cells clustered among vascular channels lined by epithelioid cells
- Rx: surgical resection after preoperative embolization or primary radiation therapy for nonoperable candidates, advanced tumors, or presence of residual disease

Acoustic Neuromas (Vestibular Schwannomas) and Other Cerebellopontine Angle Tumors

- **Acoustic Neuromas**: (*misnomer*: typically does not arise from the cochlear nerve and is not a neuroma) most common CPA tumor (≈85% of CPA tumors), arises from the schwann cells typically of the vestibular portion of CN VIII (**Vestibular Schwannoma**), indolent growth rate, very rare malignant potential
- **Cerebellopontine Angle**: space in the posterior fossa bordered anterolaterally by the petrous bone and clivus, posterolaterally by the cerebellum, inferiorly by the cerebellar tonsil, and superiorly by the cerebellar peduncles and pons
- **Neurofibromatosis Type 2 (NF2, von Recklinghausen Disease)**: autosomal dominant disorder associated with bilateral acoustic neuromas and other intracranial and spinal cord tumors (*see also* pp. 345–346)
- Acoustic Neuroma Histologic Types
  1. **Antoni Type A**: histologically parallel nuclei, uniform spindle cells, compact cells
  2. **Antoni Type B**: histologically less uniform, may have fatty or hyaline degeneration, less cellular
- SSx: progressive, asymmetric, high-tone hearing loss (most common presenting symptom); tinnitus; vertigo or imbalance; advanced tumors may present with facial nerve (**Hitselberger's sign**, reduced sensation of the EAC and conchal bowl) or trigeminal nerve involvement, papilledema, occipital headache, lower cranial nerve involvement, or ataxia

Diagnosis

- **Audiometry**: initial screening, asymmetric hearing loss with decreased SDS

- **Vestibular Tests**: normal or asymmetric caloric weakness
- **Auditory Brainstem Response**: most sensitive and specific audiological test (10–15% false negative rate), may be used for selective screening asymmetrical hearing loss or vestibular weakness (*see* page 293–294)
- **MRI with gadolinium**: enhances best imaging study, sensitive to tumors >2 mm, brightly enhance with gadolinium
- **CT with contrast**: indicated if MRI is unavailable or contraindicated, may miss tumors smaller than 10 mm, suspect with asymmetric widening of the internal auditory canal

Management

- surgical excision (subtotal excision may be considered if resection creates serious risk of neurological injury or in the elderly)
- **Stereotactic Radiation Therapy** (gamma knife or linear accelerator [LINAC] therapy): may be considered for tumors <2 cm, nonoperable candidates, prevents tumor growth (does not remove tumor), risk of facial nerve injury and hearing loss similar to surgical excision
- may consider observation in selected patients, (elderly) and nonoperable candidates, follow with serial MRIs every 9–12 months
- NF2: routine screen of all relatives with MRI of brain and spine and audiometry for bilateral acoustic neuromas, remove largest tumor first (consider placement of Auditory Brainstem Implant if hearing is sacrificed), if hearing is preserved may consider removal of second tumor, for only hearing ear may follow with serial MRIs every 9–12 months

Other CPA Lesions

- **Meningiomas**: second most common CPA tumor (≈15%), arise from cap (endothelial) cells from the tips of the arachnoid villi, presentation similar to acoustic neuromas, imaging typically shows differences (meningiomas are more homogeneous, dense, contain calcifications, and have a flat dural base), histopathology reveals large endothelial cells with uniform nuclei occurring in whorls and psammoma bodies; Rx: surgical excision, external beam radiation, gamma knife
- **Arachnoid Cysts**: thin walled sac within the arachnoid, filled with CSF, may be differentiated with MRI; Rx: incision and drainage for symptomatic lesions
- **Epidermoids**: primary or congenital cholesteatoma; may erode bone; may present with cranial nerve palsies, headache, or hearing loss
- **Hemangiomas**: (rare) capillary hemangiomas may involve the facial nerve particularly in the IAC and perigeniculate region (rather than the CPA), may erode into cochlea causing pulsatile tinnitus, may grow bone within lesion forming an Ossifying Hemangioma

- **Metastatic Lesions**: rare, primaries from glial tumors, breast cancer, lung cancer, and others
- **Malignancies**: present as more rapidly growing, earlier cranial nerve involvement, and destructive rather than expansive growth
- Others: lipomas, lymphomas, hemangiomas, gliomas, dermoids, teratomas, melanomas, paragangliomas

Surgical Approaches to the CPA

Translabyrinthine

- Indication: approach of choice for nonserviceable hearing
- Advantages: most direct route, excellent exposure, minimal cerebellar retraction, higher incidence of CSF leak than retrosigmoid/suboccipital approach, less risk of facial nerve injury
- Disadvantages: sacrifices hearing

Retrosigmoid/Suboccipital

- Indication: large tumors, tumors of the medial IAC, hearing preservation, most common approach for a vestibular neurectomy
- Advantages: provides wide exposure, hearing preservation possible, lower risk of facial nerve injury than middle cranial fossa approach
- Disadvantages: higher incidence of postoperative headaches, limited exposure to lateral IAC, higher risk of facial nerve injury than translabyrinthe approach, requires cerebullar retraction (postoperative imbalance)

Middle Cranial Fossa

- Indication: small intracanalicular lesions, serviceable hearing
- Advantages: possibility of preserving hearing
- Disadvantages: requires retraction of temporal lobe (risk of aphasia or seizure), more difficult technically, poor exposure to posterior fossa, higher risk of facial nerve injury than translabyrinthine or retrosigmoid/suboccipital approaches

Retrolabyrinthine

- Indication: not indicated for acoustic neuromas (does not allow adequate exposure); indicated for vestibular nerve section
- Advantages: preserves hearing, easier technically (shorter operative time)

- Disadvantages: limited exposure and narrow field of view (cannot access IAC lesions or porus acusticus)

Petrous Apex Lesions

- **Petrous Apex**: pyramidal shaped, portion of the temporal bone medial to the otic capsule
- SSx: headache (dural pressure), cranial compression symptoms, vertigo or imbalance, eustachian tube dysfunction and CHL (serous otitis media)
- Dx: CT or MRI of temporal bone, audiometry
- **Cholesterol Granulomas**: cystic lesion of the petrous apex, foreign body response to cholesterol crystals causing a giant cell reaction; contains brown, thick fluid; diagnosed by CT (smooth, round, expansive lesions, smooth remodeled bone, may have cyst wall enhancement) or MRI (bright images on both T_1 and T_2 weighted sequences, unlike epidermoids (cholesteatomas) which are hyperintense on T_2 weighted images only); Rx: surgical decompression and exteriorization
- **Congenital Epidermoid Cyst** (**Cholesteatomas**): cystic lesion formed from embryological entrapped epithelial remnants, slow growth, presents as a young adult; Rx: surgical removal and exteriorization
- Other Lesions of the Petrous Apex: meningiomas, glomus tumor, lymphoma, or metastatic tumors

Other Benign Neoplasms of the Temporal Bone

- **Fibrous Dysplasia**: developmental disease, medullary bone replaced with fibro-osseus tissue, usually monostotic, presents as a painless asymmetric swelling of the temporal bone, CHL, EAC narrowing, "ground glass" expansive mass of CT; Rx: excision
- **Histocytosis X**: granulomatous disease of unknown etiology, characterized by a proliferation of histiocytes, localized and chronic disseminated forms, presents with chronic otorrhea and granulation tissue; Rx: excision, radiation therapy, chemotherapy, or multimodality therapy (depends on histology type and sites involved)
- **Embryonic Tumors**: dermoids, teratomas, chordomas

Malignant Tumors of the Temporal Bone

- numerous pathways of tumor invasion that allow easy spread of tumor (typically presents in an advanced stage)

- SSx: EAC mass or granulation tissue, chronically draining ear, insidious CHL, aural fullness, bloody otorrhea, pain, facial nerve and other cranial nerve palsies, vertigo
- Dx: CT of temporal bone, biopsy, full head and neck work-up (*see* pp. 219–225), audiogram (baseline hearing)

Types

- **Squamous Cell Carcinoma** (**SSC**): more commonly arises from the auricle or EAC; Rx: wide surgical resection with consideration of adjunctive radiation therapy (for advanced disease) versus primary radiation (for unresectable disease or inoperable candidates)
- **Basal Cell Carcinoma**: often involves the auricle, may extend into EAC and temporal bone; Rx: local resection (with negative margins)
- **Adenoid Cystic Carcinoma**: uncommon tumor in the temporal bone, may arise from ectopic salivary gland tissue in middle ear, may invade neural tissue causing pain, histologically may have skip areas (high recurrence), unpredictable incidence of late metastasis; Rx: resection with consideration of adjunctive radiation therapy versus primary radiation
- **Rhabdomyosarcoma**: most common temporal bone malignancy of childhood, may arise from tensor tympani or stapedius muscles; Rx: chemotherapy and radiotherapy (surgical management rarely indicated)
- Others: Ceruminous Adenocarcinoma, Adenocarcinoma, Malignant Melanoma, Carcinoid Tumor, Verrucous Carcinoma, Giant Cell Tumor, Chondrosarcomas, Osteosarcomas, Metastasis

Temporal Bone Resection

- **Sleeve Resection**: indicated for tumors of the concha, periauricular, or cartilaginous canal regions with sparing of the bony EAC, parotid, and TMJ; removes bone lateral to TM
- **Lateral Temporal Bone Resection**: indicated for tumors lateral to the TM (may involve outer layer of the TM); removes mastoid bone, TM, incus, and malleus (may spare otic capsule and facial nerve for select tumors)
- **Total Temporal Bone Resection** *(controversial)*: indicated for tumors that involve the middle ear space, mastoid cavity, medial layer of the TM, or deep temporal bone; removes temporal bone lateral to the internal acoustic canal (subtotal resection often implies sparing of the petrous apex); results in anacusis, initial vestibular dysfunction, facial nerve paralysis, and CHL

Otosclerosis (Otospongiosis)

- abnormal resorption and deposition of bone in all 3 layers of the otic capsule (endosteal, endochondreal, and periosteal) and ossicles
- Pathophysiology: increased osteoclastic activity results in perivascular bony resorption forming fibrotic spaces (**lytic phase**), osteoblasts within the fibrotic spaces producing immature bone (**bone production phase**), cycling of resorption and bone formation results in otosclerotic bone (**remodeling phase**)
- may be an autosomal dominant trait with varying degree of penetrance
- most common involved site is anterior to oval window (**fissula ante fenestrum**), second most common site is the border of the round window (**Round Window Otosclerosis**)
- ≈10% of Caucasians have histological evidence of otosclerosis (only <1% develop clinical otosclerosis)
- frequent bilateral involvement (not necessarily associated with a hearing loss, up to 85%, varies widely among studies)
- pregnancy is associated with acceleration of otosclerosis
- **Carhart notch**: characteristic depressed bone conduction threshold at 2000 Hz which typically improves after stapedectomy causing **overclosure** of the air-bone gap (postoperative air conduction thresholds are better than preoperative bone conduction thresholds) suggesting that the cause of this audiological phenomenon is artifactual
- SSx: slowly progressive conductive hearing loss beginning between 20–40 years old, tinnitus, **Schwartze sign** (red hue behind tympanic membrane from hyperemia of the promontory mucosa from increased vascularity, represents active phase)
- Dx: clinical history, audiogram (CHL, normal speech discrimination, Carhart notch), tympanometry (may have an A_S tympanogram), and tuning fork exam, confirmation of otosclerosis is done intraoperatively
- Histopathology: active lesions reveal spongy bone seen as blue with staining (**blue mantles of Manasse**), hypercellularity, active osteocytes and osteoblasts, increased resorption spaces, and increased vascular channels; inactive lesions reveal resorption spaces filled with collagen and osteoid, sclerotic bone, and narrowed vascular spaces
- **Cochlear** (**Labyrinthine**) **Otosclerosis**: presents as an SNHL, rarely associated with vertigo

Differential Diagnosis

- **Ossicular Fixation**: may be congenital, secondary to infection or trauma, or a result of bony ankylosis or ossification of suspensory ligaments; Rx: surgical reconstruction

- **Paget's Disease** (**Osteitis Deformans**): (rare) autosomal dominant with variable penetrance, excessive absorption of bone with fibrovascular replacement forming weak haphazard trabecular bone (mosaic bony changes), similar histologically to otosclerosis pattern except begins in periosteal layer, diffuse involvement of the skull, and typically does not involve the stapes footplate; Rx: hearing amplification, medical management to inhibit bone resorption (eg, mithramycin, calcitonin)
- **Osteogenesis Imperfecta Tarda** (**van der Hoeve Syndrome**): (rare) autosomal dominant pattern characterized by increased bone resorption and abnormal remodeling, presents in childhood (tarda form), presents clinically with multiple fractures, blue sclera, may result in stapes fixation; Rx: may consider stapedectomy for stapes fixation
- **Osteopetrosis** (**Albers-Schönberg Disease**): (rare) dominant or recessive forms, results in ossicular anomalies (the otic capsule is resistant to the disease), thickened bones of the skull base, blindness (optic nerve atrophy), absent paranasal sinuses, choanal atresia, facial paralysis, hepatosplenomegaly, brittle bones; Rx: hearing amplification

Nonsurgical Management

- **Hearing Aid/Observation**: may be considered for those who do not want to undergo elective stapes surgery, an only hearing ear, failed surgical management, high-risk patients, and nonoperable candidates (elderly)
- **Medical Management**: sodium fluoride (may prevent progression of hearing loss); vitamin D and calcium (*controversial*)

Stapedectomy/Stapedotomy

- Indications: conductive hearing loss (>20 dB ABG, negative Rinne with 512 Hz tuning fork) secondary to otosclerosis with adequate cochlear reserve and good speech discrimination (initial stapedectomy should be carried out on the poorer hearing ear, the better ear may be addressed 6–12 months later)
- Contraindications: only hearing ear, active infection, suspected active endolymphatic hydrops
- **Stapedectomy**: totally or partially removes stapes footplate with placement of a prosthesis from the incus to the vestibule
- **Small-Fenestra Stapedotomy**: creates a fenestration in the footplate for placement of a prosthesis, fenestrations may be completed with a microdrill system or laser (CO_2 or KTP)

- **Complications**: SNHL (overall 1–3% may be profound), facial nerve injury (<1%), chorda tympani nerve injury (taste disturbance), displaced prosthesis or loose wire syndrome, tinnitus with dizziness

Intraoperative Considerations in Stapes

- **Perilymphatic Gusher**: (rare) may occur secondarily to a congenitally widened cochlear aqueduct, risk of permanent SNHL; Rx: seal vestibular opening with a graft, place prosthesis, pack and close, manage for perilymph fistula, may consider spinal tap or lumbar drain
- **Facial Nerve Dehiscence**: facial nerve may block access to oval window; Rx: if significant may cancel procedure, otherwise may consider gently retracting nerve superiorly (*controversial*)
- **TM Perforation**: may occur intraoperatively; Rx: small perforations may be repaired with placement of fascia, perichondrium, or fat graft; larger perforations may require deferring the stapedectomy
- **Floating Footplate**: freely mobile footplate after the stapes superstructure is removed, risk of depressing the footplate into the vestibule; Rx: if control hole is in place may attempt to remove (creating a control hole may be difficult) or abort and wait for the footplate to refix and reoperate later (laser techniques minimize floating footplates)
- "**Biscuit Footplate**": primary focus of otospongiosis causes a thickened footplate; Rx: utilize microdrill diamond to "blueline," then usual technique
- **Persistent Stapedial Artery**: depending on size may use laser or bipolar cautery to remove small artery then proceed
- **Associated Ossicular Chain Fixation**: After separation of incudostapedial joint, it is essential to evaluate for stapes or lateral chain fixation, perform ossiculoplasty technique or mobilize lateral chain

Trauma to the Ear and Temporal Bone

Barotrauma

- Pathophysiology: ambient pressure changes (such as from diving or flying) result in pressure differentials affecting the air-containing spaces of the temporal bone which may cause otalgia (pressure differential of 60 mm Hg), eustachian tube dysfunction (pressure differential of 90 mm Hg), or middle ear hemorrhage or TM perforation (pressure differential of 100–500 mm Hg)

- SSx: acute pain, hemotympanum, bloody otorrhea, acute hearing loss (SNHL or CHL), dizziness, tinnitus
- Dx: clinical exam and history
- Rx: analgesics, anti-inflammatory agents, prophylactic oral and topical decongestants

Types of Barotrauma

- **Middle Ear Squeeze**: on ascent the eustachian tube may close, thus failing to relieve pressure in the middle ear, as ambient pressure increases may cause the TM to rupture or middle ear hemorrhage resulting in hemotympanum (*theoretical*)
- **Round Window Rupture**: may occur as the diver attempts a Valsalva maneuver: the TM snaps into a neutral position pulling the stapes causing negative pressure in the cochlea (implosion of the round window) or as the diver attempts a Valsalva maneuver there is an increase in CSF pressure that is transmitted through the perilymph causing a rupture of the round window (*theoretical*)
- **Inner Ear Decompression Sickness** (**Caisson Disease, "The Bends"**): with increased pressure (diving), nitrogen becomes more soluble and enters the blood and body fluids; if decompression occurs rapidly, small gas emboli may form and enter the cerebral circulation leading to blindness, deafness, paralysis, or death; Rx: recompression (hyperbaric oxygen)

Temporal Bone Fractures

Initial Evaluation

- temporal bone fractures are associated with severe traumatic forces, initial exam includes a complete trauma work-up (*see* pp. 443–449)
- Focused History and Physical Examination: determine cause (blunt versus penetrating, lateral versus occipital); assess facial nerve (partial versus complete, immediate versus delayed); hearing loss (tuning forks if stable); dizziness (nystagmus); presence of other neurological deficits and cranial nerve palsies; clean EAC with suction (irrigation is contraindicated) to evaluate for CSF leak, hemotympanum, EAC lacerations; Battle's sign (ecchymosis over the mastoid process)
- High-Resolution CT of Temporal Bones: imaging of choice for temporal bone fractures, examine site and type of fracture (*see* Table 6–4), site of potential facial nerve injuries, and otic capsule involvement (Indications for CT of temporal bone are controversial, may not change clinical management)

TABLE 6-4. Characteristics of Longitudinal and Transverse Temporal Bone Fractures*

Characteristic	Longitudinal Fractures	Transverse Fractures
Fracture Line	parallel to long axis of petrous pyramid	perpendicular to long axis of petrous pyramid
Vector Force	temporoparietal blow	frontal or occipital blow
Relation to Otic Capsule	fracture remains anteriomedial to otic capsule	fracture may transect otic capsule and IAC
Hearing Loss Type	CHL (ossicular disruption, blood in the EAC)	SNHL (labyrinthine or nerve injury)
Signs and Symptoms	blood in EAC, Battle's sign, EAC step-off, rare vestibular signs	hemotympanum, nystagmus, CSF otorrhea

* **Pure longitudinal and transverse fractures** are not common, **most fractures follow mixed types and mixed planes**

- Audiogram: assess for CHL (suggestive of ossicular discontinuity) versus SNHL (cochlear injury)
- Arteriography: may be considered when injury to the carotid artery may be suspected (fracture extends to foramen lacerum on CT)

Management

Facial Nerve Injury

- higher risk of facial nerve injury occur with transverse or mixed fractures (30–50% incidence) due to the perpendicular path of the fracture with respect to the facial nerve
- most common site of injury of the facial nerve in temporal bone fractures is at the perigeniculate region (labyrinthine and horizontal segments)
- delayed facial nerve paralysis may occur secondary to post-traumatic edema and ischemia with or without temporal bone fractures
- with complete facial paralysis serial ENoG or EMG testing may be utilized to assess nerve integrity and function
- management includes coritcosteroids for any degree of facial nerve injury, surgical exploration indications are similar to idiopathic facial nerve paralysis (*see* p. 366)

Hearing Loss

- SNHL is more common with transverse or mixed fractures through the otic capsule
- CHL is more common with longitudinal fractures from ossicular injury, TM tears, or blood or spinal fluid in the EAC
- post-traumatic profound hearing loss has poor prognosis for recovery
- surgical exploration and repair may be considered for persistent conductive hearing loss
- Ossicular Injury: **Incudostapedial Joint Separation** (most common ossicular injury with temporal bone trauma), **Fractured Stapes Crura**, **Incus Dislocation**; Rx: ossicular chain reconstruction
- TM Injury: more common with longitudinal fractures; Rx: small TM tears typically heal spontaneously, may consider paper patch procedure
- SNHL: may occur from a fractured labyrinth, perilymph fistula, inner ear concussion, auditory nerve injury, or acoustic trauma

CSF Leak

- SSx: clear otorrhea or rhinorrhea, salty taste, "halo sign" (fluid dropped on gauze separates into a clear outer ring with blood in the center)
- Dx: fluid analysis (CSF typically has a glucose level >30 mg/cc, chloride level approximately 124 mEq/liter, and the presence of β-**transferrin**), contrast cisternography (metrizamide or iodophendolate) with CT, interthecal radioactive isotopes or fluorescein with measurements of the radioactivity of cotton pledgets after placing at site of leak or visualization of fluorescein under ultraviolet light (more sensitive but does not localize leak)
- Initial Conservative Management: bed rest, head elevation, osmotic and diuretic agents (mannitol, acetazolamide, furosemide), may consider prophylactic antibiotics (*controversial*)
- Persistent Drainage: after 2–3 days of conservative management may consider a lumbar drain or surgical exploration with closure

Other Complications

- **EAC lacerations**: EAC lacerations are almost always associated with an underlying fracture, complete closure of EAC may occur and must be repaired to avoid trapping skin
- **Cholesteatoma**: late complications from entrapped squamous epithelium from the EAC into the middle ear space
- **Dizziness**: typically self-limiting; may be related to inner ear injury (postconcussion syndrome), injury to the vestibular labyrinth may cause a complete unilateral vestibular deficit (manage with suppressant therapy and vestibular rehabilitation), must also consider perilymphatic fistulas

Tympanic Membrane Perforations

- Pathophysiology: **acute/chronic suppurative otitis media** (most common cause), persistent perforation after extrusion of a **pressure equalization tube**, **trauma** (hand blow to the ear, barotrauma, diving, water skiing, explosion, forceful irrigation, slag burns), **iatrogenic**, **cholesteatoma** (associated with peripheral perforations)
- spontaneous closure of a perforated TM results in a **monomeric membrane** (*misnomer*: actually has two layers, outer epidermal and inner mucous)
- Types
  1. **Central**: perforation does not involve the annulus, typically infectious

2. **Marginal**: involves the annulus, less likely to resolve spontaneously, higher association with cholesteatomas
3. **Subtotal**: large defect completely surrounded with an intact annulus

- SSx: CHL, tinnitus
- Dx: otoscopic exam, audiogram
- Rx: keep ear dry, may consider a tympanoplasty for a persistent perforation (*see* pp. 317–318)

PEDIATRIC AND FAMILIAL HEREDITARY HEARING LOSS

Evaluation for Hearing Loss in the Pediatric Patient

History

- Character of Hearing Loss: age of onset, progression of hearing loss, communication skills
- Prenatal and Perinatal History: term of delivery, birth weight, prenatal infections, bilirubinemia, Apgar score, maternal drug and alcohol abuse, complications (intensive care unit, ECMO)
- Contributing Factors: syndromic features (33–50% of congenital hearing loss is syndromic); family history of hearing loss; history of neurologic (seizures), cardiac (Jervell and Lange-Nielsen), thyroid (Pendred), renal (Alport's), sickle cell, infections (recurrent otitis media, meningitis), or other congenital disease; delayed development (growth history); surgical history (otologic, neurologic); medications (ototoxic); recent trauma
- Associated Symptoms: delayed speech development, imbalance or gait disturbances, vision problems, other neurologic complaints
- NOTE: most common prenatal cause of hearing loss is **intrauterine infection**; common causes of perinatal hearing loss are **hypoxia, hyperbilirubinemia, infection,** and **medication toxicity**; most common postnatal cause of hearing loss is **meningitis**

Physical Exam

- Otoscopy/Microscopy/Pneumatoscopy: inspect EAC, tympanic membrane, pneumatoscopy
- Inspection and Palpation: inspect auricle, periauricular pits, and other facial malformations

- Other Physical Exam: orofacial deformities (palate and lip deformities, mandible and maxilla abnormalities), goiter (Pendred), ocular exam (Stickler, Usher, Osteogenesis Imperfecta), telecanthus and white forelock (Waardenburg), other syndromic defects (limbs, phalanges, café-au-lait spots, etc)

Testing Hearing in Infants and Children

- Indications: universal screening of neonates may be employed in the future, otherwise, should screen for high risk factors (TORCH infections or bacterial meningitis, family history, presence of other head and neck abnormalities, birth weight <1500 g, neonatal hyperbilirubinemia, Apgar score <4 at 1 minute or <6 at 5 minutes (asphyxia), prolonged stay at the neonatal intensive care unit or ECMO)
- objective measures such as ABR, immittance testing, and otoacoustic emissions are the mainstay of testing
- **Behavior Observation Audiometry**: narrow band of tones (warble tones) are introduced via a speaker and the examiner observes for a response (eye widening, startle, head turn, etc) to provide a means for grossly estimating an infants' auditory thresholds
- by 3 years old may consider conventional testing

Ancillary Tests (may be considered for select cases)

- **CT of Temporal Bones**: may be considered to evaluate for inner ear disorders, cholesteatoma, and osteodysplasias
- **Complete Blood Count**: may suggest leukemic process, labyrinthitis, active inflammation
- **Treponemal Studies**: Lyme titers and VDRL/FTA-ABS (luetic labyrinthitis)
- **Perchlorate Test and Thyroid Function Tests**: indicated if suspect Pendred syndrome
- **Lipid Profile**: evaluate for hyperlipedemia (associated with hearing loss)
- **Immune Function Tests**: may suggest an autoimmune disorder (Cogan's)
- **Electrocardiogram**: screen for Jervell and Lange-Nielsen syndrome, Rifsum syndrome (retinitis pigmentosa, hypertrophic peripheral neuropathy, SNHL)
- **Urine Analysis, Blood Urea Nitrogen, Serum Creatinine**: screen for Alpert's and Branchio-oto-renal syndromes

- **TORCH Studies**: IgM assay which investigates common intrauterine infections (**T**oxoplasmosis, **O**ther [syphilis], **R**ubella, **C**ytomegalovirus, **H**erpes simplex

General Management Concepts

- if suspect syndromic hearing loss consider genetic evaluation (chromosomal analysis) and counseling
- hearing amplification devices should be offered early to prevent speech and developmental delays
- serial audiograms should be administered to evaluate for progression
- surgical correction may be feasible for some conductive hearing losses (eg, stapes fixation, auricular and external ear canal abnormalities, and chronic otitis media)

Acquired Prenatal Hearing Loss

Congenital Rubella

- rare since the use of the vaccine and prenatal testing
- Pathophysiology: maternal rubella causes atrophy of the organ of Corti, loss of hair cells, and thrombosis within the stria vascularis
- Otologic SSx: ossicular and cochlear disorders, severe to profound SNHL, may cause a delayed endolymphatic hydrops
- Other SSx: cardiac malformation, congenital cataracts, anemia, mental retardation, deformities of the lower extremities, microcephaly, thrombocytopenia
- Dx: culture virus from urine, throat, or amniotic fluid; anti-rubella IgM

Congenital Syphilis

- Pathophysiology: maternal *Treponema pallidum* infection that crosses the placenta
- congenital syphilis is often fatal
- Otologic SSx: deafness occurs within first 2 years of life or may present in a delayed form that manifests at second or third decade of life, **Hennebert's sign** (positive fistula test in delayed congenital syphilis), may present with endolymphatic hydrops
- Hutchinson's Triad: abnormal central incisors (Hutchinson's teeth), interstitial keratitis of the eye, deafness
- Dx: clinical history and exam, VDRL (nonspecific), FTA-ABS (specific), audiogram
- Rx: long-term penicillin, ampicillin, tetracycline, or erythromycin; consider corticosteroids for associated hearing loss

Congenital Cytomegalovirus

- rare since the use of the vaccine and prenatal testing
- Pathophysiology: spread from maternal primary CMV
- Otologic SSx: ≈10% hearing loss (mild to profound, unilateral SNHL), may be progressive
- Other SSx: hemolytic anemia, microcephaly, mental retardation, hepatosplenomegaly, jaundice, cerebral calcifications
- Dx: serum anti-CMV IgM and the presence of intranuclear inclusions ("owl eyes") in renal tubular cells found in urinary sediment

Common Inner Ear Dysmorphologies

Michel's Aplasia

- Pathophysiology: autosomal dominant trait that results in complete failure of the development of the inner ear
- SSx: anacusis, normal middle and outer ear
- Dx: CT reveals a hypoplastic petrous pyramid and an absent cochlea and labyrinth
- Rx: may consider vibrotactile devices for bilateral involvement

Mondini Aplasia

- Pathophysiology: autosomal dominant causing developmental arrest of the bony and membranous labyrinth
- SSx: progressive or fluctuating unilateral or bilateral hearing loss (may not have a hearing loss)
- associated with an increased risk of perilymphatic gushers and meningitis from a dilated cochlear aquaduct
- Dx: CT reveals a **single-turn curved cochlea** with cystic dilation of the cochlea (no interscalar septum which divides the cochlear turns), semicircular canals may be absent or wide, wide vestibular aqueduct
- Rx: amplification, cochlear implant

Scheibe Aplasia

- Pathophysiology: autosomal recessive disorder that results in **partial or complete aplasia of the pars inferior** (cochlea and saccule) and normal pars superior (semicircular canals and utricle)
- associated with other diseases such as Usher syndrome and Waardenburg syndrome
- SSx: SNHL
- Dx: difficult to diagnose by CT since primarily a membranous defect, definitive diagnosis may only be determined by histological examination

- Rx: amplification

Alexander Aplasia

- Pathophysiology: autosomal recessive disorder that results in an abnormal cochlear duct
- SSx: mild high frequency loss
- Dx: difficult to diagnose by CT since primarily a membranous defect, definitive diagnosis may only be determined by histological examination
- Rx: amplification

Common Autosomal Recessive Causes of Congenital Hearing Loss

Usher Syndrome

- most common cause of congenital deafness
- Pathophysiology: uncertain etiology, primarily an autosomal recessive trait (also may be inherited as an autosomal dominant or X-linked recessive) resulting in a variable expression of SNHL and progressive retinitis pigmentosa
- Otologic SSx: congenital SNHL (from degeneration of the organ of Corti and spiral ganglion cells)
- Other SSx: **progressive retinitis pigmentosa** (delayed tunnel vision and blindness), vestibular dysfunction, mental retardation, cataracts
- Types
  I. profound SNHL, no vestibular response, legally blind by early adulthood, most common form
  II. moderate to severe SNHL, legally blind by mid-adulthood, normal vestibular function
  III. progressive SNHL, varied progression in blindness, progressive vestibular dysfunction

Dx: electroretinography

Pendred Syndrome

- Pathophysiology: defect in tyrosine iodination
- Otologic SSx: severe to profound SNHL, normal middle and outer ear, associated with a Mondini deformity
- Other SSx: **multinodular goiter** at 8–14 years old from failure of iodine organification
- Dx: positive perchlorate test (decreased perchlorate discharge)

- Rx: exogenous thyroid hormone (suppress goiter growth, no effect on hearing), thyroidectomy typically not required

Jervell and Lange-Nielsen Syndrome

- Pathophysiology: uncertain etiology resulting in severe SNHL and cardiac defects
- Otologic SSx: bilateral SNHL
- Other SSx: **cardiac abnormalities**, recurrent **syncope** and sudden death
- Dx: electrocardiogram (prolonged QT, large T waves)
- Rx: β-blockers, hearing amplification

Goldenhar Syndrome (Hemifacial Microsomia, Oculoauriculovertebral Spectrum)

- Pathophysiology: unknown origin, affects the development of first and second arch derivatives
- Otologic SSx: preauricular appendages; pinna abnormalities; atresia of the EAC; ossicular malformation or absence (CHL); abnormal development of the facial nerve, stapedius muscle, semicircular canals, and oval window (SNHL)
- Other SSx: **ocular** (epibulbar dermoids, **colobomas** of the upper eyelid) and **vertebral** (fusion or absence of cervical vertebrae) abnormalities, facial asymmetry, mild mental retardation
- Dx: clinical history and exam

Common Autosomal Dominant Causes of Congenital Hearing Loss

Waardenburg Syndrome

- Pathophysiology: may be from abnormal tyrosine metabolism
- Otologic SSx: unilateral or bilateral SNHL, may have vestibular dysfunction
- Other SSx: **pigmentary abnormalities** (heterochromic iriditis, white forelock, patchy skin depigmentation), craniofacial abnormalities (dystopia canthorum [widely spaced inner canthi], synophrys [confluent eyebrows], flat nasal root)
- Types
  I: presence of telecanthus, 20% have SNHL
  II: no telecanthus, 50% have SNHL
  III: associated with a unilateral ptosis and skeletal abnormalities
- Dx: clinical exam and history, family history

Stickler Syndrome (Progressive Arthro-Ophthalmopathy)

- Pathophysiology: uncertain, variable expression
- Otologic SSx: progressive SNHL (may be mixed)
- Other SSx: ocular abnormalities (myopia, retinal detachment, cataracts), Marfanoid habitus (tall and thin), arthritic abnormalities (joint hypermobilization, early arthritis), Pierre Robin sequence (micrognathia, glossoptosis, cleft palate)
- Dx: clinical exam and history, family history

Branchio-oto-renal Syndrome (Melnick Fraser Syndrome)

- Pathophysiology: abnormal development of branchial arches and kidneys
- Otologic SSx: pinna deformities; preauricular ear pits, fistulas, or tags; varied hearing loss; may have an associated Mondini deformity
- Other SSx: varied renal abnormalities (agenesis, mild dysplasia)
- Dx: renal involvement may be asymptomatic and only detectable with pyelography or renal ultrasound

Treacher Collins Syndrome (Mandibulofacial Dysostosis)

- Pathophysiology: uncertain origin
- typically normal intelligence (may have developmental delays secondary to hearing loss)
- Otologic SSx: auricular deformities, atresia or stenosis of EAC, preauricular fistulas, malformed ossicles (**CHL**), bony plate replacement of the TM, widened aqueduct, aberrant facial nerve
- Other SSx: mandibular abnormalities (mandibular hypoplasia, "fishmouth"), downward slanting palpebral fissures (antimongoloid slant), coloboma of lower eyelid, palate defects
- Rx: bone conduction aids, may consider surgical correction of aural atresia

Neurofibromatosis (Von Recklinghausen Disease)

- Pathophysiology: NF1 gene localized to chromosome 17; NF2 gene localized to a defect in the long arm of chromosome 22
- Otologic SSx: retrocochlear hearing loss (acoustic neuromas)
- Other SSx: café-au-lait spots (giant melasomes >1.5 cm); neurofibromas are most commonly cutaneous although may involve

the CNS (mental retardation), viscera, orbit (optic gliomas), or peripheral nerves; Lisch nodules (hamartomas of the eye); groin and axillary freckling; associated with pheochromocytomas
- Types
  NF-1: (**Classic Neurofibromatosis** or **von Recklinghausen Disease**) numerous café-au-lait spots, neurofibromas, approximately 5% risk of unilateral acoustic neuromas
  NF-2: (**Central Neurofibromatosis**) rare, associated with bilateral acoustic neuromas
- Dx: **NF-1** requires 2 of the following characteristics (>6 six café-au-lait spots, 2 or more neurofibromas or 1 plexiform neurofibroma, axillary or groin freckling, optic nerve glioma, distinct bony lesions, first-degree relative with NF-1); **NF-2** requires bilateral acoustic neuromas (neurofibromas exclusively involve the vestibulocochlear nerve), and first degree relative with an acoustic neuroma or 2 neurofibromas and other intracranial and spinal cord tumors, gliomas, schwannomas, or meningiomas

Apert Syndrome (Acrocephalosyndactyly)

- Pathophysiology: sporadic and autosomal dominant forms, affects middle and inner ear
- Otologic SSx: **stapes fixation** (primarily a CHL), patent cochlear aqueduct, large subarcuate fossa
- Other SSx: lobster claw hands (syndactyly), midface abnormalities (hypertelorism, proptosis, saddle nose, high arched palate), craniofacial dysostosis, trapezoid mouth (down-turned corners)

Crouzon Disease (Craniofacial Dysostosis)

- Pathophysiology: unknown origin, affects the skull, maxilla, eye, and external ear
- Otologic SSx: atresia and stenosis of the EAC (predominantly **CHL**), ossicular deformities
- Other SSx: cranial synostosis (premature cranial suture closure), small maxilla, exophthalmos, parrot nose, short upper lip, mandibular prognathism, hypertelorism

Common Sex-Linked Causes of Congenital Hearing Loss

Alport Disease

- Pathophysiology: X-linked (or autosomal dominant) trait resulting in abnormal Type IV collagen formation in the glomerular basement

membrane resulting in progressive renal disease, also associated with an SNHL
- Otologic SSx: slowly progressive SNHL (bilateral degeneration of organ of Corti and stria), presents at first decade of life
- Other SSx: **progressive nephritis** (hematuria, proteinuria, chronic glomerulonephritis, uremia), ocular disorders (myopia, cataracts)
- Dx: urinalysis, blood urea nitrogen, serum creatinine
- Rx: dialysis and renal transplant

Otopalatodigital Syndrome

- Pathophysiology: unknown etiology
- Otologic SSx: ossicular malformation (CHL)
- Other SSx: palatal defects, digital abnormalities (broad fingers and toes), hypertelorism, short stature, mental retardation

VESTIBULAR PATHOLOGY

Evaluation of the Dizzy Patient

History

- a specific diagnosis may be made in a majority of dizzy patients by an adequate history and physical examination
- **dizziness** is a term used to describe any of a variety of sensations that produce spatial disorientation
- Describe Dizziness: **vertigo** (illusion of rotational, linear, or tilting movement such as "spinning," "whirling," or "turning" of the patient or the surroundings), **disequilibrium** (sensation of instability of body positions, walking, or standing described as "off-balance" or "imbalanced"), **oscillopsia** (inability to focus on objects with motion, such as reading a sign while walking, seen with bilateral or central vestibular loss), **lightheadedness** (sense of impending faint, presyncope), **physiologic dizziness** (motion sickness, height vertigo), **multisensory dizziness** (diabetes, aging resulting in partial loss of multisensory systems)
- Onset and Duration of Symptoms: seconds to minutes (BPPV, VBI, epilepsy, arrhythmia), hours (Ménière's, migraine), days (vestibular neuritis, labyrinthitis), or constant (central causes); frequency and time of the day; initial and last spell
- Character of Dizziness: associated otologic factors (hearing loss, aural fullness, tinnitus), intensity and fatigability, precipitating factors (head movement or motion, stress, diet), systemic complaints (nausea, vomiting), central symptoms (numbness, weakness, diplopia, blurred vision), *see* Table 6–5
- Contributing Factors: medications (neuroleptics, antihypertensives, ototoxic medications, sedatives); medical history (hypertension, cardiac

TABLE 6–5. General Characteristics of Peripheral and Central Causes of Vertigo

Characteristic	Peripheral	Central
Intensity	severe	mild
Fatigability	fatigues, adaptation	does not fatigue
Associated Symptoms	nausea, hearing loss, sweating	weakness, numbness, falls more likely
Eye Closure	symptoms worse with eyes closed	symptoms better with eyes closed
Nystagmus	horizontal, may be unilateral, rotary	vertical, bilateral
Ocular Fixation	suppresses nystagmus (may not suppress during acute phase)	no effect or enhances nystagmus

arrhythmias and ischemia, diabetes, thyroid disorders, vascular disease, otological problems, depression, neurologic disease, migraines, or premenstrual syndrome); recent head trauma, loud noise exposure, flying, diving, or heavy lifting; new eyeglasses; family history of hearing loss, neurological disease, or otologic diseases

- Associated Symptoms: falls, confusion, weakness, weight loss, nervousness, headache
- *see* Table 6–6 for differential diagnosis of peripheral, central, and systemic vertigo

Physical Exam

- H & N and General Physical Exam: general medical exam including vital signs with particular evaluation for general neurological disorders, cardiovascular and peripheral vascular disorders (hypertension, carotid bruits)
- Otoscopy: pneumatoscopy (induce nystagmus or dizziness), inspection of TM and middle ear (otitis media, presence of fluid, masses)
- Vestibular Testing: eye motility (check pursuit with convergence and divergence) test using **Frenzel lenses** (prevents ocular fixation that may suppress nystagmus), spontaneous and gaze-evoked nystagmus, Dix-Hallpike maneuver (*see below*), head-shake (head motion in horizontal plane for 20–30 seconds, then suddenly stopped to evaluate for nystagmus), fistula test (*see* p. 356), head thrust, caloric testing may also be provided at bedside
- Neurological Exam: cranial palsies, vestibulospinal reflexes (eg, Romberg's test, gait, past pointing test, Fukuda) lateralizing signs (eg, weakness, paresthesias), tuning forks

TABLE 6–6. Common Causes of Peripheral, Central, and Systemic Vertigo

Peripheral Vertigo

- **Benign Paroxysmal Positional Vertigo**
- **Ménière's Disease**
- **Vestibular Neuronitis**
- **Perilymphatic Fistulas**
- **Cerebellopontine Angle Tumors**
- **Otitis Media**
- **Traumatic Vestibular Dysfunction**

Central and Systemic Vertigo

- **Multiple Sclerosis**
- **Other Neurological Disorders (stroke, seizures, middle cerebellar lesions, parkinsonism, pseudobulbar palsy)**
- **Metabolic Disorders (hypo/hyperthyroidism, diabetes)**
- **Medications and Intoxicants (psychotropic drugs, alcohol, analgesics, anesthetics, antihypertensives, anti-arrhythmics, chemotherapeutics)**
- **Vascular Causes (vertebrobasilar insufficiency, basilar migraine syndrome, vascular loop compression syndrome)**

Vestibular Testing

Electronystagmography (ENG) Battery

- **ENG**: battery of tests (oculomotor evaluation, positional/positioning testing, and caloric testing) that record eye movements typically by utilizing corneoretinal potentials (*see below*); provides information regarding localization of lesion (peripheral versus central, side of lesion); dependent on anatomy of the ear canal and temporal bone, only induces a low frequency response to caloric stimuli; difficult to perform in children
- **Corneoretinal Potential**: an electrical voltage between the cornea and the retina exists with ocular axis changes (does not change with torsional ocular motion), ENG records changes in the corneoretinal potential by placing electrodes around the eyes to record eye movement

Oculomotor Testing

- **Fixation Tests**: **spontaneous nystagmus** may be induced with eyes centered and head upright and having subject fix on an object with eyes open and eyes shut (or lights off), **gaze-evoked nystagmus** may be induced by having the subject fix at an object

20 and 30 degrees to the left and right of center for 30 seconds each; **rebound nystagmus** may occur after prolonged gaze holding after the eye is returned to primary position

- **Saccadic Systems**: saccades allow shifting of gaze rapidly from one object to another maintaining the image of the target upon the fovea; saccadic tests evaluate CNS components of the vestibular system; cerebellar or brainstem injury may cause ocular **dysmetria** (overshooting or undershooting of eye rotation)
- **Ocular Pursuit Systems**: allows ocular fixation on moving objects and maintains image to be kept on the fovea; tested with **Sinusoidal-Tracking Tests** by having the patient follow a spot moving in a sinusoidal pattern, at faster speeds the eyes may not be able to "keep up" causing saccadic eye jerks; saccadic eye jerks at low velocity (rotational frequencies <0.1–0.3 Hz or target velocities <30°/s) suggests a central nervous system pathology
- **Optokinetic Systems**: allows fixation on a moving field (maintains image on the whole retina rather than specifically on the fovea as in saccades and pursuit systems); tested clinically by having the patient stay still and moving the environment (a series of black and white stripes on a moving field that encompasses the whole field of vision); brainstem disease may cause bilateral reduced gain, cerebellar lesions may induce ataxia, peripheral lesions may demonstrate asymmetry

Positional/Positioning Nystagmus Testing (Dix-Hallpike maneuver)

- **Positional Testing**: tests for nystagmus evoked by a new **static** head position (positional nystagmus is maintained as long as head remains in the evoked position)
- **Positioning Testing**: tests for nystagmus evoked by the **action** of motion of the head
- **Dix-Hallpike Maneuver**: positioning vestibular test designed to stimulate the posterior SSC (test for BPPV, *see* p. 355); induction of nystagmus is a hallmark for benign paroxysmal positional vertigo
- Dix-Hallpike Maneuver Technique: patient sits upright with head at 45° angle horizontally; patient then moves to a head-hanging position at approximately 30° with 1 ear down; eyes are observed for nystagmus (rotary to the downward side, delayed 2–15 seconds, transient [10–60 seconds] associated with vertigo) for at least 20 seconds; patient is returned to the upright position; eyes are observed again for nystagmus for at least 20 seconds

Caloric Testing

- only test that evaluates vestibular function in each ear independently, determines unilateral versus bilateral vestibular weakness

- Technique: in supine position, head is inclined to 30° to bring the horizontal canal into a vertical position, each ear is irrigated with cool and warm water (or air), nystagmus is recorded with eyes open and closed
- Theoretical Normal Response: **cool** water or air (30° C) to **right** ear with patient in upright position causes flow in the ampufugal direction which decreases the electrical activity of the ipsilateral vestibular nerve with a corresponding increase in electrical activity of the opposite vestibular nerve resulting in **left**-beating **nystagmus** (warm water or air [40° C] produces the opposite response); the pneumonic **COWS**, "**C**ool **O**pposite, **W**arm **S**ame", represents the direction of nystagmus with warm and cool water
- **Maximum Slow Phase Velocity**: determined by dividing the duration by the amplitude of the slow phase; standard measure of caloric response intensity
- **Directional Preponderance**: denotes that the nystagmus response in a particular direction is weaker than the evoked response in the opposite direction; determined by comparing the duration or velocity of right-beating nystagmus from both ears with left-beating nystagmus from both ears; **>20–25%** difference between sides may suggest a unilateral weakness
- **Unilateral Caloric Weakness**: denotes that the response of one side to a stimulus is reduced in comparison to the other side; determined by comparing the duration or velocity response from left and right ears; approximately **>20–25%** difference between sides suggest a unilateral peripheral weakness
- **Bilateral Weakness**: suggested when the total caloric maximum slow phase velocity from each ear (all 4 irrigations) is **<12–24°/second**
- **Postcaloric Fixation**: determined by dividing the maximum slow phase velocity with fixation and without fixation; an inability to suppress nystagmus suggests a central lesion

Rotation Tests (Rotational Chair)

- **Sinusoidal (Slow) Harmonic Acceleration Test:** patient is seated in the chair, eye movements are recorded while patient (chair) is rotated along the horizontal plane; evaluates the vestibulo-ocular reflex; measures gain (ratio of slow phase eye velocity to head velocity), **phase angle** (compares peak responses of the slow component of the eye with the peak velocity of the head, determines the timing relationship between stimulus and response), and **symmetry**
- Indications: procedure of choice to evaluate bilateral vestibular dysfunction, may detect mild vestibular dysfunction undetected by traditional ENG testing, may follow progress of vestibular compensation

- abnormalities are primarily seen at low frequencies (abnormal phase and gain reduction) and high frequencies (asymmetry)
- normal vestibulo-ocular response results in similar slow eye phase velocity and chair velocity
- acute unilateral peripheral lesions typically reveal low frequency phase leads and high frequency asymmetry (the absence of asymmetry suggests vestibular compensation, eg, acoustic neuroma)
- bilateral vestibular disease typically demonstrates reduced gain at low frequencies but normal gain at high frequencies

Posturography

- **Posturography**: measures postural stability under varying combinations of changing the visual field references and support structures
- evaluates proprioceptive, visual, and vestibular systems
- controversial applications in diagnosis, may be used to confirm malingering and for vestibular rehabilitation

Management Concepts

- Safety: avoid heights, ladders, driving, operating heavy machinery
- Acute Vestibular Suppression: indicated for intolerable symptoms but may delay central compensatory mechanisms in the long term; common medical therapies include phenothiazine, meclizine, diazepam, transdermal scopolamine, corticosteroids, and antiemetics
- Vestibular Rehabilitation: indicated for chronic complaints, consists of a series of positional tasks, head movements, and oculomotor exercises to facilitate central compensation
- Surgical Management: may be indicated for specific diagnoses

Vestibular Disorders

Ménière's Disease (Endolymphatic Hydrops)

Signs and Symptoms

- episodic vertigo lasting minutes to hours
- episodic fluctuating SNHL (usually unilateral), recovery between episodes may be incomplete resulting in a progressive SNHL (initially at lower frequencies)
- tinnitus and episodic fullness associated with or without the hearing loss
- classic Ménière's Disease presents with all of the above symptoms (vertigo, hearing loss, tinnitus, and aural fullness), however, Ménière's Disease may also present as any combination of the above symptoms (*see Variants below*)

Diagnosis and Other Causes of Endolymphatic Hydrops

- based on clinical history, physical examination, and audiological findings (initial low-frequency SNHL) with exclusion of other causes of hearing loss and vertigo is adequate for diagnosis and initiating empirical therapy
- vestibular testing may reveal unilateral weakness on affected side
- **Electrocochleography**: may exhibit marked negative summation potential from basilar membrane distortion (hydrops) and a larger SP amplitude/AP amplitude ratio (SP/AP ratio >0.5, normal =0.2)
- **Glycerol (dehydration) test**: oral glycerol ingestion or mannitol essentially acts as an osmotic diuretic that in the presence of active hydrops may improve symptoms temporarily within 30–60 minutes
- Other Causes of Endolymphatic Hydrops: allergy, mumps, syphilis, hypothyroidism, Mondini's aplasia, trauma, viral or bacterial infection

Pathophysiology Theories and Histological Findings (Controversial)

- classically patients with Ménière's Disease present histologically with raising of Reissner's membrane and dilation of the endolymphatic spaces (endolymphatic hydrops)
- altered glycoprotein metabolism may result in dysregulation of inner ear fluid causing abnormal osmotic pressure and quantitative volume differences between perilymph and endolymph resulting in endolymphatic hydrops
- fibrosis of endolymphatic duct and sac impairs endolymph absorption and may result in overdistention of membranous labyrinth (endolymphatic hydrops)
- membranous labyrinth distention may cause microtears (rupture) which mixes endolymph and perilymph resulting in some permanent damage to hair cells and instant vertigo, the tear then spontaneously seals, after 2–3 hours the inner ear fluid re-equilibrates with resolution of the vertigo, repeated ruptures may cause progression of the SNHL
- new theory suggests abnormal regulation of an endolymphatic sac hormone, "saccin," may cause excessive production of fluid in sac with retrograde filling

Variants

- **Cochlear Hydrops**: isolated cochlear variant characterized by hearing loss, aural fullness, tinnitus, without vertigo
- **Vestibular Hydrops**: isolated vestibular variant characterized by episodic vertigo without hearing loss or tinnitus
- **Lermoyez Syndrome**: rare, initially presents with increasing tinnitus, hearing loss, and aural fullness with sudden relief after a spell of vertigo

- **Crisis of Tumarkin (Drop Attack)**: sudden loss of extensor function causing a "drop attack" without loss of consciousness and with complete recovery
- **Delayed Endolymphatic Hydrops**: loss of hearing later followed by typical Ménière's symptoms

Medical Management

- **Dietary Restrictions**: first-line therapy; avoid fluid shifts by restricting salt, alcohol, monosodium glutamate, and caffeine; evenly space out meals (avoid binge eating or skipping meals); encourage evenly spaced water consumption
- **Diuretics**: first-line therapy; encourages constant renal output (must avoid dehydration which would exacerbate symptoms)
- **Vestibular Suppressants**: may be considered for symptomatic treatment
- **Corticosteroids**: may be considered for acute exacerbations
- **Allery Management**: *see* pp. 28–33
- **Stress Reduction**: Ménière's Disease symptoms are exacerbated by stress

Surgical Management of Vertigo

- **Endolymphatic Sac Surgery**: first choice for failed conservative management of episodic vertigo (*controversial*); may consider wide bony decompression of endolymphatic sac or endolymphatic shunt to the subarachnoid space or mastoid cavity; preserves auditory and vestibular function, low morbidity, less successful than obliterative procedures, rate of success varies from 60–80% (however, 70% usually recover spontaneously)
- **Vestibular Nerve Section**: allows individual section of vestibular nerve fibers before becoming associated with cochlear fibers (preserves hearing); higher risk of hearing loss and postoperative dizziness than endolymphatic sac surgery, requires a craniotomy
- **Transtympanic or Intratympanic Aminoglycoside Injections**: typically use gentamicin or streptomycin; ideal drug type, dosage, and means of delivery have not been standardized; risk of SNHL
- **Labyrinthectomy**: ablative procedure (contraindicated if other ear has reduced vestibular function) that may be considered for nonservicable hearing (>50–60 dB HL or <50% speech discrimination) with failed conservative management; high rate of success (up to 90%); must take into account risk of development of Ménière's disease in the opposite nonsurgical ear

Benign Paroxysmal Positional Vertigo (BPPV, Cupulolithiasis)

- most common cause of peripheral vertigo
- Causes: spontaneous, post-traumatic, and postviral (labyrinthitis, vestibular neuronitis)
- typically self-limiting, may have recurrent episodes
- SSx: recurrent episodes of brief (lasting seconds to minutes) positional vertigo (turning over in bed, getting up, turning the head, bending over, looking up); may be associated with nausea and prolonged light-headedness; induced positional nystagmus is torsional (rotary to the downward side), typically exhibits a latency of 2–15 seconds with a crescendo and decrescendo of nystagmus associated with vertigo, usually fatigable, and transient (seconds)
- Dx: clinical history and exam (Dix-Hallpike Maneuver)

Theories of Pathophysiology

- **Canalithiasis Theory**: free-floating debris (dislodged otoconia) in the endolymph of the posterior canal moves when placed in a dependent position, the inertial drag of the endolymph causes displacement of the cupula resulting in latent vertigo which resolves when the debris settles
- **Cupulolithiasis Theory**: debris (calcium carbonate) adheres to the cupula of the semicircular canal resulting in an ampulla that is gravity sensitive (objections to theory include no account for the transient nature of vertigo and the torsional nystagmus exhibited in BPPV)

Management

- education, reassurance, and observation
- **Particle Repositioning Maneuver (Epley's Maneuver)**: series of head positionings completed in the office, based on "repositioning" free-floating particle in the posterior canal, requires patient to be upright after repositioning for 48 hours
- home vestibular positional exercises induce vertigo to stimulate vestibular compensation, probably works by replacement of SSC crystals back into the vestibule
- antivertiginous medications typically are not useful due to the sporadic and brief nature of the vertigo
- **Singular Neurectomy**: transection of the nerve to the posterior semicircular canal may be considered for rare intractable BPPV that is refractory to conservative measures (rarely indicated)
- **Posterior Semicircular Canal Occlusion**: occludes ampullated end to prevent movement of endolymph, may also be considered for rare intractable BPPV that is refractory to conservative measures (rarely indicated)

Labyrinthine Concussion

- Pathophysiology: posttraumatic disorders of inner ear function without fracture of the labyrinth
- SSx: self-limiting acute vertigo, recoverable SNHL and tinnitus, normal otoscopic and radiologic findings, may result in residual BPPV-like symptoms
- Dx: clinical history and physical exam, audiology
- Rx: reassurance and observation, may consider symptomatic medications (mild sedatives)

Perilymph Fistula

- Pathophysiology: abnormal fistula opening between inner and middle ear (or mastoid cavity) results in progressive hearing loss and vertigo
- Causes: barotrauma (Valsalva maneuver, straining, sneezing, head trauma, birth trauma), iatrogenic (stapedectomy), congenital (associated with Mondini dysplasia and abnormalities of the stapes or round window), spontaneous
- SSx: sudden SNHL with vertigo subsequent to a traumatic event, **Fistula test** (nystagmus that occurs from pressure pneumatoscopy that causes stimulation of the membranous labyrinth, 10–40% sensitivity)
- Dx: **no definitive preoperative test**, exploration often not definitive, may visualize fluid at or near the oval or round window
- Rx: bed rest with head elevation and avoidance of any straining (sneezing with mouth open, stool softeners) for 5–10 days; for persistent symptoms or progression of hearing loss consider exploration surgery with grafting site of leak
- NOTE: perilymphatic fistula diagnosis and management is quite controversial, some neurologists may even question its existence

Wallenberg Syndrome (Lateral Medullary Syndrome)

- Pathophysiology: embolic event or thrombosis of the ipsilateral vertebral or posterior inferior cerebellar artery (PICA) resulting in an infarction of the lateral medullary region of the brain stem, **spares** cochlear nucleus
- SSx
  1. acute **vertigo** (spontaneous nystagmus, nausea and vomiting)
  2. **ataxia** from incoordination of the ipsilateral limbs (falls toward lesion)
  3. ipsilateral **Horner Syndrome** (anhydrosis, ptosis, miosis) from damage to the sympathetic fibers

4. ipsilateral palatal paresis (**dysphagia**) and vocal fold paralysis (**dysphonia**) from destruction of the nucleus ambiguus
5. ipsilateral numbness of the face from involvement of the spinal tract of the trigeminal nerve
6. **contralateral** loss of pain and temperature sensation from injury to the crossed spinothalamic fibers

- Dx: cerebral angiogram, CT (wedge-shaped infarct)
- Rx: per cerebrovascular accident protocols

Vertebrobasilar Insufficiency (VBI)

- Pathophysiology: compression of the vertebral artery compromises flow to the posterior and anterior inferior cerebellar arteries
- SSx: transient vertigo with neck hyperextension or excessive rotation, "**4 D's**" (**dizziness, diplopia, dysphagia, drop attacks**); also associated with dysarthria, headaches, hallucinations, ataxia, and hemiparesis (normal neurological exam between attacks)
- Dx: clinical history and physical exam, radiography (evaluate for cervical spine disease)
- Rx: anticoagulation (antiplatelet medication)

Cogan Syndrome

- Pathophysiology: unknown, autoimmune etiology, produces hydropics similar to Ménière's Disease
- SSx: **interstitial keratitis** (nonreactive VDRL, blurriness, rapidly progresses to blindness), **episodic vertigo**, **bilateral fluctuating SNHL** (associated with tinnitus), disease progresses over months
- Dx: clinical history and physical exam, elevated CSR
- Rx: high dose oral corticosteroids (usually resolves hearing and vestibular dysfunction), may consider cyclophosphamide, azanthropine, disease progresses over months
- **Vogt-Koyanagi-Harada Syndrome**: similar to Cogan syndrome (hearing loss, vertigo), also associated with granulomatous uveitis, depigmentation of hair and skin, aseptic meningitis, and loss of eyelashes

Other Vestibular Disorders

- **Basilar Migraine Syndrome**: migraine headache accompanied by symptoms of brainstem lesions including vertigo; vertigo may also present as an aura; Rx: migraine therapy
- **Vestibular Epilepsy**: dysequilibrium that may present as an aura or as a petit mal seizure, symptoms vary in severity and may present episodically similar to Ménière's disease

- **Multiple Sclerosis** (**MS**): demyelinating disorder of the CNS may result in "plaques" within the central vestibular system causing vertigo; may present initially with vertigo (10–15%)
- **Labyrinthine Apoplexy**: thrombosis of the internal auditory artery resulting in acute vertigo, hearing loss, and tinnitus
- **Subclavian Steal Syndrome**: occlusion of the subclavian artery proximal to the vertebral artery results in reverse flow of the vertebral artery in favor of the ipsilateral arm resulting in intermittent vertigo, occipital headaches, blurred vision, upper extremity pain, supraclavicular fossa bruit, blood pressure differential between arms, and a weak radial pulse
- **Hyperinsulinemia/Diabetes**: may contribute to acute vertiginous attacks or Ménière's-like complaints

THE FACIAL NERVE
Anatomy and Physiology

Anatomy

Intracranial Segment

- segment from brainstem to internal auditory canal (IAC)
- **Nervus Intermedius** (**Nerve of Wrisberg**): parasympathetic and sensory root of the facial nerve
- motor root joins with the nervus intermedius in the CPA/IAC to form the common facial nerve

Intratemporal Segments

1. Meatal

- segment from the porus acusticus to the meatal foramen of the IAC to the fundus
- the facial nerve transverses in the **anterior superior** quadrant of the IAC separated by the **falciform crest** inferiorly and **Bill's bar** posteriorly (other quadrants include the superior vestibular nerve [superior posterior], inferior vestibular nerve [inferior posterior], and the cochlear nerve [inferior anterior])
- the meatal segment is ensheathed within an extension of the meninges

2. Labyrinthine

- segment from fundus to geniculate ganglion
- narrowest segment of the fallopian canal (**0.68 mm** diameter)
- **Geniculate Ganglion**: located at the **first genu**, houses cell bodies of sensory cells and taste cells from the anterior ⅔ of the tongue and palate

- **Greater (Superficial) Petrosal Nerve**: first branch (branches off of geniculate ganglion), carries preganglionic parasympathetic fibers to the lacrimal gland

3. Tympanic (Horizontal Segment)

- segment from geniculate ganglion to the second genu (inferior to the horizontal canal)
- courses above the oval window and stapes
- **most common site of dehiscence** (**40–50%**)

4. Mastoid (Vertical Segment)

- segment from the second genu to the stylomastoid foramen
- branches to the **Stapedial Muscle** and the **Chorda Tympani** (preganglionic parasympathetic to the submandibular and sublingual glands and special sensory taste fibers)

Extratemporal Segments

- **Postauricular Nerve**: branch to the external auricular and occipitofrontalis muscles
- **Nerve to the Stylohyoid**
- **Nerve to the Posterior Digastric**

Pes Anserinus: branching point of the extratemporal segments in the parotid gland, most commonly divides into two divisions, the **temporozygomatic** and **cervicofacial** branches (many variants)

1. **Temporal**: innervates the frontalis, corrugator supercillii, procerus, and upper orbicularis oculi muscles
2. **Zygomatic**: innervates the lower orbicularis oculi muscles, abundant anastomotic supply with buccal branch
3. **Buccal**: innervates the zygomaticus major and minor, levator anguli oris, buccinator, and upper orbicularis oris muscles (smile); abundant anastomotic supply with the zygomatic branch
4. **Mandibular**: innervates the lower orbicularis oris, depressor anguli oris, depressor labii inferioris, and mentalis muscles
5. **Cervical**: innervates the platysma muscle

Nerve Fiber Components

- **Endoneurium**: surrounds each nerve fiber (axons), tightly adherent to the Schwann cell layer, provides the endoneural tube for regeneration, poorer prognosis for regeneration when disrupted

- **Perineurium**: surrounds endoneural tubules, provides tensile strength, maintains intrafunicular pressure, and protects from infection
- **Epineurium** (**Nerve Sheath**): outer layer, contains the vasa nervorum for nutrition

Facial Nerve Components

Branchial Motor *(Special Visceral Efferent)*

- **premotor cortex—motor cortex**—*corticobulbar tract* → **bilateral facial nuclei** (pons) → muscles of facial expression
- fibers that innervate the forehead receive bilateral innervation from the upper motor neurons and fibers that innervate the lower face receive contralateral fibers only from the upper motor neurons
- also supplies stapedius (**stapedial reflex**), stylohyoid, posterior digastric, and buccinator muscles

Parasympathetic *(General Visceral Efferent)*

1. superior salivatory nucleus (pons) — nervus intermedius — **greater (superficial) petrosal nerve** — through facial hiatus — middle cranial fossa—joins the *deep petrosal nerve* (sympathetic fibers from cervical plexus) — through pterygoid canal (now called *nerve of the pterygoid canal* or **vidian nerve**) — pterygopalatine fossa → **spheno/pterygo-palatine ganglion** — *postganglionic parasympathetic fibers* — joins the *zygomaticotemporal nerve* (V_2) → **lacrimal gland** (also innervates the seromucinous glands of nasal and oral cavity)
2. **superior salivatory nucleus** (pons) — *nervus intermedius* — **chorda tympani** — carried on *lingual nerve* → **submandibular ganglion** — *postganglionic parasympathetic fibers* → **submandibular** and **sublingual glands**

Sensory *(General Sensory Afferent)*

- supplies sensation to the auricular concha, postauricular skin, wall of the EAC, and part of the tympanic membrane
- cell bodies housed in the **geniculate ganglion** (also holds the cell bodies of the taste fibers)

Taste *(Special Visceral Afferent)*

- bilateral **postcentral gyrus** → **nucleus solitarius** (gustatory nucleus) — *tractus solitarius* — *nervus intermedius* → **geniculate ganglia** (taste fibers and sensory cell bodies) — **chorda tympani** → joins *lingual nerve* → anterior ⅔ of the tongue and hard and soft palate

- NOTE: **iter chordae posterior** (canal in which the chorda tympani enters into the middle ear space after branching from the facial nerve); **canal of Huguier** or **iter chordae anterior** (canal in which the chorda tympani exits the middle ear)

Sunderland Nerve Injury Classification

- Class I (**first degree**) **injury** (**Neuropraxia**): loss of axoplasmic flow from compression of the axon results in only a conduction block, complete recovery anticipated
- Class II (**second degree**) **injury** (**Axonotmesis**): axon disrupted, endoneurium preserved, wallerian degeneration occurs distal to site of injury, complete recovery anticipated (axon regenerates through an intact neural tube at 1 mm/day)
- Class III (**third degree**) **injury** (**Neurotmesis**): neural tube (axons, myelin sheath, and endoneurium) disruption, wallerian degeneration occurs distal to site of injury, unpredictable outcome (loss of endoneural tubules results in high risk for **synkinesis**, the contraction of multiple muscle fibers simultaneously, if regrowth occurs)
- Class IV (**fourth degree**) **injury**: violates perineurium
- Class V (**fifth degree**) **injury**: complete transection, risk of a neuroma from nerve sprouts outside of nerve sheath

Evaluation of Facial Nerve Paralysis

History

- Character of Facial Paralysis: onset, duration, and progression of paralysis
- Contributing Factors: recent infection or illness, trauma (birth trauma in neonates), surgery (otologic, parotid, or neurologic surgery); recent tick bites or outdoor activity; history of syphilis, HIV, tuberculosis, or herpes infections; toxin exposure (lead); history of otologic, neurologic, diabetic, or vascular disorders; previous history of facial nerve paralysis
- Associated Symptoms: fever, facial pain, hearing loss, aural fullness, otalgia, vertigo, other neurologic deficits, change in taste sensation, vision changes, drooling, cheek biting, epiphora, dysacusis, pain (auricular, postauricular, or facial)
- "Think KITTENS" for differential diagnosis (*see* Table 6–7)

Physical Exam

- Facial Nerve: observe facial symmetry at rest and with movement, paresis vs paralysis, hemifacial spasms, and facial tics at rest; determine unilateral versus bilateral weakness, eye closure, quality of **Bell's**

TABLE 6-7. Differential Diagnosis of Facial Nerve Paralysis: KITTENS Method

(**K**) Congenital	**I**nfectious and **I**diopathic	**T**oxins and **T**rauma	**T**umor (Neoplasms)	**E**ndocrine	**N**eurologic	**S**ystemic/ PSychological
Möbius syndrome Myotonic dystrophy	Idiopathic facial paralysis Melkersson-Rosenthal syndrome Ramsay-Hunt syndrome Otitis media/mastoiditis Necrotizing otitis externa Meningitis Lyme disease Tetanus TB, HIV, EBV, syphilis	Head trauma Temporal bone trauma Iatrogenic injuries Birth trauma	Parotid tumors Facial neuromas Acoustic neuromas Cholesteatoma Gliomas Meningioma Temporal bone tumors	Diabetes mellitus Pregnancy Hyper-thyroidism	Guillain-Barré Multiple sclerosis Myasthenia gravis Stroke	Sarcoidosis Amyloidosis Hyperostoses (Paget's disease, osteopetrosis)

phenomenon (globe turns up and out during attempts to close eyes), tear production, corneal reflex, and visual acuity
- House-Brackmann Grading: (*see* Table 6–8) used to evaluate recovery of paralysis (eg, recovery from Bell's palsy, preoperative versus postoperative results)
- Other Head and Neck Assessment: evaluate for mass or fluid in the middle ear, presence of vesicles in the EAC and concha, **Hitselberger sign** (hypesthesia of the sensory division of the facial nerve at the superior posterior concha), other cranial nerve involvement, other lateralizing signs (hemiparesthesias, hemiparalysis, aphasia), parotid masses

Ancillary Studies

Electrodiagnostic Tests

Electromyography (EMG)

- electrodes inserted into muscle, measures muscle response to **voluntary** contraction

TABLE 6–8. House-Brackmann System of Grading Facial Nerve Paralysis*

Grading	Function
I	**normal function**
II	**mild dysfunction**: weakness on close inspection, normal symmetry and tone at rest, moderate to good facial function (slight mouth asymmetry, complete eye closure with minimal effort)
III	**moderate dysfunction**: obvious weakness, and/or asymmetry (not disfiguring), contracture, and/or hemifacial spasms, normal symmetry and tone at rest; moderate facial function (weak mouth and forehead function, complete eye closure with effort)
IV	**moderately severe dysfunction**: disfiguring asymmetry and/or obvious weakness, normal asymmetry and tone at rest, incomplete eye closure, no forehead motion, asymmetric mouth motion with maximal effort
V	**severe dysfunction**: barely perceptible motion, asymmetry at rest, incomplete eye closure, no forehead motion, slight mouth motion
VI	**total paralysis**

*From House JW and Brackmann DE. Facial nerve grading system. *Arch Otolaryngol—Head & Neck Surg.* 1985; 93:146–147.

- useful to demonstrate the existence of functional motor units
- fibrillations from deinnervated nerve appear after 1–2 weeks
- polysynaptic signals indicate reinnervation
- presence of voluntary action potentials indicates at least partial continuity of nerve

Electroneuronography (ENoG)

- essentially an evoked EMG (muscle response is recorded by electromyogram)
- records muscle response via electrodes after stimulation of the facial nerve with a transcutaneous impulse at the stylomandibular foramen
- objectively compares muscle compound action potential amplitudes (related to intact motor axons) and latencies from the paralyzed and normal sides
- in an acute setting ENoG is typically not useful until after 24–72 hours when wallerian degeneration occurs
- can distinguish neuropraxia versus more severe injuries
- valuable in determining prognosis for idiopathic paralysis

Other Ancillary Studies *(may be considered in select cases)*

- **Audiogram**: indicated for all intratemporal injuries and for preoperative baseline hearing
- **Topognostic Tests**: (Schirmer's test, stapedial reflex, salivary flow test) in general are not useful and have been replaced with electrophysiological testing
- **CT of Temporal Bones**: may detect temporal bone lesions and fractures
- **MRI of Temporal Bones**: most sensitive imaging for examining the intratemporal segment of the facial nerve, gadolinium enhancement may reveal acoustic or facial neuromas and other tumors
- **Treponemal Studies**: Lyme titers and VDRL/FTA-ABS
- **Complete Blood Count**: may suggest inflammatory process
- **ACE level**: may suggest active sarcoidosis

Congenital Facial Palsy

Birth Trauma

- most common cause of unilateral facial paralysis in the neonate
- Pathophysiology: facial nerve is at risk for injury during delivery as it courses the underdeveloped mastoid process

- Risks: forceps delivery, prolonged delivery, large infant
- SSx: asymmetric crying facies, hemotympanum, periauricular ecchymosis
- Dx: EMG (preserved neuromuscular activity suggests inherited or developmental etiology), electrophysiological testing, ABR (evaluate for associated hearing loss)
- Rx: observation (usually recovers)

Congenital or Developmental Causes of Facial Paralysis

- **Möbius Syndrome**: wide spectrum of abnormalities secondary to central brain stem and peripheral neuromuscular defects resulting in bilateral or unilateral **facial** and **abducens** nerve palsies; other signs and symptoms include club foot (talipes equinovarus), tongue weakness, mixed hearing loss, mental retardation, external ear deformities, and ophthalmoplegia
- **Albers-Schoenberg Disease**: autosomal recessive inheritance, disorder of bone metabolism resulting in osteopetrosis of the bony IAC causing compression of nerves (CN III, VII, and VIII)
- **Congenital Unilateral Lower Lip Palsy** (**Asymmetric Crying Facies**): more common congenital causes of facial paralysis, presents with hypoplasia of the depressor anguli oris muscle, associated with cardiac defects (≈10%)

Infectious and Idiopathic Causes of Facial Palsy

Idiopathic Facial Paralysis (Bell's Palsy)

- **most common cause of facial paralysis**
- Pathophysiology: facial paralysis or paresis may be due to impaired axoplasmic flow from edema of the facial nerve within the fallopian canal secondary to a herpes simplex virus type 1 (HSV-1) infection (may be reactivated from dormancy in geniculate ganglion), also possible etiologies include immunological or vascular (ischemia) pathology
- Risks: diabetes mellitus, pregnancy, past history (7–12% recur)
- SSx: acute, **unilateral** paresis (≈⅓) or paralysis (≈⅔) of the face, rapid onset (<48 hours); may have a viral-like prodrome 3–4 days prior to paralysis, postauricular pain, dysgeusia
- approximately 70–85% will have full recovery by 6 months; approximately 15–30% will have incomplete recovery

- favorable prognosis is associated with the presence of any facial movement
- poorer prognosis is associated with complete facial paralysis (ENoG reveals >90% weakness) within 2 weeks of onset
- Dx: diagnosis is based on exclusion of other causes of facial nerve paralysis as well as clinical history and exam

Initial Management

- Medical Management: oral antivirals (acyclovir) and corticosteroids with taper for 10 days for acute phases
- Eye Protection: artificial tears, ocular ointment at night, eye patch at night
- follow progression with serial exams (may consider ENoG testing)

Surgical Decompression

- Indications: (*controversial*) Gantz et al suggest consideration of surgical decompression if ENoG reveals >90% weakness within two weeks after onset and no voluntary movement on EMG (*Laryngoscope* 109:1177–1188, 1999)
- most effective if performed within 2 weeks of onset
- typically requires a transmastoid and middle cranial fossa approach with decompression of the tympanic segment, geniculate ganglion, labyrinthine segment, and meatal foramen

Herpes Zoster Oticus and Ramsay Hunt Syndrome

- **Ramsay Hunt Syndrome**: essentially herpes zoster with a facial nerve palsy, may also affect other cranial nerves
- Pathophysiology: primary infection or reactivation of herpes simplex virus (HSV), remains dormant in the cranial nerves and the cervical plexus
- 30–50% risk of residual facial weakness after an acute episode (higher risk than Bell's palsy)
- SSx: acute peripheral facial palsy, painful vesicular lesions in the concha or EAC (often misdiagnosed as external otitis media), alterations in taste, may have hearing loss and vertigo, may involve other cranial nerves
- Dx: clinical history and physical exam, complement fixation and serum titers confirm diagnosis
- Rx: antivirals (acyclovir) and corticosteroids for 10 days, analgesics, and eye protection, rare to consider surgical facial nerve decompression (*see above*)

Melkersson-Rosenthal Syndrome

- Pathophysiology: unknown etiology
- SSx: chronic or recurrent edema of the face (defining feature), recurrent unilateral or bilateral facial motor dysfunction, fissured tongue, cheilitis granulomatosa
- facial swelling and facial paralysis begin in childhood and early adolescence
- Dx: clinical history and exam, lip biopsy reveals dilated lymphatics and granulomatous changes with giant cells, may have elevated ACE levels during attacks
- Rx: empiric management with corticosteroids, may consider surgical decompression (*controversial*)

Lyme Disease

- Pathophysiology: *Borrelia burgdorferi* (spirochete) transmitted by a tick
- SSx: 10% of patients with Lyme disease have ipsilateral or bilateral facial nerve involvement after 1–4 weeks incubation period (higher risk with tick bite of the head), initial erythema migrans (bull's-eye rash), flu-like symptoms, meningitis, multiple neuropathies, cardiac conduction disorders, meningoencephalitis, swollen joints
- facial paralysis resolves in 6–12 months (full recovery anticipated)
- Dx: identification of a tick bite, antibodies in serum or CSF (serum antibodies are less sensitive and specific than CSF)
- Rx: parenteral penicillin, ceftriaxone, or cefotaxime for severe cases; for rash only consider oral therapy (penicillin, erythromycin, or tetracycline), rare to consider surgical facial nerve decompression (*see* p. 366)

Otitis Media

- Pathophysiology: in acute otitis media, toxic effects from infectious spread into the nerve sheath results in facial nerve dysfunction; in chronic otitis media, facial nerve paralysis may occur from compressive effects from a cholesteatoma or from granulation tissue
- SSx: progressive unilateral facial palsy with suppurative otitis media
- Dx: clinical history and physical exam (otoscopic exam), CT/MRI may reveal cholesteatoma or soft tissue compression
- Rx: *see* p. 322

Malignant Otitis Externa

- Pathophysiology: facial nerve injury from the effect of temporal bone osteomyelitis

- SSx: progressive unilateral facial palsy with a malignant otitis externa
- Dx and Rx: *see* pp. 312–313

Facial Nerve Trauma

Penetrating and Blunt Trauma

- Causes: penetrating injury to the extratemporal facial branches or blunt trauma resulting in a temporal bone fracture
- Dx: assess facial nerve integrity (*as above*) and possible temporal bone fractures (CT)
- Rx: *see* pp. 335–338 *for evaluation and management of temporal bone fractures*; penetrating injury resulting in total facial nerve paralysis should be explored and repaired with an end-to-end anastomosis (*see below*) within 48–72 hours while the distal branches may be stimulated; however, injuries medial to a line perpendicular to the lateral canthus usually recover spontaneously and do not require exploration; incomplete facial palsy may be closely monitored with serial electrodiagnostic testing at 3–7 days and 4–8 weeks; if wound is contaminated or there is significant tissue loss may consider identifying distal and proximal ends of the facial nerve with plans for a second stage procedure within 3–4 weeks

Iatrogenic Injuries

- tympanic and vertical segments are the most commonly injured segment in otologic surgery (most common site of facial nerve dehiscence)
- facial nerve injury may also occur with salivary gland surgery, neck dissections, rhytidoplasties, and branchial cleft excisions

Management

- intraoperative injury may be addressed with primary anastomosis or cable grafting
- for immediate postoperative facial palsies wait for local anesthetic to wear off (2–3 hours) then reevaluate
- for postoperative paralysis (with intraoperative confirmation of facial nerve preservation) may consider corticosteroids and follow progression with serial electrodiagnostic testing
- facial weakness or delayed onset of facial paralysis usually results in complete recovery
- may consider re-exploration for complete paralysis in select cases

Other Causes of Facial Nerve Paralysis

Neoplasms

- SSx: tumors may present as an acute ipsilateral facial paralysis or progression of facial paralysis
- **Parotid Tumors**: most common malignancy to cause facial nerve paralysis, mucoepidermoid tumors are the most common parotid tumor to cause facial dysfunction although adenoid cystic carcinoma has a higher rate of neural involvement
- **Schwannomas** (**Facial Neuromas, Acoustic Neuromas**): (uncommon) large tumors may cause facial paralysis
- Other Neoplasms: cholesteatomas, hemangiomas, glomus jugulares or tympanicum, meningiomas, metastatic carcinomas, leukemia, rhabdomyosarcomas

Neurological Disease

- **Stroke**: presents with an acute, forehead sparing facial paralysis associated with other lateralizing neurological signs
- **Guillain-Barré Syndrome**: common cause of acute **bilateral** facial nerve palsy, associated with generalized weakness, central nervous and autonomic dysfunction
- **Myasthenia Gravis**: autoimmune disease with antibodies against the acetylcholine receptor at the neuromuscular junction which causes progressive motor weakness with repetitive function; associated with ptosis, difficulty chewing, talking, weakness, and thymic tumors

Facial Nerve Repair and Reanimation

Introduction

- Goals of Facial Rehabilitation: protect eye from corneal exposure, provide a balanced smile (cosmesis), resolve drooling, improve mastication
- no irreversible procedure that interrupts the continuity of the facial nerve should be considered if there is a possibility of spontaneous recovery
- if paralysis has occurred <12–18 months (before significant muscle atrophy or loss of motor endplates) may consider end-to-end anastomosis (preferred), interpositioning, crossover grafting, and upper eyelid gold weight implants
- if paralysis has occurred >12–18 months with significant muscle atrophy or the facial nerve cannot be grafted may consider musculofacial transpositions, static procedures, and upper eyelid gold weight or spring implants

End-to-End Anastomosis (Neurorrhaphy)

- Indications: best choice when tension-free closure is possible and motor endplates are intact (recent injury), provides best chance of rehabilitation (facial muscle movement with the least synkinesis)
- Technique: microsurgical anastomosis of the epineurium (nerve sheath) or perineurium (if feasible), avoidance of tension is essential (may require releasing proximally and distally or rerouting at the mastoid segment)
- ideally completed before three days (distal branches may be stimulated up to 3 days prior to transection)

Interposition Grafting

- Indications: method of choice if unable to achieve a tension free end-to-end anastomosis and motor endplates are intact
- Technique: nerve cable graft interposed between nerve endings, greater auricular or sural nerves most common nerve grafts
- typically provides resting muscle tone and spontaneous facial expression

Hypoglossal-Facial Nerve Crossover

- Indications: proximal stump unavailable (eg, temporal bone tumors), distal segment intact, and functioning motor endplates (within 12–18 months of injury, evaluated with EMG or muscle biopsy) or direct end-to-side grafting with a mobilized facial nerve
- Technique: connects hypoglossal nerve to distal segment of facial nerve via a cable graft
- restores some voluntary motion and resting tone typically by 6 months
- may result in significant synkinesis, requires patient reeducation of motor coordination
- **Crossface Nerve Graft**: connects the branches of the opposite facial nerve to distal segment of facial nerve via a cable graft (*controversial*)

Static Procedures

- Indications: may be used as an adjunctive procedure to enhance facial symmetry by providing static support
- Types: fascial and allograft slings, browlifts, rhytidoplasty, canthoplasty
- simpler techniques than dynamic procedures

Dynamic Procedures

- Indications: unavailable facial nerve or atrophic facial muscles
- **Temporalis Muscle Transposition**: reanimates the mouth (may also be used for the eye)
- **Masseter Muscle Transposition**: reanimates the mouth
- **Free Nerve Muscle Grafts**: 2-stage procedure, initial procedure creates a sural nerve anastomosis to the facial branch of the unaffected side with distal graft end left free in preauricular region of the paralyzed side, after 6–12 months a pectoralis minor graft or gracilis graft is transposed and anastomosed to the nerve graft

Ocular Rehabilitation Techniques

- **Tarsorrhaphy**: partial suturing of the eyelids, permanent procedure, does not reanimate, results in visual field deficits and cosmetic deformity
- **Upper Eyelid Gold Weight Implant**: enhances closure by utilizing gravity, may be used with reversible or irreversible paralysis, risk of implant extrusion or migration, contraindicated with glaucoma (may aggravate ocular hypertension)
- **Lateral Canthoplasty/Lid Shortening Procedures**: indicated to correct senile ectropion from lower lid laxity

CHAPTER

7

Facial Plastic and Reconstructive Surgery

R. Pasha and Richard L. Arden

(continued)

WOUND HEALING

Stages of Wound Healing

Inflammatory (Substrate) Phase

- **initial stage**

Hemostasis

- initial vasoconstriction for 10–15 minutes (thromboxane A_2) followed by vasodilation (histamine, serotonin)
- endothelial cells contract → expose collagen, fibronectin, and laminin → forms platelet plug
- initiates coagulation, kinin, and complement cycles

Inflammatory (Cellular)

- **Polymorphic Neutrophils** (**PMNs**): appear within 6 hours, maximum cellular influx at 24 hours
- **Macrophages**: essential for wound healing (regulatory function), predominant cell type by day 3 and 4
- **Fibroblasts**: appear by 48 hours, maximum cells at 15 days; predominant producer of collagen, elastin, and fibronectin (may differentiate into **myofibroblasts** which are important for wound contraction)

Proliferative Phase

- **2–24 days**

Re-epithelialization

- begins with basal epithelial cell differentiation and separation from basement membrane (may be stimulated by epidermal growth factor)
- initial cellular detachment from loss of desmosomes, pseudopod formation
- migration in "leap frog" pattern with fibronectin and others at 12–21 μm/hr (moist environment aids in migration)

Neovascularization

- stimulated by hypoxia, acidity, and lactate
- **Angiogenesis**: migration of epithelial cells into perivascular spaces, forms channels and capillary buds

Collagen Deposition

- initial deposition of Type III collagen, later forms Type I collagen
- maximum deposition at 2–3 weeks
- Collagen Synthesis: α chains → hydroxylation of proline and lysine (requires vitamin C and iron) → combine into a helix → glycosylation occurs → secreted by fibroblasts as **procollagen** → cleavage to **tropocollagen** → aggregates into fibrils → combines into **collagen fiber**

Collagen Types

I	most common, bone, tendon, **late** scar, fascia, skin
II	hyaline cartilage (chondrocytes)
III	skin, uterus, arteries, **early** scar
IV	basement membranes

Wound Contraction

- mediated by myofibroblasts
- maximal at 12–15 days
- contracts at 0.6–0.75 mm/day

Remodeling (Maturation) Phase

- increase in Type I collagen and more parallel alignment of collagen fibers results in increased tensile strength with decreased scar dimensions
- **3 weeks**: **15%** original tensile strength (highest level of collagen)
- **6 weeks**: **60%** original tensile strength
- **6 months**: **70–80%** original tensile strength

Compromised Wound Healing

Causes

- Local Factors: infection, irradiated tissue, contamination, hematomas, neoplasms, wound desiccation
- Medications: corticosteroids, NSAIDs, chemotherapy, immunosuppressive agents
- Medical Condition: diabetes, severe malnutrition, smokers, peripheral vascular disease, hypothyroidism, Ehlers-Danlos syndrome, osteogenesis imperfecta, immunodeficiency
- Technical Factors: traumatic handling of tissues, poor incision design, tension closure, poor hemostasis

Management

1. **Assess Medical Condition**
   - assess nutrition (albumin and prealbumin, total lymphocyte count, transferrin, nitrogen balance), consider parenteral nutrition
   - assess diabetes (fasting glucose) and hypothyroidism (TSH, T4)
2. **Treat Infection**
   - culture and sensitivities with appropriate antibiotics (oral versus parenteral)
   - topical antibiotics (moist environment aids in re-epithelization)
3. **Local Wound Care**
   - debridement and irrigation
   - occlusive wound dressing changes (wet to dry for debridement)
4. **Hyperbaric Oxygen (HBO)**
   - beneficial for poorly healing tissue secondary to hypoxia
   - creates a steep oxygenation gradient which aids in oxygen delivery
   - Complications: pneumothorax, seizures, middle ear barotrauma (may require a myringotomy prior to initiating HBO)
   - Absolute Contraindications: untreated pneumothorax, select medications (eg, cisplatin, doxorubicin, disulfiram, mefenamic acid)
   - Relative Contraindications: emphysematous blebs, eustachian tube dysfunction, sinusitis, seizure disorder, history of thoracic surgery

INCISION/EXCISION PLANNING AND SCAR REVISIONS

Incision and Excision Planning

Introduction

- initial proper planning is essential for achieving good results in excising lesions of the head and neck
- ideal angle for fusiform closure (*see below*) is **≤30°** otherwise consider a graft, flap, or adjunctive M-plasty (*see below*)
- ideally, excision should be elliptical with the long axis of the excision along a RSTL (*see below*), natural facial crease, aesthetic unit junction, or hairline
- long straight incisions (≥2 cm) are the most noticeable
- proper undermining is essential in all excisions to minimize tension along wound edges
- the principle of halves (placing sutures in a bisecting fashion) avoids "bunching" of tissues

- slight eversion of wound edges allows for a flatter scar by accounting for scar contraction

Skin Line Types

- **Relaxed Skin Tension Lines (RSTLs)**: generally run parallel to wrinkles (exceptions include the lateral orbital region) and perpendicular with the contractions of muscles, best demonstrated in upright position with active facial muscle contraction
- **Langer's Lines**: relaxation creases found on cadavers (rigor mortis), usually perpendicular to RSTLs, not included in planning elective excisions/incisions
- **Lines of Maximum Extensibility (LME)**: run perpendicular to RSTL

Scar Revisions

Introduction

- Ideal Scar: level to skin, no anatomical distortion, matching color, narrow, parallel to RSTLs, placed at junction of facial aesthetic units or nonvisible areas (eg, pre/post-trichial)
- Timing of Scar Revision: may revise at 6–12 months (final phases of scar contracture and maturation) or at 6–9 weeks (maximal fibroblastic activity); if obviously misplaced or misdirected may consider earlier revision
- may preserve deep scar and subcutaneous tissue to preserve strength unless the revision contributes to the deformity
- broken and irregular scars are less noticeable
- may consider dermabrasion or chemexfoliation (*see below*)
- corticosteroids, retinoids, silicone gel sheeting, and emollient creams may be considered while awaiting scar revision or after scar revision

Hypertrophic Scars and Keloids

- **Hypertrophic Scar**: overactive inflammation and collagen synthesis resulting in an enlarged scar confined to the boundaries of surgical trauma, typically flattens and softens over time, often precipitated by closure under tension, infection, or wound dehiscence
- **Keloid**: deposition of collagen that extends beyond the limits of the scar, does not regress over time
- keloids and hypertrophic scars are histologically similar under light microscopy (differences are demonstrated with scanning electron microscopy)

- Risks: darker complexion, sebaceous skin, wounds closed under tension, genetic predisposition, certain areas (eg, deltoid, sternum, over bony prominences)
- SSx: pruritic (from histamine release during active inflammation), pain, erythema, induration
- Rx: gentle massage with intralesional corticosteroid injections (triamincinolone acetonide) with repeat injections every 3 weeks for 3 consecutive times; if no response may consider excision (with corticosteroid injections), pressure delivery devices, silicone gel applications, radiation therapy (may be effective but must weigh risk of malignant transformations)

Excisional Techniques

Fusiform Excision (Fig. 7–1)

- elliptical excision with bilateral advancement for a linear closure
- Advantages: simplest technique
- Disadvantages: must be able to be designed parallel to RSTL, ideal closure angle ≤30°
- Common Uses: most common excisional technique, ubiquitous

M-plasty (Fig. 7–2)

- M-plasty modification at the ends of a fusiform to close a larger lesion with an ellipse and limit extent of a normal skin excision
- Advantages: shortens wound length, avoids extension of scars across an anatomical border
- Disadvantages: slightly more complex, diverges closure lines at ends away from RSTLs
- Common Uses: larger lesions

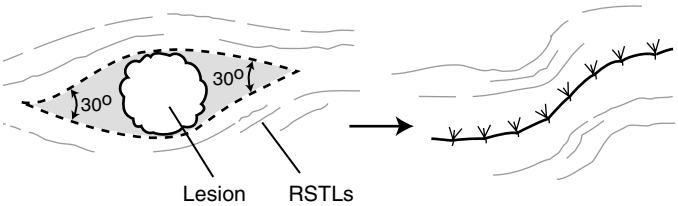

FIGURE 7–1. Fusiform excision for a circular lesion designed in order to allow final scar to parallel RSTLs; 30° angles are used at each end.

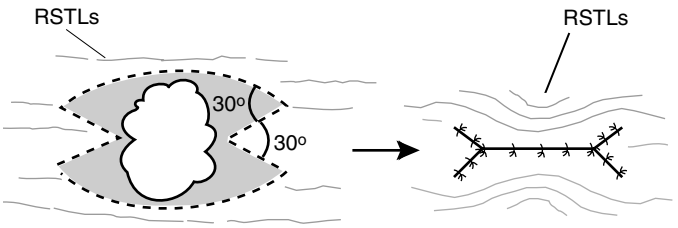

FIGURE 7–2. Fusiform excision with M-plasty modification on each end to allow closure of a large lesion with an ellipse; 30° angles are used to form each angle of the M-plasty.

Shave Excision

- removal of superficial lesion from skin surface
- Advantages: rapid, simple
- Disadvantages: may leave residual scar, predisposes to a locally recurrent lesion
- Common Uses: small elevated scars

Scar Repositioning

- repositioning of scar using fusiform excisions to a more favorable location (eg, junction of two aesthetic facial units, melolabial crease, hairline, preauricular crease)
- Advantages: scar placed in a more favorable position
- Disadvantages: requires sacrifice of tissue between scar and favorable site
- Common Uses: scars near a facial landmark

Serial Partial Excision

- removes part of scar with serial advancement flaps of normal adjacent skin
- concurrent tissue expanders decrease number of required excisions
- Advantages: may advance to hair bearing areas or aesthetic unit junctions
- Disadvantages: requires multiple procedures and longer time commitment
- Common Uses: large width scars, stellate lesions, skin grafts

Irregularization Techniques

Z-plasty (Fig. 7–3)

- two flaps classically constructed with 2 same-degree angles and 3 equal limbs; peripheral limbs run parallel with the RSTL

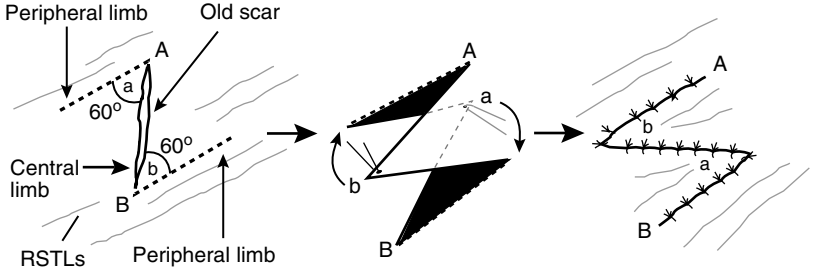

FIGURE 7–3. Classic Z-plasty technique utilizing 60° angles. The peripheral limbs run parallel with the RSTLs. (Adapted with permission from Lore JM. *An Atlas of Head and Neck Surgery.* 3rd ed. Philadelphia, Pa: W. B. Saunders; 1988.)

- Advantages: rotates the long axis of a scar to a favorable position (parallel to RSTLs or along an anatomical line), spreads force of contraction in many directions to minimize distortion, most powerful leveling technique
- Disadvantages: lengthens scar
- Common Uses: contracted scars (causes distortion of landmarks), misdirected scars (crosses RSTLs or anatomical lines), webs across concavities, edges of trap door deformities, repositioning of facial landmarks (eg, pulled up eyebrow)
- angle determines gain of length: **30° angle → 25% gain in length; 45° → 50%; 60° → 75%**
- final direction of central limb predicted by connecting free ends of peripheral limbs
- Z-plasty angle <20° increases risk of tip necrosis, angle >75° increases risk of a dog ear deformity

W-plasty (Fig. 7–4) **and Geometric Broken-Line Closure** (Fig. 7–5)

- **W-plasty**: excises scar with mirrored W-pattern, each leg <6 mm with 60° angles (regularly irregular)
- **Geometric Broken-Line Closure**: random, irregular patterns (triangles, rectangles, semicircles, and squares between 5–7 mm) with mirrored opposite side, more effective in camouflaging scar than W-plasty (irregularly irregular)
- Advantages: avoids straight-lined scar (less noticeable)

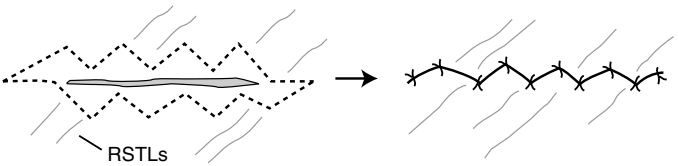

FIGURE 7–4. W-plasty technique utilizes a regularly, irregular mirrored W-pattern to camouflage the scar by avoiding a long single straight line.

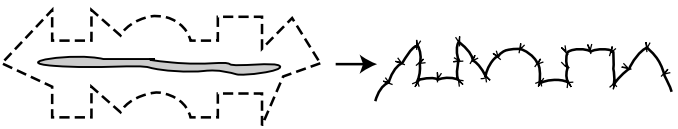

FIGURE 7–5. Geometric broken line closure technique utilizes mirrored random patterns to camouflage a scar.

- Disadvantages: more complex closure, time consuming, requires excision of small amount of normal tissue
- Common Uses: long scars not parallel to RSTLs, convex lines (mandible border, forehead, cheek)

HEAD AND NECK RECONSTRUCTIVE FLAPS

Vascular Anatomy of the Skin

Segmental and Axial Vessels

- large named vessels that run under muscle
- vascular pedicle for myocutaneous flaps

Perforators

- penetrate muscle to supply muscle
- enter skin through musculocutaneous or direct cutaneous vessels that supply the dermal-subdermal plexus

Musculocutaneous Vessels

- dominant blood supply to skin
- ubiquitous
- small variable size of perfusion area
- run perpendicular to skin
- basis of **random flaps** (most local facial skin flaps)

Direct Cutaneous Vessels

- supplementary blood supply to skin
- limited number
- larger size of perfusion area
- run parallel to skin
- associated with a vein
- basis of **axial flaps** (eg, paramedian forehead flap)

Local Skin Flaps

Introduction

- almost all local flaps in the head and neck are random patterned
- flaps are elevated and undermined in the intermediate subdermal plane (depending on bulk required at recipient site)
- estimated length to width ratio of local flaps in the face is 4:1 and 2:1 in the neck (reduce by half for irradiated tissue)

Types: defined by direction of tissue movement

- **Advancement Flaps**: linear movement (eg, monopedicle, bipedicle, Y–V advancement flaps)
- **Rotational Flaps**: radial movement
- **Transposition Flaps**: angular movement (eg, rhomboid, Dufourmental, bilobed flaps, Z-plasty)
- **Interpolated Flaps**: flap passed over or under a bridge of skin, separates donor site from defect (eg, subcutaneous island flap, paramedian forehead flap)

Single Advancement Flap (Fig. 7–6)

- advancement flap placed over defect, long axis oriented parallel to RSTLs, should not be longer than 2–3 times the width
- **Bilateral Advancement Flap**: flaps placed on opposing ends of defect (for larger defects)
- typically requires **Burrows Triangles** (*see* Figure 7–6) to prevent dog-ear formation
- Advantages: simple, avoids unwanted movement or deformity of facial structures (eg, brow, lateral canthus)
- Disadvantages: restricted flexibility
- Common Uses: forehead, lateral lip, eyelid

Rotational Flap (Fig. 7–7)

- semicircular flap that moves in a radial pattern along a defined arc with a fixed pivot point, shares a common side with the defect

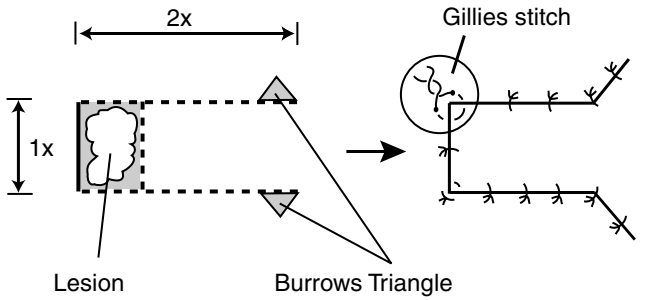

FIGURE 7–6. Illustration of the single advancement flap. Burrows triangles are utilized on the ends to prevent dog-ear deformity. Ideally, the length should not be longer than 2-3 times the width. A Gillies stitch, which is constructed similar to a mattress suture, is utilized to reduce tension at the corners.

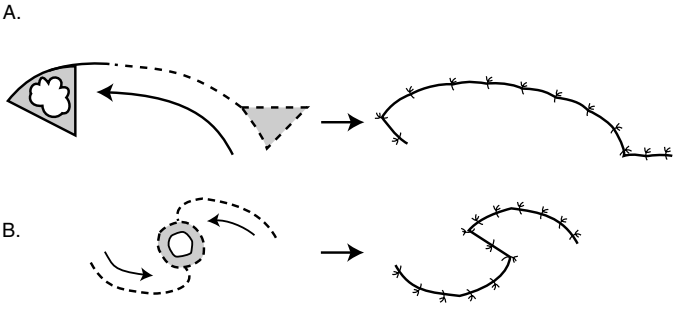

FIGURE 7–7. A. Rotational flap to close a triangular defect. Triangular excision at opposite end of defect allows for a flat closure. **B.** Bilateral rotational flaps utilized to close an elliptical defect.

- Advantages: allows closure of large lesions by recruiting lax skin over a greater distance
- Disadvantages: requires a wider base than the advancement flap, requires extensive undermining and long peripheral incision (4–5× diameter of the defect)
- Common Uses: cheek, triangular defects

Classic Rhomboid (Limburg) (Fig. 7–8)

- transposition flap constructed typically from equal length segments around two 120° and two 60° angles
- **Dufourmental Flap**: defect diamond is created similarly; however, the extending segments are constructed with more acute angles (<120° and <60° angles), allows improved blood supply to flap base and shares closing tensions
- Advantages: better distribution of tension (tension is away from defect, *see* Figure 7–8), reliable, may be designed so the final closure will be parallel to the RSTLs
- Disadvantages: forces a facial defect into an arbitrary design
- Common Uses: cheek, temple

Bilobed (Fig. 7–9)

- transposition flap that recruits lax tissue (secondary flap) from a nearby site that allows primary flap to effect closure of the defect
- primary flap should be slightly smaller than the defect, secondary flap should be ½–¾ the width of the primary flap (except on nasal tip where defect:primary flap = 1:1)
- Advantages: distributes tension evenly

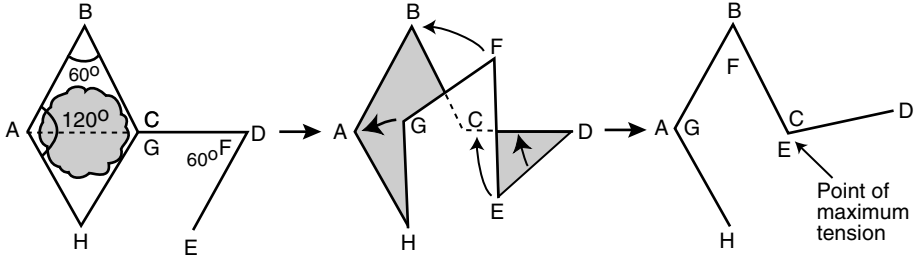

FIGURE 7–8. Classic rhomboid flap utilizes 120° and 60° angles to allow the point of maximal tension to be away from the defect. (Adapted with permission from Lore JM. *An Atlas of Head and Neck Surgery*. 3rd ed. Philadelphia, Pa: W.B. Saunders; 1988.)

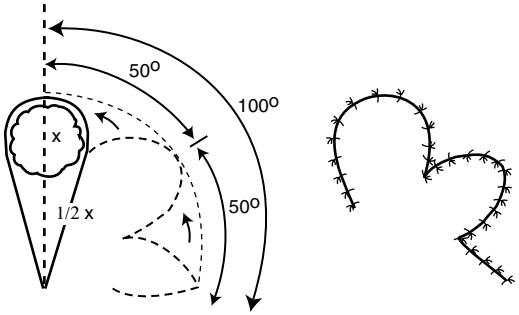

FIGURE 7–9. Bilobed flaps are designed with a primary flap and a smaller secondary flap to distribute the tension of wound closure more evenly.

- Disadvantages: risk of pin cushioning, lengthy incision that rarely exploits ideal RSTLs
- Common Uses: nasal tip/dorsum, lateral cheek

Regional Pedicled Flaps

Introduction

- **Delay Phenomenon**: surgically enhanced viability of flap by incising and partially undermining, but not transposing flap until 2–3 weeks, allows flap conditioning in preparation for transfer by increasing A-V shunting, increases vessel size and number, reorients vessels

Complications

- **Vessel Insufficiency and Injury**: prevented with atraumatic technique (avoid vasospasm), avoid tension and compression at tunnel, careful postoperative monitoring, prophylactic antibiotics (especially for intraoral contamination)
- **Partial Flap Necrosis**: skin island most susceptible (epidermolysis); Rx: debridement after adequate demarcation, surviving soft tissue and granulation may allow adequate seal, aggressive treatment of infection or fistula if present
- **Fistula**: increased risk with flap necrosis, wound infection, soilage by saliva, tight wound closure, immunocompromised or malnourished states; Rx: aggressive antibiotics coverage and wound care (packing), medialization of tract (away from carotid artery), may require another flap for closure
- **Hematomas and Seromas**: may present at donor site

Pectoralis Major

- Type: myocutaneous
- Advantage: reliable ("the workhorse"), excellent reach (up to lateral canthus), one stage procedure, potential for simultaneous harvesting, easy to harvest
- Disadvantage: may be bulky, potential for breast deformity in women, potential hair transfer, loss of pectoralis function (may be significant with concurrent ipsilateral injury to CN XI)
- Arterial supply: pectoral branch of thoracoacromial artery
- Common Uses: internal and external defects of the oral cavity, oropharynx, hypopharynx
- relatively contraindicated with ipsilateral radical mastectomies
- double paddle may be created by using the lateral thoracic artery
- incision is made along the inframammary crease in women to camouflage closure line

Pectoralis Major Anatomy

- Vascular Anatomy
  1. second portion of the axillary artery → thoracoacromial artery → pectoral branches
  2. second portion of the axillary artery → lateral thoracic artery
  3. first portion of the axillary artery → internal mammary artery
- Origin: clavicle, sternum, lower ribs, and rectus abdominis fascia
- Insertion: intertubercular groove of humerus. deltoid tuberosity, deep fascia of arm
- Action: medial rotator and adductor of upper extremities
- Innervation: lateral and medial pectoral nerves

Trapezius

- Type: myocutaneous
- Advantage: 3 forms allows for versatility, relatively flat and thin flap, 1-stage procedure
- Disadvantage: relatively limited arc of rotation, significant donor site morbidity (weakness of upper extremities, may require skin graft for closure), weaker blood supply, awkward positioning
- Common Uses: oropharyngeal and hypopharyngeal defects, lateral neck, posterior face

Flap Designs (Fig. 7–10)

1. **Superiorly Based (Upper) Trapezius Flap**: vascular supply from the occipital artery and paraspinal perforators, reliable flap, limited arc of rotation, donor site may require a skin graft

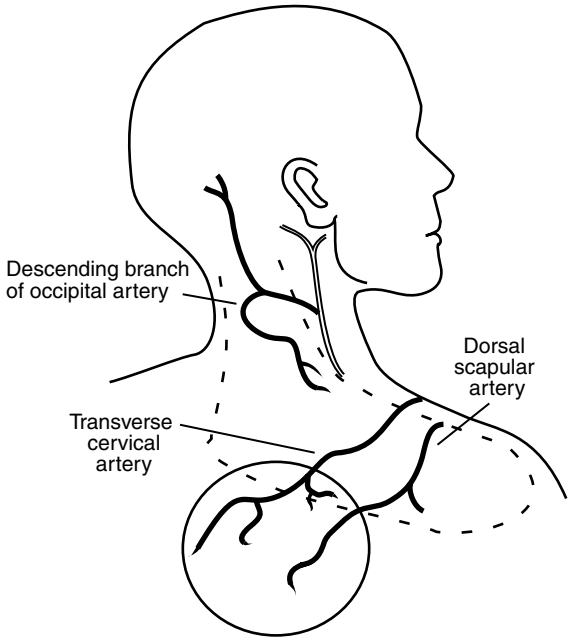

FIGURE 7–10. Diagram of the superiorly based (upper) trapezius flap (*dotted line*) based on the occipital artery and the inferior (lower) trapezius island flap (*solid line*) based on the transverse cervical and dorsal scapular arteries.

2. **Lateral Island Trapezius Flap**: vascular supply from the superficial branches of the transverse cervical artery, may cover defects of the oropharynx, posterior oral cavity, and hypopharynx
3. **Inferior (Lower) Trapezius Island Flap**: vascular supply from the descending branches of the transverse cervical artery and the dorsal scapular artery, provides a long pedicle, most commonly used trapezius flap

Latissimus Dorsi

- Type: myocutaneous
- Advantage: large amount of available skin and soft tissue, long vascular pedicle with extended arc of rotation (to vertex of scalp), less hair transfer, potential for bilobed skin islands, out of irradiated field

- Disadvantage: requires repositioning (lateral decubitus), propensity for seroma formation at donor site, may be bulky in large patients, requires extended tunneling between pectoralis major/minor
- Arterial supply: thoracodorsal artery
- Common Uses: similar to pectoralis major flaps (although not as common)

Sternocleidomastoid

- Type: myocutaneous
- Advantage: donor site located close to defect
- Disadvantage: tenuous blood supply to skin (not a true axial flap), rotation limited by accessory nerve
- Arterial supply: perforating vessels from the occipital, superior thyroid, and thyrocervical arteries (at least 2 of the 3 sources should be preserved)
- Common Uses: small defects in anterior lateral oral cavity and lateral oropharynx

Platysma

- Type: myocutaneous or muscle only
- Advantage: donor site located close to defect, thin and pliable skin
- Disadvantage: inconsistent and weak blood supply (not a true axial flap) from facial artery branches
- Arterial supply: random perforators from external carotid system
- Common Uses: small intraoral defects

Deltopectoral

- Type: fasciocutaneous
- Advantage: strong blood supply, large amount of donor tissue available, tunnel forms a favorable fistula (inferiorly located)
- Disadvantage: requires second stage procedure for detachment (6–8 weeks), requires skin graft at donor site
- Arterial supply: first 4 perforating vessels of internal mammary artery (the second vessel is the largest vessel)
- Common Uses: similar to pectoralis major flaps

Microvascular Free Flaps

Introduction

- Advantage: single-stage procedure, excellent perfusion, ability to preselect tissue characteristics of donor tissue for given recipient site defect, potential two-team approach, possible functional restoration

(sensation/motor), improved ability for spatial positioning of donor tissue
- Disadvantage: requires microsurgical expertise and specialized instrumentation, may require longer operating time, possible color and texture mismatch
- **5–15**% failure, most occur in first 24–72 hours; salvage rate on reexploration (≈**50**%)
- neovascularization complete after **8 days** (except jejunal flaps, serosa prevents neovascularization)
- vascularized muscle reduces to ≈20% of its mass, vascularized fat reduces to 40–60% of its mass (fat required for tissue bulk)
- may monitor flap clinically by examining color, turgor, capillary refill, temperature, and quality of dermal-mucosal bleeding (additional information affordable by monitoring devices [*eg,* laser Doppler])
- revisions should be completed as soon as possible to avoid no-reflow phenomenon, survival time may be extended with free radical scavengers, or NSAIDs
- Critical Time of Warm Ischemia: jejunal flaps (≈**2 hours**), skeletal flaps (~**6 hours**), cutaneous flaps (≈**8 hours**)
- Preoperative Evaluation for Microvascular Free Flaps: history of arterial lines or peripheral vascular disease; consider duplex studies for radial forearm and fibular free flaps; consider arteriograms for Ankle-Brachial Index (ABI) < 1.0 or Toe-Brachial Index (TBI) <0.7

Microvascular Anastomoses Pearls

- do not remove atherosclerotic plaques, may lead to thrombosis from intimal damage
- avoid intraoperative and postoperative hypothermia, hypovolemia, and hypotension
- avoid **vasospasm** by minimizing trauma to vessels
- avoid intraluminal introduction of adventitia (thrombogenic)
- artery and vein ideally should align in same direction
- vessels for anastomosis should be similar in size and ideally oriented longitudinally in the neck (avoid kinking)
- avoid stasis and collagen deposition by irrigating with heparin and creating a near-perfect anastomosis
- may consider aspirin for 10 days postoperatively
- vascular pedicle is susceptible to damage from infection and salivary contamination (prophylactic antibiotics, watertight seal from oral cavity and pharynx)
- bone scans may be done within 1 week to assess bone perfusion for osseous flaps

Osseous Free Flaps

Radial Forearm

- Type: fasciocutaneous or osseocutaneous (less common)
- Advantage: thin skin (≈2/3 radial forearm skin transferable), typically hairless, sensate potential, rapid harvest, long vascular pedicle
- Disadvantage: only 40% circumferential radial bone may be transferred (potential for pathological fractures), bone does not accept endosseous implants, donor site requires a skin graft, superficial branch of the radial nerve may be damaged causing numbness to snuff box, sacrifices a main artery
- Neurovascular Transfer: radial artery and venae comitantes, superficial forearm vein, lateral and medial antebrachial cutaneous nerve, superficial branch of radial nerve
- Common Uses: moderately large defects that require a thin flap (oral cavity, face), may be tubed for pharyngeal reconstruction, limited segmental mandibular defects
- preoperatively must check collateral circulation with Allen test (assessing turgor of hand after manual occlusion of radial artery to integrity of deep palmer arches) or Doppler studies

Lateral Arm

- Type: fasciocutaneous or osseocutaneous
- Advantage: thin skin, typically hairless, sensate potential, may close donor site primarily, does not sacrifice major artery, may include humerus, quick harvest
- Disadvantage: difficult to close primarily if >6 cm, unattractive scar at donor site, sensory deficit on extensor surface of forearm, difficult positioning for two-team approach, shorter vascular pedicle than a radial forearm free flap
- Neurovascular Transfer: profunda brachii artery, posterior (lower lateral) cutaneous nerve of the arm
- Common Uses: similar to radial forearm flaps

Scapula

- Type: osseocutaneous or fasciocutaneous (or bone only)
- Advantage: color and texture matches facial skin well, typically hairless, 8–14 cm of bone length available, allows primary closure of donor defect (≤12 cm)
- Disadvantage: requires detachment of shoulder girdle muscles resulting in significant weakness, requires lateral decubitus positioning restricting ease of two-team approach, smaller volume bone stock for endosseous implants

- Vascular Transfer: circumflex scapular artery
- Common Uses: complex through and through bone and soft tissue defects, facial defects, mandible defects
- **Megaflap**: large flap with bone and soft tissue components including latissimus and serratus muscles on 1 common pedicle (subscapular)

Fibula

- Type: osseocutaneous (or bone only)
- Advantage: excellent bone reconstruction (longest bone graft [25 cm], consistent diameter, good segmental periosteal supply allows for close osteotomies), moderately thin fasciocutaneous flap, sensory potential (lateral sural cutaneous nerve)
- Disadvantage: bone requires multiple osteotomies for mandibular reconstruction, skin graft may be required at donor site for osseocutaneous harvest (skin paddle >6 cm), variable and tenuous blood supply to skin paddle, septocutaneous perforators makes for a precarious skin island (should include musculocutaneous perforators through soleus muscle)
- Neurovascular Transfer: endosteal and periosteal branches of the peroneal artery, lateral cutaneous branch of the peroneal nerve
- Common Uses: mandible reconstruction for bone only or bone with limited soft tissue requirements

Iliac Crest

- Type: osseomyocutaneous or primary bone flap
- Advantage: excellent bone reconstruction (intrinsic shape reduces need for osteotomies for hemimandibular reconstruction, large cancellous component favors endosseous implants and bone healing), concealed scar placement, allows for primary closure of large flap
- Disadvantage: bulky soft tissue that cannot be thinned because of retention of obligatory muscle cuff and abdominal fat thickness, large amount of dissection required (painful donor site), temporary hip weakness with paresthesia, risk of inguinal hernia, poor color match to face
- Vascular Transfer: deep circumflex iliac artery and vein
- Common Uses: mandible reconstruction with larger soft tissue requirements (floor of mouth, hemitongue)

Soft Tissue Free Flaps

Rectus Abdominus

- Type: myocutaneous
- Advantage: large soft tissue availability, reliable, flexible flap design, long vascular pedicle, potential motor innervation

- Disadvantage: risk of ventral hernia (especially below arcuate line), poor color match to facial skin
- Vascular Transfer: deep inferior epigastric artery and vein
- Common Uses: massive soft tissue defects

Latissimus Dorsi

- Type: myocutaneous
- Advantage: massive soft tissue with large surface area, bulky, may be split into 2 islands, reinnervation potential maintains bulk, closed primarily
- Disadvantage: poor color match, potential shoulder disability when included with radical neck dissection or pectoralis muscle flap, awkward positioning for two-team approach, may have delayed healing from mobility of donor site, requires 1 week immobilization, higher risk of seroma formation
- Neurovascular Transfer: thoracodorsal artery and 9–11 intercostals (lower ⅓), thoracodorsal nerve
- Common Uses: massive defects

Lateral Thigh

- Type: fasciocutaneous
- Advantage: intermediate thickness, may close donor site primarily, potentially sensate
- Disadvantage: more difficult to harvest, occasional hair bearing, higher risk of seroma formation
- Neurovascular Transfer: third perforator of profunda femoris artery, lateral femoral cutaneous nerve
- Common Uses: large sensory skin requirements

Jejunum

- Advantage: similar diameter to the esophagus, tolerates radiation, operation is extrathoracic
- Disadvantage: laparotomy required, total flap necrosis dangerous (risk of carotid blowout), limited tolerance for warm ischemia (2 hours), inferior speech rehabilitation quality following puncture (compared to radial forearm), requires 3 visceral anastomoses
- Neurovascular Transfer: superior mesenteric arcade
- Common Uses: esophageal and long circumferential pharyngeal defects
- Contraindicated: ascites, carcinoma below thoracic inlet, inflammatory bowel disease, significant abdominal adhesions

GRAFTS, IMPLANTS, AND EXPANDERS

Introduction

- autogenous grafts, in general, are preferred over synthetic grafts or allogenic tissue (cadaveric) to avoid graft rejection and foreign body reactions
- Ideal Implants: inert, noninflammatory, resist foreign body reaction, noncarcinogenic, resist mechanical stress, sterilizable, easy to sculpt, easily removed, inexpensive, resist extrusion and mobility

Graft Types and Materials

Bone

- Advantages: does not warp, living tissue
- Disadvantages: requires bone-to-bone contact
- requires direct contact with bone to avoid resorption with fibrosis (except for nasoseptal and turbinate bone for uncertain reasons), soft tissue coverage, and immobility (8–12 weeks)
- **Cortical Bone**: outer layer, provides strength
- **Cancellous Bone**: inner, trabecular layer, contains more osteocytes and osteoblasts
- Phases of Osteogenesis
  1. **Osteoid Formation**: primarily a function of transplanted osteoblastic cells in **cancellous** bone, occurs for 1 month, forms framework
  2. **Remodeling**: function of osteoblasts, osteoclasts, and fibroblasts (require pleuripotent cells), begins at 2 weeks, mediated by **bone morphogenic protein** (BMP) found in **cortical** bone
- Common Uses: mandible reconstruction, midfacial defects, craniofacial surgery, rhinoplasty

Cartilage

- Advantages: easy to shape, maintains structural integrity, easy to harvest and store, does not require direct contact with cartilage or bone, minimal resorption and displacement
- Disadvantages: requires soft-tissue coverage for diffusion of nutrients (an attached perichondrium prevents resorption)
- Types
  1. **Hyaline**: most common, contains only type II collagen; found in articular joints and the nasal septum
  2. **Elastic**: contains elastic fibers for flexibility; found in the ear, epiglottis, and parts of the larynx

3. **Fibrocartilage**: sparse ground substance; more dense than hyaline cartilage; found in the pubic symphysis, attachments of tendons and ligaments, and intervertebral disks

- Common Donor Sites: nasal septum, auricular concha, rib
- Common Uses: rhinoplasty, otoplasty, orbital reconstruction

Dermal, Skin, and Fat

Split-Thickness Skin Graft (STSG)

- contains epidermis and variable amount of dermis (thin STSG [Thiersch]: 0.008–0.010 inch; thick STSG: 0.016–0.018 inch)
- Advantages: better survival, easy to mesh, does not require closure of donor site
- Disadvantages: contracts more than FTSGs, poorer color match/texture match, less durable (particularly over bone), donor defect
- Common Uses: defects with chance of recurrent tumor or for temporary coverage prior to definitive reconstruction

Full-Thickness Skin Graft (FTSG)

- contains entire epidermis and dermis
- Advantages: better color match, allows for hair growth, less contracture, resists trauma
- Disadvantages: higher graft failure, slower revascularization, requires primary closure at donor site
- Common Donor Sites: postauricular and preauricular skin (thin, hairless, similar facial skin color), upper eyelid (from excess skin), supraclavicular region, melolabial skin (thick, similar nasal skin color)
- Common Uses: lateral surface of the auricle, eyelids

Dermal Grafts

- contain dermis after removal of epithelium and subcutaneous fat
- Advantages: resistant to infection, may be buried under skin, capable of reepithelialization, limited resorption
- Disadvantages: loss of thickness at donor site (although more cosmetically acceptable than STSGs)
- Common Uses: intraoral coverage, vessel coverage, or subcutaneous implantation for soft tissue bulk, dural coverage

Fat (see p. 429–430)

Common Synthetic Materials

- **Silicone Rubber**: low reactivity and rejection, autoclavable, resists infection, easy to sculpt, high extrusion rate (especially in the nose)

- **Mersilene** (polyester fiber): resists infection, minimal degradation, allows for tissue ingrowth
- **Acrylics**: (eg, polymethylmethacrylate) rigid, able to be molded in situ, does not allow tissue ingrowth
- **Metals**: gold (low tissue reactivity, malleable), titanium (anticorrosive, osseointegration, light weight, low tissue reactivity), stainless steel (strong, anticorrosive, no osseointegration)
- **Medpore** (perforated polyethylene): contains pores that are interconnected allowing soft tissue ingrowth, resistant to infection, harder than silicone, autoclavable
- **Calcium Hydroxyapatite**: type of ceramic, bioreactive properties (can conduct osteogenic cells), resists infections

Tissue Expanders

Introduction

- produces additional tissue adjacent to defect
- Advantages: avoids distant flaps (poor color and texture match, donor site morbidity), allows for hair-bearing skin
- Disadvantages: requires multiple procedures, deformity from expansion device
- Common Uses: face and neck, scalp (alopecia), preauricular region (otoplasty), concurrent procedure with serial excisions

Biomechanical Properties

- Biomechanical Components: collagen (stores energy), elastin (determines elastic property), glycosaminoglycans (bind collagen and elastin fibers), hyaluronate (type of GAG that binds fluid to form a gel)
- **Biological Creep**: property of conventional tissue expanders, permanent changes in microanatomy, increases epithelial mitotic activity and subdermal vasculature, results in a net increase in surface area
- **Mechanical Creep**: no change in microanatomy, displaces fluid and extracellular substances, realigns collagen fibers by breaking bonds with glycoaminoglycans and elastin fibers (eg, rapid intraoperative expansion), no net increase in surface area
- **Stress Relaxation**: decrease in stress when skin is held under constant pressure

Types of Injection Ports

- **Self-contained**: risk of rupturing implant, less dissection required
- **Remote**: variable-length connector tubing separates expander from injection port, requires more tissue dissection to place, essentially eliminates risk of expander rupture

- **External Valve**: port outside of patient, pain is avoided because there is no percutaneous injection (excellent for children)

Techniques

- base should be **2.5 times** the area of the defect (the **rectangle** expander has the best expander volume-to-skin area)
- shape based on site of expansion (not shape of flap)
- expansion should only form slight blanching, not pain or intractable blanching

Conventional Tissue Expansion Technique

- expand over 4–6 weeks of use
- relies on **biological creep**

Rapid Intraoperative Tissue Expansion

- expansion and deflation of prosthesis in operating room over 3-minute intervals (typically 3 cycles)
- no true net gain of skin surface area
- relies on **mechanical creep**

Complications

- **Infection**: may consider a trial of antibiotics, drainage, and irrigation before removing implant
- **Extrusion and Leakage**: may not necessarily require removal, may place on antibiotics and continue process
- **Neuropraxia or Vascular Compromise**: from placement over a nerve or vessel
- **Bone Resorption**: from placement of expander near bone
- **Hematoma**
- **Skin Necrosis**

FACIAL RECONSTRUCTION TECHNIQUES

Mandibular Reconstruction

Introduction

- immediate reconstruction prevents soft tissue contraction (compromising function and cosmesis) and mandibular segment displacement from muscle vector forces

- in general, the more anterior the defect the greater the functional and cosmetic morbidity and the more difficult to reconstruct
- high, limited, posterior defects may compensate well without reconstruction (less loading requirements)
- a proximal segment that contains only a coronoid process, condyle, and portion of the ramus may benefit from a coronoidectomy to avoid upward and medial displacement from vector force of the temporalis and pterygoid muscles
- Goals of Mandibular Reconstruction: mandible contour restoration, mastication, speech, sensation of lower lip and oral cavity

Reconstruction Methods

Osseous Free Flaps

- Indications: most reliable method for optimal restoration of the mandible
- Advantages: only reconstruction technique that allows for osseointegrated implants, best functional restoration
- Disadvantages: requires microsurgical expertise and specialized instrumentation (*see* Microvascular Free Flaps for general advantages and disadvantages of each composite free flap transfer)
- Common Types: fibular, iliac crest, scapula

Bone Grafting Technique *(*see *Graft Types and Material* above*)*

- Indications: limited defects
- Advantages: tolerates higher level of stress and strain than other non-vascularized neomandibles
- Disadvantages: requires jaw immobilization for weeks or months, high rate of infection, resorption, and exposure
- Donor Sites: rib, iliac crest

Regional Pedicled Flaps

- Indications: limited posterior defects
- Advantages: provides soft tissue bulk for mandibular contour, may resist some muscle force of remaining mandibular segment, quick
- Disadvantages: does not allow osseointegrated implants, poor functional outcome, high rate of exposure and bone necrosis, minimal bone volume
- Common Types: pectoralis major (with rib), trapezius (with spine of scapula), latissimus dorsi (with rib), deltopectoral, sternocleidomastoid, temporalis, platysma

Soft Tissue Flap with Reconstruction Plate

- Indications: limited defects (<5 cm), rapid repairs (poor medical status), or the elderly (less opportunity for extrusion)
- Advantages: quick, avoids donor defect
- Disadvantages: poor oral functioning, high rate of extrusion and fractures (especially with defects >5 cm and anterior defects)

Lip Reconstruction

Introduction

- first address bony and dental defects to allow for favorable projection of the lip
- proper soft tissue contour of the lip is achieved with precise apposition of the white line (vermillion cutaneous border) and maintenance of lip symmetry and bulk

Minor Lip Defect Reconstruction

- defects <⅓ of lip (more leeway allowed for lower lip than upper lip)
- **Vermillion Flap**: axial pattern orbicularis oris flap based on the labial artery
- **Labial Mucosal Advancement Flap**: indicated after shave excisions (vermillionectomy)
- **Primary Closure**: indicated with full-thickness defects, may consider V-shaped or M-plasty excision of lower lip, generally achieved without distortion for upper lip defects ≤¼ or lower lip defects ≤⅓

Defect Involves ½–⅔ Lip

Gillies Fan Flap (Fig. 7–11)

- orbicularis oris myocutaneous flap
- Advantage: allows for defect repair of up to 80%
- Disadvantage: delayed muscle function

Karapandzik Labioplasty (Fig. 7–12)

- modifies the Gillies Fan Flap
- Advantage: preserves neurovascular supply to lip for better function, simple design
- Disadvantage: unsightly circumoral scar, risk of microstomia

Abbe-Estlander Flap (Fig. 7–13)

- transfers full-thickness segment of lip to opposing lip

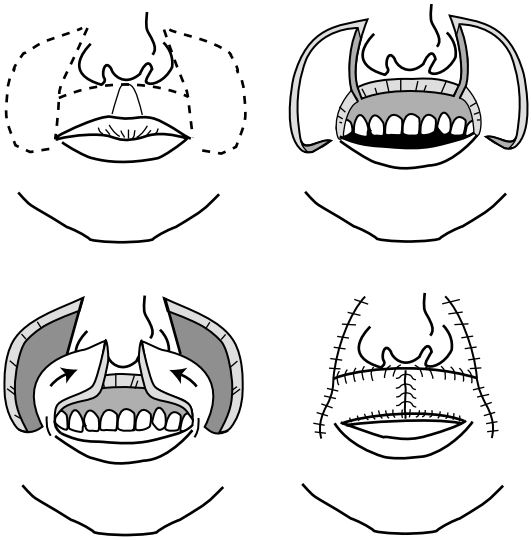

FIGURE 7–11. Gillies fan flap is based on an inferior medial pedicle constructed from full-thickness cheek flaps.

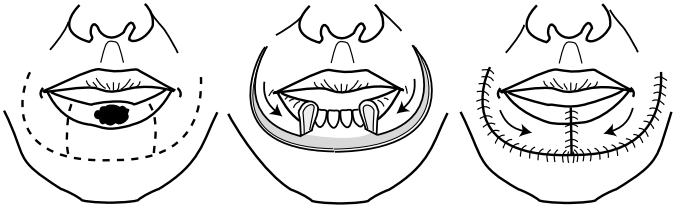

FIGURE 7–12. Karapandzik labioplasty requires circumoral incisions for closure of defects involving greater than ½ of the lip.

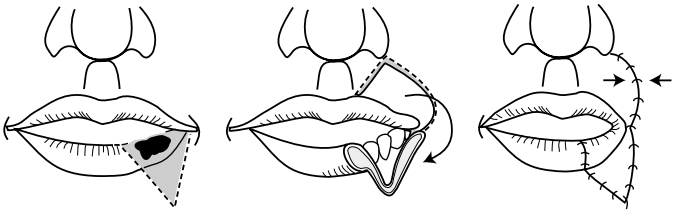

FIGURE 7–13. Abbe-Estlander flap allows for repairs of defects involving the commissure of the lip.

- Advantage: may be used for commissure defects (Estlander), good color match
- Disadvantage: 2-stage procedure (separation of flap pedicle and commissuroplasty completed later), reduces stomal size, requires an intact labial artery

Defect Involves >⅔ Lip

- **Bernard-Burow Flap**: advancement flap from cheek, indicated for midline lower lip defects
- **Nasolabial Flaps**: inferiorly designed, indicated for lateral defects, bilateral nasolabial flaps required for defects >80%
- **Johanson Technique**: may be used to close defects from <1 cm to entire lip, bilateral lateral advancement flaps with stepwise inferolateral skin incisions, preserves neuromuscular bundle and oral commissure, however loses sensation to the lip
- Total Reconstruction Techniques: radial forearm free flap, fascial sling with mucosal advancement flap, may also consider cutaneous and myocutaneous flaps

Auricular Reconstruction

Auricular Reconstruction Principles

- determine region of defect (helix, scapha, antihelix, concha, lobule, external auditory canal)
- assess cartilaginous defect (cartilaginous defects of the concha, triangular fossa, or scapha may not require cartilaginous reconstruction if the helical rim is intact)
- skin of equivalent thickness and color include the remaining auricular skin and the adjacent retroauricular (mastoid skin), postauricular, and preauricular region (skin expansion techniques have been used to gain additional skin)
- sculptured autogenous cartilage donor sites include the conchal region of the same or opposite ear (to avoid collapse the helical rim must be intact) or costal cartilage (especially for total auricular reconstruction); homologous cartilage tends to resorb

Reconstruction Techniques Based on Region

Helical Rim

- **Advancement Flaps**: utilizes retroauricular skin or skin from the posterior surface of the ear to advance over helical rim, donor defect repaired with skin graft, may consider a cartilage strip graft for support

- **Rotational Flap**: utilizes the adjacent portion of the helix (cartilage and skin) with primary closure of donor defect, may be considered for smaller defects
- **Tubed Flaps**: multistaged procedure that utilizes tubed skin constructed from the adjacent periauricular region, first stage produces the tubed flap, second stage consists of severing and transferring flap to the helical rim

Helix, Scaphoid, and Triangular Fossa

- **Helical Chondrocutaneous Advancement Flap**: for small marginal defects, advances a chondrocutaneous flap by excising a wedge of antihelix and scaphoid fossa and advancing mobilized segment(s) after incising anterior skin and cartilage for primary closure, creates a smaller ear (especially with large defects)
- **Conchal Chondrocutaneous Rotation Flaps**: utilizes a composite graft pedicled from the concha for larger defects, skin graft used to repair donor defect
- **Postauricular Turnover Flaps**: utilizes a pedicled flap with the base at the defect border and extended to the posterior surface of the ear and the postauricular skin, flap is redraped over a cartilage graft, skin graft used to repair postauricular donor defect
- **Free Transfer Composite Grafts from Opposite Ear**: transfers composite skin and cartilage, half the size of the defect, from the mirrored region of the opposite ear with primary closure

Concha

- **Postauricular/Retroauricular Skin Flaps**: utilizes a postauricular pedicled flap tunneled through the cartilage with a second-stage division 2–3 weeks later
- **Postauricular Myocutaneous Flaps**: single-stage procedure utilizes a postauricular pedicled myocutaneous flap that is transposed to the anterior surface with primary closure of the donor defect

Total Auricular Reconstruction

(Brent B, *Plast Reconstr Surg.* 1992;324:355–373)

- total auricular reconstruction requires multiple stages
- **Total Ear Resurfacing**: requires coverage with a thin flap, temporalis fascia flap most commonly used
- **Costal Cartilage Graft**: utilized due to ability to withstand absorption and trauma, maintains growth with opposite ear, sculptured from film pattern from opposite ear, initially inserted in proper position within a skin pocket with tension free coverage (may

consider intraoperative expansion techniques), secondary procedure detaches surgically created auricle with a skin graft to construct a sulcus, additional stage evacuates the helical sulcus and defats the scapha
- **Lobule Transposition**: may be performed as a second-stage procedure; if a lobular remnant is present, may unfurl and "splice" the remnant into position; if no remnant available, may create lobule with original framework
- **Tragal Reconstruction**: if available may construct tragus from an arched shaped composite graft from the conchal surface of the normal ear; if donor ear is not available may consider a conchal flap doubled on itself

Complications

- **Hematoma**: most worrisome complication since may result in loss of cartilage, must investigate severe pain by removing dressings; Rx: immediate reopening with removal of clot and controlling bleeding, antibiotics, pressure dressings, may consider suction and drainage for 2–3 days
- **Chondritis**: presents with pain, may result in loss of cartilage; Rx: aggressive antibiotic regimen, drainage and debridement if required
- **Skin Necrosis and Cartilage Exposure**: may be secondary to excessive tension, hematoma, or infection; Rx: small areas may be treated with topical antibiotics, debridement should be limited to obvious necrotic tissue, cartilage exposure requires coverage (may consider FTSG)
- **Hypertrophic Scars and Keloids**: *see above* for management, avoid by providing a tension-free closure
- **Pneumothorax and Atelectasis**: rare potential risk from rib harvest

Nasal Reconstruction

Reconstruction Principles

- the immobility of skin in the lower nose limits the use of primary closure (risk of rotating the nasal tip or distorting the alae)
- consider nasal subunits as concave shapes (soft triangles, nasal side walls, depression of the supra-alar facets) and convex shapes (dorsum, nasal tip, alar lobules) (Fig. 7–14)
- concave units may be replaced with skin grafts since they do not contract into a bulge
- convex units may be replaced with skin flaps since they tend to contract into a bulge
- nasal lining should be replaced with mucosa when possible
- if a wound involves >50% of a nasal subunit consider replacing the entire subunit versus patching the region

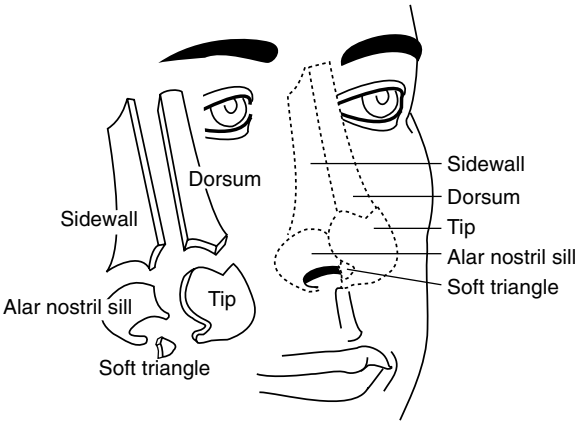

FIGURE 7–14. Illustration of the Nasal Subunits.

Management

1. examine condition of skin, general nutrition, general medical condition, smoking history
2. examine location and extent of missing subunits and external skin (ie, thick versus thin, concave versus convex)
3. plan superficial defect donor sites
   - Small Thick-skinned Regions (<1.5 cm): bilobed local flap, nasalis myocutaneous flap
   - Large Thick-skinned Regions (>1.5 cm): consider local pedicled flaps (*see below*)
   - Thin-skinned Regions: consider full-thickness skin grafts (eg, preauricular, postauricular, or supraclavicular donor sites)
   - may consider healing by secondary intention for smaller defects in concavities (eg, alar facial groove, nasoalar crease, medial canthus)
4. address bone and cartilage replacements
   - Lateral Support: consider septal and conchal cartilage free grafts, septal hinge flap (middle ⅓)
   - Midline Support: consider cantilever bone graft (attached to nasal radix to tip of nose), chondromucosal (composite) septal flaps
5. address nasal lining
   - donor sites from nasal vestibule and nasal septum (lower and middle ⅓)
   - bipartite septal flaps (lower ⅓)
   - split or full thickness skin grafts (vascularized recipient site)
   - orbicularis oculi myocutaneous pedicled flaps (medially based)

Regional Pedicled Flaps for Nasal Reconstruction

- **Sliding Nasal Dorsal Flap** (**Modified Reiger**): single-stage V-Y advancement glabellar flap continuous with nasal dorsal skin, useful for midnasal and nasal tip wounds, nasal scars tend to be more visible (requires back cut)
- **Direct Cheek Advancement and Rotational Flaps**: capable of closing large lateral nasal defects, major disadvantage is violation of principle of segmental facial composition

Nasolabial Flap

- based on random blood supply from facial artery
- transfer from cheek tissue lateral to melolabial crease
- Advantage: abundant mobile skin, scar may be hidden within melolabial crease
- Disadvantage: may result in some facial asymmetry (flattening)
- **Superiorly Based Design**: indicated for defects of the lower two-thirds of the nose (lateral nasal wall, nasal ala)
- **Inferiorly Based Design**: indicated for defects of upper lip, nasal sill, and columella
- **Subcutaneous Island Design**: indicated for whole subunit alar surface replacements

Paramedian Forehead Flap

- Advantage: reliable, good color and texture match, reliable blood supply, single pedicle allows for rotation (flap may be twisted to use skin as the internal lining of the nose),
- Disadvantage: long vertical forehead scar, 2-stages required, length limited by supraorbital rim and anterior hairline, interferes with eyeglasses (for 2–3 weeks)
- Arterial supply: supratrochlear artery (axial pattern) and branches of the facial artery (random pattern)
- Common Uses: large full-thickness defects of the lower two-thirds of the nose

INTRODUCTION TO FACIAL AESTHETIC SURGERY

Facial Anatomic Landmarks

- **Trichion**: midline at hair line
- **Glabella**: prominence in midsagittal plane, above root of nose

- **Nasion**: (**N**)asofrontal suture, superior to sellion
- **Sellion**: (**S**)oft tissue, deepest point of the nasofrontal angle
- **Radix**: root of the nose (contains the sellion and nasion)
- **Rhinion**: cartilaginous-bony junction (**thinnest skin** of nose)
- **Supratip Break**: just cephalic to tip defining point
- **Tip Defining Point**: 2 points representing the highest point of the crural arch (medial cephalic portion of the lateral crura)
- **Infratip Lobule**: portion of lobule inferior to the tip defining point
- **Infratip Break**: junction of the columella and lobule
- **Alar Crease**: most posterior portion of nose
- **Stomion**: embrasure of lips
- **Mentolabial Sulcus**: dip in chin
- **Pogonion**: anteriormost border of chin
- **Menton**: inferiormost border of chin
- **Gnathion**: intersection of the perpendicular lines from pogonion and menton (not on face)
- **Cervical Point**: menton and neck intersection
- **Tragion**: supratragal notch of ear

General Facial Assessment

- oval face shape is ideal
- width-length ratio of face should be 3:4
- rule of **facial thirds** (relates height): trichion → glabella = glabella → subnasale = subnasale → menton
- rule of **lower facial thirds** (relates height): ⅓ (subnasale → stomion) and ⅔ (stomion → menton)
- rule of **facial fifths** (relates width): intercanthal distance = width of base of nose
- skin scarring, pigmented lesions

Effects of Aging on the Face

- Skin: thinned epidermis, decreased subcutaneous fat, dermis loses elastic fragments (elastosis), decreased melanocytes and Langerhans' cells, decrease in Type I collagen (mature) and increase in Type III collagen (immature), reduced vascular supply (especially in smokers)
- Skeletal: thinned skull and mandible (especially in the edentulous patient) resulting in an excess skin envelope, laryngeal skeleton and hyoid descends
- Forehead Midface: weakened supportive matrix between SMAS (*see below*) and overlying fat pad results in deepened melolabial crease, hypotonic facial muscles results in festooning and facial rhytids

- Neck: jowling, chin ptosis, ptotic submandibular glands, platysma banding from loss of tone resulting in loss of cervicomental angle

Poor Candidates for Cosmetic Facial Plastic Surgery

- unrealistic expectations
- multiple physician visits
- psychiatric instability (eg, depression, anxiety)
- smokers have >12× increased risk of skin sloughing and scarring

Preoperative Photography

- **Frankfort Horizontal**: horizontal line from inferior border of external auditory canal (or top of tragus) to infraorbital rim, reference plane used for photos and analysis
- Recommended Film and Settings: 35 mm, slower film (ASA <100), focal length 90–105 mm, medium blue background most common, 2 light sources to reduce shadowing effects
- Rhinoplasty Views: left and right lateral, ¾ oblique, lateral with smile, frontal, and base
- Blepharoplasty, Rhytidoplasty, and Forehead/Brow Views: full frontal face, lateral, ¾ oblique, close-up eyelids (straight and upward gaze, eyes closed), and facial animation (smiling, raising eyebrows)

RHINOPLASTY

Surgical Nasal Anatomy

Nasal Tip Support

Major Tip Support

1. direction, strength, and resiliency of **lower lateral (alar) cartilage**
2. medial crura attachment to the **inferior septal angle**
3. attachment of the **lower and upper lateral cartilage**

Minor Tip Support

- interdomal ligament between lower lateral cartilage
- sesamoid complex
- skin/soft-tissue envelope (SSTE): overlying skin/muscle and its attachment to the alar cartilage
- anterior nasal spine
- cartilaginous septal dorsum
- membranous septum

Anatomical Terms

- **Nasal Length**: distance from nasofrontal angle to tip defining point of domes, determined mainly from the length, width, and direction of the lateral crura
- **Scroll**: continuous roll of the cephalic margin of the lateral crus that overlies the upper lateral cartilage (may be trimmed to reduced bulbousness of the tip)
- **Keystone Area**: region of overlap of the upper lateral cartilage by nasal bones
- **Angle**: junction of the medial and lateral cartilage (narrowest portion of alar cartilage)
- **Dome**: concavity formed by the inner surface of most superior extent of the lateral crus (lateral to angle)
- **Sill**: continuation of alar margin toward base of columella

Initial Consultation and Analysis

Introduction

- Hx: nasal trauma, cocaine abuse, smoking, granulomatous disease, sinusitis, nasal obstruction, previous surgery, bleeding disorders
- assess motivational factors for surgery
- preoperative photography (*as above*)
- **Non-Caucasian Nose**: usually address nasal tip projection, nasolabial angle, dorsal hump, and alar base, risk of over-projected look, limited by maximum projection

Internal Nose

- evaluate functional nasal airway (resistance, nasal valve)
- assess for septal deflection/deformities and turbinate hypertrophy

External Nose

Skin

- ideal skin is not too thick or thin, minimal sebaceous glands
- Thicker Skin: more postoperative edema, more subcutaneous scarring (increased risk of a pollybeak deformity)
- Thinner Skin: reveals minor irregularities

Profiles

- Nasofrontal Angle: angle from external nose and forehead, ≈**120°** (approximately at the level of the upper eyelid folds)
- Columellar-Labial Angle: male ≈90–100°, female ≈95–110°

- Dorsal Hump: may be straight, slight hump (males), slight scoop (females), suggestion of dorsal prominence at rhinion
- Dorsal Height: rule of **facial thirds**, nasion to subnasale = middle ⅓ of facial length (*see above*)
- Dorsal Width: rule of **facial fifths**, nasal width = ⅕ facial length or intercanthal distance (*see above*)
- Frontonasoorbital Line: frontal view should reveal a smooth line from eyebrows along the lateral edge of the dorsum then diverging slightly at the tip

Nasal Tip

- **Double Break**: characterizes a refined nasal tip, **supratip break** (separates dorsum from lobule) located 1–3 mm above the tip defining point and an **infratip break** between the infratip lobule and columella
- **Light Reflex**: two symmetrical reflections defined by the **tip defining points** (5–6 mm apart)
- **Tip Defining Point**: should lie 0.28 cm anterior to line connecting nasofrontal angle to the upper lip vermillion-cutaneous junction
- **Tip Rotation**: determined by **columellar-labial angle**
- **Tip Projection**: distance between facial plane and tip of the nose, **3:4:5 rule** = columella:base:dorsum
- **Tip Recoil**: tip of nose is depressed and degree of resistance is assessed, adequate recoil required for manipulation of lower lateral cartilage

Lobule and Columella

- alar rim should arch 2–3 mm above columella (**columellar show**)
- alar-columellar margin forms a "gull in flight" outline
- basal view should reveal an isosceles triangle
- lobular height should be ⅓ of total height on basal view

Chin

- chin should align with a vertical line dropped from the vermilion border of the lower lip (in women may be slightly behind this line)
- *see* p. 431

Surgical Techniques

Introductory Points

- minimize reduction of upper lateral cartilage to maintain an adequate nasal valve

- avoid transection of nasal dome to maintain alar spring
- preserve nasal tip support, especially caudal septal cartilage
- always err on the side of conservation of tissue rather than resection
- tip rhinoplasty generally should be performed first to determine nasal profile (to determine amount of hump to remove)

Incisions

- **Intercartilaginous**: disrupts attachment of lower and upper lateral cartilage, scarring may result in nasal obstruction since incision involves nasal valve
- **Marginal**: follows caudal margin of the lower lateral cartilage, may be combined with intercartilaginous incisions to form a **Bipedicle Flap** (**Intact Strip**)
- **Trans/Intracartilaginous**: splits lower lateral cartilage, lessens risk of nasal obstruction
- **Transcolumellar**: used for open rhinoplasty

Approaches

Nondelivery Approach

- Types
  1. **Cartilage Splitting** (**Trans/Intracartilaginous**): indicated for minimal refinement of nasal tip
  2. **Retrograde Approach**: indicated for minimal refinement of nasal tip with thick skin, uses an intercartilaginous incision
- Advantages: no major tip support affected, preserves intact caudal rim, one incision, no external scar, reduced surgical trauma and edema
- Disadvantages: minimal exposure of lower lateral cartilage, limited to conservative tip work, risk of asymmetrical incisions

Delivery Approach

- combines intercartilaginous and marginal incisions
- allows delivery of lower lateral cartilage as a bilateral pedicled chondrocutaneous flap
- Advantages: visualize entire lower lateral cartilage, good access to tip and nasal dome, no external scar
- Disadvantages: compromises major support by removing the attachment of the alar cartilage and the upper lateral cartilage, causes more edema (postoperative distortion), requires 2 incisions

Open Rhinoplasty

- combines marginal and transcolumellar incisions

- Indications: extensive tip work, revisions, non-Caucasians, cleft lip, crooked nose, major reconstructions, academic teaching
- Advantages: affords maximal nondistorted exposure, allows accurate placement and direct suture ability for grafting, does not disrupt major tip support mechanisms
- Disadvantages: compromises one minor support by separating the skin/soft tissue envelope (SSTE) from the lateral cartilage, external scar, more postoperative edema, sensory tip disturbances

Nasal Tip Projection and Rotation

- **Nasal Tripod Model**: predicts changes in nasal length, tip projection, and tip rotation in relation to changes in the 3 legs (legs of the tripod include the conjoined medial crura and the two lateral crura)
- should maintain a minimum of 6–8 mm of alar cartilage for support

Tip Correction Methods

Complete Strip Technique

- reduces volume of the cephalic margin of the lower lateral cartilage leaving the caudal margin intact from the foot of the medial crus to the pyriform aperture
- results in a slight cephalic rotation, accentuates domes, and decreases the convexity of the lower lateral cartilage
- Advantages: better preserves tip projection (intact medial crus), allows for more symmetrical healing, resists tip retrodisplacement
- Disadvantages: flaring of nostrils may result from any tip rotation

Interrupted Strip Technique

- "interrupts" the lower lateral cartilage
- indicated for tips requiring maximal rotation, overprojected tips from excessively large alar cartilage, wide boxy tips
- contraindicated in thin skinned patients (subtle irregularities cannot be camouflaged)
- **Rim Strip**: interrupts anterior border of lower lateral cartilage lateral to dome and proximal to pyriform (loss of tip support), provides maximal tip rotation
- **Lateral Crural Flap**: similar to rim strip procedure; however, leaves triangular portion of lateral crura at pyriform to prevent alar margin retraction and pinching, limits supra-alar definition
- **Dome Division**: complete interruption of lower lateral cartilage near dome, corrects asymmetric tips, allows increased or decreased projection, and narrowing of tip

Methods to Increase Tip Projection

- suture the medial crura together (transdomal or interdomal sutures)
- may increase cephalic rotation
- permanent septocolumellar suture
- **Supradomal Graft or Shield Graft**: adds 1–2 mm additional projection
- **Columellar Strut**: placed between medial crura (does more to maintain projection than actually increase projection)
- **Plumping Grafts**: placed at base of columella, changes the columellar-labial angle with an illusion of increased projection

Methods to Decrease Tip Projection (Retrodisplacement) Techniques

- complete transfixion of caudal septum
- full-thickness excision and suture of segments of the lateral and/or medial crura
- detachment of lateral crura

Methods to Increase Tip Rotation

- reduce caudal septum edge
- shorten lateral crura (must avoid excessive reduction to prevent valve collapse)
- augment the premaxilla
- removal of dorsal hump, augmenting the columellar-labial angle, and rotating the infratip lobule causes an apparent increase in tip rotation
- augment medial crura (ie, tripod concept)
- also see tip correcting methods above

Methods to Decrease Tip Rotation

- excise caudal septum near spine and shorten medial crura
- dorsum augmentation causes an apparent decrease in tip rotation
- consider an infratip button graft for a retruse infratip lobule

Nasal Hump

- most nasal hump deformity is caused from the cartilaginous dorsum
- nasal skin is thinnest at mid-dorsum (**rhinion**) therefore must create a slight middorsal hump to avoid saddle nose deformity
- an illusion of increased nasal length is achieved by leaving a small dorsal hump
- Dorsal Augmentation Graft Materials
  1. **Septal Cartilage**: does not resorb, ample supply, low infection rate

2. **Conchal Cartilage**: more difficult to sculpt
3. **Split Calvarium**: may be too rigid
4. **Costal Cartilage**: may warp
5. **Allografts**

Medial and Lateral Osteotomies

- osteotomies reduce open roof defects caused by removal of a nasal hump, straighten a crooked nose, and flatten convex nasal bones
- Medial Osteotomy: completed first, frees nasal bones from perpendicular plate of ethmoid
- Lateral Osteotomy: low, curved (low to high) cut (along nasomaxillary groove), leaves a triangular wedge at pyriform ledge superior to level of inferior turbinate to protect airway
- intermediate osteotomies may be utilized to correct the deviated nose with one sidewall longer than the other or to straighten an excessively wide or convex nasal bone
- damage to the lacrimal crest is rare
- to avoid nasal obstruction, for short nasal bones or when dorsal hump reduction is >3 mm consider **spreader grafts** (placed between the septum and upper lateral cartilage), osteotomies may considerably narrow the nasal valve

Postoperative Care

- Nasal Splints: remove after 1 week
- Intranasal Packing: remove after 1–2 days
- Septal Splints: helps prevent synechiae, remove after 1 week
- avoid strenuous activity
- cold packs to reduce swelling
- oral antibiotics while nasal packing in place

Revision Rhinoplasty and Complications

- **10–15%** of rhinoplasties require revision work
- typically wait 1 year for revisions
- Immediate and Short-term Postoperative Complications: epistaxis, infection, septal perforation, septal hematoma, skin necrosis, transient epiphora

Late Postoperative Defects and Complications

Pollybeak Deformity

- supratip prominence resulting in a convexity of the lower ⅔ of nasal dorsum

- Types: soft tissue versus cartilaginous defects
- Causes
  1. excess resection of nasal dorsum resulting in increased dead space and soft-tissue scarring resulting in a **paradoxical pollybeak deformity**
  2. inadequate removal of cartilaginous dorsum
  3. loss of nasal tip support
  4. insufficient lowering of dorsal septal borders
  5. shortened columella
  6. excessive excision of domes of alar cartilage
- Management Techniques
  1. supratip massage
  2. resect prominent tissue or scar for soft-tissue pollybeak deformities
  3 graft dorsum
  4. augment lower lateral cartilage
  5. steroid injections
  6. columellar struts to maintain desired projection and rotation

Retracted and Notched Ala

- notched or retracted intranasal tip
- Causes: excess lower lateral cartilage excision, excess vestibular skin removal (soft triangle), improper closure of a marginal incision
- Rx: auricular cartilage placed in lateral crura, composite graft if skin required, avoided by leaving 2–3 mm inferior alar rim

Columellar Defects

- **Retracted Columella**: deficient columella show resulting in an acute nasolabial angle (<90°), caused by excessive resection at base of columella or caudal septum; Rx: plumping graft at columella base, premaxillary implant cartilage, composite graft if skin required
- **Hanging Columella**: caused by persistent overly convex medial crura or abnormally elongated caudal septum

Saddling

- concavity of nasal dorsum from excess removal of nasal dorsum or an open roof deformity
- Rx: lateral ostomies to close open roof for mild deformities, dorsal implant or graft for more severe deformities

Nasal Obstruction

- transient nasal obstruction should be expected from mucosal edema and crusting for 2 weeks

- **Persistent Septal Deformity**: preoperatively may not have been obstructing; Rx: septoplasty
- **Intranasal Synechia**: more common with concurrent turbinectomies; Rx: prevented by avoiding mucosal injuries and placement of septal splints, managed with lysis with Z-plasty, intralesional corticosteroids, septal splints, or mucosal flaps
- **Narrowing of the Nasal Vault**: may occur from overresection of caudal end of upper lateral cartilage or lower lateral crura (**nasal tip pinching**), overaggressive infracture of nasal bones, high intercartilaginous incisions; Rx: revision with placement of spreader grafts

Tip Defects

- **Tip Ptosis**: caused by loss of tip support, unfavorable healing factors, uncorrected depressor septi, or false tip projection from surgical tip edema; Rx: reconstruct tip, address depressor septi
- **Bossae**: knob-like protuberances in the dome area caused by irregular scarring; Rx: add a small alar graft to opposite side or shave excision
- **Pinched Tip**: caused by excessive reduction of dome, failure to maintain intact rim strip, internal adhesions, or excision of vestibular skin

Midnasal Asymmetry

- Causes: subluxation or asymmetric removal of upper lateral cartilage, deviated septum
- Rx: onlay graft, septoplasty

OTOPLASTY

Introduction

Initial Evaluation

- determine auriculocephalic angle (auricular protrusion from the skull normally 20–30°) and distance of helical rim to mastoid skin (2.0–2.5 cm)
- evaluate slope of ear (normally ≈20° from vertical or approximately the slope of the nasal dorsum)
- measure vertical height (normally ≈6 cm)
- evaluate the auricular components and convolutions (helix, antihelix, scaphoid fossa, fossa triangularis, lobule)
- compare ear to opposite side

- evaluate for other associated defects (eg, branchial arch deformities, cleft palate, urogenital and cardiovascular malformations)
- prior to reconstruction consider CT of temporal bone (for microtia/anotia) and audiological evaluation
- preoperative photodocumentation
- Common Syndromes with Auricular Malformations: Treacher Collins Syndrome, Hemifacial Microsomia, Goldenhar Syndrome, Down Syndrome

Common Auricular Defects

- **Protruding Ear**: auriculocephalic angle >30°, caused from loss of formation of the antihelical fold or overgrowth or protrusion of the conchal cartilage
- **Cryptotia**: pocket ear deformity
- **Cup Ear Deformity**: constriction of the helix and scapha; Rx: requires unfurling of the helical rim by dividing the helical-scaphoid cartilage to allow for expansion followed by redraping of skin over the excess cartilage
- **Colobomata**: clefts
- **Lobule Deformities**
- **Microtia and Anotia**

Surgical Management

Correcting the Protruding Ear

- best age for reconstruction is at 5–6 years old when ear is at 85% of adult size and before the age of social stigmatization
- **Suture Techniques** (**Mustardé**): mattress sutures placed along the scapha through a posterior incision to create an antihelical fold with resulting reduction of the auriculocephalic angle, does not address conchal bowl, simple technique
- **Cartilage Sculpting Techniques**: reshaping (scoring, thinning) or splitting of auricular cartilage to weaken the cartilage surface to create a convexity for the antihelical fold
- **Farrior Technique**: combination suture and cartilage sculpting techniques, removes wedges of cartilage from posterior surface to allow bending to supplement the mattress sutures (additionally may place a wedge of cartilage anteriorly to help form the antihelical fold)
- **Concha Setback Technique** (**Conchomastoid Suture Technique of Furnas**): sutures the conchal bowl to the mastoid periosteum to reduce the auriculocephalic angle

Complications

- similar to Auricular Reconstruction (Hematomas, Infection, see p. 404)
- **Telephone Ear Deformity**: "telephone" appearance (upper and lower poles of the auricle protrude on anterior view); may occur from overcorrecting the middle antihelix, improper placement of mattress sutures, inadequate skin or cartilage excision; Rx: revision with possible skin excision and suturing the root of the helix to the temporal fascia
- **Malposition and Reprotrusion**: varied causes including suture splitting, overcorrection, and undercorrection; Rx: revision

THE AGING FACE

Blepharoplasty

Anatomy

- **Orbital Septum**: continuous with periosteum, houses orbital fat, barrier to both neoplastic and inflammatory invasion, upper eyelid septum fuses with **levator aponeurosis**, lower lid septum fuses with **capsulopalpebral fascia**
- **Medial Canthal Tendon**: constructed from medial heads of the pretarsal and preseptal muscles, attaches to the **lacrimal crest**
- **Lateral Canthal Tendon**: constructed from lateral heads of the pretarsal and preseptal muscles, attaches inside **orbital tubercle of Whitnall**
- **Superior Palpebral Sulcus**: formed from insertion of **levator aponeurosis** to the lid skin
- **Whitnall's Ligament**: dense fibrous connective tissue that functions as a fulcrum for levator muscle
- **Lockwood's Ligament**: dense fibrous connective tissue that functions as a fulcrum for lower lid
- Superior Tarsal Plate Height: 8–9 mm
- Inferior Tarsal Plate Height: 4–5 mm
- The East Asian Eye: the insertion of the levator aponeurosis may attach lower in the lid than in Caucasians resulting in a less developed superior palpebral sulcus (superior tarsal crease), the orbital fat tends to be more fibrous

Layers Above Eyelid Crease (Fig. 7–15)

1. skin (devoid of subcutaneous fat)
2. orbicularis muscle (comprised of orbital [outer, thicker], preseptal, and pretarsal [inner, thinner] bands)

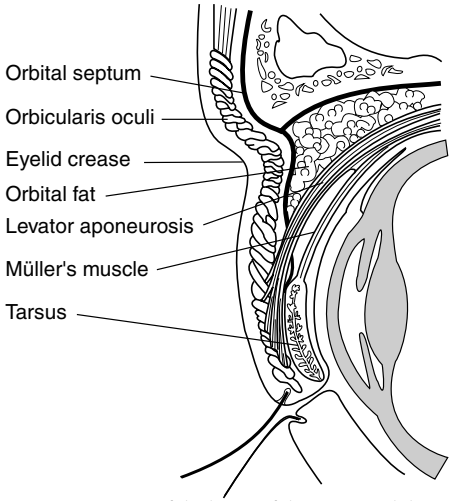

FIGURE 7–15. Cross-section of the layers of the upper eyelid.

3. orbital septum
4. orbital fat
5. levator aponeurosis
6. Mueller's muscle (sympathetic smooth muscle)
7. tarsal portion

Orbital Fat Compartments *(lies beneath orbital septum)*

1. Lower Medial: lighter color, denser, separated by inferior oblique muscle
2. Lower Central: darker than medial compartment
3. Lower Lateral: separated by fascial barrier
4. Upper Medial: separated by superior oblique muscle
5. Upper Central: medial to lacrimal gland (which replaces most of the upper lateral compartment)

Indications

- **Blepharochalasis**: rare condition of unknown cause, may be familial, characterized by recurrent attacks of lid edema resulting in thin, redundant eyelid skin and a sunken appearance from fat herniation, more common in young women, may consider excision of redundant skin once attacks abate
- **Pseudoherniation**: orbital fat protrudes through a lax orbital septum behind orbicularis muscle, baggy lids

- **Dermatochalasis**: acquired draping of excess skin over lids, usually from actinic damage, thins eyelid skin, allows for prolapse of orbital fat, progressive ptosis occurs with aging
- **Orbicularis Hypertrophy**: may be cause of residual bagginess, obvious with squinting

Blepharoplasty Does Not Correct

- crows feet (lateral rhytids) caused by the orbicularis muscle insertion to skin; Rx: temporal lift, collagen injection, botulism toxin injections, or peels
- asymmetric brow or brow ptosis; Rx: requires a brow lifting procedure
- malar or cheek pads (fluid); Rx: corrected with cheek implants or deep plane facelifts
- fine wrinkles and hyperpigmented lesions; Rx: consider a chemical peel or another resurfacing procedure

Causes of False Pseudoherniation of Fat

- prominent orbital rim
- allergic edema
- lacrimal gland ptosis
- orbicularis oculi hypertrophy
- malar or cheek bags
- myxedema
- festooning (hammock-like bags in the lower eye lid from folds in the orbicularis oculi, must consider hypothyroidism)

Evaluation

History

- Hx: family history of baggy eyes, fluid retention, thyroid disorders, allergic dermatitis, ophthalmic history (corrective lenses, dry eyes, tearing), collagen vascular diseases, previous keloids, bleeding disorders
- assess motivational factors for surgery
- preoperative photography (*as above*)

Ophthalmic Evaluation

- check visual acuity (Snellen eye chart), visual field tests, motility, ocular tension, measure scleral show, proptosis, fundoscopic exam
- **Normal Bell's phenomenon**: assesses adequate protection of the globe, evaluated by forcing the lids open and observing the upward rotation of the globe to protect the cornea

- **Schirmer Test**: Schirmer strip (filter paper) is draped over the lower lid margin (lateral to the cornea), the distance between the leading edge of wetness and the lid is measured after 5 minutes (abnormal is < 5 mm with topical anesthesia)

Evaluate Brow and Upper Lid

- examine for brow ptosis (brow should be at orbital rim in males or just above orbital rim in women), if present patient at risk of worsening ptosis with upper lid blepharoplasty (consider repositioning brow with brow lift procedures before blepharoplasty)
- the superior orbital sulcus (eyelid crease) should be <10 mm from lid margin and below bony margin of superior orbital rim (should also note symmetry)
- evaluate for fat herniation, skin redundancy, ptotic lacrimal gland, and muscle hypertrophy
- evaluate skin type, thin skin requires conservative resection to avoid a retracted or "hollow" look, thick skin results in an ill-defined superior orbital sulcus and requires considerable amount of fat and muscle excision
- determine presence and degree of ptosis by measuring the margin-reflex distance (MRD), the distance between the corneal light reflex and the upper lid margin (normally between 3.0–4.5 mm)
- access levator functioning by measuring the upper lid margin excursion in downward and upward gaze (normally >12 mm)
- evaluate for the presence of **lagophthalmos** (incomplete closure of eye) and **lateral hooding**

Evaluate Lower Lid

- the inferior orbital sulcus (eyelid crease) should be <5–6 mm from lid margin (a second crease may be located inferiorly corresponding to an area of tissue thickness)
- evaluate fat herniation (have patient look superiorly), skin redundancy, and muscle hypertrophy
- palpate inferior orbital rim, may imitate fat herniation
- **Lid Distraction Test** (**Snap Test**): tests lateral lower lid laxity; outwardly displace the lower lid and observe for a normal snap, if >10 mm displacement or if lid settles slowly patient at risk of ectropion or scleral show, patient requires a lid tightening procedure at the same time as the blepharoplasty
- **Lid Retraction Test**: tests lateral lower lid laxity; inferiorly displace the lower lid, if the puncta moves > 3mm indicates a lax canthal tendon and suggests the need of a tendoplication to avoid ectropion or scleral show

- examine for skin crepe (fine wrinkles), festooning, malar bags, and lateral orbital rhytids (crow's feet), which can **not** be corrected with blepharoplasty

Surgical Procedures

Brow Lift Procedures

- should be completed first
- (*see also* pp. 428–429)

Addressing the Upper Lid

- completed before addressing the lower lid
- mark estimated skin to be excised by bringing together redundant skin with forceps while patient is in an upright position (pinch technique), correct amount should begin to move the eyelashes upward but not move the lid margin, ideally lid should leave a slight (1–2 mm) lagophthalmos intraoperatively, lower incision begins at the superior lid crease (hooding may be addressed by extending the incision lateral to the orbital rim)
- a strip of orbicularis muscle may be excised in the heavy lid with thick skin, allows for better definition of lid crease
- open the orbital septum and excise the pseudoherniated fat (amount typically determined by excising the fat that extrudes out of the septum when placing gentle digital pressure on the globe)
- ptotic lacrimal gland may be addressed by plicating to the periosteum
- accentuation of the superior eyelid crease may be considered by plicating the skin margin of the inferior skin incision to the levator aponeurosis

Addressing the Lower Lid

- Approaches: skin-muscle flap (most common, more avascular dissection), skin flap (allows for independent modification of skin and muscle, more skin trauma, increased risk of vertical eyelid retraction), and transconjunctival (indicated for excessive fat herniation with minimal skin redundancy, no external scar, avoids eyelid retraction, may be combined with a chemical peel or laser resurfacing to address rhytids)
- excise redundant skin (marked similarly to the upper lid)
- excise orbicularis oculi if needed
- excise pseudoherniated fat (medial compartment typically requires more aggressive excision)

- **Repositioned Orbital Fat Technique**: releases the orbital fat by opening the orbital septum and allowing the pseudoherniated fat to be repositioned inferiorly with the suborbicularis oculi fat (SOOF), allows for a more subtle change (avoids hollowing of the orbit)

Postoperative Care

- cold compress
- limit physical activity
- avoid downward gaze, may cause skin flap to slide
- contacts may be used after postoperative day 10

Complications

- **Milia**: inclusions cysts, most common complication; Rx: pinpoint cautery and marsupialization
- **Hematoma**: more common in lower lid; Rx: consider reopening for major hematomas with removal of clot and controlling bleeding; for minor hematomas may evacuate through incisions or wait 7–10 days to allow for liquification then aspirate, antibiotics, pressure dressings
- **Blindness**: can occur up to 5 days postoperatively, usually from retrobulbar hematoma from lower eyelid fat removal (intraocular pressure >80 mm Hg increases risk of blindness), prior to ocular symptoms typically presents with pain; Rx: urgent decompression with a lateral canthotomy and orbital decompression
- **Subconjunctival Ecchymosis**: usually only a temporary cosmetic problem; Rx: self limiting, typically resolves in 3 weeks
- **Lagophthalmos**: normally present for a short period postoperatively, persistent lagophthalmos is caused by excess upper lid excision or sutured orbital septum with skin closure; Rx: often resolves, ophthalmic drops, night taping of lids, consider a full-thickness skin graft for persistent lagophthalmos
- **Scleral Show**: 1 mm considered a normal variant, caused by excess lid reduction or excess laxity, may resolve with edema in 1–2 weeks; Rx: postoperative squint exercises, upward massage, superotemporal taping
- **Ectropion** (eversion): caused by excessive lower lid skin or muscle excision, lid contracture, lateral laxity; Rx: initially treat conservatively with tape closure, cold compress, squinting exercises, and perioperative corticosteroids; if no resolution consider lid lengthening with a full-thickness skin graft from the upper eyelid (best color match) or a full-thickness skin graft from postauricular skin; if from lateral lid laxity a horizontal lid-shortening procedure is required
- **Ptosis**: caused by levator aponeurosis dehiscence from edema, hematoma, excess cautery, or infection; Rx: intraoperative

aponeurosis injury should be corrected immediately, transient ptosis may be caused by edema (resolves within days), may consider a Mueller's muscle resection or other ptosis correcting procedures
- **Pseudoepicanthal Folds (Webbing)**: caused by extending incision too medial (<1 mm of punctum); Rx: Z-plasty
- **Epiphora**: caused by eversion of punctum, lacrimal pump injury, disregard for eyelid laxity, or removal of too much skin beneath punctum; Rx: transient epiphora from edema typically resolves within the first few days; persistent epiphora may be addressed with a punctoplasty, horizontal eyelid shortening procedure, or dacryocystorhinostomy
- **Dry Eyes**: avoided by assessing tearing function (eg, Schirmer's test); Rx: topical wetting agents and ophthalmological ointments at night
- **Ocular Palsy**: may be caused by too much fat excision in the nasal pocket causing superior oblique or inferior rectus injury; Rx: diplopia usually resolves in 6 months, may release scar tissue
- **Conjuctival Lymphedema**: skin and muscle flaps may compromise lymphatics; Rx: usually resolves in 1–2 months
- **Chemosis** (edema of the conjunctiva): affects bulbar conjunctiva; Rx: usually resolves in 6 weeks, may consider blephamide ophthalmic drops
- **Persistent Lower Fat Pocket**: lateral compartment is the most common; Rx: revision

Rhytidoplasty, Forehead Lifts, Brow Lifts, and Liposurgery

Anatomy

Layers of Facial Planes

- skin
- subcutaneous fat
- SMAS (*see below*)
- mimetic muscles
- deep facial fascia
- deep elements (eg, facial nerve, parotid duct, buccal fat)

Facial Nerve Relationships

1. **Frontal Branch**: courses along line from earlobe to zygomatic arch at point midway between tragus and lateral canthus, over zygomatic arch then penetrates deep temporal fascia into the undersurface of the superficial temporal fascia

2. **Zygomatic Branch**: courses 1 cm below the zygomatic arch in a superior and medial direction from tragus to lateral canthus
3. **Marginal Branch**: exits the parotid 1 cm below the mandibular angle, deep to the platysma within the submandibular gland fascia (superficial to the facial vein)

NOTE: the facial nerve innervates facial muscles deeply except the buccinator, levator angular oris, and mentalis muscles

Superficial Muscular-Aponeurotic System (SMAS)

- translates facial muscle action to dynamic action in the facial skin
- continuous with platysma, superficial temporal fascia, perioral, nasolabial, and periorbital muscles
- adherent to **parotidomasseteric fascia** (continuation of deep cervical fascia in the neck and deep temporal fascia above zygomatic arch), dermis of the cheek, and zygomatic arch
- located above parotid fascia, facial nerve, and facial artery
- plicated for lift in rhytidoplasty
- **Melolabial Crease** (**Nasolabial Fold**): insertion of muscles from the zygoma and the SMAS

Osseocutaneous Retaining Ligaments: Skin Attachments to Bone

- Zygomatic Ligament: attaches from zygomaticomaxillary suture (McGregor's patch) to overlying malar skin
- Mandibular Ligament: attaches from anterior mandible to overlying parasymphyseal skin

Fascia-Fascia Retaining Ligaments: Skin Attachments to Fascia

- Parotid Ligament: attaches to overlying skin
- Masseter Ligament: supports soft tissue of the medial cheek superiorly over the mandibular body (attenuation results in jowls)

Malar Fat Pad

- triangular-shaped fat pad, superficial to SMAS, adherent to overlying skin
- volume of fat pad does not change with relative changes in body adiposity
- with aging loss of malar support results in inferior migration of the malar fat pad about the melolabial crease resulting in the illusion of a deepened melolabial crease

Evaluation

History

- history of sun exposure (most common factor in premature aging from actinic damage from **UVA**)
- smoking history (increases skin slough 12×)
- history of recent weight loss (if patient is losing weight should wait until weight is stable)
- complete medical history including thyroid disorders, allergic dermatitis, collagen vascular diseases, previous keloids, and bleeding disorders

Diseases of Premature Aging

- **Cutis Laxa**: autosomal recessive, associated with hernias, emphysema, aneurysms; rhytidoplasty **helpful**
- **Progeria**: autosomal recessive, associated with growth retardation, early atherosclerosis; rhytidoplasty **not helpful**
- **Werner Syndrome**: autosomal recessive, high pitched voice, diabetes, osteoporosis; rhytidoplasty **not helpful**

General Assessment

- ideal candidates are in their 40–50's, good bony framework, strong cheekbones and chin, normally placed hyoid bone, thin face, good elasticity, and stable weight
- screen for precancerous lesions
- must have favorable medical and psychological conditions
- preoperative photography (*as above*)

Middle, Lower, and Neck Assessment

- evaluate for well demarcated mandibular line and jowling (loss on contour at the angle of the mandible)
- assess for deepened melolabial crease, chin ptosis, increased distance from ciliary margin to malar crescent
- **Turkey Gobbler Deformities**: loss of cervicomental angle (>120°) from laxity in platysma (does not decussate across midline), addressed with liposuction and approximating platysma at midline
- **Ptotic Submandibular Glands**: addressed with platysma slings

Rhytidoplasty Surgical Techniques

- mark patient in the upright position
- liposuction may be used for cervicofacial, submental, periparotid, and jowl regions, allows for shorter flaps and less risk of hematomas
- chemical peels should not be completed concurrently

- laser resurfacing techniques may be considered 6–12 months postoperatively

Superficial Plane Technique

- original technique described in late 1960s
- addresses redundant skin only
- Advantages: simple
- Disadvantages: does not address deep elements, interrupts vascular perforators to skin, higher risk of skin sloughing

Deep Plane Technique (Hamra, 1990)

- subSMAS dissection from midface to medial melolabial crease, elevates cheek fat pad attached to skin by zygomaticus muscles, provides vertical lift
- Advantages: addresses melolabial crease and midface (malar fat pad) by separating SMAS from fixed bony attachments of zygomaticus muscles, provides an excellent jaw line, preserves structural integrity of facial blood supply
- Disadvantages: lengthy anterior dissection, persistent malar edema

Composite Technique

- combines a deep plane technique with an orbicularis oculi muscle flap
- Advantages: repositions zygomaticus major to original position, healthy and thicker flap (less chance of sloughing, especially with smokers), more avascular dissection
- Disadvantages: persistent malar edema, requires some blind blunt dissection at curve of maxilla, unnatural tension of overlying skin of temple area

Subperiosteal Technique

- variations of subperiosteal undermining of orbital rim, zygomatic arch, maxilla, and zygoma body
- Advantages: maintains perforators to skin, excellent repositioning of lateral canthus, addresses midface without causing unnatural tension on skin (places traction on periosteum)
- Disadvantages: causes prolonged facial edema, increases intermalar distance, limited improvement of jowling, technically more challenging

Multiplane Technique

- combines an extended SMAS dissection and a deep plane lift (allows dissection in subcutaneous plane to release zygomatic ligaments and skin of midface)

- Advantages: separate dissection of SMAS allows better redraping of skin, more skin may be excised than deep plane lifts, may address melolabial crease better
- Disadvantages: similar to composite techniques, more compromise of vascular supply than deep plane lifts with separate SMAS and skin dissections
- **Triplane Technique**: 3 levels of dissection (subcutaneous in temple and neck, sub-SMAS in lower cheek, subperiosteal in midface), allows maximal correction of jowl while limiting the degree of sub-SMAS dissection to lower cheek, protects the facial nerve

Addressing the Melolabial Crease

- Deep Plane and Multiplane Rhytidoplasty (*see above*)
- Direct Fold Excision
- Melolabial Crease Liposuction
- Collagen Injection

Addressing the Neck

- redrape neck skin for minor defects
- "best necklift is a facelift"
- liposuction to sculpt and redefine cervical lines (usually approached from submental incision)
- address platysma with a submental tuck (reconstruct midline by anchoring to the mandible) or vertical excision with midline plication
- correct ptotic submandibular glands with a platysma sling

Forehead and Brow Surgical Procedures

- incision of the procerus, corrugator, and frontalis muscles creates a smoother look (do not excise)
- **Forehead Arteries**: superficial temporal artery (from ECA), zygomatico-orbital artery (from superficial temporal artery), supraorbital artery (from ophthalmic artery), and supratrochlear artery (from ophthalmic artery)
- **SCALP**: **S**kin, sub**C**utaneous, galea **A**poneurosis, **L**oose areolar tissue, **P**ericranium
- Causes of Horizontal Rhytids in the Forehead: frontalis muscle, procerus muscle, and the vertical segment of glabellar lines (corrugator supercillii muscle causes vertical rhytids)
- Indications for Forehead and Brow Lifts: elevate ptotic and asymmetrical brows, address upper eyelid hooding, reduce glabellar and forehead creases

Techniques

- **Coronal Lift**: standard for generalized brow ptosis (affords best exposure), incision placed above hairline, dissection in supraperiosteal and subgaleal plane, risk of raising anterior hairline, contraindicated with male pattern baldness or high frontal hairlines in females, does not correct brow asymmetries
- **Pretrichal Lift**: incision at anterior hairline, indicated for high hairlines (especially in women) and long vertical forehead height
- **Midforehead Lift**: incision above brow, indicated for expansive foreheads (balding hair) especially in men, candidates should have prominent forehead creases with thin, dry skin to hide the skin incision, indicated for primary brow ptosis but not for forehead rhytids, risk of unsightly scar
- **Direct Brow Lift**: incision directly above brow, useful for unilateral brow ptosis, does not allow resection of muscle, effect may be temporary, scar may be unsightly
- **Endoscopic Forehead Lift**: incisions in coronal scalp, less risk of nerve injury
- **Supratarsal Lift**: access through upper blepharoplasty incision, suspends brow directly

Liposurgery

- hereditary and hormonal factors, diet, and exercise contribute to localized adipose tissue which may persist despite exercise and dieting
- adipocytes increase in number until puberty, weight gain in the adult is secondary to hypertrophy of the adipocyte and **not** an increase in the number of cells

Lipoinjection

- 75% of adipocytes that are acquired atraumatically and injection with large cannulas (>18 gauge) will persist after 1 year
- Common Uses: cheek hollows, angle of the mandible, glabella frown line, oral commissure

Liposuction

- can **not** be used for dermal defects such as rhytids
- ideally should maintain a minimum of 2 mm of subdermal fat
- Advantages: quick, short postoperative course, minimal risk of nerve and vascular damage, minimal scarring, reduced soft tissue trauma
- Disadvantages: in general not used for generalized weight loss
- Common Uses: improve contour of submandibular (jowls), submental (wattles), cheek, malar, and parotid regions, defat pedicled or free flaps, lipoma excision

- Contraindicated: collagen vascular diseases, bleeding disorders, endocrine disorders

Complications

- **Temporal Alopecia and Unfavorable Hair Lines**: avoid poorly placed incisions or tension; Rx: typically temporary, for persistence may consider scar excision for affected region or punch grafting
- **Hematoma**: most common early complication (15%) in rhytidectomy and suction lipectomy, *see* p. 423 *for management*
- **Skin Slough and Necrosis**: higher risk for smokers, longer and thinner flaps, and hematomas; prevented by maintaining dermal-subdermal plexus (avoid overaggressive liposuction) and a tension-free closure; Rx: topical debridement if required, topical and oral antibiotics, should allow healing by secondary intention (requires several months of wound care)
- **Pixie Earlobe**: elongated earlobe from excessive inferior tension at lobule; Rx: V-Y advancement flap
- **Chronic Pain**: may be secondary to neuromas or nerve entrapment by a buried suture; Rx: nerve block, excision of neuroma, removal of buried suture
- **Hyperpigmentation/Hypervascularity**: thin-skinned patients are higher risk for telangiectasia development, pigment changes may occur from postinflammatory melanocyte activation; Rx: usually resolve, sunblock, corticosteroid creams, may consider chemoexfoliation techniques to blend skin
- **Nerve Injuries**: in general, nerve injury may be the result of transection, electrocautery injury, traction and compression injury (neuropraxia), and infection
  1. Greater Auricular Nerve: most common (7%), most commonly injured below tragal notch at anterior border of SCM, protect by leaving fascia overlying SCM
  2. Frontal Branch of Facial Nerve: most commonly injured as it lies superficially over the zygomatic arch
  3. Marginal Mandibular Branch of Facial Nerve: most commonly injured at anterior margin of flap elevation over mandible
  4. Supraorbital and Trochlear Branches: results in paresthesia and itching, avoided with dissection in the subgaleal plane
- **Cobra Neck Deformity and Other Contour Deformities**: cobra neck occurs from excess liposuction in anterior neck region; Rx: digastric and platysma plication or dermal grafts, select depression may be injected with autogenous fat
- **Infection**: in general rare due to abundant vascular supply

- **Postoperative Nausea:** common in forehead lifts; Rx: self limiting, antiemetics

Chin and Malar Augmentation

Evaluation

- chin should be at or just behind a line perpendicular to Frankfort horizontal and the vermilion border of lower lip (or nasion)
- must consider effects from adjunctive procedures (rhinoplasty, blepharoplasty, liposuction)
- ideally the malar region should appear full and should not fall below 5 mm posterior to melolabial groove
- evaluate microgenia (diminished chin eminence requiring augmentation or sliding genioplasty) versus retrognathia (retrodisplaced mandible requiring sagittal split osteotomies)

Surgical Procedures

Chin Implants

- chin implants typically gain up to 70% projection (after settling and bone resorption)
- may insert chin graft externally through submental crease or intraorally, graft is placed subperiosteally or above periosteum
- Complications: infection, displacement (must be removed and replaced), improper size (wait about 3 months), mental nerve injury, extrusion

Sliding Genioplasty

- moves anteriormost mandible forward
- indicated for more severe cases (eg, retrognathia, insufficient vertical mandibular height, hemifacial atrophy, failed implant)
- more difficult, requires intraoral incision, mandibular osteotomy, advancement of segment with plates or wires
- Complications: lip incompetence (if mentalis is not approximated correctly), tooth injury, mental nerve injury

Malar Implants

- may be approached intraorally (canine fossa) or subcilliary (more accurate)
- Complications: similar to chin implants

Facial Resurfacing

Introduction

Skin Layers

- Epidermal Layers: stratum corneum (dead cells), granular layer, prickle layer, basal layer
- Dermis Layers: papillary dermis, reticular dermis
- Subcutaneous Tissue: contains fat and fibrous tissue

Peel Postoperative Healing

- 5 Days: epidermis regenerates
- 1 Week: epidermis loosely attached to dermis
- 2 Weeks: new collagen deposits, youthful look
- 1 Month: pigment returns
- 6 Months: epidermis at normal thickness
- 10 Months: dermis normalizes

Chemoexfoliation

- Indications: postacne scars, scar revision, actinic keratosis, seborrheic keratosis, rejuvenate wrinkled and aged skin, pigment irregularities, address photoaging, texture, decrease oiliness, size of pores
- Depth of Penetration Factors: chemoexfoliation agent, skin thickness, sebaceous gland density, use of retinoic acid or lactic acid, use of degreasing agents or pre-peel agents, occluded or unoccluded
- Contraindications: active herpetic lesions, pustular acne, history of keloids, collagen diseases
- Postoperative Care: avoid sun for 3 months (sunscreen), skin emollients to keep moist, resume topical tretinoin

Preoperative Planning

- Assess Skin: darker and oilier skins have higher risk of pigment changes; evaluate for keloids, precancerous lesions, and herpetic lesions
- consider agents that remove keratin to enhance peels (eg, Lac-Hydrin, retinoic acid)
- curette actinic keratosis and seborrheic keratosis before peel
- must remove surface oils before peeling (acetone)
- preoperative photodocumentation

Retinoic Acid

- corrects epidermal dysplasia, reduces actinic keratosis, increases vascularization, disperses melanin granules

- Indications: freshening effect, enhance other peels
- Advantages: can be completed at home, does not induce skin injury
- Disadvantages: may cause drying and irritation, limited effect

Trichloroacetic Acid Peels (TCA)

- Indications: superficial and medium-depth peels (depending on percentage of solution)
- Mechanism of Action: coagulation necrosis limits injury to epidermis and some papillary dermis (up to the upper reticular dermis)
- Advantages: less risk of hypopigmentation, no significant systemic toxicity
- Disadvantages: deep peels less effective than phenol peels

Phenol Peels

- Indications: deep dermal peel
- Mechanism of Action: liquification necrosis resulting in deep dermal injury (dilution results in deeper penetration)
- Advantages: addresses severe pigmented lesions and deep wrinkles, typically requires only one-time therapy
- Disadvantages: systemic effects (requires preoperative evaluation of cardiac, renal, and hepatic function prior to therapy and cardiac monitoring to evaluate for phenol toxicity, *see below*), can **not** be used in conjunction with rhytidoplasty (increases chance of skin necrosis), requires premedication sedation, longer healing process
- Contraindications: areas treated with external beam radiation (may not have adnexal structures); renal, cardiac, or hepatic disease
- Baker's Solution: chemical or peel solution; contains liquid phenol (88% phenol concentrate), distilled or tap water, liquid soap (aids in emulsifying the solution), croton oil (enhances the keratolytic and penetrating action)
- consider tape occlusion for deeper wounding or without taped occlusion to avoid increased risk of scarring
- avoid applying to more than 25% of face and wait 10 minutes between peels to avoid cardiac toxicity
- postoperative swelling occurs for 2 days and crusting for 1 week

Dermabrasion

- limit abrasion to the papillary dermis (pinpoint bleeding), must avoid the parallel white fibers of the reticular layer which houses adnexal structures required for healing (injury to the epidermis and papillary dermis does **not** result in scarring)

- preoperative topical tretinoin 2 weeks prior to dermabrasion and postoperative occlusive dressings will shorten healing from 7–10 days to 5–7 days
- freezing skin prior to abrading allows for a rigid surface and preservation of anatomical markings
- Indications: acne scars, adjunctive procedure with irregularization techniques (scar revisions), actinic keratosis, seborrheic keratosis, facial wrinkles, tattoos, rhinophyma
- Advantages: quick, local anesthesia, may be repeated, more precise level of depths
- Disadvantages: higher risk of scarring, more bleeding
- Risks of Bad Outcome: keloid, hypertrophic scar history, vitiligo, active herpes
- Postoperative Care: occlusive dressing, consider short course of oral corticosteroids to reduce edema, acyclovir for 5 days, sunblock (especially in hyper/hypopigmented skin), resume topical tretinoin

Laser Resurfacing

- adjustment of the power (watts) determines the energy delivered to tissues
- power density is determined by power (watts) divided by spot size (area)
- the concept of **selective thermolysis** is determined by the absorbance of the skin's constituents, specifically the chromophores, oxyhemoglobin, and melanin, which have their own absorbance spectra; water absorbs infrared laser energy which causes the thermal effect of CO_2 lasers
- Indications: acne scars, scar revisions, actinic keratosis, seborrheic keratosis, facial wrinkles, rhinophyma, tattoos, cutaneous vascular lesions, keloids

Common Laser Types

- **CO_2 Laser**: invisible beam (aiming beam must be used in conjunction with laser), primarily absorbed by water, excellent for cutting, coagulation, or ablating (depending on area of focus)
- **Nd:YAG Laser**: near-infrared beam, invisible (requires an aiming beam), primarily absorbed by pigmented tissues, deeper penetration, more scatter than CO_2 lasers, may use a fiberoptic carrier, indicated for cutaneous lesions (port-wine stains, telangiectasias, hemangiomas, etc)
- **KTP Laser**: visible laser, primarily absorbed by oxyhemoglobin, may use a fiberoptic carrier, indicated for cutaneous lesions and as a cutting tool

- **Argon Laser**: visible, broad-band blue light, primarily absorbed by oxyhemoglobin, depth of penetration between CO_2 and Nd:YAG lasers, similar indications as the Nd:YAG laser
- **Flashlamp-excited Dye Laser**: visible, yellow light, vascular sensitive, less scarring and hypopigmentation than Nd:YAG and argon lasers, indicated for cutaneous vascular lesions

Lasers of Choice for Tattoo Colors

- **Purple/Violet**: Q-switched ruby (694 nm)
- **Green/Blue**: Alexandrite (755 nm)
- **Black**: Q-switched Nd:YAG (1064 nm)
- **Red**: Frequency-doubled Nd:YAG (532 nm)

Complications

- **Phenol Toxicity**: cardiac toxicity (must monitor for arrhythmias), neural (muscle weakness, slowed respiration), hepatic and renal toxicity, headache, nausea, hypotension, coma, and death; Rx: avoid by evaluating cardiac, renal, and hepatic function prior to phenol peel; avoid single large surface areas of peel; preoperatively should hydrate patient; if occurs, remove peel and address arrhythmia, hypotension, etc
- **Hypopigmentation**: normal sequela after deep peels (typically resolves), may result from melanocytic injury from deep dermabrasion; Rx: sunscreen, makeup, consider tattooing if severe, psoralens (increases pigmentation)
- **Hyperpigmentation**: more common in superficial peels; Rx: sunscreen, consider tretinoin, corticosteroid, and hydroquinone creams
- **Melasma**: hyperpigmention in face (common with estrogen, birth control pills, pregnancy)
- **Milia**: small epidermal cysts, common 1 month postoperatively; Rx: mild abrasive cleaners, unroofing, tretinoin therapy
- **Scarring**: higher risk for deep peels, perioral and chin peels, keloid formers; Rx: compression, massage, corticosteroid injections, silicon gel sheeting
- **Ectropion**: may occur with phenol eyelid peels
- **Prolonged Erythema**: should resolve after 1 month; Rx: topical corticosteroids and tretinoin for prolonged erythema
- **Herpes Simplex Outbreak**: reactivation from quiescent herpetic infection (HSV-2) in the trigeminal ganglion, occurs within 24 hours; Rx: perioperative high-dose acyclovir for prophylaxis, topical and oral acyclovir for active outbreak

- **Telangiectasias**: may become more prominent after peel; Rx: electrocoagulation/laser
- **Persistent Rhytids**: may repeat peel

CLEFT LIP AND PALATE

Introduction

Anatomy

- **Primary Palate (Premaxilla)**: anterior to incisive foramen, formed first, wedge shaped, houses four incisors
- **Secondary Palate**: non-premaxilla portion of hard palate and the soft palate

Soft Palate Muscles

- Tensor Veli Palatini: (CN V_3) opens auditory tube and depresses and tenses soft palate
- Levator Veli Palatini: (pharyngeal plexus) elevates palate
- Musculus Uvulae: (pharyngeal plexus) moves uvula upward and forward
- Glossopalatine: (pharyngeal plexus) draws palate down, narrows pharynx
- Palatopharyngeous: (pharyngeal plexus) draws palate down, narrows pharynx

Embryology

Frontonasal Prominence

- first phase (**4–5 weeks**)
- medial
- anterior labial component → philtrum
- medial component → midline alveolus
- posterior component → palate anterior to incisive foramen (**primary palate**)
- also contributes to nasal anatomy

Maxillary Prominence

- second phase (**8–9 weeks**)
- lateral
- **secondary palate**
- upper jaw
- fusion begins rostral → caudally beginning at incisive foramen

Mandibular Prominence: lower jaw

Epidemiology

- second most common malformation (club foot is the most common)
- cleft lip and palate occurs in 1/1000 births, cleft palate occurs in 1/2000 births
- Risks: Asians, Native Americans, males have more cleft lips but females have more isolated palates
- 45% both cleft lip and palate; 30% secondary palate only; 25% lip only
- cleft lips (with or without associated cleft palate) and isolated cleft palates occur in distinct genetic lines

Evaluation and Associated Problems

1. Define Type of Defect

- unilateral, bilateral, median
- complete (extension into nasal floor) or incomplete (submucosal)
- primary (anterior to incisive foramen) or secondary (posterior to incisive foramen)

2. Examine for Other Defects

- nasal deformities
- facial defects (telecanthus, maxillary/malar hypoplasia)
- otologic defects (abnormal pinna, atresia)
- facial nerve paralysis
- synostoses

3. Determine if Syndromic

Genetic Evaluation

- isolated cleft palates have 8% association with a syndrome
- **Stickler Syndrome** (autosomal dominant): cleft palate, retinal detachment, cataracts, early arthritis
- **Treacher Collins Syndrome** (autosomal dominant): cleft palate, malformation of the malar and other facial bones, eyelid colobomas, middle ear ossicular abnormalities
- **Pierre Robin Sequence**: micrognathia, cleft palate, glossoptosis
- **Apert Syndrome** (autosomal dominant): cleft palate, acrocephaly, fused digits, stapes fixation
- 200+ other syndromes

Teratogens: ethanol, thalidomide

Environmental: maternal diabetes, amniotic band syndrome

4. Evaluate Feeding Assistance

- often have difficulty with a seal requiring a special nipple (preemie, Lamb's, Mead-Johnson cross-cut, and McGovern's nipples)
- may require frequent rests and burps
- may benefit from a palatal prosthesis

5. Evaluate Otological Disease and Audiology

6. Speech Evaluation

7. Plan Surgical Repair

- 3 months: tip rhinoplasty, cleft-lip repair, close nasal floor, myringotomy tubes
- 1 year: palate repair
- 5 yrs: columellar lengthening (bilateral clefts)
- 8–16 yrs: orthodontal
- 10 yrs: alveolar cancellous bone grafts
- 14 yrs: definitive rhinoplasty and orthognathic surgery

Cleft Lip

Unilateral Cleft Lip Defect

- orbicularis muscle is directly superiorly in complete clefts or hypoplastic in incomplete clefts
- floor of nose communicates with oral cavity
- maxilla is hypoplastic on cleft side
- lower lateral cartilage is inferior on cleft side
- nasal ala is retro-displaced on cleft side
- nasal columella is displaced toward normal side
- dome is lowered, horizontal nares on cleft side
- cartilaginous nasal septum and nasal spine are deflected away from cleft
- alveolar defect passes through developing dentition

Bilateral Cleft Lip Defect

- floor of nose communicates with oral cavity bilaterally

- central part of alveolar arch is rotated forward and upward
- prolabium skin for the lip is underdeveloped
- central lip has no muscle or vermilion
- nasal columella is short, nasal tip is widened, cartilaginous nasal septum and nasal spine are deflected forward

Treatment

Lip Adhesion

- completed at 2–4 weeks for severe clefts
- reduces tension when definitive repair is completed
- produces an incomplete cleft
- scar band may interfere with definitive repair
- definitive repair is postponed to 4–6 months

Cleft Lip Repair

- repair at 10–14 weeks (**rules of 10**: >10 weeks, >10 lb, Hbg >10)
- correct cupid bow, approximate orbicularis oris muscle, symmetrical repair of vermilion, create nasal floor and vestibular sill, placement of nasal alar base and columella
- bilateral defects may be staged within months

Common Techniques

- **Millard Repair**: rotation-advancement technique for unilateral lip repair, limited tissue loss, reconstructs the philtrum and nasal sill, may be difficult to close large clefts
- **Randall-Graham**: utilizes the interposition of triangular flaps
- **Rose-Thompson**: straight line repair, simple design, favorable suture line, risk of vertical line contracture, difficult to close large clefts

Cleft Palate

Associated Defects

Eustachian Tube Dysfunction

- hypoplastic levator and tensor veli palatini (ineffective eustachian tube dilator) muscles → eustachian tube more horizontal, hypercompliant allows for reflux
- leads to chronic otitis media

Velopharyngeal Insufficiency

- incomplete closure of soft palate against the posterior pharyngeal wall

- Sx: hypernasal speech, nasopharyngeal regurgitation
- Rx: coronal pattern pharyngoplasty, sagittal pattern pharyngeal flap, circular pattern pharyngeal flap, posterior wall augmentation

Submucosal Clefts

- a minor form of a secondary cleft palate defect causing a midline diastasis of the levator muscles, mucosa remains intact, deficient musculature of the palate causes a blue streak (zona pellucida)
- associated with a bifid uvula and loss of posterior nasal spine (palatal notch)
- higher risk of Velopharyngeal Insufficiency after adenoidectomies

Treatment

- repair at 10–14 months (after lip repair)
- may require orthopedic movement of premaxilla or to realign maxillary segments

Common Techniques

- **V-Y pushback**: indicated for incomplete cleft, provides additional palate length
- **Two Flap Palatoplasty**: indicated for complete clefts, adequate closure of the cleft alveolus
- **Four Flap Palatoplasty**: two flap technique, however uses short incomplete clefts
- **Schweckendick's Primary Veloplasty**: closes only soft palate
- **Furlow Palatoplasty**: opposing Z-plasty, elongates soft palate

CHAPTER

8

Head and Neck Trauma

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continued

EVALUATION OF THE HEAD AND NECK TRAUMA PATIENT

Resuscitation (ABCs)

Airway and Cervical Spine

- anoxia can result in death in 4–5 minutes
- establish airway without mobilizing cervical spine (immobilize neck, maintain axial traction, avoid hyperextension)
- do **not** precipitate an airway crisis, any attempt at endotracheal intubation should also have a backup plan for an emergent surgical airway
- initially suction blood, clots, and secretions from the oropharynx (remove foreign bodies and teeth)
- overcome pharyngeal collapse with forward traction of the tongue, jaw lift, chin lift, or reduction of an unstable mandible or maxillary fracture
- masked ventilation may provide adequate oxygenation until able to secure an airway
- **Nasopharyngeal or Oral Airway**: indicated for a conscious or obtunded patient or as a temporary airway for an obstruction at the level above the tongue base and nasopharynx (nasal airways are better tolerated in conscious patients)
- **Nasotracheal Intubation**: generally better tolerated in an awake patient and can be accomplished without extension of the neck, intubation approach of choice for severe mandibular fractures or oral obstruction
- **Oral Intubation**: has a slightly higher risk of cervical spine injury; however, should be considered for patients with midfacial or basilar injuries; Jackson sliding laryngoscope allows for better visualization in difficult intubations
- **Fiberoptic Intubation**: should be considered in any potential "difficult airway"
- **Needle Cricothyrotomy**: establishes a temporary airway until a more secure surgical airway can be established (may be maintained for 30 minutes until hypoventilation results in toxicity), a #12 or larger intravenous catheter is placed through the cricothyroid membrane, the needle is removed leaving the sheath which is connected via intravenous tubing to oxygen (1 second of oxygen injection, 4 seconds of exhalation)
- **Cricothyrotomy**: enter airway via a vertical skin incision and a horizontal incision through the cricothyroid membrane with placement of an endotracheal tube
- **Urgent Tracheostomy**: may be considered in an emergent situation in pediatric patients, laryngeal trauma, or tracheal disruption (laryngeal blunt trauma may require a surgical airway to avoid worsening the airway; however, penetrating laryngeal trauma may undergo fiberoptic endotracheal intubation due to lower risk of laryngeal skeletal injury)

Breathing

- severe head trauma commonly results in decreased respiratory drive (narcotics, alcohol, and other medications may also depress respiratory drive)
- after establishing a secure airway, ease of ventilation and maintenance of oxygenation should be evaluated
- hypoventilation may be caused by tension or sucking pneumothoracies, hemothorax, or severe pulmonary contusions
- tension pneumothorax occurs with a tear in the pleura or tracheobronchial tree resulting in escape of air without means of egress; Rx: needle thoracocentesis and chest tube placement
- sucking pneumothorax results from a larger defect in the chest wall allowing exchange of air through opening; Rx: occlusion of defect with positive pressure ventilation

Circulation and Hemorrhage Control

- identify and control hemorrhage with direct pressure or packing (severe oral bleeding that does not resolve with pressure may require oral packing with rolled gauze after establishment of an airway)
- essential to recognize shock (inadequate perfusion of end organs) by tachycardia, hypotension, skin pallor, lethargy, decreased urine output; assess class of hemorrhage (*see* Table 8–1)
- adult blood volume is approximately 7% of total body weight (5 liters in a 70 kg man)
- establish circulation with 2 large-bore intravenous lines (minimum 18-gauge), resuscitate with crystalloid solutions (normal saline or lactated Ringer's solution) initially with 2 liters in adults and 20 ml/kg in children
- replacement of blood loss with crystalloid solution requires 3 times the volume of the estimated blood loss
- packed red blood cells are indicated for blood volume >30% (hemorrhage class III and IV), specified blood should be utilized (available usually <15 minutes), for emergencies Type O blood ("universal donor") may be used (Rh negative for females)
- for severe venous injuries may place in Trendelenburg position to avoid air embolism (also appropriate for shock)

Disability

- determine Glasgow Coma Scale (GCS, *see* Table 8–2)
- GCS <8 suggests severe head injury; 9–12 moderate head injury, >13 mild head injury

TABLE 8–1. Hemorrhage classes

Class	Loss of Blood	Symptoms	Treatment
Class I	10% (500 cc)	asymptomatic	maintain urine output of 30–60 cc/hour
Class II	20% (1000 cc)	mildly anxious, narrowed pulse pressure	replace blood loss with crystalloid fluids (3× estimated blood loss)
Class III	30–40% (1500–1700 cc)	tachycardia, diaphoresis, hypotension, confusion	rapidly bolus with crystalloid fluids, replace blood loss with packed red blood cells
Class IV	>40%	lethargy, coma, tachycardia, diaphoresis, hypotension	rapidly bolus with crystalloid fluids, replace blood loss with packed red blood cells, address acid/base/electrolytes

TABLE 8–2. Glasgow Coma Scale (GCS)

	Response	Score
Eye Opening	spontaneous	4
	responds to speech	3
	responds to pain	2
	no eye opening	1
Verbal Response	oriented	5
	confused	4
	inappropriate	3
	incomprehensible sounds	2
	none	1
Motor Response	moves to command	6
	localizes to pain	5
	withdrawals from pain	4
	decorticate posturing (flexion)	3
	decerebrate posturing (extension)	2
	no movement	1

Exposure and Secondary Survey

- remove clothing for complete physical exam
- complete history (**AMPLE**: **A**llergies, **M**edications, **P**ast illnesses, **L**ast meal, **E**vents preceding injury)
- Screening Films: chest, C-spine (must visualize C1–7, T1), and pelvic x-rays
- Initial Labs: type and screen (type and cross for severe blood loss), complete blood count, electrolytes, coagulation studies, urine analysis, ethanol level, pregnancy test
- Other Tests as Needed: electrocardiogram, sickle prep, arterial blood gas, urine toxicology, cardiac enzymes, serum amylase, thoracic and lumbar spine films, diagnostic peritoneal lavage
- Infection Prophylaxis: tetanus status, antibiotics for facial fractures (penicillin, clindamycin), antibiotic ointment to wounds
- Consults: Maxillofacial/Dental, Ophthalmology, Neurosurgery/Neurology, Trauma Surgery/Orthopedics, Vascular Surgery, Cardiothoracic Surgery

History and Physical Exam

Head and Neck History

Nature of Injury

- When: elapsed time of injury
- Where: motor vehicle, pedestrian, etc
- What was the mechanism of injury: assault (fists, bat, etc), penetrating trauma (gunshot wound, stab wound), blunt injury (motor vehicle accident), vector of force (anterior versus lateral impacts), fall
- Motor Vehicle Accidents: speed of injury, restrained, air bag deployed, condition of windshield and steering wheel
- Gunshot Wound: caliber of gun, distance fired, number of gunshots

Review of Systems

- Ocular: change in vision (pre-injury visual acuity), diplopia
- Otological: dizziness, change in hearing (pre-injury visual hearing level), otalgia
- Nasal: nasal obstruction, epistaxis, rhinorrhea
- Laryngeal: hoarseness, difficulty breathing, odynophagia
- Oral: malocclusion, pre-injury missing teeth, dysarthria
- Neurological: loss of consciousness, numbness, weakness
- General: facial pain, shortness of breath, chest tightness

Head and Neck Physical Exam

Otologic

- Inspection: (otoscopic exam) foreign bodies, hemotympanum, CSF otorrhea, tympanic membrane perforation, Battles' sign (mastoid ecchymosis)
- Palpation: auricular hematoma
- Assess Function: hearing, balance

Ocular

- Inspection: (fundoscopic exam) enophthalmos, proptosis, ptosis, extraocular range of motion, "raccoon eyes" suspect basilar skull fracture (cribiform fracture), eyelid lacerations, telecanthus
- Palpation: palpate bony rim (crepitus, tenderness, step-offs, mobility), periorbital soft tissue, assess entrapment (**forced duction test**, apply topical optic anesthetic, use forceps on conjunctiva to move globe), assess medial and lateral canthus (**lid torsion test**, tests for medial canthal ligament by palpating ligament while pulling lid laterally)
- Assess Function: visual acuity, nystagmus, hypesthesia of infraorbital and supraorbital nerves, assess lacrimal system (epiphora), pupil response (equal, reactive, consensual response, Marcus Gunn pupil)

Nasal

- Inspection: epistaxis, septal hematoma, septal dislocations, mucosal tears, external deformities, edema, ecchymosis, open fractures
- Palpation: palpate for nasal fractures (crepitus, tenderness, step-offs, mobility)
- Assess Function: CSF leak (**halo sign**, peripheral clear zone with a central pigmented spot appears when fluid placed on a gauze)

Mandible and Oral Cavity

- Inspection: floor of mouth hematoma, dental exam (missing teeth, condition of dentition, dental fractures), palatal fracture, intraoral and lip lacerations
- Palpation: palpate for mandible fractures (tenderness, step-offs, mobility)
- Assess Function: hypesthesia of lower lip (inferior alveolar nerve) and chin (mental nerve), occlusion

Face and Facial Skeleton

- Inspection: lacerations, hematomas, ecchymosis
- Palpation: bimanual palpation for facial fractures (crepitus, tenderness, step-offs, mobility), mobility of maxilla
- Assess Function: cranial nerve exam, sensory defects, salivary gland and ductal injuries

Larynx and Neck

- Inspection: lacerations, hematomas, ecchymosis, vocal fold mobility
- Palpation: crepitus, tenderness
- Assess Function: hoarseness, stridor (auscultate), airway obstruction

Head and Neck Radiologic Exams

Facial Plain Films

- largely been replaced by computer tomography (except for the mandible, *see below*)
- European Fifth (Hyperextended Base View): views posterior table of frontal sinus
- Water's: views orbital rim, nasal bones, zygoma, and medial and lateral maxillary wall
- Lateral Face: views frontal sinus
- Caldwell: views orbital wall and frontal sinus
- Submental Vertex: views zygomatic arch

Plain Film Mandible Series and Panorex

- Posterior Anterior Projection: views symphyseal region

- Lateral Oblique: views ramus, body, and angle
- Towne's: views condyle and coronoid process
- Panorex: most informative mandibular film, requires patient to be able to sit erect, poor view of symphyseal region and the lingual surface of the mandible

Computed Tomography (CT)

- most informative radiographic exam for head and neck trauma
- axial and coronal facial CT with bone and soft tissue windows, 2–3 mm sections; orbital cuts (coronal) if suspect orbital fractures, CT of head for intracranial injury, CT of neck for laryngeal trauma
- Inspect Areas of Concern: skull base, orbit (walls, floor, retrobulbar hematoma, muscle entrapment), vertical buttresses, horizontal beams, zygomatic arch, bony palate, mandible (condyles), soft tissue (fluid in sinuses, subcutaneous air, hematomas, etc)

Special Radiologic Exams

- **Angiography**: indicated if risk of vascular injury
- **Magnetic Resonance Imaging**: may be considered for intracranial injury and complications
- **Modified Barium Swallow and Esophagram**: evaluate esophageal tears and aspiration

MANDIBULAR FRACTURES

Introduction

- most common in young males (ages 18–30)
- Causes: assault (most common cause), motor vehicle accidents, sports, falls, and gunshot wounds
- Most Common Fracture Sites: (in order of occurrence) condylar neck, angle (especially vulnerable with an unerupted third molar), parasymphysis region near the mental foramen
- Risks: impacted teeth, osteoporosis, edentulous areas, pathologic lytic lesions

Classification

Classification by Site

- **Symphyseal/Parasymphyseal**: between canines, mental foramen weakens parasymphyseal region
- **Body**: between canines and anterior attachment of masseter

- **Angle**
- **Ramus**: protected by the "sling" of pterygoid and masseter muscles
- **Coronoid Process**: protected by zygoma, rarely fractured
- **Condyle**: neck (**weakest point**) and head (TMJ)
- **Alveolar Process**: teeth-bearing region

Classification by Favorability (Fig. 8–1)

- favorability is determined by direction of fracture line and result of muscle force on the mandibular segments
- **Favorable**: vector forces from muscles pull fragments together
- **Horizontally Unfavorable**: vector force of masseter and temporalis muscles pulls fragments apart (angle)
- **Vertically Unfavorable**: vector force of anterior muscles and pterygoid muscles pulls fragments apart (body and symphyseal fractures)

Anterior Muscles

- weaker force
- mylohyoid, geniohyoid, genioglossus, platysma, anterior digastric muscles
- muscle action depresses and retracts (opens mandible)

Posterior Muscles:

- stronger force
- **Temporalis Muscle**: raises and retracts mandible
- **Masseter Muscle**: raises and retracts mandible
- **Medial Pterygoid Muscle**: attaches to medial ramus, raises mandible
- **Lateral Pterygoid Muscle**: attaches to condylar neck, **protrudes** and depresses mandible

Classification by Type of Fracture

- Open versus Closed: **Simple** (closed wound, skin and mucosa intact), **Compound** (open wound, exposed bone)
- Fracture Pattern: comminuted, oblique, transverse, spiral, greenstick (incomplete fracture through only one cortical surface)
- Pathologic: fractures secondary to bone disease (eg, osteogenic tumors, osteoporosis)

Dental Evaluation

- 32 permanent (secondary) teeth (8 incisors, 4 canines, 8 bicuspids or premolars, 12 molars), numbering begins with right third maxillary molar (#1) to left third maxillary molar (#16) then from left third mandibular molar (#17) to right third mandibular molar (#32)

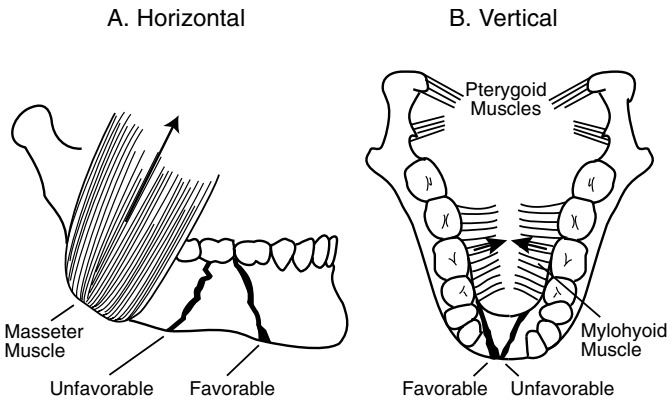

FIGURE 8–1. A. *Horizontal* favorable and unfavorable fractures of the mandible are determined by the action of the masseter muscle. **B**. *Vertical* favorable and unfavorable fractures of the mandible are determined by the actions of the pterygoid and mylohyoid muscles.

- 20 primary deciduous teeth (8 incisors, 4 canines, 8 molars) lettered from A to L

Dental Classification

- **Class I**: teeth on both sides of fracture line, dentulous
- **Class II**: fracture has teeth on one side (partially dentulous)
- **Class III**: completely edentulous

Angle's Classification

- **Class I**: normal bite, the mesiobuccal cusp of the maxillary first molar lines the buccal groove of mandibular first molar
- **Class II**: retrognathic (**overbite**)
- **Class III**: orthognathic (**underbite**)

Management

Management Concepts

- Goals: restore occlusion, establish bony union, return of function, avoid TMJ ankylosis
- repair within first week, delayed repair increases complications (especially without prophylactic antibiotics)

- when placing into occlusion must match wear of the facets of the mandibular and maxillary teeth (do not assume Class I occlusion, recreate the premorbid occlusion if present prior to injury)
- **Compression Stress:** force on lower border of mandible, pushes segments together, compensated with a rigid compression plate placed inferiorly
- **Tension Stress:** force on upper border of mandibular angle and body that causes distraction (separation), compensated by placing a thinner, monocortical miniplate superiorly or place in Maxillo-Mandibular Fixation (MMF)
- in general favorable fractures may only need closed reduction, open fractures tend to require open reduction
- Postoperative Care: antibiotics (penicillin, clindamycin), analgesics, oral hygiene (hydrogen peroxide rinses), soft diet

Indications to Remove Tooth

- tooth within fracture line interferes with occlusion
- infected tooth within fracture line
- fractured tooth, nonviable tooth, or exposed pulp
- Disadvantage: an extracted tooth leaves a point of entry for infection and a point of weak fixation (dental occlusion prevents upward drift at angle), second and third molars also make up a significant portion of mandibular height

Maxillo-Mandibular Fixation (MMF)

- Indications: initial stabilization of occlusion prior to exposure of the fracture (provides a tension band), favorable minimally displaced fractures, selected condylar fractures
- Methods: arch bars (stronger), interdental eyelet wires (Ivy Loops), or circumferential wiring of dentures or teeth
- requires an intact maxilla
- typically MMF may be removed after 2–8 weeks depending on age and type of injury (eg, children 3–4 weeks, adults 4–6 weeks, elderly >8 weeks), condylar fractures 1–2 weeks, body and angle fractures 4–6 weeks)
- rubber bands may be used for easy removal and resetting, allows for exercise
- Complications: airway compromise, dental injury, weight loss, loss of fixation (malunion, nonunion), TMJ dysfunction, aspiration

Open Reduction Internal Fixation (ORIF)

- Indications: unfavorable and comminuted fractures, elderly, poor pulmonary reserve (unable to endure MMF), noncompliant patients

(alcoholics), pregnant women, multiple fractures, bilateral fractures, or seizure disorders
- MMF needed to establish occlusion prior to placement in ORIF
- Approaches:
  1. **Transoral**: avoids injury to marginal mandibular nerve and no external scar; easy access to symphyseal, parasymphyseal, and body regions (more posterior fractures may be approached intraorally with percutaneous drilling techniques)
  2. **External**: indicated for more posterior fractures which cannot be accessed intraorally or severely comminuted fractures

Management by Type

- **Coronoid, Greenstick, Unilateral Nondisplaced Fractures**: observation with soft diet, analgesics, oral antibiotics, and close follow-up; physiotherapy exercises for 3 months (may consider MMF for severely displaced coronoid fractures)
- **Favorable, Minimally Displaced Noncondylar Fractures**: may consider closed reduction and 4–6 weeks of MMF

Displaced Fractures

- **Symphyseal and Parasymphyseal Fractures**: tend to be vertically unfavorable from the force of anterior muscle groups
- **Body Fractures**: almost always unfavorable due to the obliqueness of the fracture and pterygoid and masseter muscle pull
- **Angle Fractures**: in general have the highest complication rate by site because of location posterior to dentition, thin-walled bone, and vector forces from the masseter muscle
- **Ramus Fractures**: isolated ramus fractures are rare (protected by the masseter muscle), displacement is typically minimal, ORIF may be considered for multiple fragments or marked displacement

General Management Options for Displaced Fractures

- **Compression and Tension Plates**: an inferiorly placed compression plate and a superiorly placed monocortical tension miniplate provides stable fixation for early removal of MMF and early oral intake (may also consider using two miniplates)
- **Compression Bar with MMF**: compression plate placed inferiorly with MMF for tension forces
- **Eccentric Dynamic Compression Plate (EDCP)**: indicated when a tension plate or MMF cannot be used, single plate provides compression and tension forces
- **Interosseous Wires with MMF**: wires placed on either side of the fracture to provide fixation with MMF

- **Lag Screws**: bicortical screws; drill through superficial cortex, through fracture line, and into deep cortex of opposite bone, then overdrill proximal segment; brings fracture together; indicated to stabilize fixation for oblique symphyseal or body fractures
- **External Fixation**: indicated for "difficult" fractures (comminuted, multiple, osteomyelitis, elderly), requires two fixation points on either side of an unstable area

Condylar Fractures

- for nondisplaced fractures (typically high condylar fractures) may consider observation with close follow-up, soft diet, analgesics, oral antibiotics, and physiotherapy exercises for 3 months
- displaced fractures (typically subcondylar fractures) may be addressed with closed reduction by pushing the jaw down while rotating the chin upward (may consider MMF for severe displacement for 2 weeks followed by rubber band fixation)
- Open Reduction with Fixation (External Pin Fixation, Interosseous Wires) Indications: condyle displaced into middle cranial fossa, lateral extracapsular displacement of condyle, unable to obtain good occlusion (no occluding teeth), foreign body in TMJ, bilateral condylar fractures with gross displacement, other associated fractures in the mandible or maxilla
- Bilateral Condylar Neck Fractures: bilateral neck fractures are at risk of airway compromise (may need intubation or tracheostomy), associated with anterior bite deformity, consider MMF for 2–3 weeks if minimal displacement; however must encourage movement of jaw to prevent TMJ ankylosis

Comminuted Fractures

- in general should retain as many fragments as possible
- General Management Options: IMF for 6 weeks, reconstruction plate, external fixation, bone grafts (for significant nonviable bone)

Pediatric Patients

- in general prefer conservative management (soft diet, observation with follow-up, joint exercises), usually can tolerate some malocclusion which adjusts with growth
- may treat with MMF for 3 weeks only for open bite deformity or severe trismus (TMJ ankylosis may lead to facial distortion from altered joint growth)
- may need direct skeletal wires, primary (deciduous) teeth are difficult to wire

- pediatric subcondylar fractures are more prone to ankylosis and mandibular growth abnormalities and therefore require long-term follow-up
- consider lower jaw splints or lingual splints if significantly displaced

Edentulous Patients

- edentulous patients typically have atrophic mandibles from decreased loading effects from loss of dental support (also secondary to osteoporosis in the elderly)
- ORIF with plates should be procedure of choice to prevent reduction of jaw opening
- other options include applying arch bars directly to dentures (circumdenture wires), transosseous wires, or external fixation

Surgical Complications

- **Chin and Lip Hypesthesia**: inferior alveolar or mental nerve injury (the mental nerve is a continuation of the inferior alveolar nerve), **most common complication**; spontaneous regeneration occurs in the majority of cases
- **Osteomyelitis**: increased risk from poor oral hygiene, devitalized teeth, infected teeth within fracture line, improperly repaired fracture (nonunion), loose hardware; Rx: initially should remove any unstable hardware followed by long-term intravenous antibiotics to avoid bone loss; infected teeth should be removed; surgical debridement of nonviable bone should be replaced with fixation with later consideration of bone graft for large defects
- **Malunion**: healing of bone in malalignment caused by inadequate immobilization, inaccurate reduction, infection, gross loss of bone, compromised blood supply, malnutrition, osteogenesis imperfecta, or osteopetrosis; Rx: address underlying cause (infection, malnutrition, etc), consider orthodontic realignment for subtle defects or osteotomy with repositioning and refixation
- **Nonunion**: failure of bone to produce osteogenic tissue, nonosteogenic matrix produced (**fibrous union**); Rx: initial excision of fibrous tissue and nonviable bone with refixation (may require bone grafting)
- **Plate Exposure**: similar causes as malunion; Rx: address underlying cause (infection, malnutrition, etc), oral antibiotics, local application of antibiotic ointment and 3% hydrogen peroxide, plate should be retained as long as possible then removed with completion of healing
- **Marginal Mandibular Nerve Injury**: increased risk with external approaches; Rx: direct anastomosis if noticed intraoperatively
- **Necrosis of Condylar Head** (**Aseptic Necrosis**): compromised vasculature to the condylar head with damage to the condylar neck; Rx: debridement of necrotic bone with reconstruction

- **TMJ Ankylosis**: defined as inability to open jaw beyond 5 mm between incisors, in children may cause facial deformities from growth disorders occurring at joint; Rx: if fibrotic ankylosis has occurred treat with passive jaw opening exercises, if skeletal ankylosis occurs must resect with reconstruction of the TMJ
- **Dental Injury**: may be secondary to improper placement of arch bars and MMF

MAXILLARY FRACTURES

Introduction

- Causes: assault, motor vehicle accidents, sports, falls, and gunshot wounds
- the matrix of the maxilla absorbs energy with impact, thereby protecting the orbit, intracranial contents, and nose from total destruction
- severe maxillary fractures are associated with high incidence of intracranial and orbital injuries
- sinusitis is a potential complication and is generally avoided with prophylactic antibiotics and early removal of nasal packing; persistent sinusitis may require surgical intervention to address structural abnormalities

Classification

Buttress System (Fig. 8–2)

Vertical Buttresses: sturdy, developed to withstand load from mastication

1. **Naso-Maxillary** (**NM**)
2. **Zygomatico-Maxillary** (**ZM**): bears the strongest load, begins above maxillary first molars
3. **Pterygo-Maxillary** (**PM**): projects into skull base
4. **Nasal Septum**: midline support

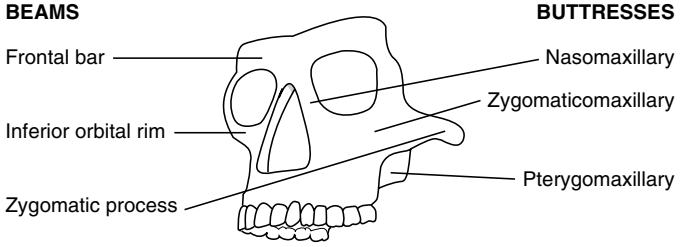

FIGURE 8–2. Facial skeleton buttress system of horizontal buttresses and vertical beams.

Horizontal Beams: weaker, reinforces vertical buttresses and provides width and projection of the face

1. **Frontal Bar**: essential support, made from superior orbital rim and glabellar area, suspends the naso-maxillary and zygomatico-maxillary struts
2. **Inferior Orbital Rims**
3. **Maxillary Alveolus and Palate**
4. **Zygomatic Process**
5. **Greater Wing of the Sphenoid**
6. **Medial and Lateral Pterygoid Plates**
7. **Mandible**

Le Fort Classification (Fig. 8–3)

- based on patterns of fractures ("lines of minimal resistance"), classified according to the highest level of injury
- in many cases Le Fort classification is incomplete for maxillary fractures
- Le Fort fractures may present in many combinations or on one side (hemi-Le Fort)

Le Fort I (Low Maxillary)

- transverse maxillary fracture (upper alveolus becomes separated from upper maxilla) typically caused by a low anterior-to-posterior force

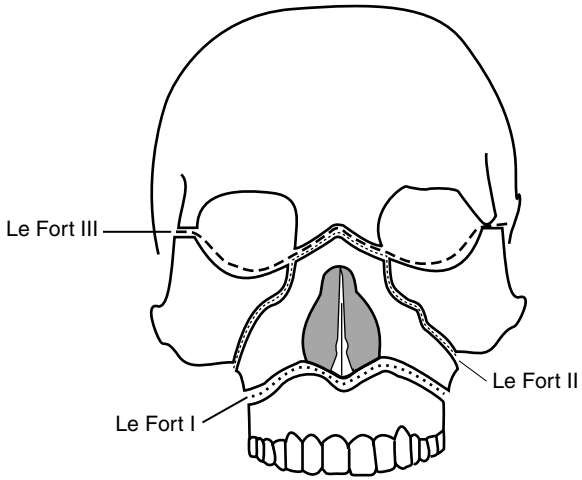

FIGURE 8–3. Classification of Le Fort fractures: Le Fort I or Low Maxillary, Le Fort II or Pyramidal, Le Fort III or Craniofacial Disjunction.

- involves anterolateral maxillary wall, medial maxillary wall, pterygoid plates, septum at floor of nose

Le Fort II (Pyramidal)

- caused typically from a superiorly directed force against the maxilla or an anterior to posterior blow along the Frankfort plane
- involves nasofrontal suture; orbital foramen, rim, and floor; frontal process of lacrimal bone, zygomaxillary suture; lamina papyracea of ethmoid, pterygoid plate; and high septum

Le Fort III (Craniofacial Dysjunction)

- separates facial skeleton from base of skull, typically caused by high velocity impacts, motor vehicle accidents, oblique forces
- involves nasofrontal suture; medial and lateral orbital wall and floor; zygomaticofrontal suture; zygoma and zygomatic arch; pterygoid plates and nasal septum

Management

Principles

- Goals of Reconstruction: reestablish midfacial height, width, and projection; restore soft tissue contour; restore function and occlusion
- Exposure: gingival labial degloving, coronal incision, periorbital incisions (transconjunctival, subciliary incisions, subtarsal)
- Timing: most favorable operative time is within first 24–48 hours prior to significant facial edema; however, direct fixation techniques allow repair up to 2 weeks after injury prior to the onset of significant scarring, fibrosis, and bony resorption
- Panfacial Fractures: "**work from a stable base**"; begin with MMF and management of associated mandible fractures; work lateral (zygoma and ZM buttress to establish anterior projection) to medial to restore buttress system; may consider repairing smaller components (eg, NOE) first then assemble them all together
- Postoperative Care: antibiotics, analgesics, soft diet

Techniques

- **Plate Fixation** (**Miniplates**): miniplates allow earlier removal of MMF, earlier return of function, and have biomechanical advantages over wire techniques; however, may be more sensitive to errors than interosseous wire fixation

- **Interosseous Wire Fixation**: may be considered for less displaced fractures (with 4–6 weeks of MMF), adequate for infraorbital rim
- **Bone Grafts**: indicated for significant bone loss

Management by Le Fort Classification

- **Le Fort I**: generally may be reduced (digitally or with disimpaction forceps) and placed in MMF; may consider fixation of ZM buttress; edentulous patients may require splints, circummandibular wires, or circumzygomatic fixation
- **Le Fort II**: stabilization of the ZM buttress with fixation essential after placing in MMF, may also consider fixation of the nasofrontal process or inferior orbital rim
- **Le Fort III**: usually requires coronal flap for adequate exposure for exploration and miniplate fixation

Palatal Fractures

- palatal fractures must be reduced for a proper dentoalveolar complex positioning
- fracture lines typically paramedian due to the stability of the vomer at midline
- after reduction (Rowe forceps, bone hooks) open reduction may be considered for anterior defects with miniplate fixation
- posterior defects usually may be repaired by closed reduction with MMF (although an open approach may be considered with transosseous wires or plates)
- edentulous patients may require reduction with placement of upper denture or splint (with or without MMF), may also consider miniplate fixation

Surgical Complications

- **Malunion, Nonunion, Plate Exposure**: address similar to mandible fractures (*as above*)
- **Palpable or Observable Plates**: avoided with thinner miniplates; Rx: remove after healing
- **Forehead or Cheek Hypesthesia**: injury to supraorbital or infraorbital nerves; spontaneous regeneration occurs in the majority of cases
- **Osteomyelitis**: address similar to mandible fractures (*as above*)
- **Dental Injury**: may be secondary to reduction techniques or improper placement of arch bars and MMF

ZYGOMATICOMAXILLARY AND ORBITAL FRACTURES

Zygomaticomaxillary Complex (Trimalar) Fractures

Introduction

- Sx: subconjunctival and periorbital ecchymosis, eyelid edema, epistaxis, cheek hypesthesia, diplopia, hypophthalmos, enophthalmos, trismus
- rarely occur in children <5 years old (maxillary sinus not pneumonized)

Four Sutures Involved in Zygomaticomaxillary Complex Fractures

1. **Zygomaticofrontal Suture**: usually fractures cleanly
2. **Zygomaticomaxillary Suture**: at the face of the maxilla, high risk of comminution
3. **Zygomaticotemporal Suture**: zygomatic arch, typically fractures at midpoint or produces a double fracture
4. **Zygomaticosphenoid**: broad suture line

Management

- stabilizing the zygomatic arch allows accurate anterior projection of the malar prominence
- stabilization of the zygomaticomaxillary complex requires a minimum of 2-point fixation (usually ZF and infraorbital rim)
- **Closed Reduction**: indicated for noncomminuted, simple fractures
- **Open Reduction**: indicated for trismus, orbital complications (eg, enophthalmos, diplopia), and facial asymmetry; fixation techniques include miniplates and less commonly interosseous wiring

Common Approaches to the Zygoma

- Incisions: coronal incision (visualize the lateral orbital rim and zygomatic arch), periorbital incisions (transconjunctival, subciliary incisions, subtarsal; visualize the orbital rim and floor), eyebrow incisions (visualize frontozygomatic suture), gingivobuccal incision (visualize the maxillary face)
- **Gillies Approach**: indicated for isolated arch fractures with no comminution; incision behind the temporal hairline is carried through the superficial temporalis fascia, the superior auricular muscle, and the deep temporalis fascia; approach zygoma by placing elevator below the deep temporalis fascia and above the temporalis muscle to avoid injuring the temporal branch of facial nerve (underside of superficial temporalis fascia)

- **Intraoral Approach** (**Keen**): incision at gingivobuccal sulcus, tunnel lateral to the maxilla and under the zygomatic arch
- **Coronal, Hemicoronal, or Extended Pretragal Approaches**: indicated for multiple complex facial fractures (eg, Le Fort III, frontal sinus); access to full length horizontal arc to repair a displaced middle arc segment
- **Lateral Brow Approach**: allows reduction of the zygomatic arch

Surgical Complications: similar to Orbital Floor Fractures (*see below*)

Orbital Fractures

Introduction

- Seven Orbital Bones: frontal, lacrimal, ethmoid (lamina papyracea weakest portion), maxilla, zygoma, sphenoid, palatine bones (Fig. 8–4)
- Optic Canal Contents: optic nerve (CN II), ophthalmic artery
- Superior Orbital Fissure Contents: trochlear nerve (CN IV), lacrimal and frontal divisions of V_1, supraorbital vein, occulomotor and abducens (CN III and CN VI), nasociliary division of V_1
- Inferior Orbital Fissure Contents: zygomaticofacial and zygomaticotemporal divisions of V_2, inferior ophthalmic vein
- SSx: enophthalmos (from increased orbital volume, >2–3 mm pathologic), hypophthalmos, exophthalmos, proptosis, entrapment, diplopia

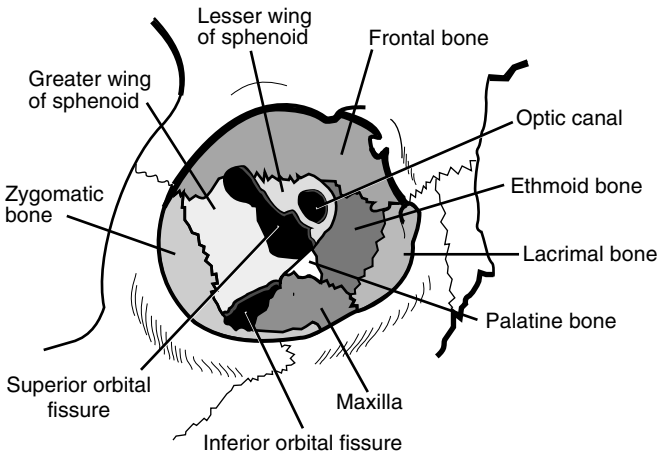

FIGURE 8–4. Anatomy of the bony orbit.

(from entrapment or enophthalmos), hypesthesia of infraorbital nerve, pseudoptosis (enopthalmos causes upper eyelid to droop)

Theories of Orbital Floor Injury

- **Hydraulic Theory**: force to orbital region → ↑ intraocular pressure → fractures floor
- **Buckling Theory**: force on inferior rim → directly fractures floor

Types

- **Pure**: involves the central area of a wall or floor only
- **Impure**: extension of the fracture to include an orbital rim
- **Junctional**: involves floor and medial wall

Traumatic Optic Neuropathy

- Pathophysiology: injury to the optic canal and superior orbital fissure (SOF) results in compressive injury to involved nerves
- SSx: ophthalmoplegia (optic nerve), ptosis (V_1), pupillary dilation (CN III), anesthesia of upper eyelid and forehead (V_1)
- Dx: visual acuity, Marcus Gunn pupil (afferent injury), high-resolution CT
- sudden visual loss carries a poor prognosis (unsalvageable)
- Rx: for progressive loss consider high-dose corticosteroids and osmotic diuresis (mannitol), if no improvement may consider orbital or optic nerve decompression; if CT reveals bony impingement may undergo decompression urgently

Management

- Indications for Surgical Intervention: enophthalmos and/or hypophthalmos (>2–3 mm), mechanical entrapment, diplopia, dehiscence of intraorbital tissue, high risk of enophthalmos and/or hypophthalmos (large floor defects)
- Contraindications for Surgical Intervention: hyphema, retinal tear, globe perforation, only seeing eye, sinusitis, frozen globe
- Ophthalmological Evaluation: essential to evaluate for serious and potentially blinding problems (eg, hyphema, dislocated or subluxed cataractous lenses, retinal holes or detachment, or optic nerve contusion)
- Timing: ideally should be completed 7–10 days after swelling has subsided, delayed repair may reveal bone resorption and scar contracture
- Technique: expose orbital rim, medial orbital wall, and floor posteriorly to the junction of the infraorbital canal and inferior orbital fissure; elevate soft tissue from the floor defect; reduce fracture; for significant defects or difficult reductions may reinforce floor with floor grafts (eg, polyethylene [Marlex], gelfilm, or bone for larger defects), typically may repair up to 5 mm of enophthalmos without straining optic nerve

Approaches

1. **Subciliary Incision** (**Infraciliary**): incision placed 2–3 mm below the cilia of the lower eyelid (may extend into "crow's feet"), allows exposure to orbital floor and rim, risk of ectropion and scleral show
2. **Transconjuctival Incision**: allows exposure to orbital floor and rim, risk of entropion, no external scar, limits exposure
3. **Lynch Incision** (**Frontoethmoidal**): allows exposure to medial wall
4. **Brow Incision**: allows exposure to posterolateral wall
5. **Subtarsal Incision**: incision placed 5–7 mm below the cilia of the lower eyelid in a crease line
6. **Caldwell-Luc** (**Transantral**) **Approach**: indicated for severely comminuted and posterior fractures, completed in conjunction with an approach from above

Surgical Complications:

- **Postoperative Blindness**: may be caused by injury to orbital nerve, retrobulbar hematoma, and retinal artery occlusion; Rx: consider a urgent lateral canthotomy for a retrobulbar hematoma, also may consider removal implant, controlling hemorrhage, and administration of corticosteroids and diuretics
- **CSF Leak**: high risk with Le Fort II and III fractures; Rx: *see* p. 338 *for management*
- **Persistent Enophthalmos and Diplopia**: causes may be from inadequate repair, scarring, or motor nerve injury; Rx: must re-explore and establish volume/contact relationship, release scar
- **Ectropion**: risk in subciliary incisions from scarring or injury to the tarsal plate; Rx: conservative management, massage, corticosteroid injections, oculoplastic surgery (lysis of scar, split-thickness skin grafts) if persists
- **Entropion**: risk in transconjunctival incisions; Rx: oculoplastic surgery
- **Epiphora**: damage to canalicular system; Rx: dilation and irrigation, or dacrocystorhinostomy
- **Cheek Hypesthesia**: injury to infraorbital nerves; spontaneous regeneration may occur
- **Extrusion of Grafts**: alloplastic materials have reduced incidences; Rx: reexploration with replacement of graft
- **Malunion, Nonunion, Plate Exposure**: address similar to mandible fractures (*as above*)
- **Osteomyelitis**: address similar to mandible fractures (*as above*)
- **Palpable or Observable Plates**: avoided with thinner miniplates; Rx: remove after healing

FRONTAL SINUS AND NASO-ORBITOETHMOID FRACTURES

Frontal Sinus Fractures

Introduction

- SSx: laceration of the forehead, forehead swelling (may be confused with a subgaleal hematoma), palpable frontal defect, frontal pain, epistaxis, forehead hypesthesia
- Complications of Frontal Sinus Fractures: **mucoceles** (from entrapped mucosa), cosmetic deformity, chronic sinusitis, CSF leak, epiphora, dystopia (if involves the roof of the orbit), intracranial infections (meningitis, brain abscess)
- Approaches: coronal (best exposure, most cosmetically acceptable), bilateral sub-brow incision ("butterfly" approach), through an existing laceration, "open sky" approach (medial canthal incisions are connected with a horizontal incision)
- frontal sinus fractures are rare in children (frontal sinus does not appear until age 5–6 years old)
- frontal sinus anterior wall is thick, posterior wall and floor are thin

Management

Anterior Table Fractures

- Linear, Minimally Displaced: conservative measures (low risk of mucosal entrapment and cosmetic deformity), observation
- Depressed Fractures "Trap Door": explore, remove mucosa from fractured edges (may consider cutting burs), reduce fracture, may place interosseous wires
- Comminuted or Unstable Fractures: explore, inspect posterior wall and nasofrontal recess, remove mucosa from fractured edges, reduce fracture, thin plate fixation for support, if <1 cm of total frontal bone fragments are missing may consider skin covering only, if >1 cm fragments are missing may use a split calvarial bone graft or mesh (eg, titanium) to reconstruct

Posterior Table Fractures

- high risk of intracranial injury (intracranial hematomas), dural tears, CSF leak, frontal nasal ostium injury, and entrapped mucosa (future risk of mucocele formation)
- Isolated Nondisplaced Posterior Table Fracture: rare, conservative measures (observation), if sinus fluid does not clear within 6 weeks may consider exploration

- Displaced Posterior Table Fracture: must explore frontal sinus via an **osteoplastic flap** (*see* pp. 50–51); for more extensive fractures (comminution, dural tears, CSF leak) consider a subcranial approach (with Neurosurgery consultation); repair dural tears and CSF leaks, inspect nasofrontal recess, and remove mucosa (may consider cutting burs); obliterate cavity with fat, muscle (temporalis muscle galea flap), or bone; reduce fracture and repair anterior table as above
- Comminuted, Contaminated, or Through-and-Through Fractures: considered a neurosurgical emergency, consider cranialization (allows frontal lobes to fill sinus cavity), eliminates sinus although risks contamination of the cranial cavity

Nasofrontal Recess Injuries

- most common cause of traumatic **mucocele** formation
- presents with persistent sinus fluid >10 days postinjury
- Nasofrontal Recess Reconstruction: indicated for limited fractures; widen the ostium and consider placement of a stent or reconstruct with adjacent nasal mucous membrane lining
- Sinus Septectomy: removal of the intersinus septum, indicated for injury of one ostium, allows drainage of damaged side to the opposite nasofrontal recess
- Sinus Obliteration: indicated for injury to both ducts or severe complicated fractures

Surgical Complications

- **Mucoceles, Mucopyoceles**: from entrapped mucosal lining in the fracture, erodes into surrounding bone, may not appear until many years postoperatively, confirmed with CT; Rx: re-explore with an obliterative procedure (*see also* pp. 50–51)
- **Sinusitis**: may be secondary to frontonasal recess injury, postsurgical contamination, or mucoceles; Rx: consider re-exploration to consider obliteration (or reobliteration), repair of nasofrontal recess, or debridement with culture; aggressive antibiotic regimen (must avoid intracranial extension)
- **Forehead Contour Deformity**: may be the result of poor reduction or slipped bone flap; Rx: consider re-exploration, correction of the defect with recontouring, cranial grafts, or alloplastic grafts
- **Intracranial Infections**: increased risk with dural tears, wound contamination, or cranialization procedures (*see* p. 330 *for management*)
- **Osteomyelitis**: address similar to mandible fractures (*as above*)
- **CSF Leak**: (*see* p. 338)

- **Forehead Hypesthesia**: higher risk with sub-brow incisions from injury to the supraorbital nerves, return of function should be expected
- **Forehead Paralysis**: from injury of the frontal branch of the facial nerve, return of function may occur

Naso-orbitoethmoid (NOE) Fractures

Introduction

- **NOE**: frontal process of maxilla, nasal bones, and orbital space
- Common Fracture Patterns:
  1. nasal bones and frontal process of the maxilla is telescoped behind the frontal bone
  2. comminuted fracture with fragments into the orbital space, cranial fossa, and nasal vault
- SSx: flattened nasal dorsum, periorbital swelling, epistaxis, nasal obstruction, medial canthal ligament injury (telecanthus, epiphora, bowstring sign, *see below*), CSF rhinorrhea
- **Bowstring Sign**: tests the integrity of the medial canthal ligament, grasp medial eyelid near lash line and pull laterally, normally should snap back
- **Pseudohypertelorism** (**Traumatic Telecanthus**): widen intercanthal distance (<22 mm in infants and <32–35 mm in adults or the distance of the width of one eye), initially presents with rounding of the medial canthus from releasing the medial canthal ligament, later results in eversion of the lacrimal papilla and the appearance of flattened nasal bones
- **Fluorescein Dye Test**: assess nasolacrimal duct by placing dye within the eye and check presence of dye on nasal pledgets placed intranasally for 1–5 minutes

Anatomy

Medial Canthal Ligament (MCL)

- the MCL is an extension of the tarsal plates which attaches to the medial orbital wall
- MCL receives contributions from the tendinous portion of the preseptal and pretarsal parts of the orbicularis oculi muscle, superior suspensory ligament (**Whitnall**), and the inferior suspensory ligament (**Lockwood**)

Lacrimal Collecting System

- **Puncta**: located at the medial aspect of the upper and lower eyelids, picks up tears
- **Canaliculi**: upper and lower canaliculi form the common canaliculus
- **Lacrimal Sac**: located in the lacrimal fossa, insertions of the MCL straddle the lacrimal sac and act as a pump (sac is compressed with eyelid shortening such as a blink)
- **Lacrimal Duct**: enters into the medial maxilla and exits into the inferior meatus

Planum Sphenoidale

- radiologic term describing the area of the floor of the anterior cranial fossa that is anterior to sella turcica and posterior to fovea ethmoidalis and cribriform plate
- component of the lesser wing of the sphenoid
- laterally forms the roof of optic foramen (high risk of optic nerve injury)

Management

- first reconstruct medial orbital wall prior to repair of the MCL
- must consider associated injuries: ocular injury (Ophthalmology consult), CSF leak, nasoseptal deformities, lacrimal duct injuries, eyelid lacerations
- may attempt closed reduction if MCL and lacrimal system is intact (rarely works)
- Telescoping Nasal Bones and Frontal Process of the Maxilla: requires open reduction via an open sky or coronal incision, reduction and maintaining reduction of NOE fractures are difficult, secure fixation with miniplates or interosseous wires

MCL Repair

- first priority is to repair MCL, the lacrimal system may be reconstructed as a second operation
- must attempt to recreate the pull of MCL by reattaching in the direction of its initial vectors

Classification and Management of MCL Injuries

Type I: MCL remains attached but bone fragment is detached or ligament has been severed completely; Rx: attempt to wire bone fragments with attached MCL to stable bone or release lateral canthal ligament then reattach the MCL to the medial wall of the orbit posterior to the lacrimal fossa (slight overcorrection is required)

Type II: comminuted medial orbital wall fracture; Rx: attach MCL with wires transnasally to the opposite side medial orbital wall (may also consider releasing the lateral canthal ligament to allow for some overcorrection)

Type III: bilateral medial orbital wall fractures; Rx: attach MCL with wires transnasally to the opposite MCL (transnasal canthoplasty), may also consider wiring both MCLs to the opposite frontal process

Lacrimal System Injury

- the lacrimal system may be explored using an operating microscope and lacrimal dilators, stents may be placed to allow primary closure of tears (the lower canaliculus is the primary drainage system)
- **Dacryocystorhinostomy**: indicated for injury distal to the sac
- **Conjunctivorhinostomy or Conjunctivodacryocystorhinostomy**: indicated for injuries proximal to the sac after failed recannulization

Surgical Complications

- **Persistent Telecanthus**: avoided with accurate attachment of MCL; Rx: reexploration with repair
- **Persistent Epiphora**: injury to the lacrimal system, assess with fluorescein dye test; Rx: repair as described above
- **Frontal Sinusitis**: may result from injury to the nasofrontal recess; Rx: (for management *see* pp. 50–51)
- **Scleral Show**: may occur with low placement of transnasal wires; Rx: reattachment of wires at higher plane or may consider Z-plasty
- **Osteomyelitis**: address similar to mandible fractures (*as above*)
- **CSF Leak**: may occur from reduction of naso-orbital injury or associated cribiform plate fracture and dural tears; Rx: *see* p. 338 *for management*

NASAL FRACTURES

Introduction

- most common facial bone fracture
- anterior impacts may result in tip fractures, flattened nasal dorsum, splayed nasal bones, and septal deformities
- lateral impacts may result in a depression of the lateral nasal bone, "C-" or "S-shaped" nasal dorsum deformities, medial maxillary wall fractures, and septal deformities
- dislocated quadrangular cartilage inferiorly or "C-shaped" deformity superiorly are common septal abnormalities from trauma

- children usually have dislocated or greenstick fractures and have a higher risk of septal hematomas and fractures (septum is more rigid than the cartilage of the anterior nose)
- comminutions are more common in adults
- SSx: palpable deformity, epistaxis, edema, nasal obstruction
- Dx: clinical diagnosis (physical exam), radiographs (including CT) may be considered if additional fractures are suspected)
- Complications of Nasal Fractures: medial canthal ligament injury, lacrimal duct injury, cribiform plate fracture (CSF leak, anosmia), septal hematoma may cause saddle nose deformity if untreated, persistent epistaxis

Management

Initial Management

- preoperative photography/x-ray may be considered for medicolegal documentation
- septal hematomas (unilateral or bilateral purple compressible bulge) requires immediate evacuation of hematoma, nasal packing, and antibiotic prophylaxis
- open fractures must be cleaned then given antibiotics

Surgical Management

- generally nasal bone depressed or deviation may undergo closed reduction within first 3 hours (before swelling) or between 3–10 days (after swelling and before healing)
- Closed Reduction: indicated as an initial trial for most nasal fractures; should generally be avoided for cribriform plate fractures with CSF leaks (to avoid nasal packing); reduction is done with elevation and lateral motion using a Boise elevator or Ashe forceps, nasal tip should also be addressed by elevating at the anterior portion of the septum, the septum should be inspected for adequate reduction and evaluation for septal hematomas, internal and external splints should be applied (may also consider nasal packing)
- Open Reduction with Internal Fixation (Septorhinoplasty): indicated for failed closed techniques or extensive injuries that involve the maxilla or frontal bones; open septorhinoplasty techniques are utilized, fixation is achieved with interosseous wire (plates should be avoided), bone grafts may be considered for severe comminution
- Pediatric Nasal Fractures: generally should be treated conservatively to avoid damage to growth centers

- Postoperative Care: ice for 24 hours, prophylactic antibiotics, analgesics, nasal splints, nasal packing

Surgical Complications and Associated Injuries

- **Persistent Deformity**: failed closed reduction should be addressed with an open rhinoplasty approach after healing (3–6 months)
- **Nasal Obstruction**: may occur from synechiae, scar contracture, or structural deformity obstructing the nasal vault
- **Septal Hematoma**: failure to recognize may result in septal necrosis (saddle nose deformity); Rx: *see above*
- **Septal Perforations and Deviations**: may result from open septorhinoplasty techniques
- **Cribriform Plate Fracture**: may result in anosmia or CSF leak

PENETRATING HEAD AND NECK TRAUMA

Penetrating Neck Trauma

Introduction

- low risk of serious injury if platysma muscle is not penetrated
- although overall uncommon, pharyngoesophageal injury is the most commonly missed injury to the neck

Symptoms

- 5–15% of aerodigestive injuries are asymptomatic
- Laryngotracheal Injury: hoarseness, stridor, airway obstruction, subcutaneous emphysema, pain, hemoptysis
- Esophageal or Hypopharyngeal Injury: dysphagia, odynophagia, hematemesis, subcutaneous emphysema
- Vascular Injury: shock, hematoma, diminished pulses, stroke (hemiplegia)

Management Principles

- Pharyngoesophageal Injury: primary closure if <24 hours, otherwise consider drainage (diversion) procedure, reconstruction with muscle transposition, or esophagectomy
- Vertebral Artery Injury: embolization preferred due to difficult exposure and control, for failed embolization may consider surgical repair
- Carotid Injury: associated with high mortality (10–20%); primary repair treatment of choice, otherwise may consider patch grafting, by-pass grafting, or ligation (ligation should not be considered if sus-

pect a stroke to avoid hemorrhagic stroke with revascularization); intimal injuries require repair if obstructing
- Laryngeal Injuries: *see* pp. 472–475

Management by Zones

Zone I: sternal notch to cricoid cartilage

- high risk of injury to great vessels, trachea, and lungs
- difficult region for exposure and control
- Management: arteriography of arch, great vessels, carotids, and vertebrals (may also be used for balloon occlusion or temporary preoperative control); esophagram with esophagoscopy, direct laryngoscopy, and bronchoscopy

Zone II: cricoid cartilage to angle of mandible

- high risk of injury to carotid sheath (carotid artery, internal jugular vein) and aerodigestive system
- more easily accessible and more easy to control
- Management
  1. **Mandatory Surgical Exploration**: gunshot wounds that cross midline, obvious serious injury (stridor, active hemorrhage, absent carotid pulse), active bleeding, air bubbling through wound, arteriography not available, intoxicated patient
  2. **Elective Surgical Exploration**: more accurate than diagnostic tests, up to 50–70% of elective neck explorations are negative
  3. **Selective Management**: arteriography, esophagram with esophagoscopy, direct laryngoscopy, and bronchoscopy

Zone III: angle of mandible to base of skull

- high risk of injury to distal carotid artery, parotid gland, and pharynx
- difficult region for exposure and control
- Management: arteriography (balloon occlusion), esophagram with esophagoscopy and direct laryngoscopy

Penetrating Facial Trauma

Management by Divisions

Division I: above supraorbital rim

- high risk of intracranial complications and frontal sinus injuries
- Management: complete neurological exam, facial and brain CT, Neurosurgical consultation for intracranial complications, manage frontal sinus fractures as described above

Division II: supraorbital rim to commissure of lips

- consider parotid duct and facial nerve injuries (*see* p. 477)
- disrupted globes require Ophthalmology evaluation for possible repair, otherwise consider enucleation to prevent **sympathetic ophthalmoplegia** (autoantibodies stimulated by globe injury may result in blindness in opposite seeing eye)
- Management: complete neurological, oropharyngeal, and ophthalmological exam; CT of brain, orbits, and paranasal sinus; manage maxillary fractures as above

Division III: commissure of lips to hyoid bone

- **Mandibular Angle Plane** (**MAP**): vertical plane from angle of mandible to base of skull, if injury crosses this plane must consider carotid sheath injury
- Management: management similar to neck division I and III (exploration versus endoscopy), panorex, angiography (if wound crosses MAP)

LARYNGEAL TRAUMA

Introduction

Introduction

- blunt trauma has a higher risk of skeletal fractures than penetrating injuries
- SSx: dysphonia, subcutaneous air, dysphagia, cough, increasing stridor or dyspnea, subcutaneous emphysema, hemoptysis, laryngeal pain and tenderness, neck deformity
- Mechanisms of Injury: motor vehicle accidents, assaults, "clothesline injury," strangulation, penetrating injuries (gunshot wounds, knife)
- Complications: airway compromise, laryngeal stenosis, vocal fold immobility (aspiration, dysphonia)
- pediatric laryngeal fractures are rare because of elasticity of cartilage and higher position of the larynx in the neck; however, children have higher risk of soft tissue injury

Types of Laryngeal Injuries

- endolaryngeal tears, edema, and hematomas
- arytenoid cartilage subluxation (dislocation): presents as a fixed cord
- cricoarytenoid joint injuries: may damage recurrent laryngeal nerve
- cricoid fractures: high risk of airway compromise and injury to recurrent laryngeal nerve
- hyoid bone fractures: may risk airway compromise

- cornus of thyroid cartilage may lacerate pharyngeal mucosa
- **Cricotracheal Separation**: trachea tends to retract substernally and the larynx tends to migrate superiorly, high mortality, risk of "clothesline" injuries, may have bilateral recurrent laryngeal nerve paralysis
- Pharyngoesophageal Tears
- Recurrent Laryngeal Nerve Injury

Management

Establish Airway and Stabilize Cervical Spine (ABCs)

- must establish airway while protecting the cervical spine
- in blunt trauma premature endotracheal intubation is avoided to prevent an airway crisis (fiberoptic intubation may be attempted)
- a surgical airway is a safe method to establish an airway without obscuring diagnostic viewing and potentiating an airway crisis (should be completed under local anesthesia)

Diagnosis

- Physical Exam: soft tissue edema or hematoma, laryngeal tenderness and crepitus, subcutaneous emphysema, laryngeal tenderness
- Fiberoptic Nasopharyngoscope: first-line diagnostic test, allows visualization of the endolarynx with minimal risk to airway, evaluate vocal fold mobility (fixation, arytenoid dislocation or avulsion), endolaryngeal lacerations, airway patency, laryngeal edema and hematomas
- CT of Neck: diagnostic test of choice to evaluate laryngeal fractures (hyoid bone, thyroid and cricoid cartilage); however, not indicated if injury is severe enough that exploration would occur regardless of CT result (CTs have replaced contrast laryngograms which may compromise a marginal airway)
- Roentgenograms of the Neck: largely been replaced with CT
- Esophagram: best to begin with a water soluble contrast to avoid barium-sulfate-induced mediastinitis
- Direct Laryngoscopy and Esophagoscopy: may be considered after airway has been established to evaluate the endolarynx (allows palpation of arytenoids)

Medical Management

- Indications for Medical Management Only: smaller soft tissue injuries (hematomas, lacerations), single nondisplaced fracture (*controversial*), stable laryngeal skeleton with an intact endolarynx

- hospitalization for at least 24 hours for observation with tracheostomy set at bedside
- nothing by mouth with hydration
- elevate head of bed, voice rest, humidified air
- prophylactic antibiotics, antireflux protocol, systemic corticosteroids

Surgical Management

- Indications for Surgical Management: large lacerations, airway obstruction, disrupted anterior commissure, exposed cartilage, progressive subcutaneous emphysema, fractured or dislocated laryngeal skeleton, dislocated arytenoids, vocal fold immobility
- Timing: ideally should be repaired within 2–3 days to avoid infection and necrosis
- Endoscopic Repair: may attempt smaller mucosal disruptions and repositioning of arytenoids

Open Reduction and Repair

- Approach: midline thyrotomy or infrahyoid laryngotomy
- repair mucosal injuries to provide adequate internal covering of cartilage, approximate tissues well to reduce potential of scarring and granulation tissue formation (may require local flaps or grafts)
- may reposition subluxed arytenoids (or remove for severe disruption)
- laryngeal fractures should be reduced and immobilized
- consider placing a keel or silastic stent for unstable or comminuted fractures, disrupted anterior larynx, or massive mucosal injuries; stents increase risk of granulation tissue formation, infection, and wound necrosis
- Montgomery T tubes may be considered for complex tracheal and subglottic injuries (eg, subglottic resections, severe cricoid fractures)
- keels may be used to prevent anterior web formation for isolated anterior commissure mucosal disruptions, keels (which possess low tissue reacting potential) are interposed between the anterior thyroid cartilage and the anterior commissure
- repair recurrent laryngeal nerve with microsurgical primary anastomosis (prevent muscle atrophy, does not restore mobility of vocal fold since there is a mixture of adductor and abductor innervations)

Complications of Surgical Management

- **Granulation Tissue**: high risk if cartilage left exposed at points of contact of the laryngeal stents; Rx: laser excision, laryngeal dilation
- **Stenosis**: avoided with proper fixation of the laryngeal skeleton; Rx: initially should attempt dilation otherwise may require a revision thyrotomy with excision of scarred tissue with reconstruction, severe stenosis may require tracheal resection or supraglottic laryngectomy

- **Vocal Fold Paralysis**: *see* pp. 112–117, for complete discussion on management
- **Displaced Stents**: may occur during coughing; Rx: for respiratory distress may extract the tube from a tracheostomy otherwise may access with bronchoscopy

SOFT TISSUE TRAUMA

Introduction

Anatomy of the Skin

Epidermis

1. **stratum corneum**: no nucleus, keratin in cytoplasm, loosens then desquamates
2. **stratum granulosum**: 3–5 layers thick, flattened, keratohyalin granules
3. **stratum spinosum**: basophilic cells
4. **stratum basale**: single layer of cuboidal cells at basal lamina

Epidermal-Dermal Junction: "blueprint" for overlying skin, must be reestablished in repair

1. **rete pegs**: epidermal projections into dermal layer
2. **papillae**: dermal, vascularized projections into epidermal layer

Dermis: provides nutrition and support

1. **papillary dermis**: collagen and elastin fibers
2. **reticular dermis**: denser layer

Subcutaneous Layer: contains fat and fibrous tissue

General Management of Soft Tissue Trauma

Local Anesthesia

- lidocaine [Xylocaine] maximum dose 4 mg/kg or 7 mg/kg if combined with epinephrine (epinephrine helps control bleeding and prolongs the therapeutic effect)
- excess local injection distorts tissue
- nerve blocks diminish volume of local anesthesia required and minimize tissue distortion

Cleaning and Debriding Wound

- wash with a mild antibacterial soap
- irrigate contaminated wounds with copious saline or antibiotic solutions (high-pressure irrigation)

- remove embedded particles, asphalt, or tattooing with a brush or dermabrader
- remove tar, grease, or other petroleum products with ether or acetone
- preserve all facial tissue unless unequivocally dead (facial soft tissue tends to heal well because of excellent blood supply)
- avoid the use of hydrogen peroxide (causes a toxic exothermic reaction)

Antimicrobial Therapy

- prophylactic antibiotics should cover skin flora (gram positives)
- oropharyngeal exposure requires prophylactic antibiotics that cover mixed flora including anaerobes (penicillin, clindamycin)
- topical antibiotics help provide moisture which aids in reepithelialization and migration during the proliferation stage of wound healing
- Tetanus Prophylaxis: in general tetanus toxoid may be given for any wound if no immunization in past 10 years or as a booster for high risk injuries; tetanus immunoglobulin may be given concurrently (at different site and with a different syringe) for tetanus-prone wounds

Specific Soft Tissue Injuries

Hematomas and Ecchymosis

- small asymptomatic hematomas may be observed (typically resorb)
- larger hematomas must be evacuated to avoid pressure necrosis and infection
- early hematomas initially are gelatinous and cannot be aspirated (must undergo incision with drainage to evacuate), after 1 week hematomas become liquefied and may be aspirated

Lacerations

- facial wounds tend to resist infections and may be closed primarily up to 2–3 days ("golden hours" <8 hours) due to excellent blood supply
- always utilize atraumatic techniques (skin hooks, multitoothed forceps)
- reapproximate normal landmarks first (around the nose and eyes)
- subcutaneous closure removes dead space, decreases tension, and helps with alignment
- may need to freshen edges for better approximation and healing
- undermining reduces tension at dermal-subcutaneous junction
- evert all edges (bevel wound edges outward) to allow for a flatter, less noticeable scar

Lip Injuries

- may close primarily up to 1/2 the upper or lower lip
- should begin with a nonabsorbable "key stitch" above vermilion border
- reconstruct in 3 layers including the orbicularis oris muscle
- use absorbable suture in mucosal and muscular layers
- for larger defects consider local flaps (*see* pp. 400–402)

Trap-Door Deformities

- Definition: "U-shaped laceration," slanted, often with a beveled edge
- wound congestion occurs when base is at an antigravitational position
- avoid pincushion deformity
- Rx: create a perpendicular edge to the flap, undermine surrounding tissue to allow closure in layers; if defect is small may excise flap and close primarily; for larger defects may require later revision with Z-plasty

Parotid Duct Injuries

- must be considered with vertical lacerations posterior to the anterior border of the masseter muscle at line from tragus to upper lip (may have an associated buccal branch of the facial nerve injury)
- Dx: cannulate duct if suspect injury
- Rx: place a stent intraorally though Stensen's duct out the distal cut end, thread through proximal end (locate by palpating the parotid gland to express saliva), repair primarily with microscopic anastomosis around stent, remove stent after 10–14 days, may require repeated dilations; may also consider ligating duct or rerouting the duct intraorally
- Complications: sialoceles and fistulas

Eyelid and Lacrimal Injuries

- consider Ophthalmology consult for underlying ocular injury prior to any repair
- full-thickness lacerations of the eye margin require 3-layer closure initially at the "gray line" (anterior aspect of the tarsal plate), the posterior lid margin, and the tarsal plates (avoid sutures in the orbicularis oris muscle to prevent ectropion)
- may close primarily up to 1/4 of the lid without causing entropion; >1/4 of lid may require skin grafts (split-thickness graft to upper lids and full-thickness grafts to lower lids, full-thickness grafts may be obtained from opposite eyelid)

- a lateral canthotomy may add an additional 10 mm of length
- completely avulsed eyelids should be reattached
- lacrimal duct injuries must be considered with injuries medial to puncta or through medial canthus, injury requires cannulation of duct and microscopic primary repair over a silastic stent

Auricular Trauma

- **Hematomas:** usually are a result of blunt trauma (wrestling, boxing) and should be evacuated aggressively (incision and drainage) and sandwiched with dental rolls for pressure to avoid perichondritis and "cauliflower ear," oral antibiotics should be given while rolls in place (2 weeks)
- **Lacerations**: initial irrigation with conservative debridement; may reapproximate cartilage by placing stitches in the perichondrium; cartilage exposure requires coverage by primary closure, wedge excision, local flaps, or bury in a postauricular pocket (for later reconstruction)
- **Helical Rim Defects**: may be closed primarily if <2 cm, if >2 cm may use a chondrocutaneous advancement flap
- **Avulsions**: partial avulsions usually survive with at least a 1–2 mm skin pedicle, complete auricular avulsion requires attempted replantation (microvascular techniques), may require a meatoplasty to avoid external auditory canal stenosis

Human and Animal Bites

- in general, human bites are considered one of the most contaminate injuries; however, in the face early human bites may be closed primarily with a drain after aggressive irrigation; delayed closure or healing by secondary intention may be considered for old human bites (>8 hours) to avoid infection
- early animal bites may be closed primarily after vigorous irrigation and debridement
- broad-spectrum antibiotics, tetanus booster (*as above*), also may consider rabies immunoglobulins

Burns of the Head and Neck

Introduction

- "Rules of 9's": used to determine total body surface area (TBSA): arm 9%, leg 18%, anterior and posterior trunk 18% each, head 9%, palm 1%

- Burn Types
  1. **Superficial** (first degree): mild, erythema
  2. **Partial-Thickness** (second degree): penetrate into dermis and adenexa, blisters, painful
  3. **Full-Thickness** (third degree): irreversible deep dermal injury, damages nerve endings (painless), necrotic

Initial Management

- ABCs: consider early endotracheal intubation if suspect inhalation injury, avoid tracheotomies through burn regions
- History: burn agent (fire, chemical, electrical, scald), time of burn, open versus closed space (closed spaces have increased risks for inhalation injuries)
- Initial Wound Management: wash chemical burns, brush off powder, neutralization of chemical burns is not indicated (the reaction itself may become exothermic), apply surface cream (*see* Table 8–3)
- Ancillary Tests: complete blood count, electrolytes, arterial blood gasses (with carboxyhemoglobin), chest x-ray, electrocardiogram, urinalysis (myoglobin), renal panel, pregnancy test
- Medications: reflux regimen, may consider prophylactic antibiotics, tetanus prophylaxis, corticosteroids are not indicated
- provide volume according to **Parkland Formula** (volume/day = TBSA (%) $\times$ Kg $\times$ 4 cc, $^1/_2$ volume given first 8 hours, the remaining volume given over 16 hours), consider Foley catheter to record urine output
- assess for carbon monoxide poisoning (headaches, cherry-red skin color, mental status changes, carboxyhemaglobin >10% in a nonsmoker or >20% in a smoker)
- all significant burns of the face require hospital admission for management and observation

TABLE 8–3. Surface Creams for Burn Injuries

Agent	Comments
Silver Nitrate	broad spectrum, may cause electrolyte imbalance, painless, poor penetration of eschar
Silverdene	painless, no electrolyte problems, may cause neutropenia, broad spectrum (including fungal)
Mafenide Acetate	penetrates eschar, broad spectrum (covers *Pseudomonas*), painful, may cause hyperchloremic acidosis
Bacitracin Ointment	indicated for most superficial injuries and facial wounds

Inhalation Injury

- significant cause of death in burned patients
- Causes: closed space exposure to chemicals, carbon monoxide, toxins in smoke (ammonia, sulfur dioxide, etc.), and steam; direct thermal injury is rare due to glottic reflex closure to heat)
- SSx: facial burns (amount of facial surface burn does not correlate with severity of inhalation injury), singed nasal hairs, soot in mouth or nose, hoarseness, wheezing, carbonaceous sputum
- Dx: direct laryngoscopy and bronchoscopy, may consider radionucleotide xenon scans
- Complications: upper airway obstruction, pulmonary edema, chemical tracheobronchitis, carbon monoxide poisoning
- Rx: monitoring, supplementary oxygen (may require intubation), aggressive pulmonary toilet, serial arterial blood gasses, bronchodilators, **corticosteroids are contraindicated** (increases mortality)

Facial Burns

- superficial and partial thickness injuries require only local wound care (initial wash with soap and bacitracin ointment)
- deeper partial-thickness injuries and full-thickness injuries should be managed conservatively initially with local care, consider waiting up to 10 days before excising necrotic tissue
- skin grafting should be planned according to the aesthetic units of the face and placed over a well perfused granulation base
- may consider the use of tissue expanders (*see* pp. 397–398)
- pressure dressings (pressure masks), massage, and corticosteroid injections reduce hypertrophic scarring
- physical therapy prevents contracture
- **Auricular Burns**: delicate, thin tissue requires gentle cleansing and avoidance of pressure to the ears (doughnut ring); Rx: conservative initial debridement; cover exposed cartilage with flap closure (postauricular flap) to avoid suppurative chondritis; if extensive may remove auricle and place in an abdominal pocket for later reconstruction
- **Oral Burns**: oral commissure often involved with electrical injuries (pediatrics); Rx: initially should manage with conservative local care, may delay debridement for up to 10 days to allow areas to demarcate into regions of necrosis, to avoid microstomia from contracture may consider placing an oral stent
- **Eyelid and Ocular Burns**: risks ectropion, lid contraction, keratitis, and cataracts (electrical burns); Rx: Ophthalmology consult; keep eye moist with artificial tears and lubricants; eyelid full-thickness burns require initial evaluation for adequate lid support (tarsal plate) prior to

reconstruction; may consider split-thickness graft to upper lids and full-thickness grafts to lower lids; may require lid release for contractions or tarsorrhaphy

FOREIGN BODY AND CAUSTIC INGESTION

Foreign Body Ingestion and Aspiration

Foreign Body Ingestion

- most common objects in adults are fish bones, dentures, and meat (most common objects in pediatrics are coins)
- 95% of esophageal foreign bodies are located at the cricopharyngeus (other common sites are the gastroesophageal junction and the indentation from the aortic arch and left mainstem bronchus)
- SSx: dysphagia, drooling, weight loss, chest pain, fever
- Dx: chest x-ray to identify object, barium swallow should be avoided since it may obscure the field with endoscopy
- Complications: esophageal perforation, mediastinitis, pneumomediastinum, pneumothorax, aspiration

Management

- obtain description of object
- Rigid Esophagoscopy: indicated for foreign bodies that remain in the esophagus for >24 hours, large object in the esophagus, any batteries; if possible may obtain similar object in order to determine a strategy for instrumental removal; always check for a second foreign body; Fogarty catheters may be used for distal objects
- if no endoscopy indicated then consider following with abdominal films and straining stool
- Long-term Management: consider oral corticosteroids for presence of edema and prophylactic antibiotics, close follow-up

Foreign Body in the Larynx and Tracheobronchial Tree

- most common aspirated objects are peanuts (also food particles, metallic objects, balloons)
- SSx: choking, stridor, chest pain, wheezing, hoarseness, audible slap
- Dx: chest x-ray with inspiratory and expiratory phases (evaluates air trapping)
- Complications: airway compromise, subglottic laryngeal edema, pneumonitis, pneumothorax, chronic lung infection, bronchiectasis, bronchial suppuration, permanent parenchymal loss

Management

- avoid digital manipulation, back slapping, Heimlich maneuver (**unless completely obstructed**)
- obtain description of object
- preparation for bronchoscopy is essential (must coordinate with anesthesia), no preoperative anesthesia
- Rigid Bronchoscopy: light sedation with short-term muscle relaxants are used, jet ventilation may be considered, intraoperative corticosteroids may be used to minimize edema, if possible may obtain similar object in order to determine a strategy for instrumental removal, always check for a second foreign body
- Thoracotomy: indicated if unable to remove with bronchoscopy
- Long-term Management: consider oral corticosteroids for presence of edema and prophylactic antibiotics, close follow-up, may require multiple endoscopies

Caustic Ingestion

Introduction

- alkaline ingestion (pH >12.5) causes liquefaction necrosis (more severe damage to esophagus) and is often odorless and tasteless (initially better tolerated)
- acidic ingestion causes coagulation necrosis, rapid transit results in skip areas and more severe damage to the stomach
- liquids burn more distal; solids (powder) burn more proximal (solids cause instant pain and irritation therefore often expelled early)
- severity of external and oropharyngeal injury does not correlate with extent of esophageal and gastric injury
- Common Alkali Agents: (**<3.8%** NaOH) clinitest tablets, alkali batteries, bleaches, household ammonia, hair straighteners
- SSx: drooling, mouth pain, stridor, dysphagia, chest or abdominal pain, oral injury (burn)
- Complications: stricture (circumferential burns), pneumonia, tracheoesophageal fistulas, laryngeal edema, mediastinitis, perforations, esophageal carcinoma (increases risk by 1000)

Stages of Esophageal Caustic Injury

- dusky submucosal edema within first 24 hours
- ulceration may appear (alkali injuries) within 24 hours
- submucosal inflammation (gray coagulum) with thrombosis at 2–5 days
- sloughing of superficial layer at 5–7 days

- fibrosis of deep layers and formation of scars and strictures (collagen deposition begins during proliferative phase at 1 week with contraction beginning at 2-4 weeks)

Management Principles

- evaluate airway (may require an emergent tracheostomy)
- identify agent, determine pH (alkali if >**12.5**), amount, and concentration (call poison information center)
- Initial Management: remove granules and powder with water, consider initial dose of corticosteroids (should not be given if severe esophageal burns suspected, increased risk of perforation), administer intravenous antibiotics, maintain nothing by mouth with intravenous hydration, do not induce emesis, do not neutralize (causes a harmful exothermic reaction)
- Evaluate for Complications: chest x-ray (mediastinitis), abdominal series (gastric perforation), arterial blood gas (acid-base disturbance)
- Direct Laryngoscopy with Esophagoscopy: some specialists recommend universal endoscopy 24–48 hours after injury has declared itself, others observe and scope only for oral burns, symptoms, or clear history of significant volume (endoscopy is never wrong)

Management Based on Endoscopic Findings

First Degree Burns

- Endoscopic Presentation: hyperemia and edema of mucosa
- Immediate Management: observation overnight
- Long-term Management: reflux regimen, follow-up after 2 weeks (consider esophagram if symptomatic at follow-up)

Noncircumferential Second Degree Burn

- Endoscopic Presentation: injury to mucosa, submucosa, and muscle layers
- Immediate Management: observation overnight
- Long-term Management: broad-spectrum antibiotics for 1 week, oral corticosteroids 2–3 week taper, reflux regimen
- Follow-up: esophagram at 1 and 3 month follow-up (evaluate for esophageal stricture formation)

Circumferential Second Degree Burn

- Endoscopic Presentation: circumferential injury to mucosa, submucosa, and muscle layers

- Immediate Management: observation overnight, if viability in question may rescope within 24 hours, may consider nasogastric tube placement, feeding tube, or silastic stenting
- Long-term Management: broad-spectrum antibiotics for 1 week, reflux regimen, may consider oral corticosteroids for 3 weeks (*controversial,* may mask infection and increase risk of perforation), may consider **Lathyrogenic agents** (β-aminopropionitrile, acetylcysteine, penicillamine) which reduces collagen cross-linking
- Follow-up: esophagram after 1 month from injury then every 3 months × 4 (evaluate for esophageal stricture formation)

Third Degree Burn

- high mortality
- Endoscopic Presentation: full-thickness burn, black coagulum, pleural and mediastinal involvement
- Immediate Management: may require esophagectomy/gastrectomy with exploratory laparotomy to remove necrotic tissue, for minimal necrosis may consider nasogastric tube placement or silastic stents, overnight monitored bed, second look at 24 hours
- Long-Term Management: long-term broad-spectrum antibiotics, reflux regimen, oral corticosteroids are contradicted (masks infection and increases risk of perforation), may consider lathyrogenic agents
- Follow-up: esophagram after 1 month from injury then every 3 months × 4 (evaluate for esophageal stricture formation)

Strictures

- Immediate Management: serial dilation, esophagectomy for severe cases
- Follow-up: monitor with esophagrams after 1 month then every 3 months × 4 for circumferential, third degree injuries, and all symptomatic patients

Appendix A

BRANCHIAL APPARATUS

Components of the Branchial Apparatus

1. **Branchial Arch:** composed of a cartilaginous bar, brachiometric nerve, muscular component, and aortic arch artery
2. **Branchial Grooves** or **Clefts:** external, lined with ectoderm
3. **Branchial Membrane:** formed between branchial groove and pouch
4. **Branchial Pouches:** internal, lined with endoderm, contains a ventral and dorsal wing

Branchial Arches

I (Mandibular Arch)

- **Meckel's Cartilage:** malleus head and neck, incus body and short process, anterior malleal ligament, mandible (formed from intramembraneous ossification)
- **CN V_3**: muscles of mastication, tensor tympani, tensor veli palatini, mylohyoid, and anterior digastric muscles
- **Maxillary Artery**
- Hillock of His: 1. tragus; 2. helical crus; 3. helix

II (Hyoid Arch)

- **Reichert's Cartilage**: manubrium of malleus, long process and lenticular process of the incus, stapes (except vestibular part of footplate), styloid process, stylohyoid ligament, lesser cornu and upper half of hyoid
- **CN VII**: muscles of facial expression, stapedius, stylohyoid, and posterior digastric muscles
- **Stapedial Artery** (degenerates)
- Hillock of His: 4. antihelix crus; 5. scapha; 6. lobule

III

- Cartilage: greater cornu and lower half of the hyoid
- **CN IX**: stylopharyngeus muscle, superior and middle constrictors
- **Common Carotid and Internal Carotid Arteries**

IV

- Cartilage: thyroid and cuneiform cartilage
- **Superior Laryngeal Nerve**: cricothyroid muscles and inferior pharyngeal constrictors
- **Aorta** (left); **Proximal Subclavian Artery** (right)

V/VI

- Cartilage: cricoid, arytenoid, and corniculate cartilage
- **Recurrent Laryngeal Nerve**: intrinsic laryngeal muscle (except cricothyroid muscle)
- **Ductus Arteriosus and Pulmonary Artery** (left); **Pulmonary Artery** (right)

Branchial Pouches

I: eustachian tube, middle ear (mastoid air cells), inner tympanic membrane
II: supratonsillar fossa, palatine tonsils, middle ear
III: epithelial reticulum of thymus, inferior parathyroids
IV: parafollicular cells (C-cells) of thyroid, superior parathyroids

Branchial Clefts (Grooves)

- **I:** external auditory canal, outer tympanic membrane (dorsal part)
- **II-V:** obliterates

Appendix B

CRANIAL NERVES
CN I — Olfactory

Olfaction

- neurosensory cells (in olfactory epithelium) → lateral olfactory stria → lateral olfactory area of temporal bone
- neurosensory cells (in olfactory epithelium) → medial olfactory stria (lesser contribution) → frontal lobe (limbic system)

CN II — Optic

Vision

- ganglion cells of the retina — optic nerve → lateral geniculate body (thalamus), pretectal area (midbrain), primary visual cortex (occipital lobe)

CN III, IV, VI — Oculomotor, Trochlear, Abducens

Somatic Motor

- **CN III**: levator palpebrae superioris, superior rectus, medial rectus, inferior rectus, and inferior oblique rectus muscles (**oculomotor nucleus**)
- **CN IV**: superior oblique muscle (**trochlear nucleus**)
- **CN VI**: lateral rectus muscle (**abducens nucleus**)

Parasympathetic

- **Edinger-Westphal nucleus** — *preganglionic parasympathetic fibers* (**CN III**) → **ciliary ganglion** — *postganglionic parasympathetic fibers* → ciliary muscles and sphincter pupillae muscles
- NOTE: sympathetic fibers to the globe and sensation from V_1 also pass through the ciliary ganglion

CN V — Trigeminal

Branchial Motor

- CNV_3 (foramen ovale) → muscles of mastication, tensor tympani, tensor veli palatini, mylohyoid, and anterior digastric muscles (**masticator trigeminal nucleus**)

Sensory

- **CN V_1**: lacrimal (also carries parasympathetic fibers from facial nerve), frontal, nasociliary, and meningeal branches
- **CN V_2**: zygomatic, infraorbital, pterygopalatine, and meningeal branches
- **CN V_3**: buccal, auriculotemporal, lingual, inferior alveolar, and meningeal branches
- **trigeminal ganglion** (**Meckle's cave** in middle cranial fossa) → **trigeminal nucleus**

CNVII — Facial Nerve

(*see* pp. 358–361)

CNVIII — Vestibulocochlear Nerve

Balance

- vestibular nerve → ipsilateral and contralateral pontomedullary (4) vestibular nuclei

Hearing

- cochlear nerve → cochlear nucleus → superior olivary nuclei → lateral lemniscus → inferior colliculus → thalamus (medial geniculate body) → auditory cortex at sylvian fissure of temporal lobe (Brodmann's area 41)
- see also p. 292

CNIX — Glossopharyngeal

Taste

- taste from posterior ⅓ → nucleus solitarius

Branchial Motor

- stylopharyngeus muscle (**ambiguus nucleus**)

Parasympathetic

- **inferior salivatory nucleus** (medulla) — **glossopharyngeal nerve** (**Jacobson's nerve**) — **lesser (superficial) petrosal nerve** → **otic ganglion** → *postganglionic parasympathetic fibers* → **parotid gland**

Sensory

- visceral sensation from carotid body (chemoreceptors for oxygen tension) → **nucleus of the tractus solitarius**
- sensation from posterior ⅓ of tongue, external auditory canal, and tympanic membrane

CN X — Vagus

Branchial Motor (ambiguus nucleus)

- **Recurrent Laryngeal Nerve**: all intrinsic laryngeal muscles except cricothyroid muscle (also sensory to laryngeal mucosa inferior to glottis)
- **Superior Laryngeal Nerve** (**External Branch**): cricothyroid and pharyngeal constrictors
- muscles of the pharynx (except stylopharyngeus), levator veli palatini, uvulae, palatopharyngeus, palatoglossus, salpingopharyngeus, and pharyngeal constrictors muscles

Parasympathetic

- **dorsal motor nucleus** — *preganglionic parasympathetic fibers* → smooth muscle innervation to thoracic and abdominal viscera, secretory glands of the pharynx and larynx

Sensory

- **Superior Laryngeal Nerve** (**Internal Branch**): sensory to laryngeal mucosa above glottis
- **Auricular Branch** (**Arnold's Nerve**): sensory from postauricular skin, external auditory canal, tympanic membrane, and pharynx
- visceral sensory from pharynx, larynx, and viscera → **nucleus of the tractus solitarius**

CN XI — Accessory

Branchial Motor

- sternocleidomastoid and trapezius muscles (**accessory nucleus**)

CN XII — Hypoglossal

Somatic Motor

- intrinsic muscles of the tongue (except palatoglossus muscle), styloglossus, hyoglossus, and genioglossus muscles (**hypoglossal nucleus**)

Appendix C

COMMONLY PRESCRIBED DRUGS IN OTOLARYNGOLOGY

Krista Piekos

Abbreviations in Appendix

APP	antipseudomonal penicillin	ml	milliliter
ASP	antistaphylococcal penicillin	mo	month
cap	capsule	MU	million units
d	day	PO	by mouth
ER	extended release	PR	by rectum
GI	gastrointestinal	q	every
gm	gram	R	renal metabolism/excretion
H	hepatic metabolism/elimination	SQ	subcutaneously
HR	combined hepatic/renal elimination	SR	sustained release
IM	intramuscular	tab	tablet
IV	intravenous	TD	transdermal
kg	kilogram	U	units
LFTs	liver function tests	UTI	urinary tract infection
mg	milligram	y	years old
min	minute		

TABLE C–1. Single-Agent Antitussives, Expectorants, and Decongestants

Drug	Category	Brand Names	Adult Dose (max/day)	Pediatric Dose (max/day)	Side Effects/ Considerations
Benzonatate	antitussive	Tessalon Perles	100mg q8 (600mg)	<10y: not recommended >10y: 100mg q8 (600mg)	drowsiness, headache, constipation, do not chew/crush, **H**
Codeine	antitussive	Codeine sulfate	10–20mg q4–6 15–30mg q4–6 (IV/IM/SC) (120mg)	2–6y: 2.5–5mg q4–6 (PO/IM/SQ) (30mg) 6–12y: 5–10mg q4–6 (PO/IM/SQ) (60mg)	drowsiness, constipation, orthostatic hypotension, dry mouth, **H,**
Dextromethorphan	antitussive	Benylin DM, Robitussin Pediatric	15–30mg q4–8 (120mg) 60mg q12 (ER) (120mg)	2–6y: 2.5–7.5mg q4–8 15mg q12 (SR) (30mg) 6–12y: 5–15mg q4–8 30mg q12 (SR) (60mg)	nausea, vomiting, drowsiness, **H**
Guaifenesin	expectorant	Robitussin, Organidin NR, Humibid LA	200–400mg q4 (2400mg)	2–6y: 50–100mg q4 (600mg) 6–12y: 100–200mg q4 (1200 mg)	nausea, vomiting, headache, rash, H

(continued)

TABLE C–1. *(continued)*

Drug*	Category	Brand Names	Adult Dose (max/day)	Pediatric Dose (max/day)	Side Effects/ Considerations
Hydrocodone w/homatropine)	antitussive	Hycodan	5–10mg q4–6 (90mg)	2–12y: 2.5mg q4–6 (30mg) >12y: 5mg q4–6 (60mg)	hypotension, sedation, GI upset, **H**
Phenylpropanolamine	decongestant	Rhindecon	25mg q4 or 50mg q8 75mg q12 (SR) (150mg)	2–5 y: 6.25mg q4 (37.5mg) 6–12 y: 12.5mg q4 (75mg)	hypertension at high doses, palpitations, insomnia, **H**
Pseudoephedrine	decongestant	Sudafed, Actifed Allergy, Efidac	30–60mg q4–6 120mg q12 (12 Hour) 240mg qd (Efidac) (240mg)	3–12mo: 3 drops/kg q6 (4 doses) 1–2y: 7 drops/kg q6 (4 doses) 2–5 y: 15mg q4–6 (60mg) 6–12 y: 30mg q4–6 (120mg)	tachycardia, palpitations, arrhythmias, insomnia, tremor, **H**

TABLE C–2. Single Agent Antihistamines

Drug	Brand Name	Adult Dosing	Pediatric Dosing	Onset (hr)	Duration (hr)	Comments
Azatadine	Trinaline	1–2mg q12	<12y: not recommended	2–4	12–24	moderate sedation, caution in combination with MAO inhibitors, **HR**
Brompheniramine	Dimetapp Allergy	4mg q4–6 8mg q8–12 (ER)	<6y: not recommended 6–12y: 2mg q4–6	3–9	12–30	low sedation, **HR**
Cetirizine	Zyrtec	5–10mg q24	2–5y: 2.5mg q24 6–12y: 5–10mg q24	4–8	8–9 (adults) 4–5 (children)	non sedating, **HR**
Chlorpheniramine	Chlor-Trimeton	4mg q4–6 8–12mg q12 (ER)	<2y: not recommended 2–6y: 1 mg q6 6–12y: 2 mg q6	1–2	20	low sedation, **HR**
Clemastine	Tavist	1tab q12	<6y: not recommended 6–12y: ½ tab to 1 tab q12	4–6	12–24	moderate sedation, **HR**

(continued)

TABLE C–2. *(continued)*

Drug	Brand Name	Adult Dosing	Pediatric Dosing	Onset (hr)	Duration (hr)	Comments
Diphenhydramine	Benadryl, Benylin	25–50mg q4–6	>10kg: 12.5–25mg q6–8	0.25–1	4–7	high sedation, also available IV/IM, **HR**
Fexofenadine	Allegra	60mg q12 180mg q24	<6y: not established 6–12y: 30mg q12	1–3	12–16	nonsedating Allegra D also available, **HR**
Hydroxyzine	Atarax, Vistaril	25–100mg q4–8 25–100mg q6–24 (IM)	<6y: 6.25–12.5mg q4–6 0.5–lmg/kg q4–6 (IM) 6–12y: 12.5–25mg q4–6 0.5–lmg/kg q4–6 (IM)	0.5	4–6	high sedation, not for SQ or IV, **H**
Loratadine	Claritin	10 mg q24	<6y: not recommended 6–12y: 10mg q24	1–3	24	nonsedating also available, Reddi Tabs, syrup, Clariton D-12 or D-24, **HR**

TABLE C–3. Common Combination Cough and Cold Products

Brand Name	Antihistamine	Decongestant	Analgesic	Expectorant	Antitussive	Adult Dose
Actifed	TRI	PE	—	—	—	1 tab or 10ml q4–6
Actifed with Codeine	TRI	PE	—	—	CO	10ml q4–6
Advil Cold & Sinus	—	PE	IB	—	—	1–2 caplets q4–6
Benadryl Allergy/Cold	DI	PE	APAP	—	—	1–2 tabs q4–6
Benylin Multisymptom	—	PE	—	G	DM	20ml q4
Chlordrine SR	CH	PE	—	—	—	1 tab q8–12
Chlor-Trimeton	CH	—	—	—	—	1 tab q4–6
Chlor-Trimeton 12 Hour	CH	PE	—	—	—	1 tab q12
Comtrex	CH	PP	APAP	—	DM	1–2 tabs q4–6
Contac 12 Hour	CH	PP	—	—	—	1 cap q12
Contac Day & Night	DI	PE	APAP	—	—	1 cap q6
Deconamine	CH	PE	—	—	—	1 tab or 5–10ml q6–8
Deconamine SR	CH	PE	—	—	—	1 cap q12
Deconsal II	—	PE	—	G	—	1–2 tabs q12
Dimetapp	BR	PP	—	—	—	1 tab or 10ml q4

(continued)

TABLE C–3. Common Combination Cough and Cold Products

Brand Name	Antihistamine	Decongestant	Analgesic	Expectorant	Antitussive	Adult Dose
Dimetapp Extentabs	BR	PP	—	—	—	1 tab q12
E.N.T. Tablets	CH/PY	PH	—	—	—	1–2 tabs q12
Entex	—	PH/PP	—	G	—	1 tab or 10ml q6
Entex LA	—	PP	—	G	—	1 tab q12
Entex PSE	—	PE	—	G	—	1 tab q12
Histussin D	—	PE	—	—	HY	5ml q6
Histussin HC	CH	PH	—	—	HY	10ml q4
Humibid DM	—	—	—	G	DM	1–2 tabs q12
Huumabid LA	—	—	—	G	—	1 tab q12
Hycodan w/homatropine	—	—	—	—	HY	1 tab q4–6
Hycomine	—	PP	—	—	HY	5ml q4
Naldecon	CH	PP/PH	—	—	—	1 tab q8 or 15ml q3–4
Naldecon DX	—	PP	—	G	DM	10ml q4
Novahistine	CH	PH	—	—	—	10ml q4
Ornade Spansules	CH	PP	—	—	—	1 cap q12
Phenergan VC	PRO	PH	—	—	—	5ml q4–6
Phenergan VC/Codeine	PRO	PH	—	—	CO	5ml q4–6
Robitussin	—	—	—	G	—	10ml q4
Robitussin AC	—	—	—	G	CO	10ml q4
Robitussin DM	—	—	—	G	DM	10ml q4

Brand Name	Antihistamine	Decongestant	Analgesic	Expectorant	Antitussive	Adult Dose
Robitussin CF	—	PP	—	G	DM	10ml q4
Robitussin Pediatric	—	—	—	—	DM	2.5–10mg q4
Rondec	CX	PE	—	—	—	5ml or 1 tab q6
Rondec DM	CX	PE	—	—	DM	5ml q6
Rondec Oral Infant Drops	CX	PE	—	—	—	0.25–1 ml q6
Rynatan	CH/PY	PH	—	—	—	1–2 tab q12
Sudafed	—	PE	—	—	—	30–60mg q4–6
Sudafed Cold & Allergy	CH	PE	—	—	—	1 tab q4–6
Tavist	CL	—	—	—	—	1–2 tab q12
Tavist-D	CL	PP	—	—	—	1 tab q12
Triaminic	CH	PP	—	—	—	20ml q4
Triaminic DM	—	PP	—	—	DM	20ml q4
Triaminic Infant Drop	PY/PR	PP	—	—	—	1 drop/kg q6
Tussar DM	CH	PE	—	—	DM	10ml q6
Tussionex	CH	—	—	—	HY	5ml q12
Tussi Organidin DM	—	—	—	G	DM	10ml q4
Tussi-Organidin NR	—	—	CO	G	—	10ml q4
Vanex Forte	CH/PY	PP/PH	—	—	—	1 tab q12
Vicodin Tuss	—	—	—	G	HY	5mg q6 (w/ meals)

(continued)

TABLE C–3. *(continued)*

Abbreviations

APAP—acetaminophen	BR—brompheniramine	CH—chlorpheniramine;	CL—clemastine
CO—codeine	CX—cabinoxamine	DI—diphenhydramine	DIVI—clextromethorphan
FE—fexofenadine	G—guaifenesin	HY—hydrocodone	IB—ibuprofen
LO—Ioratacline	PE—pseudoephedrine	PR—pheniramine	PH—phenylephrine
PP—phenylpropanolamine	PRO—promethazine	PY—pyrilamine	TRI—triprolidine

TABLE C–4. Topical Nasal Decongestants*

Drug	Brand Names	Adult Dose	Pediatric Dose
Phenylephrine (0.125%, 0.16%, 0.25%)	Neo-Synephrine	2–3 sprays each nostril q3–4	<2y: not recommended 2–6 y: (0.125%) 1–2 drops each nostril q4 6–12y: (0.25%) 1–2/drops in each nostril q4
Oxymetazoline HCI (0.025%, 0.05%)	Afrin, Dristan LA,	2–3 sprays/drops each nostril q12	<2y: not recommended 2–5y: (0.025%) 2–3 drops nostril q12 6–12y: (0.025% or 0.05%) 2–3 drops each nostril q12
Xylometazoline HCI (0.05%, 0.1%)	Otrivin	2–3 sprays each nostril q8–10	<2y: not recommended 2–12y: (0.05%) 2–3 drops each nostril q8–10

*Limit duration of therapy to 3–5 days to prevent rhinitis medicamentosa

TABLE C–5. Intranasal Corticosteroids

Drug*	Brand Names	Comments
Beclomethasone	Beconase, Vancenase	Beclomethasom/Vancenase available in aqueous (AQ), Vancenase available in double strength (DS)
Budesonide	Rhinocort	Available in aqueous (AQ)
Dexamethasone	Dexacort Turbinaire	—
Flunisolide	Nasarel, Nasalide	—
Fluticasone	Flonase	Approved for 4–6 years old
Mometasone	Nasonex	Approved for 3–6 years old
Triamincinolone	Nasacort, Nasacort AQ	Available in aqueous (AQ)

*Doses vary from 1 to 2 puff each nares Q-12 to Q-24.

TABLE C–6. Otic Preparations

Brand Name	Active Ingredient(s)	Comments
Auralgan	benzocaine, antipyrine	anesthetic/ceruminolytic
Bleph-10	sulfacetamide	antimicrobial contraindicated in sulfa allergy
Debrox, Murine Ear	carbamide peroxide	ceruminolytic
Domeboro Otic	acetic acid	antimicrobial
Cipro HC Otic	ciprofloxacin, hydrocortisone,	antimicrobial/antiinflammatory, not recommended <1y
Cortisporin Otic	hydrocortisone polymyxin, neomycin	antimicrobial/antiinflammatory, available as solution or suspension, may be toxic in middle ear (controversial)
Floxin Otic	ofloxacin	antimicrobial, not recommended <1 y
Vosocidin	prednisolone acetate, sulfacetamide	antimicrobial/anti-inflammatory contraindicated in sulfa allergy
VoSol Otic	acetic acid	antimicrobial
VoSol HC	acetic acid, hydrocortisone	antimicrobial/anti-inflammatory

TABLE C–7. Gastroesophageal Reflux Agents

	Drug	Brand Name	Adult Dosing	Pediatric Dosing	Comments
H_2 Receptor Antagonists	Cimetidine	Tagamet	400mg q6 or 800mg q12 300mg q6-8 (IV)	neonates: 5-10mg/kg/d q8-12 (PO/IV) infants: 10-20mg/kg/d q6-12 (PO/IV) children: 20-40mg/kg/d q6 (PO/IV)	GI upset, p450 interactions, thrombocytopenia,neutropenia, agranulocytosis, **HR**
	Famotidine	Pepcid	20-40mg q12 20mg q12 (IV)	0.5mg/kg q8-12 0.5-1mg/kg q12 (IV)	GI upset, thrombocytopenia,neutropenia, agranulocytosis, **HR**
	Nizatidine	Axid	150mg q12	not well established	GI upset, thrombocytopenia,neutropenia, agranulocytosis, **R**
	Ranitidine	Zantac	150mg q12 50mg q8 (IV)	>1mo: 1.25-2mg/kg q12 0.1-0.8mg/kg q6-8 (IV)	GI upset, thrombocytopenia, neutropenia, agranulocytosis, **R**
Prokinetic Agents	Cisapride	Propulsid	10-20mg q6 before meals	0.15-0.3mg/kg q6-8 before meals	GI upset, prolongs QT interval (ECG monitoring required), contraindicated with azole antifungals and macrolides, **HR**
	Metoclopramide	Reglan	10mg q6 5-10mg q6-8 (IV) before meals	0.125mg/kg q6 0.1mg/kg q6 (IV) before meals	GI upset, sedation, extrapyramidal symptoms, may potentiate seizures **R**

	Drug	Brand Name	Adult Dosing	Pediatric Dosing	Comments
Proton-Pump Inhibitors	Omeprazole	Prilosec	20mg q12-24	<3y: 0.5mg/kg q24 >3y: 10-20mg q24	GI upset, headache, myalgias, p450 interactions, **H**
	Lansoprazole	Prevacid	15-30mg q24	not well established	GI upset, headache, myalgias p450 interactions, **H**
Mucosal Protectants	Rabeprazole	Aciphex	20-40mg q24	not well established	GI upset, headache, myalgias, p450 interactions, **H**
	Sucralfate	Carafate	1gm q6 before meals	0.5gm q6 before meals	constipation, aluminum toxicity in ESRD, interferes with absorption of many drugs, **NA**

TABLE C–8. Antimicrobials

Antibiotic	Brand Name	Adult Dose	Pediatric Dose	Comments
Amikacin	Amikin	5-7.5mg/kg q8-12 or 15-20mg/kg q24 (IV)	5-7.5mg/kg q8-12 (IV)	nephrotoxicity, ototoxicity, monitor serum levels **R**
Amoxicillin	Amoxil	250-500mg q8 (PO)	6.6-13.3mg/kg q8 (PO)	GI upset, rash, **R**
Amoxicillin/ Clavulanate	Augmentin	500mg q8, or 875mg q12 (PO)	6.6-13.3mg/kg q8 (PO)	GI upset, **R**
Ampicillin	Omnioen, Principen	250-500mg q6 (PO) 500mg-2gm q4-6 (IV/IM)	6.25-25mg/kg q6 (PO/IV/IM)	GI upset, CNS toxicity, **R**
Ampicillin/ Sulbactam	Unasyn	1.5-3gm q6 (IV)	25-50mg/kg q6 (IV)	GI upset, CNS toxicity, anaerobic coverage, neutropenia, thrombocytopenia, elevated LFTs, **R**
Azithromycin	Zithromax	500mg, then 250mg q24 x 2-5d (PO) 500mg q24 (IV)	10 mg/kg, then 5mg/kg/d q24 for days 2-5 (PO)	GI upset, arrhythmias, elevated LFTs, cholestatic jaundice, **H**
Aztreonam	Azactam	1-2gm q8-12 (IV/IM)	30-50mg/kg q8-12 (IV/IM)	GI upset, thrombophlebitis; less risk of cross-allergenicity with beta-lactams, **R**
Cefaclor-2	Ceclor	250-500mg q8 or 500mg q12 ER	6.6-13.3mg/kg q8-12 (PO)	pseudomembranous colitis, diarrhea, Stevens-Johnson syndrome, rare reports of neutropenia, hemolytic anemia, eosinophilia, **R**
Cefadroxil-1	Duricef	500-1000mg q12-24 (PO)	PO: 15mg/kg q12 (PO)	diarrhea, neutropenia, **R**

Antibiotic	Brand Name	Adult Dose	Pediatric Dose	Comments
Cefazolin-1	Ancef, Kefzol	1-2gm q8 (IV)	>1mo: 15-35mg/kg q8 (IV)	diarrhea, seizures, rare reports of neutropenia, leukopenia, thrombocytopenia, elevated LFTs, cholestatic jaundice, **R**
Cefepime-4	Maxipime	1-2gm q12 (IV)	50mg/kg q12 (IV)	diarrhea, elevated LFTs, phlebitis, **R**
Cefixime-3	Suprax	400mg q12-24 (PO)	4-8mg/kg q12-24 (PO)	rash, GI upset, neutropenia, leukopenia, thrombocytopenia, eosinophilia, elevated LFTs, **HR**
Cefmetazole-2	Zefazone	2gm q6-12 (IV)	not recommended	rash, GI upset, anemia, **R**
Cefonicid-2	Monocid	1-2gm q24 (IV/IM)	not recommended	rash, elevated LFTs, **R**
Cefoperazone-3	Cefobid	1-2gm q12 (IV/IM)	25-100mg/kg q12 (IV/IM)	diarrhea, phlebitis **HR**
Cefotaxime-3	Claforan	1-2gm q6-8 (IV/IM)	10-50mg/kg q6 (IV/IM)	GI upset, thrombocytopenia, neutropenia, elevated LFTs, **HR**
Cefotetan-2	Cefotetan	1-2gm q12 (IV/IM)	20-40 mg/kg q12 (IV/IM)	anaerobic coverage, rash, diarrhea, elevated LFTs, **R**
Cefoxitin-2	Mefoxin	1-2gm q6 (IV/IM)	>3mo: 20-25mg/kg q4-6 (IV/IM)	anaerobic coverage, diarrhea, leukopenia, thrombocytopenia, eosinophilia, **R**
Cefpodoxime-3	Vantin	100-400mg q12 (PO)	5mg/kg q12 (PO)	diarrhea, GI upset, **HR**
Cefprozil-3	Cefzil	250-500mg q12-24 (PO)	15mg/kg q12-24 (PO)	GI upset, elevated LFTs, cholestatic jaundice, **R**
Ceftazadime-3	Fortaz	1-2gm q8 (IV/IM)	30-50mg/kg q8 (IV/IM)	diarrhea, pain, eosinophilia, hemolytic anemia, **R**

(continued)

TABLE C–8. *continued*

Antibiotic	Brand Name	Adult Dose	Pediatric Dose	Comments
Ceftizoxime-3	Ceftizox	1-2gm q8-12 (IV/IM)	33-50mg/kg q6-8 (IV/IM)	anemia, leukopenia, neutropenia, thrombocytopenia, **R**
Ceftriaxone-3	Rocephin	1-2gm q12-24 (IV/IM)	50 mg/kg q24 (IV/IM)	diarrhea, leukopenia, eosinophilia, elevated LFTs, **HR**
Cefuroxime-2	Ceftin, Kefurox, Zinacef	250-500mg q12 (PO) 750mg-1.5gm q8 (IV/IM)	10mg/kg q12 (PO) 25-50mg/kg q8 (IV/IM)	anemia, eosinophilia, elevated LFTs, thrombophlebitis, neutropenia, leukopenia, **R**
Cephalexin-1	Keflex, Keftab	250-500mg q6 (PO)	6.25-12.5mg/kg q6 (PO)	diarrhea, neutropenia, **R**
Cephalothin-1	Keflin	500-2000mg q4-6 (IV/IM)	10-30mg/kg q4-6 (IV/IM)	GI upset, thrombocytopenia, **R**
Chloramphenicol	Chloromycetin	12.5-25mg/kg q6 (PO/IV)	>30d: 12.5-25mg/kg q6-12 (PO/IV)	blood dyscrasias, gray baby syndrome, bone marrow suppression, aplastic anemia, **HR**
Ciprofloxacin	Cipro	500-750mg q12 (PO) 200-400mg q12 (IV)	not recommended in children <18y due to arthropathy	GI upset, seizures, acute renal failure, interacts with antacids, iron or carafate, **HR**
Clarithromycin	Biaxin	250-500mg q12 (PO)	7.5mg/kg q12 (PO)	GI upset, headache, torsades de pointes, leukopenia, **HR**
Clindamycin	Cleocin	150-450mg q6 (PO) 600-900mg q6-12 (IV/IM)	2.5-10mg/kg q6-8 (PO) 6.25-13mg/kg q6-8 (IV/IM)	diarrhea, pseudomembranous colitis, Stevens-Johnson syndrome, **H**
Dicloxacillin	Dynapen, Pathocil	500mg q6 (PO)	4-8mg/kg q6 (PO)	diarrhea, neutropenia, leukopenia, thrombocytopenia, elevated LFTs, **R**
Dirithromycin	Dynabac	500mg q24 (PO)	not well established	hyperkalemia, GI upset, neutropenia, thrombocytopenia, increased LFTs, **H**

Antibiotic	Brand Name	Adult Dose	Pediatric Dose	Comments
Doxycycline	Vibramycin, Doryx, Vibra-tab	100-200 mg q12-24 (PO/IV)	>8y: 1-2mg/kg q12 (PO/IV)	discoloration of teeth, esophagitis, neutropenia, hepatotoxicity, **HR**
Erythromycin	Ery-Tab, Eryc, PCE, E.E.S., Eryped, Ilosone	250-500mg q6 (PO) 500-1000mg q6 (IV)	7.5-15mg/kg q6-8 (PO) 5-10mg/kg q6 (IV)	ototoxicity with high IV doses, GI upset, jaundice, p450 interactions, **H**
Gatifloxacin	Tequin	400mg q24 (PO/IV)	not recommended in children <18y due to arthropathy	GI upset, seizures, acute renal failure, interacts with antacids, iron or carafate, **HR**
Gentamicin	Garamycin	1-3mg/kg q8-12 or 4-7mg/kg q24 (IV)	1.5-2.5mg/kg q8-12 (IV)	nephrotoxicity, ototoxicity, monitor serum levels **R**
Imipenem/ Cilastatin	Primaxin	500mg q6 (IV)	15-25mg/kg q6 (IV)	seizures, GI upset, elevated LFTs, **R**
Levofloxacin	Levaquin	500mg q24 (PO/IV)	not recommended in children <18y due to arthropathy	GI upset, seizures, acute renal failure, interacts with antacids, iron or carafate, **HR**
Linezolid	Zyvox	600mg q12 (IV/PO)	not well established	GI upset, headache, elevated LFTs, **HR**
Loracarbef-3	Lorabid	200-400mg q12-24 (PO)	7.5-15mg/kg q12 (PO)	GI upset, leukopenia, thrombocytopenia, elevated LFTs, **R**
Meropenem	Merrem IV	1-2gm q8 (IV)	20mg/kg q8 (IV)	seizures, GI upset, **R**
Metronidazole	Flagyl	500mg q8 (PO/IV)	7.5mg/kg q8-12 (PO/IV)	GI upset, disulfiram reaction, **HR**
Mezlocillin	Mezlin	1.5-4gm q4-6 (IV/IM)	50mg/kg q4-6 (IV/IM)	GI upset, seizures, coagulopathy, **R**

(continued)

TABLE C–8. *continued*

Antibiotic	Brand Name	Adult Dose	Pediatric Dose	Comments
Nafcillin	Unipen	1-2q 4-6 (IV/IM)	12.5-25mg/kg q6 (IV/IM)	coagulopathy, interstitial nephritis, neutropenia, hepatotoxicity, **H**
Nitrofurantoin	Macrobid	50-100mg q6 (PO)	1.25-2mg/kg q6 (PO)	GI upset, fever, chills, primarily for urinary tract infections, **HR**
Norfloxacin	Noroxin	400mg q12	not recommended in children <18y due to arthropathy	GI upset, elevated LFTs, increases BUN and creatinine, **R**
Ofloxacin	Floxin	200-400mg q12 (PO/IV)	not recommended in children <18y due to arthropathy	GI upset, seizures, acute renal failure, interacts with antacids, iron or carafate, **HR**
Oxacillin	Bactocill	500-1000mg q4-6 (PO) 250-1000mg q4-6 (IV/IM)	12.5-25mg/kg q6 (PO) 37.5-50mg/kg q6 (IV/IM)	GI upset, neutropenia, hepatotoxicity, **R**
Penicillin G	Wycillin (procaine), Bicillin L-A (benzathine), Pfizerpen	500-1000mg q6 (PO) 1-4MU q4-6 (IV) 0.6-1.2MU q12 (IM)	6.25-12.5mg/kg q6 (PO) <7d: 25,000U/kg q12 >7d: 25,000U/kg q6-12 (IV/IM)	neurotoxicity, hemolytic anemia, interstitial nephritis, neutropenia, **R**
Penicillin V	Pen-Vee K	250-500mg q6-8 (PO)	6.25-12.5mg/kg q6 (PO)	GI upset, interstitial nephritis, seizures, **R**
Pipercillin	Pipracil	3-4gm q4-6 (IV/IM)	50mg/kg q4-6 (IV/IM)	seizures, hemolytic anemia, neutropenia, **R**
Pipercillin Tazobactam	Zosyn	3.375gm q4-6 (IV) 2.25gm q6-8 (IM)	<12: not well established	GI upset, leukopenia, serum sickness, **R**
Quinupristin/Dalfopristin	Synercid	7.5mg/kg q8 (IV)	not well established	myalgia, arthralgia, GI upset, elevated LFTs, **H**

Antibiotic	Brand Name	Adult Dose	Pediatric Dose	Comments
Rifampin	Rifadin	300-600mg q12 (PO/IV)	7.5mg/kg q12 (PO/IV)	hepatotoxicity, discoloration of saliva, sweat, urine, and feces; peripheral neuropathy, blood dyscrasias, **H**
Streptomycin	Streptomycin sulfate	1gm q12 (IM)	20-40mg/kg/d q6-12 (IM)	nephrotoxicity, ototoxicity, monitor serum levels **R**
Tetracycline	Sumycin	250-500mg q6 (PO)	>8y: 6.25-12.5mg/kg q6 (PO)	azotemia, hepatotoxicity, bone growth retardation, discoloration of teeth, photosensitivity, **HR**
Ticarcillin	Ticar	1-4gm q4-6 (IV/IM)	50mg/kg q4-6 (IV/IM)	seizures, hemolytic anemia, interstitial nephritis, **R**
Ticarcillin/ Clavulanate	Timentin	3.1gm q4-6 (IV)	50mg/kg q4-6 (IV)	CNS toxicity, hemolytic anemia, interstitial nephritis, **R**
Tobramycin	Nebcin	1-3mg/kg q8-12 or 4-7mg/kg q24 (IV)	1.5-2.5mg/kg q8-12 (IV)	nephrotoxicity, ototoxicity, monitor serum levels **R**
Trimethoprim-sulfamethoxazole	Bactrim, Cotrim, Septra, Sulfatrim	1 tab DS q12 (PO) 3-5mg/kg q6-12 (IV)	3-5mg/kg q6-12 (PO/IV)	GI upset, Stevens-Johnson syndrome, toxic epidermal necrolysis, blood dyscrasias, hepatitis, electrolyte abnormalities; increases BUN and creatinine, **R**
Vancomycin	Vancocin	10-15 mg/kg q12 (IV)	10mg/kg q6 (IV)	GI upset, interstitial nephritis, thrombocytopenia, red man syndrome, ototoxicity, **R**

TABLE C–9. Spectrum of Activity of Various Antimicrobials

		β-Lactamase Susceptible						β-Lactamase-Resistant									
		Non-APP			APP			APP			Other						
		Amoxicillin	Ampicillin	Penicillin	Mezlocillin	Piperacillin	Ticarcillin	Dicloxacillin	Nafcillin	Oxacillin	Amoxicillin/Clavulanate	Ampicillin/Sulbactam	Piperacillin/Tazobactam	Ticarcillin/Clavulanate	Aztreonam	Imipenem/Cilastatin	Meropenem
Gram Positive Cocci	Streptococcus pyogenes/ viridans (Groups A, B, C, G, F)	1	1	1	3	3	3	3	3	3	2	2	3	3	-	3	3
	Streptococcus pneumoniae	1	1	1	3	3	3	3	3	3	1	3	3	3	–	3	3
	Staphylococcus epidermidis (methicillin-sensitive - MSSE)	–	–	–	–	–	–	2	1	1	2	2	3	–	–	3	3
	Staphylococcus epidermidis (methicillin-resistant - MRSE)	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–
	Staphylococcus aureus (methicillin-sensitive- MSSA)	1	1	1	–	–	–	1	1	1	2	2	2	3	–	3	3
	Staphylococcus aureus (methicillin-resistant -MRSA)	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–
	Enterococcus faecalis	1	1	2	2	2	3	–	–	–	2	2	2	3	–	2	–
	Enterococcus faecium	1	1	2	3	3	3	–	–	–	2	2	3	3	–	3	–
Gram Positive Bacilli	Actinomyces israelii	1	1	1	–	3	–	–	–	–	2	3	2	–	–	3	–
	Bacillus anthracis	–	–	1	–	–	–	–	–	–	–	–	–	–	–	–	–
	Corynebacterium diptheriae	–	–	2	–	–	–	–	–	–	–	–	–	–	–	–	–
	Listeria, monocytogenes	1	1	1	2	2	2	–	–	–	2	2	–	2	–	3	3
	Nocardia, sp.	–	–	–	–	–	–	–	–	–	2	–	–	–	–	2	–
Gram Negative Cocci	Moraxella catarrhalis	–	–	–	–	3	–	–	–	–	1	3	3	3	3	3	3
	Neisseria gonorrhoeae	1	1	1	3	3	3	–	–	–	3	3	3	3	3	3	3
	Neisseria meningitis	3	3	1	3	3	3	–	–	–	3	3	3	3	3	3	3
Gram Negative Cocci-Bacilli	Chlamydia sp.	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–
	Bordetella pertussis	–	2	–	–	–	–	–	–	–	–	–	–	–	–	–	–
	Helicobacter pylori	–	2	–	–	–	–	–	–	–	–	–	–	–	–	–	–
	Hemophilus influenzae	3	3	–	3	3	3	–	–	–	1	2	2	2	3	3	3
	Legionella pneumophilia	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–
Gram Negative Enterobacteriaceae	Citrobacter sp.	–	–	–	3	2	3	–	–	–	–	–	2	2	2	1	2
	Enterobacter	–	–	–	1	1	1	–	–	–	–	3	2	2	3	3	3
	Escherichia coli	–	–	–	2	2	3	–	–	–	2	2	2	2	3	3	3
	Klebsiella pneumoniae	–	–	–	2	2	–	–	–	–	2	2	2	2	3	3	3
	Proteus, mirabilis	1	1	–	2	2	2	–	–	–	2	2	2	2	3	3	3
	Proteus vulgaris	–	–	–	2	2	2	–	–	–	2	2	2	2	3	3	3
	Providencia sp.	–	–	–	2	2	2	–	–	–	2	2	2	2	3	3	3
	Serratia sp.	–	–	–	2	–	2	–	–	–	–	–	2	2	3	3	3
Gram Negative Bacilli	Acinetobacter sp.	–	–	–	2	1	2	–	–	–	–	1	2	1	–	1	1
	Burkholderia (pseudomonas) cepacia	–	–	–	–	–	–	–	–	–	–	–	–	–	–	1	2
	Pseudomonas aeruginosa	–	–	–	3	1	2	–	–	–	–	–	1	1	2	2	2
	Stenotrophomonas (xanthomonas) maltophilia	–	–	–	–	–	–	–	–	–	–	–	–	2	–	–	–
Anaerobes	Bacteroides	–	–	–	3	3	3	–	–	–	2	2	2	2	–	3	3
	Fusobacterium sp.	–	–	1	2	–	–	–	–	–	–	–	2	–	–	–	–
	Prevotella sp.	1	1	1	3	3	3	–	–	–	2	2	3	3	–	3	3
	Clostridium difficile	–	–	2	2	2	–	–	–	–	–	2	–	–	–	3	3
	Clostridium perfringens	2	2	1	3	3	3	–	–	–	1	1	1	2	–	3	3
	Peptostreptococcus sp.	1	1	1	3	3	3	3	3	3	2	3	3	3	–	3	3

–: not well studied, or not clinically efficacious

1: clinically effective with good susceptibility profile. Should be first-line therapy, when appropriate.

2: literature supports use, has acceptable susceptibility profile

3: not well studied, but has some activity against the organism. Combination or alternative therapy may be required.

+: synergistic when in combination with appropriate first-line agent

U: acceptable for use in urinary tract infections only.

Cephalosporins																						Aminoglycosides				Macrolides			
1st				2nd						3rd										4th									
Cefadroxil	Cefazolin	Cephalexin	Cephalothin	Cefaclor	Cefmetazole	Cefonicid	Cefotetan	Cefoxitin	Cefuroxime	Cefixime	Cefoperazone	Cefotaxime	Cefpodoxime	Cefpozil	Ceftazidime	Ceftibuten	Ceftizoxime	Ceftriaxone	Loracarbef	Cefepime	Amikacin	Gentamicin	Streptomycin	Tobramycin	Azithromycin	Clarithromycin	Dirithromycin	Erythromycin
1	1	1	1	2	3	2	3	3	2	3	3	3	3	2	3	3	3	3	2	3	–	+	–	–	2	2	2	2
3	2	2	2	3	3	2	3	3	2	3	3	2	2	2	3	3	3	2	3	3	3	3	2	3	2	2	2	2
1	1	1	1	2	3	2	3	3	2	–	3	3	2	2	3	–	2	3	2	3	+	+	–	+	2	2	3	3
–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	+	–	–	3	3	3	2
1	1	1	1	2	3	2	3	3	2	–	2	3	3	2	2	–	2	3	2	3	+	+	–	+	2	2	3	3
–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	+	–	–	–	–	–	–
–	–	–	–	–	–	–	–	3	–	–	–	3	–	–	–	–	–	–	–	–	3	+	3	3	–	–	–	–
–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	+	–	–	–	–	–	–
–	–	–	2	–	–	–	–	–	–	–	–	3	–	–	–	3	2	–	–	–	–	–	–	–	3	3	–	3
–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	2
–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	1
–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	3
–	–	–	–	–	–	–	–	–	3	–	–	–	–	–	–	–	–	–	–	–	3	–	3	–	–	–	–	–
2	2	–	2	2	3	3	2	3	2	2	2	3	2	2	3	2	3	2	2	3	2	2	–	2	1	1	2	2
–	3	–	3	3	2	3	3	3	3	1	3	3	1	3	3	2	3	1	3	2	–	–	–	–	3	3	2	2
–	–	–	–	3	2	3	3	2	2	3	3	2	–	3	3	2	3	2	3	3	–	–	–	–	–	–	3	3
–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	1	3	–	2
–	–	–	–	–	–	–	2	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	2	2	–	1
–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	2	–	–
–	2	–	2	2	3	2	2	2	1	1	2	2	2	1	1	2	2	2	2	3	3	3	3	3	2	3	3	3
–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	2	2	2	1
–	–	–	–	3	3	3	3	3	3	3	2	2	3	–	2	–	3	3	3	2	2	1	–	1	–	–	–	–
–	–	–	–	–	–	3	3	–	3	–	2	2	–	–	2	–	2	2	–	2	3	2	3	2	–	–	–	–
2	2	2	2	3	2	2	2	2	2	3	3	3	3	3	3	–	2	3	2	3	3	3	3	3	–	–	–	–
2	2	2	2	2	2	2	1	1	1	3	2	1	–	3	1	–	1	1	3	2	3	1	–	1	–	–	–	–
2	2	2	2	2	2	2	2	2	2	2	2	2	3	3	2	–	2	2	3	3	2	3	3	3	–	–	–	–
–	–	–	–	–	2	2	2	2	–	3	1	1	3	–	1	–	1	1	–	3	2	3	–	3	–	–	–	–
–	–	–	–	–	2	2	2	2	2	2	1	3	3	–	1	–	1	1	–	3	3	3	–	3	–	–	3	–
–	–	–	–	–	–	–	2	–	–	3	2	1	–	–	1	–	2	2	–	3	2	1	–	3	–	–	–	–
–	–	–	–	–	–	–	–	–	3	–	3	2	–	–	2	–	2	2	–	3	+	3	–	+	–	–	–	–
–	–	–	–	–	–	–	–	–	–	–	2	2	–	–	2	–	2	2	–	3	–	–	–	–	–	–	–	–
–	–	–	–	–	2	–	2	2	–	–	–	3	2	–	–	–	3	–	–	–	–	–	–	–	–	–	–	–
–	–	–	–	–	–	3	–	–	3	–	–	3	3	–	–	–	–	–	–	3	–	–	–	–	–	–	–	–
–	–	–	–	2	1	–	1	1	3	2	3	3	–	2	2	–	3	–	–	–	–	–	–	–	2	2	–	–
–	–	–	–	–	–	–	2	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–
–	3	–	–	–	2	3	2	3	3	–	2	2	–	3	2	–	3	2	3	–	–	–	–	–	2	3	3	3
–	2	–	–	2	2	2	2	2	2	2	2	2	–	2	2	–	2	3	–	2	–	–	–	–	–	3	–	–

(continued)

TABLE C–9. *(continued)*

		Quinolones						Miscellaneous							
		Ciprofloxacin	Sparofloxacin	Lomefloxacin	Levofloxacin	Gatifloxacin	Ofloxacin	Chloramphenicol	Clindamycin	Linezolid	Metronidazole	Quinupristin-Dalfopristin	Rifampin	Tetracycline	Trimethoprim-Sulfamethoxazole
Gram Positive Cocci	Streptococcus pyogenes/ viridans (Groups A, B, C, G, F)	3	2	–	2	2	3	3	2	2	–	2	2	3	2
	Streptococcus pneumoniae	–	2	–	2	2	–	–	2	+	–	3	+	–	2
	Staphylococcus epidermidis (methicillin-sensitive - MSSE)	3	3	3	3	3	3	–	2	+	–	2	+	–	3
	Staphylococcus epidermidis (methicillin-resistant - MRSE)	3	3	–	3	3	3	3	2	+	–	2	+	3	2
	Staphylococcus aureus (methicillin-sensitive- MSSA)	3	3	3	3	3	3	–	2	+	–	2	+	–	3
	Staphylococcus aureus (methicillin-resistant -MRSA)	–	–	–	–	–	–	–	–	+	–	2	+	–	+
	Enterococcus faecalis	U	2	2	2	2	U	3	–	+	–	2	+	3	–
	Enterococcus faecium	–	3	–	3	3	–	3	–	–	–	2	–	–	–
Gram Positive Bacilli	Actinomyces israelii	–	–	–	–	–	3	3	3	–	–	3	–	2	–
	Bacillus anthracis	1	–	–	–	–	–	3	–	–	–	–	–	2	–
	Corynebacterium diptheriae	–	–	–	–	–	–	–	2	3	–	–	3	–	3
	Listeria, monocytogenes	3	–	–	–	–	–	3	–	–	–	3	–	3	2
	Nocardia, sp.	–	–	–	–	–	–	–	–	–	–	–	–	2	1
Gram Negative Cocci	Moraxella catarrhalis	2	2	2	2	2	2	3	–	2	–	3	2	3	1
	Neisseria gonorrhoeae	2	2	2	2	2	2	3	–	3	–	3	2	3	3
	Neisseria meningitis	3	2	–	3	3	3	3	–	3	–	3	3	3	2
Gram Negative Cocci-Bacilli	Chlamydia sp.	–	3	–	3	3	2	–	–	–	–	–	–	1	–
	Bordetella pertussis	–	–	–	3	3	–	–	–	–	–	–	–	–	–
	Helicobacter pylori	–	–	–	–	–	–	–	–	–	1	–	–	1	–
	Hemophilus influenzae	2	2	3	2	2	2	3	–	2	–	3	2	3	2
	Legionella pneumophilia	2	3	2	2	2	2	–	–	–	–	3	–	–	2
Gram Negative Enterobacteriaceae	Citrobacter sp.	2	3	2	3	3	2	3	–	–	–	–	–	–	–
	Enterobacter	1	1	1	1	1	1	–	–	–	–	–	–	–	–
	Escherichia coli	2	2	2	2	2	2	3	–	–	–	–	–	3	2
	Klebsiella pneumoniae	2	2	2	2	2	2	3	–	–	–	–	–	–	2
	Proteus, mirabilis	2	3	2	2	2	2	–	–	–	–	–	–	–	–
	Proteus vulgaris	2	3	2	3	3	2	3	–	–	–	–	–	–	–
	Providencia sp.	2	2	3	3	3	2	–	–	–	–	–	–	–	–
	Serratia sp.	2	2	3	3	3	2	–	–	–	–	–	–	–	3
Gram Negative Bacilli	Acinetobacter sp.	2	3	–	2	2	3	–	–	–	–	–	–	3	–
	Burkholderia (pseudomonas) cepacia	1	–	–	–	–	–	2	–	–	–	–	–	–	1
	Pseudomonas aeruginosa	2	–	3	3	3	3	–	–	–	–	–	–	–	–
	Stenotrophomonas (xanthomonas) maltophilia	–	3	–	–	–	–	2	–	–	–	–	–	–	2
Anaerobes	Bacteroides	–	–	–	–	–	–	3	1	–	1	3	–	3	–
	Fusobacterium sp.	1	–	–	–	–	–	3	1	–	1	3	–	3	–
	Prevotella sp.	–	–	–	–	–	–	3	1	–	1	3	–	3	–
	Clostridium difficile	–	–	–	–	–	–	3	–	–	1	3	–	3	–
	Clostridium perfringens	3	–	–	3	3	3	3	2	–	1	3	–	2	–
	Peptostreptococcus sp.	3	2	–	2	3	3	2	1	–	2	3	–	–	–

–: not well studied, or not clinically efficacious
1: clinically effective with good susceptibility profile. Should be first-line therapy, when appropriate.
2: literature supports use, has acceptable susceptibility profile
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