Head and Neck Imaging
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CHAPTER 10

The central nervous system (CNS) includes the skull, brain, spine, and spinal cord. The head and neck region includes the face, eye and orbit, nasal cavity and paranasal sinuses, ear and temporal bone, oral cavity, jaw, and neck. Modalities used for the imaging of the pediatric CNS and head and neck region include plain film/computerized radiography (PF/CR), ultrasonography (US), computed tomography (CT), multidetector CT (MDCT), magnetic resonance imaging (MRI), radionuclide imaging (RI), catheter angiography, and cerebrospinal fluid (CSF) imaging (e.g., CT myelography). Imaging modalities may be classified as structural or functional. Structural imaging modalities provide spatial resolution primarily on the basis of anatomic or morphologic data (e.g., CT). Functional imaging modalities (including molecular imaging) provide spatial resolution on the basis of physiologic, metabolic, or biologic data or markers (e.g., positron emission tomography [PET]). Some modalities may actually be considered to provide both structural and functional information (e.g., MRI, PET-CT). The technical and procedural descriptions for angiography, myelography, and other invasive and interventional modalities are detailed in other texts. In this chapter, guidelines for utilization are presented by region and modality.

HEAD AND NECK IMAGING GUIDELINES

Plain Films and Computerized Radiography
PF/CR is only occasionally used for the initial assessment of facial, orbit, sinus, or jaw trauma (e.g., panoramic [Panorex] tomography), radiopaque foreign body, or sinus infection. For the face, orbits, and sinuses, the series usually includes straight and angled frontal views (e.g., Caldwell and Waters views, respectively) in the upright position (i.e., for air-fluid levels), and occasionally lateral views. PF/CR of the petrous temporal bone or mastoid is rarely needed (e.g., cochlear implant evaluation). This may include frontal, Towne, or angled lateral (Laws or Owens) views. Lateral neck PF/CR may be performed initially to assess upper airway abnormalities (e.g., infection) or, along with frontal neck and chest PF/CR, for foreign body ingestion or aspiration. Head and spine PF/CR is also part of the skeletal survey that may be used to assess for nonaccidental injury, skeletal dysplasia, or multiple congenital anomalies/deformities.

Ultrasoundography
Real-time US is often very useful in evaluating head and neck masses with regard to size, location, and tissue characteristics. High-frequency (7.5- or 10-MHz) transducers permit the differentiation of solid from cystic elements and the detection of calcification. Oscillations may indicate the fluid nature of an apparently solid lesion (e.g., abscess). Doppler US imaging provides information about the extent of vascularity, flow direction, and pulsatility, and enables the differentiation of arterial from venous flow. This is particularly important in the assessment of vascular anomalies (e.g., hemangioma). US assessment of the thyroid gland is useful for detection and characterization of cysts and masses. Ocular US is primarily used in ophthalmology.

Computed Tomography
CT has for the most part replaced PF/CR as the primary modality for imaging of the pediatric head and neck region. In general, for imaging of the sinuses, orbits, facial bones, jaw, and temporal bones, axial and coronal (and occasionally sagittal) images using high-resolution, thin-section bone and soft tissue algorithms are necessary. This is best done using MDCT with reformatting as guided by initial frontal or lateral scout projection images.

For the most part, only none enhanced CT is needed to delineate bony, air space, and soft tissue abnormalities. Furthermore, it is the definitive procedure for detecting and confirming calcification. Enhancement with intravenously administered contrast agents may be added or substituted to demonstrate normal vascular structures (e.g., CT angiography [CTA]), abnormal vascularity, or abnormal vascular permeability (e.g., inflammatory or neoplastic neovascularity), especially if intracranial extension is suspected. CT is preferred for evaluation of the facial bones (including the mandible and temporomandibular joint) and orbits, especially in facial trauma and craniofacial malformations, because it precisely demonstrates both bony and soft tissue structures, including the intraorbital and intracranial contents. Such an approach is particularly critical in the timely evaluation of the newborn in respiratory distress in whom nasochoanal stenosis/atrophia is suspected. CT is also preferred for assessment of traumatic, neoplastic, and inflammatory involvement of the paranasal sinuses, particularly for its precise delineation of anatomy (e.g., ostiomeatal complex), bone destruction, and soft tissue changes (e.g., mucosal thickening, cyst, polyp, mucocoele).

Ophthalmologic assessment and US often suffice for the evaluation of intracranial masses for diagnosis, extent, and treatment planning (e.g., retinoblastoma). Extraocular orbital lesions are often best evaluated first with high-resolution CT. This modality often provides definitive evaluation, especially for trauma, infection, and pseudotumor. CT may also suffice for detecting associated intracranial trauma, intracranial inflammation, and hydrocephalus. Orbital involvement in craniofacial syndromes is often best delineated by three-dimensional CT (3DCT), especially for planning of reconstructive craniofacial surgery.

CT has also replaced PF/CR and polytomography for the evaluation of the petrous and mastoid portions of the temporal bone (e.g., hearing loss, facial palsy), including developmental malformations and acquired diseases—especially inflammatory, traumatic, and neoplastic processes of the inner ear, middle ear, mastoid, and external auditory canal. CT determines the extent of bony destruction associated with cholesteatoma, mastoiditis, and tumors, including skull base and intracranial extension.

CT scanning of the neck (e.g., neck mass, infection, vascular anomaly) is usually performed after bolus intravenous administration of a contrast agent, and axial sections from the clavicles to the skull base are obtained. It is the standard for the emergency evaluation of suppurative head and neck lesions (e.g., retropharyngeal abscess). The bolus technique provides a “blood pool” effect to visualize normal neck vessels and abnormal vascularity.
 Delayed post-injection imaging may demonstrate abnormal tissue enhancement (e.g., abscess). Occasionally, axial sections may be obtained prior to the enhanced study to evaluate for calcification or hemorrhage.

**Radionuclide Imaging**

One of the most important uses of RI in the evaluation of the neck in childhood is the imaging of the thyroid. Iodine 123 (I\textsubscript{123}) and technetium Tc 99m (Tc\textsubscript{99m}) pertechnetate are the agents currently used. I\textsubscript{123} is trapped and organified by the thyroid, whereas Tc\textsubscript{99m} pertechnetate is not organified. Because its biochemical behavior is identical to that of stable iodide and because it affords a higher thyroid-to-background ratio, I\textsubscript{123} is probably preferred. Common indications for thyroid RI include the identification of lymphoproliferative disorder [PTLD] in childhood and adolescence (e.g., lymphoma, post-transplant immunity in the evaluation of neoplastic processes involving the neck thyroid tissue is also essential), and the evaluation of a solitary thyroid nodule. PET-CT has also emerged as a primary modality in the evaluation of neoplastic processes involving the neck in childhood and adolescence (e.g., lymphoma, post-transplant lymphoproliferative disorder [PTLD]).

**Magnetic Resonance Imaging**

MRI is often adjunctive to CT in the assessment of head and neck lesions. However, in a number of situations MRI may be the technique of choice (e.g., vascular anomalies). In general, MRI should be considered for specific delineation of soft tissue elements, vascular components, and intracranial involvement. Disease processes and abnormalities involving the skull base are probably best evaluated with a combination of CT for bony involvement and MRI for neurovascular involvement. MR angiography (MRA) or venography (MRV) may be added to confirm vascular occlusion (e.g., dural sinus thrombosis) or to show high-flow vascular lesions such as arteriovenous malformations (AVMs) and hemangiomas. Doppler US imaging is often preferred in young infants, however, and CTCA or CT venography may be better for vascular assessment in older children, especially in the diagnosis of venous thrombosis. In petrous temporal bone abnormalities, MRI is generally reserved for the detection and delineation of tumors or complicated inflammatory conditions. Clinical indications for MRI may include retrocochlear sensorineural hearing loss, facial nerve paralysis, and vertigo.

Desirable MRI techniques include fast spin echo (FSE), inversion recovery, fat suppression, and gadolinium enhancement sequences. The volume head coil, or semivolume head and neck coil, is used to obtain sagittal T1-weighted images, axial proton density images, and axial T2-weighted images. Short T1 inversion recovery (STIR) images may be preferred, however, for the additive T1 and T2 effect and the superb fat suppression provided. Gadolinium-enhanced T1-weighted images with fat suppression are often used in one or more planes, particularly for the evaluation of tumors and inflammation. High-resolution thin-section axial and coronal T1-weighted acquisitions are often used with fat suppression and gadolinium enhancement, particularly to evaluate the orbits and internal auditory canals. Gradient echo (GE) techniques are used to enhance vascular flow, CSF flow, and magnetic susceptibility effects (mineralization or hemorrhage). Non-MRA vascular flow–enhanced studies may be done using multiple single-slice or GE techniques. A number of two-dimensional (2D) and 3D time-of-flight (TOF) and phase-contrast MRA techniques are available.

### CONGENITAL AND DEVELOPMENTAL ABNORMALITIES

See Box 10-1.

**Box 10-1. Pediatric Head and Neck Developmental Anomalies**

<table>
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<th>Primary ocular abnormalities</th>
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<td>Orbital/orbital abnormalities associated with CNS malformations</td>
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**Orbit and Globe**

**Normal Development**

The eye and orbit develop from the neuroectoderm, the cutaneous ectoderm, and the neural crest cells. The optic primordium gives rise to the optic vesicle and stalk, which become the eye (including the retina) and the optic nerve. A transitory vascular system, the hyaloid artery and its branches, forms the primary vitreous and then involutes by the 35th gestational week. The globe lies within the fat of the orbit. The outer layer of the globe is the sclera and cornea; the middle layer is the choroid, ciliary body, and iris, and the inner layer is the retina. The retina, which is the neurovisual membrane, is continuous posteriorly with the optic nerve. The refracting media include the aqueous humor, lens, and vitreous humor. The lacrimal gland lies in the superolateral orbit and secretes tears. The tears are drained from the eye by the lacrimal canals into the lacrimal sac medially and then into the nasolacrimal duct, which empties into the inferior meatus of the nasal cavity.

The orbit contains the orbital fascia, ocular muscles, globe and its appendages, and associated arteries, veins, and nerves. The optic foramen lies at the orbital apex and transmits the optic nerve and ophthalmic artery. The superior orbital fissure lies inferolaterally to the optic foramen and transmits the third and fourth cranial nerves, the ophthalmic division of the fifth cranial nerve, the sixth cranial nerve, sympathetic nerves, and the ophthalmic vein. The extraocular muscles originate at the orbital apex and insert on the globe, forming a cone about the globe and optic nerve. The orbital fascia forms the periestuem of the orbit, and its anterior reflection about the globe is the orbital septum. This septum separates the preseptal space from the postseptal space. The postseptal space is further subdivided by the muscular cone (i.e., intraconal and extraconal space). The orbital cavity grows passively in response to the growth of the globe. The globe is 75% of adult size at birth, and its growth is complete by age 7 years.

**Primary Ocular Abnormalities**

It may be difficult to distinguish anophthalmia (congenital absence of the eye) from severe microphthalmia (hypoplastic eye) or orbital hypoplasia. They result from incomplete formation or degeneration of the optic vesicle. They may coexist with congenital cystic eye. Ocular structures (lens and globe) are absent in primary anophthalmia but present in microphthalmia (Fig. 10-1). Anophthalmia may be sporadic or may occur with chromosomal syndromes and complex craniofacial anomalies. Imaging shows a poorly formed and shallow orbit containing rudimentary tissue.
Microphthalmia may be isolated or may be associated with other abnormalities (e.g., coloboma, duplication cyst, glaucoma, cataracts, septo-optic dysplasia, genetic syndromes [trisomy 1], and the TORCH (toxoplasmosis, other agents, rubella, cytomegalovirus, and herpes simplex) infections (i.e., chorioretinitis).

Coloboma refers to any congenital or acquired ocular structural defect. Typical colobomas result from failure of embryonic chorial fissure closure and are usually bilateral (e.g., autosomal dominant). They may also be sporadic and unilateral. On imaging, a small cyst is found behind the globe at the head of the optic nerve (see Fig. 10-1).

Optic nerve hypoplasia, defined as a subnormal number of axons, is a common and isolated anomaly. Particularly when bilateral, it may be associated with ocular, facial, endocrine, or CNS anomalies (e.g., septo-optic dysplasia or encephalocoele). Imaging demonstrates small optic nerves and a small chiasm (see Fig. 10-1).

Persistent hyperplastic primary vitreous (PHPV) represents persistence and hyperplasia of the embryonic hyaloid vascular system. PHPV is usually unilateral and associated with microphthalmia. Typical clinical findings include leukocoria (white pupil), microphthalmia, and cataract. Microphthalmia and the absence of calcification are important in differentiating PHPV from retinoblastoma (calcifications in a normal-sized or enlarged globe). Complications of PHPV include glaucoma (i.e., buphthalmia), recurrent hemorrhage, retinal detachment, and phthisis bulbi. Imaging demonstrates a small globe with a bandlike hyperdensity on CT or a hyperintensity on T1-weighted MR imaging (T1 hyperintensity) that extends from the lens to the posterior globe (Fig. 10-2).

Glaucoma is abnormally elevated intraocular pressure due to disordered aqueous humor flow. Primary congenital glaucoma is usually bilateral and may occur with other disorders (e.g., phakomatoses). The increase in intraocular pressure causes ocular enlargement (i.e., buphthalmos). Secondary congenital glaucoma results from intrauterine eye inflammation (e.g., rubella), trauma, or an ocular tumor (e.g., retinoblastoma).

Coats disease is a primary retinal vascular anomaly (telangiectasia with retinal and subretinal lipoproteinaceous exudates) with peak occurrence at the end of the first decade. Retinal detachment with leukocoria makes it difficult to differentiate from retinoblastoma. Characteristic imaging findings include vitreous CT hyperdensity or T1 hyperintensity but no focal mass. Calcification sometimes occurs.

Retrolental fibroplasia, or retinopathy of prematurity, is usually bilateral and asymmetric. There may be retinal detachment or leukocoria, in which case the abnormality can mimic retinoblastoma.

Ocular and Orbital Abnormalities Associated with Central Nervous System Malformations

Orbital abnormalities are commonly associated with neural tube disorders (see Chapter 8). Cephaloceles, which commonly involve the orbit or optic pathways, can be classified as sphenoidal or frontoethmoidal. Dermal sinuses and dermoid-epidermoids
(discussed later) may be associated with widening of the nasal bridge, hypertelorism, or midline anomalies (e.g., callosal hypogenesis with a lipoma). Midface hypoplasia and hypotelorism are commonly associated with the holoprosencephalies (HPE). The alobar form of HPE may also have cyclopia, ethmocephaly, cebocoephaly, or median cleft lip with hypertelorism. Septo-optic dysplasia (de Morsier syndrome) involves partial or complete absence of the septum pellucidum and optic hypoplasia. Orbital deformity is commonly associated with the craniostenoses (e.g., metopic, coronal, multiple).

Orbital abnormalities are also part of the craniofacial malformations and craniostenosis associated with disorders such as Crouzon disease and Apert, Carpenter, and Pfeiffer syndromes. Reconstructive surgery is often required to improve function and preserve vision. Treacher Collins syndrome is another example of a craniofacial syndrome with orbital/ocular abnormalities (i.e., microphthalmia, coloboma). Neuroophthalmologic involvement often occurs in the neurocutaneous syndromes and includes neurofibromatosis type 1 (NF-1) (sphenoid-orbital dysplasia, optic glioma), tuberous sclerosis (retinal neuronal hamartoma, Sturge-Weber syndrome (choroidal neovascular malformation with buphthalmos), and von Hippel-Lindau disease (retinal hemangioblastoma with retinal detachment and hemorrhage).

Migrational disorders are often associated with ocular, orbital, or optic pathway abnormalities (callosal hypogenesis, lissencephaly syndromes). Callosal hypogenesis is seen in a wide array of anomalies, including cephaloceles, dermal sinus, septo-optic dysplasia, cleft lip and palate, Apert syndrome, hypertelorism, coloboma, and Aicardi syndrome. Midface and orbital dysmorphia, as well as ocular anomalies, are frequently seen in the lissencephaly syndromes (e.g., Walker-Warburg syndrome).

Malformatives Lesions
Malformatives tumors, nonneoplastic and neoplastic, are aberrations of development. They are usually of neuroectodermal origin (e.g., dermoid-epidermoid) or mesodermal origin (e.g., lipoma). Some germ cell neoplasms (e.g., teratoma) and vascular anomalies are also included in this category. Malformatives tumors may be cystic, solid, or mixed. In the pediatric orbit, these tumors include colobomata (see Fig. 10-1), duplication cysts, nasolacrimal duct cysts, lacrimal ectopia, dermoids-epidermoids, teratomas, and (rarely) arachnoidal cysts and lipomas. It may be difficult to differentiate a coloboma from a retrolubar duplication cyst. Hydrops and arachnoidal cyst of the optic nerve sheath are exceedingly rare in the absence of suprasellar tumors or cysts.

Congenital nasolacrimal duct cyst or mucocele probably results from incomplete canalization of the duct on one or both sides. Proximal obstruction results in a lacrimal sac mucocele and manifests as a medial orbital canthal mass (dacryocystocele). Distal obstruction produces a nasolacrimal duct mucocele that extends beneath the inferior turbinate into the nasal cavity. The two types may coexist. Bilateral involvement may clinically mimic choonal atresia. The resultant cystic dilatation often resolves in early infancy. Persistence may cause nasal airway obstruction, infection, and dacryocystitis. Imaging demonstrates a medial canthus cystic mass in continuity with an enlarged nasolacrimal duct (and canal) and an intranasal submucosal cystic mass (Fig. 10-3). The latter differentiates the mucocele from other medial canthal cystic masses (e.g., dacrocyctitis, choristoma, dermoido-epidermoid, or cephalocele). With associated abscess there may be restricted diffusion on diffusion-weighted MRI (DWI), which can make it difficult to distinguish from dermoid-epidermoid.

Ectopic lacrimal gland tissue may appear as solid or cystic lesions of the orbit and may produce proptosis. Neoplastic transformation is rare (e.g., pleomorphic adenoma and adenocarcinoma).

Dermoid-epidermoid, the most common congenital lesion of the orbit, arises as a developmental sequestration of ectoderm along the sutures (Fig. 10-4). It most frequently occurs in the superolateral or medial orbit. Relatively slow growth of the cyst erodes adjacent bone (i.e., notching or scalloping) and displaces adjacent structures. CT hypodensity or fat-like TI hyperintensity may be present along with calcification or a fat-fluid level. Restricted diffusion on DWI is also a characteristic finding that may mimic abscess (see Figs. 10-3 and 10-4).

Orbital teratoma is often benign and produces proptosis in infancy. It may be cystic, multicystic, solid, or mixed. There may be orbital expansion with ocular displacement or compression. CT or MRI may demonstrate fatty features with calcification or ossification. Mesodermal dysplasias affecting the orbit include NF-1 (sphenoid-orbital dysplasia; see Chapter 8) and the skeletal dysplasias (e.g., fibrous dysplasia, craniometaphyseal dysplasia, and osteopetrosis). Fibrous dysplasia produces a characteristic “ground-glass” or sclerotic appearance of the orbit, facial bones, or skull base (Fig. 10-5).

Nasal Cavity, Paranasal Sinuses, and Face
Normal Development
The mesenchymal primordia of the face form about the stomodeum (primitive mouth) and include the frontonasal prominence, maxillary prominences, and the mandibular prominences. These structures, respectively, give rise to the forehead, nose and nasal septum; turbinates, upper lip, premaxilla, maxilla, hard palate, soft palate, uvula; mandible, lower lip, chin, and lower cheek. The nasal cavities develop and ultimately communicate with the nasopharynx and oral cavity after rupture of the oronasal membrane at the level of the choanae. The paired turbinates form from the lateral wall of each nasal cavity. Specialized olfactory

![Figure 10-3](image) Right dacryocystocele (arrows in A and C) with large right nasolacrimal canal (arrow in B) and abscess on axial CT scans (A and B) and a diffusion-weighted MR image (C).
Epithelium develops in the roof of each nasal cavity and connects with the olfactory bulbs of the prosencephalon. The paranasal sinuses form as diverticula of the walls of the nasal cavities and later become pneumatized.

The small size of the face relative to the head at birth results from the more rapid development of the brain. The nose is the major portal of air exchange, especially in the newborn. The nasal cavity and paranasal sinuses are covered with respiratory epithelium. The maxillary sinuses and ethmoid air cells are present at birth but may not be visible until 3 to 6 months of age (adult size by 10 to 12 years). The frontal and sphenoid sinuses may not be visualized until 6 years of age. The frontal sinuses, anterior and middle ethmoidal air cells, and maxillary sinuses drain into the middle meatus via the ostiomeatal complex. The posterior ethmoidal air cells and sphenoidal sinuses drain into the spheno-ethmoidal recess and superior nasal meatus. During early infancy, there may be physiologic underaeration of the paranasal sinuses owing to redundant normal mucosa. Paranasal sinus disease is characterized by decreased aeration, mucosal thickening, soft tissue masses (e.g., mucus retention cyst, polyp, mucocele, tumor), air-fluid levels, and demineralization or bone destruction.

Congenital Nasal Stenosis and Atresia
Nasal airway obstruction may be the cause of respiratory distress in the newborn and infant. An obstructive abnormality is further indicated by inability to pass nasal catheters. The differential diagnosis usually includes nasal cavity and choanal stenosis or atresia, basal cephalocele, and bilateral nasolacrimal duct cysts.
Choanal stenosis and atresia, respectively, are narrowing of the posterior nasal cavity and obstruction by an atresia plate (bony, membranous, or both). The bilateral form manifests as neonatal respiratory distress (Fig. 10-6A); the unilateral form may not manifest until later (Fig. 10-6B). There may be co-existing nasal cavity stenosis or atresia and other anomalies or syndromes, such as cleft palate, cardiovascular and abdominal abnormalities, Treacher Collins syndrome, CHARGE (colobomata, heart defect, atresia, choanal retarded growth and development, genital hypoplasia, and ear abnormalities and/or deafness) association, fetal alcohol and Apert syndrome, and Crouzon disease.

Stenosis of the entire nasal airway is usually bony and may be associated with prematurity or maxillary hypoplasia (e.g., Apert syndrome). Atresia is extremely rare. Symptoms may not arise until there is a complicating rhinitis.

Segmental atresia, or stenosis, may occur anteriorly (i.e., piriform aperture stenosis; Fig. 10-7). This may be associated with single midline maxillary incisor (mega-incisor) and midline intracranial anomalies (e.g., holoprosencephaly). Segmental stenosis may also result from maxillary hypoplasia, turbinate hyperplasia, or nasal septal deviation.

**Congenital Nasal Masses**

Congenital nasal masses resulting from defective neural tube closure include cephalocele, neuroepithelial heterotopia (nasal glioma), and dermoid-epidermoid (see Chapter 8). They may manifest as bilateral nasal obstruction and respiratory distress in the newborn and are to be distinguished from nasochoanal stenosis/atresia and nasolacrimal duct cysts (discussed earlier). Later in life they may be the cause of a nasal mass or obstruction.

The fonticulus frontalis and prenasal space are transient nasofrontal structures that involute in early gestation. The fibrous tissue filled foramen cecum remains as the only remnant (see Fig. 10-1). Persistence of these primitive structures may be associated with a dural diverticulum and protrusion of intracranial contents as a nasofrontal cephalocele or a nasoethmoidal cephalocele.

With partial or complete obliteration of the intracranial connection, the cephalocele becomes a sequestered neuroepithelial heterotopia (nasal glioma; Fig. 10-8). As the dural diverticulum regresses, incorporation of surface ectoderm may form a dermal sinus. This commonly manifests as a skin dimple or mass in the nasal region. The sinus may extend to any point from the columella to the glabella. Other associated findings include nasal

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**Figure 10-6.** Axial CT scans showing choanal atresia. A, Bilateral choanal atresia (*lower arrows*) with retained secretions plus right nasal septal deviation (*upper arrow*). B, Right unilateral choanal atresia (*arrow*) with retained secretions; compare with the normal left choanal aperture.

**Figure 10-7.** A and B, Axial CT scans showing neonatal piriform aperture stenosis (*upper arrows* in A) with bilateral open choanae (*lower arrows* in A) and a single midline mega-incisor (*arrows* in B).
or frontal bony defect, enlarged foramen cecum, dermoid, epidermoid, or lipoma (Figs. 10-9 and 10-10). The mass is often CT hypodense and calcified and has fatty T1 hyperintensity on MRI. An intracranial communication may result in recurrent meningitis, abscess, or empyema. Other rare congenital nasal masses are nasoalveolar (incisive canal) cysts, dentigerous cysts, mucous cysts, vascular anomalies, branchial cysts, hamartomas, teratoid tumors (embryoma, epignathus), and Tornwaldt cyst.

**Facial Anomalies**

**Cleft Lip and Palate**

Clefts involving the lip, alveolus, or palate are common anomalies and may be partial, complete, unilateral, or bilateral. Maxillary hypoplasia with prognathism often accompanies bilateral clefts. A complete cleft disrupts facial growth, dentition, speech, and eustachian tube function.

![FIGURE 10-8.](image)

**FIGURE 10-8.** Right sequestered nasoethmoidal cephalocele (“nasal glioma,” indicated by long white arrows) with extension via foramen cecum (short black and white arrows) on axial (A and B), coronal (G), and sagittal (D) CT scans as well as sagittal T1-weighted (E) and T2-weighted (F) MR images.
Craniofacial clefts (facial, cranial, or combined) extend along continuous axes through the eyebrow or eyelid, maxilla, nose, and lip. Facial clefts extend caudally from the lower eyelid, whereas cranial clefts extend cephalad from the upper eyelid. Associated anomalies include orbital dystopia, microphthalmos, coloboma, cephaloceles, and orbital hypertelorism. Associated syndromes include median cleft syndrome, Treacher Collins syndrome, hemifacial microsomia, amniotic band syndrome, otomandibular syndrome, and Goldenhar syndrome. The associated dysmorphia includes abnormalities of the forehead, orbits (hypertelorism, exorbitism), midface, and anterior cranial base. Cloverleaf craniofacial anomaly results from multiple craniosynostoses and is associated with extensive craniofacial deformities (Fig. 10-12). Amniotic band syndrome (congenital constrictions or amputations) manifests as facial clefts, calvarial defects, hydrocephalus, cephaloceles, or anencephaly. Hemifacial microsomia has facial asymmetry with maxillary and malar hypoplasia, macrostomia, mandibular hypoplasia (ramus and condyle), microtia (external ear atresia/stenosis and middle ear hypoplasia), microphthalmia, congenital cystic eye, and...
coloboma. **Treacher Collins syndrome** is a mandibulofacial dysostosis (autosomal dominant) characterized by bilateral zygomatic, malar, and mandibular hypoplasia (Fig. 10-13). Also common are microtia (external and middle ear hypoplasia), colobomata, and microphthalmia. **Goldenhar syndrome** (oculoauriculovertebral syndrome) is a mandibulofacial dysostosis with hemifacial microsomia, epibulbar dermoids or lipodermoids, and vertebral anomalies.

**Figure 10-11.** Pierre-Robin sequence with microretroglossia (long arrow), posterior palate defect (short arrow), and high-riding tongue (T) with narrow airway on sagittal CT scan (A) and frontal 3DCT reconstruction (B).

**Figure 10-12.** Multiple craniosynostosis (bicoronal, metopic, sagittal) with cloverleaf deformity and exorbitism on fetal axial T2-weighted MR image (A) as well as neonatal coronal (B) and axial (C) CT scans with lateral (D) and frontal (E) 3DCT reconstructions.
Developmental Variants and Anomalies of the Nose and Paranasal Sinuses

Developmental variants and anomalies may predispose to osmioatlantal complex obstruction with inflammation (Fig. 10-14). Endoscopic or open surgical procedures may be necessary for correction. Nasal septal deviation is common and is often associated with asymmetry or deformity of adjacent structures (e.g., abnormally large middle turbinate [concha bullosa], septal spur, secondary turbinate anomalies, extramural sinus pneumatization, uncinate process deviation). Extramural extension of the ethmoid cells includes pneumatization of the supraorbital ridge, superior or middle turbinate (e.g., concha bullosa), uncinate process, orbital plate of the maxilla, the sphenoid bone, agger nasi cells (anterior to the upper nasolacrimal duct), Haller cells (infraorbital; may obstruct the maxillary sinus infundibulum), and Onodi cells (sphenoid body near the optic nerve canal, internal carotid artery [ICA], cavernous sinus). Other common anomalies are maxillary sinus hypoplasia, extramural sphenoid or maxillary sinus extension, maxillary sinus septation, and accessory ostia.

Ear and Temporal Bone

Normal Development

The external and middle ear (mastoid portion of the temporal bone) are derived from the branchial apparatus, and the internal ear is derived from the neuroectoderm. The auricle and external ear (membranous and bony portions) begin development along with the mandible. The middle ear cavity expands and incorporates the tympanic membrane, eustachian tube, auditory ossicles (malleus, incus, stapes), muscles (tensor tympani and stapedius), their tendons and ligaments, the round and oval windows, and the chorda tympani nerve, and then gives rise to the attic and mastoid antrum.

The petrous portion of the temporal bone contains the inner ear and transmits the cochlear aqueduct and perilymph, ICA, internal jugular vein, and cranial nerves VII (facial) and VIII (vestibulocochlear). The facial nerve extends from the internal auditory canal into the facial nerve canal, which has a labyrinthine segment (anterior genu and geniculate ganglion within the otic capsule), a tympanic segment (horizontal course within the middle ear extending to the posterior genu and facial nerve recess), and a mastoid segment (vertical course to the stylomastoid foramen). Pneumatization of the mastoid occurs rapidly and is visible by 4 to 6 months of age. Mastoid disease is characterized by decreased aeration, mucosal thickening, edema, accumulation of fluid, bony demineralization, and bone destruction.

External Auditory Canal and Middle Ear Cavity Anomalies

External auditory canal (EAC) stenosis/ataresia is commonly associated with a malformed auricle (i.e., microtia), hypoplasia of the middle ear and mastoid, and, occasionally, mandibular hypoplasia (Fig. 10-15). The complex may be unilateral, bilateral, isolated, or associated with syndromes (e.g., hemifacial microsomia, Treacher Collins syndrome [see Fig. 10-13], Crouzon disease, and Goldenhar syndrome). EAC atresia may be partial or complete, and membranous...
or bony. Complete (osseous) atresia consists of a bony atresia plate at the tympanic membrane and fusion of the malleus to the plate. Partial (membranous) atresia consists of a soft tissue plug at the tympanic membrane (with or without fusion of the malleus).

EAC atresia may be associated with a congenital cholesteatoma (primary epidermoid) of the EAC or middle ear cavity (MEC) (Fig. 10-16). MEC hypoplasia may be mild or severe. Osseous anomalies include absence, rotation, fusion, and dysplasia. The facial nerve is often thickened, has an aberrant course, and may be exposed (dehiscence, protrusion). First branchial arch dysplasia results in unilateral (e.g., hemifacial microsomia) or bilateral (e.g., Treacher Collins syndrome; see Fig. 10-13) mandibulofacial dysostosis with consequent anomalies of the mandible, EAC, MEC, malleus, and incus. Second branchial arch dysplasia results in anomalies of the hyoid, styloid, stylohyoid ligament, and stapes.

Inner Ear Anomalies
Congenital sensorineural hearing loss is commonly associated with inner ear anomalies. A common anomaly is vestibular aqueduct dysplasia (ranging from obliteration to dilatation; Fig. 10-17). Dilated vestibular aqueduct (diameter > 1.5 mm, or greater than posterior semicircular canal [SCC] diameter) may be associated with malformations of the cochlea (e.g., Mondini syndrome [see later]), modiolus (hypoplasia), vestibule, SCCs, or cochlear nerve (e.g., hypoplasia). Another common anomaly is malformation of the lateral SCC. In other cases, the SCCs may be absent or may be malformed and incorporated into a dilated vestibule (Fig. 10-18).

Cochlear anomalies may be classified according to the stage of developmental arrest. Complete labyrinthine aplasia (Michel deformity) results in a single small cystic cavity. Other anomalies include a large common cavity (common chamber anomaly), cochlear aplasia or hypoplasia, and incomplete partition (Mondini syndrome—small cochlea with incomplete septation, i.e., less than two and one-half turns; Fig. 10-19). Malformations of the internal auditory canal consist of stenosis and atresia.

Facial Nerve Anomalies
Aberrant course of the facial nerve is usually associated with an anomaly of the external, middle, or inner ear. Lateral and anterior displacement of the mastoid segment is common in EAC and MEC anomalies (see Fig. 10-15). Anomalous facial nerve may be directly involved in ossicular malformations. Dehiscence of the facial nerve canal most often occurs in its tympanic portion at the level of the stapes and results in a conductive hearing loss (Fig. 10-20). Facial nerve hypoplasia has been described in syndromes, such as Goldenhar syndrome, VATER (abnormalities of vertebrae, anus, trachea, esophagus, and radial and renal dysplasia) association, and some trisomies. Absence of the facial nerve has been described in a few cases, including the Möbius sequence.

Neck, Oral Cavity, and Jaw
Normal Development
The branchial apparatus, which contributes to formation of the head and neck, consists of paired branchial arches, pharyngeal pouches, branchial grooves, and branchial membranes. The branchial arches form along the lateral primitive pharynx. Each arch consists of a mesenchymal core (containing neural crest cells and arterial, nerve, cartilage, and muscular elements). Each arch is separated by branchial membranes and covered externally by surface ectoderm (branchial grooves) and internally by endoderm (pharyngeal pouches). The primitive mouth (stomodeum) arises from the surface ectoderm in contact with the amniotic cavity externally and the primitive gut internally via the esophagus (after rupture of the primitive buccopharyngeal membrane). The tongue buds are mesenchymal proliferations derived from the...
first pair of branchial arches. The salivary glands begin as solid proliferations from epithelial buds. The developing thyroid gland is a diverticulum connected by the thyroglossal duct ventral to the hyoid to the tongue base at the foramen cecum. The thyroglossal duct normally involutes. The thymus and inferior parathyroid originate from the third pharyngeal pouch. The superior parathyroid glands arise from the fourth pharyngeal pouch. The laryngotracheal groove and tracheoesophageal folds form to become the ventral laryngotracheal tube and dorsal esophagus.

The neck is divided by the hyoid bone into the suprahyoid and infrahyoid regions. Three layers of deep cervical fascia divide the suprahyoid neck into eight compartments (parapharyngeal space, pharyngeal mucosal space, masticator space, parotid space, retropharyngeal space, perivertebral space, and posterior cervical space). The sternocleidomastoid muscle divides the infrahyoid neck into anterior and posterior triangles. The layers of the deep cervical fascia permit further subdivision of the infrahyoid neck into five major spaces that are continuous with corresponding spaces in the suprahyoid neck (carotid, visceral, posterior cervical, retropharyngeal, and perivertebral spaces).

The adenoids become conspicuous within the nasopharynx by 2 to 3 years of age and regress during adolescence. If no adenoidal tissue is seen in a young child, and in the absence of prior adenoidectomy, the possibility of immunodeficiency should be considered. The lymph nodes of the neck occur in contiguous groups and may be classified according to various systems. Normal nodes are usually homogeneous, similar in density to muscle, and oval or flat on CT. Contrast enhancement of lymph nodes is abnormal and may be seen in a variety of inflammatory and neoplastic processes. The major vessels of the head and neck include the common carotid arteries, which bifurcate into internal and external carotid arteries, the external jugular veins, the anterior jugular veins, and the internal jugular veins. The major nerves traversing the neck include cranial nerves IX through XII, the sympathetic chain, and the facial nerve.

The oral cavity contains the tongue and is bound inferiorly by the mylohyoid muscle. Within the oral cavity are the submandibular and sublingual spaces (separated by the mylohyoid muscle). The major salivary glands consist of the paired parotid, submandibular, and sublingual glands. Minor salivary glands also exist at several
levels. Most muscles of the suprahyoid neck attach to the mandible. The maxilla contains the maxillary sinuses.

**Branchial Anomalies**

Branchial anomalies arise from incomplete development of the branchial apparatus. These anomalies are therefore classified according to the level (arch, cleft, or pouch) of origin. Defects include branchial cysts, aberrant tissue, branchial sinus (incomplete tract usually opening externally that may communicate with a cyst), and branchial fistula (epithelial tract with both external and internal openings). Fistulae and sinuses are usually identified at birth because of drainage. They are best imaged by a contrast-enhanced fistulogram (e.g., CT).

Cysts are more common in older children and adults. They usually manifest as a soft tissue mass that may enlarge with infection. US reveals a cystic or complex mass. CT and MRI show an oval or round cystic mass. Wall thickness, enhancement, content, and surrounding edema often increase with inflammation. A first branchial cyst usually arises as an enlarging mass about the parotid gland but may occur about the pinna, in the EAC, MEA, or nasopharynx, or submandibularly above the hyoid bone. The differential diagnosis includes an inflammatory cyst, lymphatic malformation, and necrotic adenopathy. The second branchial cyst is the most common of these anomalies. It usually manifests as a mass at the mandibular angle but may occur at any site along a line from the tonsillar fossa to the anterior margin of the sternocleidomastoid muscle to the supravaculicular region (Figs. 10-21 and 10-22). The differential diagnosis includes vascular anomaly, suppurative adenopathy, paramedian thyroglossal duct cyst, laryngocele, and necrotic metastatic adenopathy. The third branchial sinus/fistula arises from the inferior pyriform sinus and extends between the common carotid artery and vagus nerve to the lower lateral neck. The fourth branchial sinus/fistula usually arises from the left inferior pyriform sinus, looping beneath the aortic arch (or subclavian artery if on the right) and then upward via the carotid bifurcation to the lateral neck.

Recurrent neck abscess or suppurative thyroiditis, particularly if it contains air, should raise the possibility of a pyriform sinus/fistula (Fig. 10-23). After treatment of the infection, a swallowing study using the appropriate contrast medium is performed to demonstrate the sinus/fistula. Other branchial anomalies are exceedingly rare but include anomalies of the thymus, thyroid (see later), and parathyroid glands. These include aberrant cervical thymus (e.g., within the neck), thymic cysts, parathyroid cysts, and aberrant parathyroid tissue.

**Thyroid Anomalies**

Thyroglossal duct cyst arises from thyroglossal duct remnants and often occurs in childhood. They are usually midline, or paramedian, and occur at any site from the tongue base to the suprasternal region. Off-midline cysts often occur near along the outer thyroid cartilage and deep to the neck muscles. The lesion is often circumcised and anechoic or hypoechoic on US, low density on CT, and either T1-isointense to hypointense or T1-isointense to hyperintense, with T2 hyperintensity on MRI (Fig. 10-24). Complex lesions with heterogeneity on imaging also occur (e.g., with infection), and a sinus tract may be present. The differential diagnosis includes dermoid, teratoma, vallecular cyst, mucous retention cyst, laryngocele (see Fig. 10-24), lymphatic malformation, and branchial

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**Figure 10-21.** Left second branchial cyst (white arrows) on axial vascular contrast-enhanced CT scan, showing enhancement of carotid and jugular vessels (black arrows). S, sternocleidomastoid muscle.

**Figure 10-22.** Axial T1-weighted (A) and T2-weighted coronal (B) MR images with fat suppression showing right second branchial cyst (long arrows), normal carotid and jugular vessels (short arrows in A) and normal left lymph nodes (short arrows in B). S, sternocleidomastoid muscle.
FIGURE 10-23. Axial (A) and coronal (B) vascular contrast–enhanced CT scans showing pyriform sinus fistula with perithyroidal abscess (long arrow) and air bubble (short arrow in B). C, common carotid artery; J, internal jugular vein; T, thyroid.

FIGURE 10-24. Thyroglossal duct cyst (TGDC) appears as an infrahyoid, extralaryngeal high intensity on sagittal (A) and axial (B) T2-weighted MR images. Laryngeal cyst (LGC) appears as a low-density, hyperintense laryngeal mass on axial CT scan (C) and T1-weighted MR image (D), also showing thyroid cartilage involvement (arrows) and extralaryngeal extension. L, larynx; T, tongue base.
anomalies. Other thyroid anomalies include hypogenesis (partial or complete) and ectopic thyroid tissue (usually near the foramen cecum at the tongue base; Fig. 10-25). In congenital hypothyroidism with absence of a normal thyroid gland, US should search for ectopic tissue. Such tissue may be shown by thyroid scintigraphy to be nonfunctional or the only functioning thyroid tissue.

**Laryngocele**
A laryngocele results from obstructive dilatation of the laryngeal ventricle and may be aerated or fluid-filled. The lesion may be internal, external, or translaryngeal. Complications include infection and airway compromise. The CT attenuation and MR intensity of the content vary with air and fluid content. There may be slight enhancement of the cyst wall. The differential diagnosis includes thyroglossal duct cyst and laryngeal mucosal cyst (see Fig. 10-24).

**Anomalies of the Oral Cavity, Tongue, and Salivary Glands**
Congenital and developmental abnormalities of the oral cavity previously described include lingual thyroid (see Fig. 10-25), thyroglossal duct cyst, and second branchial cleft cysts. Dermoid-epidermoid and vascular anomalies are also discussed elsewhere.

Agenesis of the major salivary glands is rare, causes xerostomia, and may be associated with absence of the lacrimal glands. The diagnosis can be confirmed by RI or MRI. Developmental cystic lesions of the salivary glands are uncommon. They usually involve the parotid gland and include branchial cysts and dermoid cysts. There may be painless swelling or signs of infection. These cysts are to be differentiated from lymphoepithelial cysts (e.g., acquired immunodeficiency syndrome [AIDS]). Nonobstructive sialectasis is a common anomaly of the salivary glands. Saccular ductal dilatation often involves the parotid gland. Infection exacerbates the ectasia and manifests as parotitis.

**Jaw Anomalies**
Mandibular and maxillary hypoplasia may be seen with a number of craniofacial syndromes (see earlier discussions). This includes mandibular hypoplasia with micrognathia and retrognathia (e.g., Pierre Robin sequence) associated with posterior palatal defects and aerodigestive compromise. 3D-CT is often needed for preoperative planning (see Fig. 10-11).

*Fibrous dysplasia* occurs more often in the maxilla than the mandible. It appears as a unilocular or multilocular, ill-defined, expansile CT hypodensity when fibrous tissue predominates (see Fig. 10-5). Admixture of bony matrix increases the lesion’s density. It may appear homogeneously hyperdense with bony expansion. *Cherubism* is a benign hereditary condition misnamed “congenital fibrous dysplasia.” It often appears between 2 and 5 years of age, progresses until puberty, and then regresses. The mandible and maxilla are often both involved by multiple expansile fibrous-lesions (Fig. 10-26).
Trauma

**Orbit and Globe**

Blunt and penetrating impact injuries are common in childhood. Orbit fracture may be isolated or occur with other face or cranial fractures. *Orbit floor and inferior rim fractures* rarely occur prior to maxillary pneumatization. Frontal impact may result in a *blow-out* fracture of the orbital floor near the infraorbital canal (Fig. 10-27). CT may also show herniation of orbital fat or displacement of the inferior rectus or inferior oblique muscle into the fracture and maxillary sinus (upward gaze impairment). Rarely is there upward displacement of the orbital floor fragments (blow-in fracture) with impingement on the muscles or globe.

Orbit roof and superior rim fractures may be associated with CSF leak or herniation of brain tissue or meninges into the orbit. Fracture depression may rarely impinge upon the globe (upward gaze impairment).

Medial orbital wall fractures into the ethmoid may be isolated or may be associated with an orbital floor fracture (see Fig. 10-27). Orbital emphysema most commonly occurs with this fracture type. Although orbital fat herniation can occur, muscle entrapment is rare. Orbital emphysema associated with frontal or sphenoid fractures usually indicates severe or complex injury.

Orbit and ocular complications of trauma (including surgery) include hematoma, emphysema, CSF leak, traumatic cephalocele, infection, growing fracture, retinal tear, intraocular hemorrhage, ocular rupture, enophthalmos, optic nerve avulsion, vascular occlusion, pseudoaneurysm, and carotid-cavernous fistula. Penetrating orbit injury may result in retained foreign body and secondary infection.

**Nasal Cavity, Paranasal Sinuses, and Face**

**Foreign Body**

Insertion of foreign material into the nose, including that of extrinsic origin (beans, seeds, toys, beads, plastic or metal objects, etc.) and intrinsic origin (emesis or expectoration of ingested or aspirated material, inspissated mucosal secretions, bony sequestra, and mineralized concretions [rhinoliths]), is common in young children. Secondary purulent rhinitis, sinusitis, adenoiditis, or otitis media (OM) is common. CT may be needed to identify a radiopaque object or to delineate the associated infection.

**Fractures**

Facial fractures may be related to vehicular accident, fall, recreation, or assault. They are infrequent in young children (< 5-6 years) and tend to be greenstick in type. The high craniofacial ratio predisposes to frontal, cranial, and intracranial injuries. With maturation and increased sinus pneumatization, the adult pattern becomes more common, including midface and mandibular fractures, plus fragmentation and displacement. Frequently involved structures include the nasal bones, mandible, orbit

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**Figure 10-27.** Right orbit and sinus trauma on axial (A and B) and coronal (C and D) CT scans. Findings include periorbital and malar swelling, periorbital edema, and hemorrhage (arrows on A); orbitoethmoid fracture (horizontal long arrows in B to D) and orbital floor fracture with herniation of orbital fat and inferior rectus muscle (short arrows in B to D); and maxillary sinus air-fluid level (vertical black arrow in B). Compare with normal left orbit and sinus structures.
and zygomaticomaxillary structures (see Figs. 10-27 and 10-28). Other system injuries are common, and midface fractures usually indicate severe trauma. Axial CT with coronal and sagittal reformatting is indicated and is performed in proper sequence according to injury priority.

**Nasal fracture** from minor frontal impact includes the greenstick type in younger children with splaying of the nasal bones. In older patients, such an impact usually produces bilateral distal nasal bone fractures, and the septum may be fractured and displaced. With severe impact, more extensive fractures may involve the nasal pyramid, maxilla, lacrimal and ethmoid bones, nasal septum, cribriform plate, and orbital roof (e.g., hemorrhage or CSF leak). Subchondral nasal septal hematoma may produce thickening or a mass. Surgical drainage can prevent necrosis, abscess, and perforation.

Mandibular condyle fracture, the most common mandible fracture, often results from a fall with impact to the chin. It is frequently bilateral and may be associated with a parasymphysyal fracture. Condylar head injury may result in underdevelopment or ankylosis. Condylar neck fractures typically occur in older children. Associated temporomandibular joint injury may rarely occur. The latter may require additional MRI. Asymmetric mandibular body fractures are commonly seen with side impact (e.g., assault) in adolescence (Fig. 10-28).

Other midface fractures (e.g., maxillary and zygomatic) are rare in young children but may extend bilaterally or superiorly to involve the frontal region. Associated CNS injury or CSF leak is common. In older children, the adult pattern predominates and includes isolated maxillary alveolar fractures, partial fractures of the maxilla, palatal fractures, the LeFort fractures, and lateral midface or trimalar fractures.

**Frontal bone fractures** occur most commonly in younger children, are of the greenstick type (nonpneumatized frontal sinus), and may extend into the skull or orbital roof. In older children, frontal sinus fractures may result from direct impact or from extension of a skull fracture. Intracranial injury is common. Posterior wall fracture may be associated with CSF leak, pneumocephalus, or CNS infection.

**Sphenoid fractures** rarely occur but indicate severe trauma, including other skull base fractures. CT may show sinus opacification, air-fluid level, or pneumocephalus. CTA may be indicated to evaluate for associated carotid arterial or cavernous sinus injury (e.g., carotid-cavernous fistula; see Chapter 8).

**Ear and Temporal Bone**

Petrous and mastoid trauma may result in external auditory canal hemorrhage, hemotympanum, CSF otorrhea, hearing loss, vertigo, or facial nerve palsy. CT may show opacification of the EAC,

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**Figure 10-28.** Bilateral mandibular body fractures (arrows) on axial (A) and coronal (B) CT scans as well as frontal coplanar (C) and 3D (D) CT reconstructions.
MEC, or mastoid air cells. Local pneumocephalus is common, and there may be intracranial hemorrhage or brain injury.

Fractures are classified according to their course relative to the long axis of the petrous bone. Longitudinal fractures often result from lateral impact and commonly involve the mastoid (Fig. 10-29). The result may be tympanic membrane rupture, ossicular disruption, and fracture of the tegmen tympani. The inner ear is often spared. Fractures may not heal except for fibrous union. Facial nerve paralysis may result from injury distal to the geniculate ganglion.

Transverse fractures usually result from an occipital or frontal impact and may involve the mastoid (Fig. 10-30) or the otic capsule and internal auditory canal. With the latter fracture, there may be sensorineural hearing loss and vertigo. Facial nerve paralysis is often due to injury proximal to the geniculate ganglion. Combined longitudinal and transverse, or oblique, fractures usually result in petrous bone fragmentation. Because of the incompletely developed mastoid, the facial nerve is also susceptible to trauma in the neonate and young infant.

Oral Cavity and Neck
Trauma mechanisms in the oral cavity and neck include vehicular accidents, bites, knife or gun wounds, surgery, burns, and oral penetration by foreign objects. CT may show edema, contusion, laceration, hematoma, or air. Soft tissue emphysema may result from external penetration or injury of the airway, thorax (i.e., pneumothorax), or esophagus. There may be airway compression, airway or esophageal perforation, vascular injury, retained foreign body, or subsequent infection. Intravenous contrast agents may be used to delineate vessels or to evaluate infection. Oral or esophageal contrast enhancement may show pharyngeal or esophageal penetration. Vascular injury may include laceration, transection, contusion, dissection, false aneurysm, arteriovenous fistula, thrombosis, and embolization. CTA or CT venography using MDCT may be preferred to MRAMRV, although angiography may be necessary. Salivary gland trauma may cause emphysema, hematoma, duct stricture or transection, fistula, sialocele, or subsequent infection.

Vascular Abnormalities
Vascular abnormalities of the head and neck may include variants, anomalies, and tumors. These are presented here according to category and region. The Mulliken and Glowacki biologic classification of vascular anomalies involving cutaneous and muscular tissues includes hemangiomas and vascular malformations. Hemangiomas are congenital endothelial tumors, whereas vascular malformations are endothelial-lined anomalies. They are distinguished both by clinical criteria and imaging features (high flow vs. low flow), especially on US with Doppler imaging and MRI. The orbit, parotid, face, scalp, oral cavity, and neck are frequent sites of origin.

Hemangioma is the most common tumor of the head and neck in childhood. It evolves from a cellular proliferative phase to a plateau phase, and then to an involuting phase. Proliferating hemangiomas are high-flow benign neoplasms that appear as vascular flow voids (hypointensity) on spin echo (SE) sequences, hypointensity on GE vascular flow or MRA sequences, and marked tumor enhancement on gadolinium-enhanced sequences (Figs. 10-31 to 10-33). There may be arteriovenous shunting. Involving hemangiomas demonstrate decreasing flow characteristics, decreasing tumor size, and increased fibrofatty tissue. Involvement is usually complete by age 7 to 8 years and may mimic a lipoma or low-flow malformation. Hemangiomas may be further categorized as congenital hemangioma, endangering hemangioma (e.g., orbit, airway, spontaneous hemorrhage), noninvoluting congenital hemangioma, multiple hemangioma or hemangiomatosis, kaposiform hemangioendothelioma, and Kasabach-Merritt syndrome (consumptive coagulopathy).

Vascular malformations are subclassified as capillary, arterial, venous, lymphatic, and combined. Venous malformation (VM) and lymphatic malformation (LM) are low-flow anomalies (no vascular high-flow voids) consisting of septated or cystic channels, and often with a fibrofatty stroma. VMs are characterized by phleboliths and

**Figure 10-29.** Right longitudinal mastoid fracture (arrows) on axial CT scan.

**Figure 10-30.** Right transverse mastoid fracture (arrows) on axial CT scan.

**Figure 10-31.** Orbit hemangioma (arrows) on coronal T2-weighted (A) as well as coronal (B) and axial (C) gadolinium-enhanced T1-weighted MR images, all with fat suppression, which also show intrinsic vascular high-flow voids.
Gadolinium enhancement of the cavernous blood-filled spaces (Fig. 10-34). There may also be prominent draining or anomalous veins (hyperintensity on GE vascular flow or MRV sequences). LMs may be macrocystic (e.g., cystic hygroma), microcystic, or mixed (Figs. 10-35 and 10-36). Only the septa (separating the lymph-filled spaces) show enhancement. Proteinaceous or hemorrhagic fluid-fluid levels are characteristic. Rapid enlargement may be seen with hemorrhage or infection. Common combined malformations include venolymphatic malformation (VLM) and AVM. VLMs share features of both VM and LM. It may be difficult to distinguish VLM from mixed microcystic/macrocytic LM. AVM is a high-flow anomaly without a tumor component, although reactive tissue intensities may be present. It is composed of arterial feeding vessels, vascular nidus, and venous draining vessels. In some cases there are one or more direct AV connections or fistulae without a nidus.

A number of syndromes in childhood are associated with vascular anomalies. Examples are PHACE association (posterior fossa abnormalities, facial hemangioma, arterial abnormalities, cardiovascular defects, eye abnormalities; see Fig. 10-33), Sturge-Weber syndrome (facial cutaneous capillary or telangiectatic malformation), Beckwith-Wiedemann syndrome (facial capillary or telangiectatic malformation), Klippel-Trenaunay-Weber syndrome (capillary, venous, and lymphatic malformations), Maffucci syndrome (multiple venous malformations), Rendu-Osler-Weber (hereditary hemorrhagic telangiectasia) syndrome (capillary or telangiectatic malformation), and blue rubber bleb nevus syndrome.

**Orbit and Globe**

Hemangioma is the most common tumor of the pediatric orbit (as previously discussed) and may be preseptal, extraconal, intracanal, or multicompartamental (see Figs. 10-31 and 10-33). VM or LM may occasionally involve the orbit. Other vascular abnormalities of the orbit include varices, AVM, aneurysm, angiodysplastic syndromes, and vascular occlusive disease. Primary varices are venous malformations that drain to the cavernous sinus, face, or scalp veins. Secondary varices are associated with AV shunting (e.g., intracranial AVM, carotid-cavernous or dural AV fistulae, hemangioma) or venous occlusion (dural sinus or jugular venous atresia [smallness or absence of jugular foramina], stenosis, or thrombosis). The varices may be associated with VM or an angiodysplastic syndrome (e.g., Klippel-Trenaunay-Weber). Varices appear as prominent, tortuous flow voids whose size may vary with respiration, Valsalva maneuver, or arterial pulsation. Blood pool enhancement on CT or MRI is characteristic, but hemorrhage is rare.

Aneurysms and pseudoaneurysms of the orbit are very rare in childhood (e.g., AVM, hemangioma, carotid dissection). Prominent ophthalmic arterial collateral vessels may be seen with moyamoya disease (see Chapter 8). Other vascular anomalies and abnormalities may occur in association with cervicofacial hemangiomas.
Figure 10-34. Venous malformation (VM) of the right masseter muscle appears as a high-intensity and enhancing area (arrows) on axial T2-weighted MR image with fat suppression (A) and coronal gadolinium-enhanced T1-weighted MR image (B). Low intensities within the VM represent phleboliths.

Figure 10-35. Left neck macrocystic lymphatic malformation (LM) (cystic hygroma) on coronal T2-weighted MR image with fat suppression (A) and coronal gadolinium-enhanced T1-weighted MR image (B), showing septal and rim enhancement.

Figure 10-36. Macrocystic and microcystic lymphatic malformation (arrows) on axial T2-weighted MR image with fat suppression (A) and coronal gadolinium-enhanced T1-weighted MR image (B). Subsequent axial T2-weighted MR image (C) shows enhancing septations and hemorrhagic levels.
The Wyburn-Mason syndrome is an ocular vascular malformation with orbital and intracranial components (i.e., pituitary-hypothalamic and brainstem).

Nasal Cavity, Paranasal Sinuses, and Face
Vascular abnormalities may manifest as epistaxis, nasosinus obstruction, or cosmetic deformity. The nose and nasal cavity are vascularized by terminal branches of the internal and external carotid arteries. Common causes of epistaxis in childhood are infections, allergic rhinitis, and trauma (e.g., fracture, foreign body, excoriation). Severe or recurrent epistaxis may be related to coagulopathy, neoplasia (e.g., angiofibroma), or vascular anomalies (e.g., telangiectasia in hereditary hemorrhagic telangiectasia syndrome). CT and CTA may be performed initially. MRI or angiography may be necessary, particularly if intervention is needed for control of the bleeding.

Ear and Temporal Bone
A high jugular bulb is the most common vascular anomaly of the temporal bone. There is a thin bony covering, a poorly pneumatized mastoid, and dehiscence of the floor with protrusion of the jugular bulb into the middle ear cavity (Fig. 10-37). Symptoms may include pulsatile tinnitus, headache, or conductive hearing loss. An enhancing mass is present in the middle ear at the bony defect on CT or MRI. The anomalous vein is vulnerable to trauma or surgical procedures. Atresia or stenosis of the jugular vein may occur in isolation or in Crouzon disease, achondroplasia, and other similar conditions. The foramen is absent or small, and venous collaterals are present.

Anomalies of the ICA include aberrancy and partial or complete absence. They may be asymptomatic (e.g., tinnitus, hearing loss). Aberrant ICA results from absence of the bony plate of the carotid canal. The aberrant ICA may protrude into the middle ear cavity and lie against the tympanic membrane, making it vulnerable to trauma or surgery. Partial absence of the ICA most often involves the vertical petrous segment, including atresia of that segment of the canal. Reconstitution occurs through an enlarged inferior tympanic artery. These anomalies appear as an enhancing mass in the hypotympanicum. Complete agenesis of the ICA is associated with an atretic carotid canal. Angiography shows the arterial collateralization.

Persistent stapedial artery may occur with aberrant ICA or may be isolated (see Fig. 10-37). This anomaly is suspected when there is absence of the foramen spinosum and an anterior tympanic facial canal mass (i.e., aberrant middle meningeal artery). Hemangiomas and vascular malformations (as discussed previously) may involve the auricle and EAC but are uncommon in the temporal bone.

Neck, Oral Cavity, and Jaw
LM (i.e., cystic hygroma), VM, and hemangiomas are the most common vascular anomalies arising in the neck in childhood (see Figs. 10-32 to 10-36). They may be small and localized, or large and extensive, involving many compartments, including the mediastinum.

Jugular vein and carotid artery variants, or anomalies, are also common. The internal jugular veins are almost always asymmetric, the right larger than left. Occasionally they are multiple. The external and anterior jugular veins are also asymmetric and may be multiple or absent. The term phlebectasia (i.e., varix or ectasia) describes a normal vein that appears dilated in the supine position and may cause soft tissue fullness. The pterygoid venous plexus may be asymmetric and appears on CT or MRI as a pseudomass in the parapharyngeal space. The ICA may be tortuous, swing medially, and cause a pulsatile submucosal retropharyngeal mass. An aberrant medial course is also found in the velocardio facial syndrome and must be documented before corrective palatal surgery.

Infections and Inflammatory Processes
See Box 10-2.

**Box 10-2. Pediatric Head and Neck Infections and Inflammatory Processes**

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<th>Periorbital/orbital cellulitis/abscess</th>
<th>Inflammatory pseudotumor</th>
<th>Chorioretinitis/endophthalmitis/optic neuritis</th>
<th>Acute rhinitis/sinusitis</th>
<th>Allergic rhinitis</th>
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**Figure 10-37.** A and B, Axial CT scans of left jugular vein dehiscence and persistent stapedial artery anomaly. A, Absence of the jugular bony strut (lower arrow) and of the foramen spinosum (upper arrow). B, Mass (arrow) near the anterior tympanic portion of the facial nerve canal. C, cochlea; IAC, internal auditory canal; J, jugular bulb; MC, mandibular condyle; O, foramen ovale; os, ossicles; V, vestibule.
Orbit and Globe

The orbit is a common site of infection or inflammation, whether primary or secondary (especially from the paranasal sinuses). The infecting agent is usually bacterial and less often viral, mycotic, parasitic, or tuberculous. Noninfectious or postinfectious orbital inflammation may be seen as orbital pseudotumor with myositis. Infection may also be seen after penetrating trauma, especially if there is a foreign body. Unusual inflammations include endophthalmitis, dacryoadenitis, and optic neuritis.

Suppurative Infection

The most common orbital disease of childhood is bacterial infection (e.g., staphylococcal, streptococcal, pneumococcal, or from Haemophilus). Preseptal (periorbital) cellulitis involves the eyelid and adjacent face without intraorbital (postseptal) involvement. In infants, it is commonly of hematogenous origin (e.g., Haemophilus). Postseptal (orbital) cellulitis is usually extraconal and subperiosteal, but usually manifests with a preseptal component (Fig. 10-38). It is usually associated with ethmoid sinusitis (e.g., younger children) but may occur with maxillary or frontal sinusitis (e.g., older children, adolescents).

Orbital infection (extraconal or intracranal) may also result from facial infection, from sinus or facial fracture, or from penetrating trauma with a retained foreign body. Infection may also complicate an existing anomaly (e.g., cephalocele, nasolacrimal duct obstruction, fracture, foreign body). Other complications of orbital infection may result from facial infection, from sinus or facial fracture, or from penetrating trauma, especially if there is a foreign body. Unusual inflammations include endophthalmitis, dacryoadenitis, and optic neuritis.

Other Inflammatory Processes

Orbital invasion may follow an aggressive fungal sinus infection (e.g., mucormycosis, aspergillosis), especially in immunocompromised individuals. Vascular and cavernous sinus involvement may cause thrombosis, infarction, or hemorrhage. Other complications of sinusitis which may rarely involve the orbit include mucoceles, retention cysts, papillomas, polyps, and granulomas (as discussed below).

Ocular and Optic Inflammatory Processes

Sclerosing endophthalmitis is a granulomatous uveitis due to Toxocara canis infestation. Chorioretinitis is often present, including calcification and retinal detachment. A vitreous high density or hyperintensity without a discrete mass is often present on CT or MRI. The findings are similar to those in Coats disease. Chorioretinitis may also be seen with the TORCH infections, particularly cytomegalovirus, and may have similar findings (e.g., calcifications). Optic neuritis rarely occurs in childhood (see Chapter 8). It may be viral or postviral or may be associated with inflammatory pseudotumor, vasculitis, leukemia, granulomatous disease, or juvenile multiple sclerosis. Imaging may show optic or periocular thickening with marked enhancement.

Nasal Cavity, Paranasal Sinuses, and Face

Acute Rhinitis and Sinusitis

Upper respiratory tract inflammation is very common in childhood and usually viral or allergic. Bacterial infection is usually secondary and results from swelling, obstruction, or stasis (Fig. 10-41). The infecting agents include group A Streptococcus, Pneumococcus, Haemophilus, Staphylococcus, and Moraxella catarrhalis. Acute, recurrent, and chronic sinusitis may subsequently develop because of ostiomeatal obstruction from persistent swelling or from mucociliary disorders. The difficulty is differentiating viral or allergic inflammation from secondary bacterial infection, which requires antibiotics. Persistent ostial obstruction or mucociliary impairment allows the proliferation of anaerobic microbes. Persistent nasal discharge suggests sinusitis. Other causes of rhinitis/sinusitis are obstruction from foreign body, nasochoanal stenosis/atresia,
septal deviation, polyp, and tumor. Sinus infection may occasionally be of dental origin, including periodontitis, periapical abscess, minor trauma, and surgery (e.g., perforation with oroantral fistula). Developmental bony defects and dental cysts may also provide a direct pathway for sinusitis.

Allergic Rhinitis
Allergic rhinitis is another common cause of nasal or sinus obstruction and rhinorrhea in children (Fig. 10-42). Mucostasis with ostial obstruction is often followed by bacterial infection. Differentiating infectious from allergic sinusitis is difficult because they often coexist. The involvement is usually bilateral and diffuse in allergic sinusitis. The nasal turbinates are often edematous or thickened. Nodular thickening may be present, but air-fluid levels are unusual. Polyps often result from chronic mucosal hyperplasia and are commonly multiple. Unilateral involvement suggests ostiomeatal obstruction, and an air-fluid level suggests an infectious process. Intraluminal sinonasal fungal infections (including aspergillosis) may manifest as polypoid lesions or as fungus balls in patients with atopy (Fig. 10-43). Relative CT hyperdensity or T2 hypointensity may be seen along with marked enhancement.

Subacute and Chronic Sinonasal Infection
Fungal infection tends to be seen in chronically ill or immunocompromised children (e.g., patients with cancer and transplant recipients). Mucormycosis and aspergillosis are the most common pathogens. They are aggressive and fulminant fungal infections that invade the orbit, cavernous sinus, and neurovascular structures (Fig. 10-44; see Fig. 10-39). The result may be thrombosis, infarction, hemorrhage, or abscess. Adenoidal and tonsillar hyperplasia may cause obstruction; acute obstruction may lead to purulent rhinorrhea, and chronic obstruction to alveolar hypventilation, cor pulmonale, and sleep apnea. Nasal obstruction and
Rhinorrhea may occur with cerebral palsy, familial dysautonomia, midface anomalies, or tumors.

Sinonasal obstruction and rhinorrhea are common manifestations of cystic fibrosis (Fig. 10-45). There is chronic sinusitis with mucosal thickening, mucus inspissation, and nasal polyps. Inflammatory sinonasal disease also occurs in systemic lupus erythematosus, other rheumatoid or connective tissue diseases, Wegener granulomatosis, sarcoidosis, Churg-Strauss syndrome, and atrophic rhinitis. Nasal septal or bony sinus destruction is characteristic of these conditions.

Inflammatory pseudotumor is a chronic inflammatory lesion that may result from an exaggerated immune response. These are histologically diverse masses of acute and chronic inflammation with a variable fibrous response, often a plasmacytic component, and no granulomatous elements. These pseudotumors may respond to steroid therapy. They mimic lymphoma and chloroma.

**Imaging Findings**

The imaging findings in sinonasal congestion or inflammation may not correlate with clinical sinusitis. Acute sinusitis may show sinus opacification (i.e., edema and secretions) or mucosal thickening with air-fluid levels (see Figs. 10-41 and 10-42). Air-fluid levels may also occur from trauma, barotrauma, lavage, intubation, hemorrhage, or CSF rhinorrhea. Chronic sinusitis may appear on imaging as mucosal thickening, retention cysts, polyps, sinus opacification, loss of the mucoperiosteal margin, and osteopenia or sclerosis (see Figs. 10-14, 10-42 to 10-45). Mucosal thickening, edema, and mucous secretions are CT-isointense to hypodense, T1-hypointense, and T2-hyperintense. With inspissation, the secretions may become concretions and are CT-hyperdense, T2-hypointense, and T1-hypointense or hyperintense. Tumor is often isointense, whereas fibrosis is usually hypointense on all MR sequences. Acute or subacute inflammatory mucosal thickening usually demonstrates contrast enhancement, whereas chronic, fibrotic thickening often does not. Secretions and edema usually do not enhance. Single, or unilateral, turbinate enlargement may reflect the normal nasal cycle rather than inflammation.

**Complications of Sinusitis**

Mucous and serous retention cysts result from obstruction of submucosal mucinous glands or from a serous effusion (see Fig. 10-14). Polyps result from mucosal hyperplasia (see Figs. 10-42, 10-43, 10-45). They may be solitary or multiple and usually are allergic or occur with cystic fibrosis. Antrochoanal polyps are usually solitary and arise from the maxillary antra. They often extend through the ostium into the middle meatus, enlarge the sinonasal cavity, and may also extend into the posterior choana and nasopharynx. On imaging, cysts and polyps are homogeneous soft tissue masses with an air interface. They have CT and MRI characteristics similar to those of mucosal inflammation. Polyps often appear as rounded masses associated with ostial enlargement, sinonasal expansion, and bony attenuation. Polyps are often T2-hyperintense and may show diffuse or surface enhancement. Occasionally, they are fibrous with high CT density and T2 hypointensity. There may be trapped secretions intermixed with mucoceles.
A mucocele develops from sinus ostial obstruction and results in opacification and expansion of the sinus (Fig. 10-46). An air-fluid level may suggest a mucopyocele. On CT and MRI, the peripheral enhancement of a mucocele distinguishes it from neoplasm. CT hyperdensity, T2 hypointensity, or T1 hyperintensity may represent chronic inspissated secretions, mycetoma (e.g., aspergillosis), hemorrhage, a sinolith, or an intrasinus tooth.

Orbital complications of sinusitis include preseptal peri orbital cellulitis, postseptal or orbital cellulitis, and orbital abscess (see Fig. 10-38). Intracranial complications include meningitis, empyema, abscess, thrombophlebitis, and cavernous sinus thrombosis (see Chapter 8). Osteomyelitis rarely complicates sinonasal infection but may occur with trauma, surgery, or hematogenous spread. Osteopenia progresses to bone destruction. With chronic osteomyelitis, there is irregular thickening and sclerosis with
sequestra formation. Imaging may show an irregular, mottled pattern (similar to that in radiation osteitis).

**Ear and Temporal Bone**

**Otitis Media and Mastoiditis**

Acute and chronic forms of otitis media characteristically produce a conductive hearing loss. In the acute form there is mucosal edema, effusion, and stasis. Tympanic membrane rupture, otorrhea, and middle ear atelectasis may also occur. Initially the infection may be viral or allergic. Secondary bacterial infection is common (e.g., with *Haemophilus influenzae*, *Streptococcus*, *Moraxella catarrhalis*). Middle ear cavity and mastoid air cell opacification (CT isodensity, T2 hyperintensity) usually results from eustachian tube dysfunction or obstruction, especially in infants and young children. The opacification may represent either serous or purulent acute otitis media.

Mastoiditis results from occlusion of the aditus ad antrum and may be a complication of acute OM or the result of chronic OM (e.g., cholesteatoma), CT or MRI shows patchy mastoid opacification (i.e., mucosal edema, mucus, or purulent exudates). *Coalescent mastoiditis* refers to bony trabecular osteopenia or destruction (Fig. 10-47). Complications include supplicative labyrinthitis (MRI enhancement), facial nerve palsy (MRI enhancement), subperiosseal abscess, and neck abscess (Bezold abscess).

**Gradenigo syndrome** is the triad of petrous apex mastoiditis, eighth cranial nerve palsy, and deep trigeminal pain (Fig. 10-48). Intracranial complications result from bony erosion or septic thrombophlebitis and include epidural abscess, subdural empyema, meningitis, cerebritis, cerebellitis, brain abscess (usually in the temporal lobe or cerebellum), and dural venous sinus thrombosis.

**Chronic Otitis Media and Cholesteatoma**

Chronic OM results from persistent atelectasis or tympanic perforation, recurrent infection, and chronic effusion. Granulation tissue may be soft or fibrous, contain cholesterol or hemorrhage, and may coexist with cholesteatoma (Fig. 10-49). Enhancement may be seen only on MRI.

**Cholesteatomas** are stratified squamous epithelial masses with exfoliated keratin (CT-isodense, T2-isointense to hyperintense, nonenhancing). They may be congenital (see also under “Neoplastic Processes”) or acquired. Primary acquired cholesteatomas result from eustachian or attic obstruction with tympanic membrane (superior pars flaccida) retraction. They begin in Prussak’s space and extend to the mastoid antrum and air cells, often with extension to the posterior tympanic recesses. Secondary acquired cholesteatomas arise from chronic OM with tympanic membrane perforation (pars tensa). They tend to involve the posterior recesses. Cholesteatomas may also involve the petrous apex. Complications are related to bony erosion or deformity that may involve the scutum, ossicles, mastoid, tegmen tympani, sigmoid sinus plate, facial nerve canal, or lateral semicircular canal (e.g., labryintine fistula). Restricted diffusion (DWI-hyperintense, ADC-hypointense) assists in distinguishing cholesteatoma from other inflammatory masses (e.g., granulation). Rare intracranial complications include meningitis, abscess formation, venous sinus thrombosis, and CSF rhinorhea. Labyrinthine or facial nerve involvement may result in enhancement on MRI. Other causes of conductive hearing loss in chronic otitis media (without cholesteatoma) include ossicular erosion (e.g., incus, stapes), ossicular fixation (e.g., fibrosis, ossification), and tympanosclerosis (hyalinized collagenosis).

**Cholesterol Granuloma**

Cholesterol granuloma, which may also result from middle ear or mastoid obstruction, contains hemorrhage plus cholesterol crystals. It rarely occurs in childhood and may arise at any point from the middle ear cavity to the petrous apex, or within a mastoidectomy defect. It appears as a nonenhancing soft tissue mass with sharply marginalized bone destruction. Hyperintensity on T1- and T2-weighted MRI, which is due to hemoglobin breakdown, is characteristic.

**Otitis Externa**

Otitis externa is often self-limiting. In immunocompromised patients, a severe necrotizing form may develop that extends to the middle ear and mastoid (e.g., *Pseudomonas aeruginosa*). There may be bony involvement, but rarely intracranial extension.

**The Mastoidectomy Ear**

A simple mastoidectomy results in removal of mastoid air cells but preservation of the external canal wall and ossicles. Modified radical mastoidectomy preserves the ossicular chain, the bulk of which is removed in a radical mastoidectomy. CT evaluates the surgical defects, residual debris, ossicular chain, the inner ear for possible fistula, and the facial nerve canal.

**The Neck, Oral Cavity, and Jaw**

**Pharyngeal and Retropharyngeal Infection**

Inflammatory processes are common in childhood and may manifest as fever, sore throat, jaw pain, dysphagia, drooling, stridor, or torticollis. Intrinsic airway inflammation (e.g., epiglottitis, croup)
is covered elsewhere in this book. Acute tonsillitis often occurs in older children and adolescents and may occasionally result in peritonsillar cellulitis or abscess or may extend to the parapharyngeal and retropharyngeal spaces. Parapharyngeal or retropharyngeal lymphadenitis may do the same, especially in infants and young children (Fig. 10-50). The retropharyngeal space extends from the skull base to the mediastinum and contains lymph nodes that drain the sinonasal structures and pharynx, including the eustachian tube. Unilateral or bilateral posterior pharyngeal swelling is often seen on CT. This is to be distinguished from the more midline perivertebral space edema that may follow vertebral osteomyelitis or epidural abscess. Imaging findings include thickening of the retropharyngeal soft tissues and anterior displacement of the airway. The presence of gas in the abscess, although uncommon, is diagnostic in the absence of acute trauma, foreign body ingestion, and recent surgery. There may be anteroposterior, rotary, or transverse displacement of C1 on C2, or C2 on C3, caused by intense muscle spasm or direct inflammatory ligamentous involvement. US may be used to differentiate between adenitis and abscess (i.e., fluid content), and provides guidance for aspiration. Contrast-enhanced CT will demonstrate the location and extent of the disease and often distinguishes cellulitis from suppurative adenopathy and abscess. Complications include airway encroachment, osteomyelitis, sinus or orbital involvement, internal jugular vein thrombosis, carotid artery rupture, intracranial sepsis, and mediastinal spread. The source of infection may be apparent on CT. Examples are dental infection, penetrating foreign body, and sialolithiasis.

**Lymphadenitis, Cellulitis, and Abscess**

*Lymphadenitis* is the most common cause of lymphadenopathy in childhood. It may be primary or may follow adenotonsillar, pharyngeal, or dental infection. Acute adenitis may be bilateral (e.g., systemic infection or viral pharyngitis) or unilateral (e.g., primary streptococcal or staphylococcal infection). Persistent adenitis after antibiotic therapy may be seen with Kawasaki disease or infectious mononucleosis. Subacute or chronic lymphadenitis is more typical of mycobacterial infections, cat-scratch disease, toxoplasmosis, and AIDS. Noninflammatory adenopathy raises suspicion for malignancy (e.g., leukemia, lymphoma). US, CT, or MRI shows lymphadenopathy as one or more nodular masses along the cervical lymphatic chains that are more than 1.0 to 1.5 cm in diameter. Uniform contrast enhancement is common with viral processes. Abscess formation is characteristic of
bacterial infection (Fig. 10-51). Cat-scratch disease (i.e., due to *Rochalimaea henselae*) may mimic neoplasm, including marked nodal enlargement with necrosis and adjacent edema. Mycobacterial adenitis (tuberculous or nontuberculous) is suggested by a nodal mass with central liquefaction, thick margin enhancement, and extension to the skin (Figs. 10-52 and 10-53). Calcification is common but may also be seen in other granulomatous infections, treated lymphoma, and metastatic disease. Lymphadenopathy associated with salivary gland enlargement and multiple parotid cysts is characteristic of human immunodeficiency virus (HIV)/AIDS. Sarcoidosis may produce lymphadenopathy and parotid enlargement.

Cellulitis refers to diffuse bacterial or viral inflammation with edema, swelling, and fat plane obliteration, but no distinct mass. Extensive soft tissue infiltration of multiple tissue planes, including muscle, suggests the more severe condition of fasciitis.

Abscess is a more discrete suppurative collection. US shows one or more complex masses with partially anechoic centers. Contrast-enhanced CT shows a discrete hypodense mass that may contain gas and has rim enhancement with adjacent edema. Differentiation from necrotic adenitis may not be possible. MRI demonstrates an encapsulated mass that is T1-hypointense and T2-hyperintense and shows gadolinium ring enhancement. In addition to antibiotic therapy, surgical drainage may be necessary to prevent or address complications such as airway obstruction, rupture with aspiration, mediastinal spread, and vascular involvement (e.g., jugular venous thrombosis).

Thyroid Inflammation
Hashimoto thyroiditis is the most common acquired thyroid disorder of childhood, including hypothyroidism. US, CT, and MRI show diffuse and homogeneous enlargement of the thyroid gland. Acute suppurative thyroiditis with abscess suggests a congenital pyriform sinus fistula (see Fig. 10-23). Multinodular goiter is of mixed echogenicity, density, and intensity on US, CT, and MRI.

Salivary Gland Inflammation
Acute sialadenitis is usually viral or bacterial. Suppurative sialadenitis most commonly affects the parotid gland and usually follows prior infection, dehydration, trauma, surgery, irradiation, certain medications, or duct obstruction from stone or tumor. It may also be seen in premature neonates. Rarely is there an abscess. Suppurative submandibular sialadenitis is usually related to sialolithiasis. In sialadenitis, CT and MRI show diffuse swelling of the gland with adjacent lymphadenopathy (Fig. 10-54). A calcified stone appears as a focal ductal CT hyperdensity. There may be contrast enhancement of the gland and duct walls with ductal dilatation. Thickened, enhancing duct walls indicate sialodochitis. Abscess appears as one or more "liquid" foci with rim enhancement. Sialography is contraindicated during acute infection.

In *sialolithiasis*, stones are usually solitary and arise within Wharton’s duct. Complications include obstruction, infection, stricture, mucocele, swelling, and progression to atrophy. Calcified stones are usually visible on CT. Noncalcified stones may be diagnosed only with sialography.
Ranula results from obstruction and fluid expansion of a sublingual gland duct and manifests as a unilateral mass in the floor of the mouth (Fig. 10-55). Extension below the mylohyoid muscle anterior to the submandibular gland is called a “plunging ranula.”

Chronic sialadenitis may be idiopathic or result from recurrent bacterial infection, ductal obstruction (especially submandibular gland), granulomatous disease, prior irradiation, or autoimmune disease. The diagnosis is suggested by recurrent sialadenitis with fluctuating size or progressive gland enlargement. Sjögren syndrome is an autoimmune disease that may be limited to the salivary or lacrimal glands or may also have systemic involvement. Imaging shows glandular enlargement with sialectasis. The disease may be complicated by lymphoma. HIV/AIDS, tuberculosis, or sarcoidosis may also cause salivary gland enlargement or enlargement of intraglandular lymph nodes. Glandular involvement in HIV/AIDS includes bilateral parotid enlargement with lymphocytic infiltration, lymphoepithelial cysts, and diffuse neck lymphadenopathy. Such involvement is to be distinguished from infection and lymphoma.

Sialosis is nonneoplastic, noninflammatory recurrent or chronic salivary gland enlargement. The parotid is most commonly involved including gland enlargement but normal ducts.

Osteomyelitis
Osteomyelitis of the mandible may result from direct inoculation (e.g., trauma), hematogenous origin (e.g., distant infection, vascular catheter), or contiguous spread (e.g., dental or sinus infection). Imaging may show permeative bone destruction, soft tissue edema, cellulitis, or abscess. Chronic periosteal reaction, sequestrum formation, and bony sclerosis indicate chronicity. A chronic sclerosing form may be seen and may be associated with systemic disorders.

Neoplastic Processes
See Box 10-3.

Orbit and Globe
Neoplastic processes of the orbit and globe include ocular tumors, orbital tumors, sinus or craniofacial tumors that involve the orbit, and optic pathway tumors. Pathologically, these may be neoplastic processes of mesenchymal, neural, or malformative origin. Malformative tumors are also addressed in earlier sections of this chapter. The most common benign primary orbital “tumors” of childhood are dermoid-epidermoid (Fig. 10-56), hemangioma, lymphatic malformation, plexiform neurofibroma, and teratoma. The most common primary malignant orbital tumors are leukemia (e.g., chloroma), neuroblastoma, rhabdomyosarcoma, other sarcomas, Langerhans cell histiocytosis (LCH), and lymphoma. Tumors most often arising extraconally include dermoid-epidermoid, hemangioma, lymphatic malformation, plexiform neurofibroma, teratoma, neuroblastoma, rhabdomyosarcoma, histiocytosis, and lymphoma. The most common intraconal tumors are optic nerve glioma and hemangioma.
Mesenchymal Tumors

Rhabdomyosarcoma is the most common malignant tumor of the head and neck region. Common origins are the orbit, sinuses, pharynx, temporal bone, and neck. They may also arise elsewhere and metastasize to the orbit. These aggressive, invasive neoplasms are usually of the embryonal or alveolar subtype. Like other small round cell malignancies—neuroblastoma, Ewing sarcoma, primitive neuroectodermal tumor (PNET), lymphoma, leukemia, histiocytosis—these hypercellular tumors are often large soft tissue masses that infiltrate tissue planes and cause permeative bone destruction (Fig. 10-57). There may be intracranial extension and regional or systemic metastases. Also like other small round cell tumors, rhabdomyosarcomas are often CT isodense to hyperdense and show enhancement. MRI shows T1 isointensity to hypointensity, T2 isointensity to hypointensity or occasional hyperintensity, and variable gadolinium enhancement.

Reticuloendothelial and lymphoreticular neoplasms that involve the orbit include LCH, leukemia, and lymphoma. In Langerhans cell histiocytosis, there may be solitary or multiple soft tissue masses with lytic bony destruction of the orbit, sinuses, cranial base, or calvaria (see Chapter 8). Intracranial involvement may also occur. CT usually shows isodense to hyperdense, or occasionally hypodense, masses that enhance. MRI often shows T1 isointensity to hypointensity, T2 isointensity to hypointensity or occasional hyperintensity, and variable gadolinium enhancement. There may be bone marrow replacement (e.g., T1 hypointensity, T2 hyperintensity). There may also be pituitary-hypothalamic involvement with diabetes insipidus, absence of the posterior pituitary bright spot, and hypothalamic or stalk enhancement (see Chapter 8).

Leukemic infiltration of the orbit may occur in acute lymphoblastic leukemia. Chloromas are leukemic masses and occur more often with the myeloblastic forms (Fig. 10-58). Orbital involvement by non-Hodgkin lymphoma (e.g., Burkitt lymphoma) is more common than Hodgkin lymphoma. Juvenile angiofibroma is an invasive fibrovascular mesenchymal tumor of adolescent males that arises in the nasal cavity and may involve the orbit along with other structures (see paranasal sinus tumors).

Fibromas are mesenchymal tumors that may be isolated and relatively benign. When aggressive and malignant, they range from fibromatosis to fibrosarcoma. Benign or malignant osteochondral tumors rarely involve the orbit in childhood. Examples are osteoma, osteochondroma, aneurysmal bone cyst, giant cell tumor,
osteoblastoma, Ewing sarcoma, osteosarcoma, chondrosarcoma, and fibrosarcoma, which are discussed later.

Neural Tumors
Neural tumors of the orbit include retinoblastoma, medulloblastoma, neuroblastoma, esthesioneuroblastoma, PNET, progonoma, schwannoma, neurofibroma, and plexiform neurofibroma.

Retinoblastoma is by far the most common primary intraocular malignancy. Retinoma (retinocytoma) is a benign variant. Retinoblastoma may be multifocal, bilateral, or familial. Bilateral retinoblastoma is usually hereditary and may be associated with a pineoblastoma (trilateral retinoblastoma), additional hypothalamic involvement (quadrilateral retinoblastoma), and radiation-induced or second nonocular malignancies (e.g., osteosarcoma, fibrosarcoma, rhabdomyosarcoma). Retinoblastoma is the most important lesion to be ruled out in the differential diagnosis of leukocoria or strabismus (Fig. 10-59). Calcification occurs in more than 90% of cases. CT usually shows a high-density intraocular mass that often enhances. MRI commonly shows T1 isointensity to hypointensity, T2 isointensity to hypointensity, and gadolinium enhancement. Spread may occur along the optic nerve or by lymphatic or hematogenous means. Retinoblastoma is to be differentiated from other ocular lesions, such as retinoma, Coats disease, PHPV, retrolental fibroplasia, chronic retinal detachment, sclerosing endophthalmitis, congenital cataract, coloboma, retinal hemangioblastoma, choroidal or retinal hemangioma, and medulloepithelioma.

Neuroblastoma is the most common neural tumor to invade the orbit secondarily (Figs. 10-60 and 10-61). It is usually a nodular infiltrating mass causing permeative, blastic, or spiculated bone destruction. Esthesioneuroblastoma, PNETs, and progonomas are other neural tumors that have similar imaging findings (see later). Nerve sheath tumors rarely involve the orbit but include schwannoma, neurofibroma, and...
plexiform neurofibroma (e.g., sphen-orbital dysplasia of NF-1; Fig. 10-62).

**Optic Pathway Tumors**

Optic pathway tumors are common tumors of childhood and are often associated with NF-1 (Fig. 10-63). Solitary intraorbital lesions are rare and include hamartomas, arachnoidal hyperplasia, and low-grade astrocytomas. Tumors arising from the chiasm and optic tracts range from hamartomas and low-grade astrocytomas to anaplastic astrocytomas. Often there is combined intraorbital, intracanalicular, and intracranial optic pathway involvement. Optic gliomas must be distinguished from perioptic tumors such as a schwannoma, neurofibroma, and meningioma.

**Nasal Cavity, Paranasal Sinuses, and Face**

Tumors of childhood arising in the nasal cavity, sinuses, and face may be neoplastic or nonneoplastic (e.g., dysplastic), and circumscribed, expanding, or infiltrating. They may be of mesenchymal, neural, cutaneous or mucosal origin. The extent of regional involvement, including orbital or intracranial, is important for treatment. Combined therapies may be required (e.g., surgery, chemotherapy, and radiotherapy). Mesenchymal tumors are of vascular, soft tissue, reticuloendothelial, osteochondroid, dental, and notochordal origin. Neutral tumors include those of neuroepithelial, neural crest, and nerve sheath origins. Neoplastic lesions of cutaneous or mucosal epithelial origin are rare. The most common and important tumors arising in the nasal cavity, paranasal sinuses, and nasopharynx are juvenile
Angiofibroma, rhabdomyosarcoma, neuroblastoma, LCH, chordoma, leukemia, lymphoma, and fibrous dysplasia. Vascular anomalies and tumors and cysts of dental origin are discussed elsewhere.

Vascular Tumors

Juvenile nasal angiofibroma (JNA) is a common benign but aggressive fibrovascular tumor that occurs primarily in adolescent boys. Arising from the posterolateral nasal cavity near the pterygopalatine fossa and sphenopalatine foramen, it manifests as nasal obstruction, epistaxis, facial swelling, proptosis, otitis media, or headache (Figs. 10-64 and 10-65). These are isodense or low-density masses that enhance markedly on CT. Bony expansion and erosion are common, including widening of the pterygopalatine fossa and anterior bowing of the posterolateral maxillary sinus wall. Extension often occurs into the sphenoid, maxillary, and ethmoid sinuses as well as the orbit, middle cranial fossa, and cavernous sinus. Vascular, neural, and intracranial involvement are best evaluated on MRI. The MRI findings reflect varying components of increased vascularity (flow signal voids), fibrous tissue (hypointensity), tumor matrix (marked gadolinium enhancement), edema (T2 hyperintensity), cysts, cavitation, and hemorrhage. Sinus or otomastoid obstruction with mucosal edema and retained secretions is common. Preoperative catheter angiography and therapeutic embolization often facilitate surgical excision.

Angiomatous polyp and hemangiopericytoma are very rare in childhood but may be mistaken for angiofibroma.
Soft Tissue and Reticuloendothelial System Tumors

Common malignant soft tissue tumors of the head and neck region in childhood include rhabdomyosarcoma, lymphoma, Ewing sarcoma, histiocytosis, leukemia, neural origin tumors (neuroblastoma, PNET), and fibromatous tumors.

The orbit and paranasal sinuses are common sites of origin of rhabdomyosarcoma (see Fig. 10-57). Similar to the other small “blue” round cell tumors, these are hypercellular tumors that often manifest as infiltrating soft tissue masses with bone destruction and regional or systemic metastases. They are often isodense to hyperdense on CT with iodinated contrast enhancement, and T1-isointense to hypointense, T2-isointense to hypointense (or occasionally hyperintense), with variable gadolinium enhancement on MRI.

Langerhans cell histiocytosis is a reticuloendothelial disorder histologically characterized by tissue infiltration with reticulum cells, histiocytes, plasmocytes, and leukocytes (see Chapter 8). The involvement may be isolated (formerly eosinophilic granuloma), or there may be dissemination with cutaneous, visceral, and bony involvement.

Lymphoma is another common malignant tumor of the head and neck region in childhood. Hodgkin disease often manifests as cervical lymphadenopathy and spreads contiguously along nodal chains. Non-Hodgkin lymphoma is often widespread with noncontiguous nodal involvement. The origin may be in the nasopharynx, sinuses, adenotonsillar region (Waldeyer ring), or salivary glands. Head and neck lymphomas may be associated with childhood AIDS or PTLD (see Chapter 8).

Leukemia rarely involves the nasal cavity or paranasal sinuses. Such involvement is more often due to infection or hemorrhage. Occasionally, a chola (e.g., in myeloblastic leukemia) is seen as an osseous or soft tissue mass that may expand or destroy bone. It is often CT-isodense to hyperdense and T2-isointense to hypointense, and shows enhancement.

Fibromatous tumors are mesenchymal neoplasms that may be isolated and benign (solitary fibroma) or aggressive and malignant (fibromatosis, fibrosarcoma). The fibromatous tumor is a locally infiltrating pseudoneoplastic process characterized by fibroelastic proliferation. Desmoid tumor is a well-differentiated form with no tendency to metastasize. Wide tissue infiltration and recurrence after resection is common. Progression to fibrosarcoma may occur despite therapy. In other forms there may be widespread visceral and bony involvement without metastases (e.g., congenital or infantile form). The juvenile form usually involves musculoskeletal structures but not the viscer. Imaging shows an infiltrating soft tissue mass or masses that involve bone. CT and MRI may show isodensity to hypodensity and hypointensity, respectively, in the more fibrous forms. There may be minimal or no enhancement. CT hypodensity, T2 hyperintensity, and contrast enhancement may be seen in the more aggressive, malignant forms.

Osseous and Chondroid Tumors

Osseous and chondroid tumors may arise from the facial bones or from the skull base and may secondarily involve the nasal cavity, sinuses, and nasopharynx.

Osteoma, a benign osseous neoplasm, is rare in childhood, but most often arises in the frontal or ethmoid sinus. It may be asymptomatic or associated with headache, sinus obstruction, CSF rhinorrhea, or Gardner syndrome. The imaging appearance depends on the histologic subtype (cortical, cancellous, or fibrous), varying from a sclerotic lesion to a soft tissue density.

Osteochondroma is a benign osteocartilaginous exostosis that may arise from the mandible, maxilla, sphenoid bone, zygoma, or nasal septum. Multiple lesions occur in familial cases and in Ol- lier disease. They may arise after radiotherapy. Malignancy (e.g., osteosarcoma or chondrosarcoma) is rare except in familial cases. Imaging shows a miniature metaphysis, growth plate, and carti- laginous cap that are continuous with the bone of origin. Malignant degeneration is indicated by a disorganized appearance and involvement of the parent bone.

Fibrous dysplasia is an idiopathic and benign fibro-osseous disorder that may be monostotic, polyostotic, or part of the McCune-Albright syndrome. The maxilla and mandible are most frequently involved, unilaterally or bilaterally (Fig. 10-66). There may be encroachment upon the neurovascular foramina, orbit, nasal structures, or sinuses (see Fig. 10-5). Lesion growth may continue after skeletal maturation, but conversion to sarcoma is rare. CT and MRI findings include inhomogeneous soft tissue density or intensity, a “ground-glass” appearance, or sclerotic bony thickening. Marked enhancement may mimic neoplasm. Ossifying fibroma is a circumscribed fibrous neoplasm that progressively ossifies. The CT and MRI appearance may mimic those of fibrous dysplasia, although this tumor tends to be expansile, grows faster, and recurs. Cementifying fibroma is another fibro-osseous tumor that is aggressive and tends to recur.

Giant cell tumor, giant cell reparative granuloma, aneurysmal bone cyst, and osteoblastoma (Fig. 10-67) are benign osseous tumors rarely arising in this region in childhood. These often have overlapping pathologic findings, and combined lesions are well known. On CT and MRI, these lesions often are lytic and expansile and have a bony matrix or calcification, cortical erosion, soft tissue mass, and a thin calcified shell. Cavitation, cyst formation, and hemorrhage may be observed. Moderate contrast enhancement is common. A multiloculated appearance with fluid-fluid levels is
suggestive of aneurysmal bone cyst. The latter finding, however, has also been reported with lymphatic malformation, venolymphatic malformation, and telangiectatic osteosarcoma.

As previously discussed, cherubism is an autosomal dominant disorder with progressive giant cell tumor involvement of the mandible and maxilla in childhood (misnomer “congenital fibrous dysplasia”—see Fig. 10-26).

Chondrosarcoma is a malignant bone neoplasm of cartilage origin that may arise de novo, from an osteochondroma, or following radiotherapy. It is occasionally found in the sphenoid bone or sphenoorbital synchondrosis. On CT and MRI, this expansile mass is often of nonspecific soft tissue density, intensity, and enhancement. Chondroid matrix calcifications may not be present.

Chordoma is rare tumor that arises from intraosseous notochordal remnants in the skull base near synchondroses. These tumors are locally invasive, destroy bone, and may metastasize. The chordoid form of chordoma may be indistinguishable from chondrosarcoma on imaging.

Osteosarcoma, fibrosarcoma, and Ewing sarcoma are other rare mesenchymal neoplasms that arise in this region as primary or secondary neoplasms (e.g., after radiation therapy for retinoblastoma). These invasive tumors spread regionally and metastasize. Osteosarcoma may appear as a soft tissue mass with bony destruction and spiculated periosteal bone reaction, or as a partially calcified or ossified osteoid matrix mass. Fibrosarcoma and Ewing sarcoma produce soft tissue masses and permeative bony destruction, but no osteoid or chondroid matrix elements.

Neural Tumors

Neuroepithelial or neural crest tumors include neuroblastoma, PNET, esthesioneuroblastoma, retinoblastoma, and progono mas. Neuroblastoma, the most common of these tumors, may arise in or involve the skull base, nose, sinuses, or orbit, usually as part of metastatic disease (see Figs. 10-60 and 10-61). Esthesioneuroblastoma (olfactory neuroblastoma) is a very rare tumor that arises from the olfactory groove and produces extensive destruction of the sinuses, orbit, and adjacent skull base and extends intracranially. Primitive neuroectodermal tumors are rare extra-CNS malignancies of primitive neuroepithelial origin. They are characterized by small round cell infiltrations similar to those of neuroblastoma. Progono mas are rare retinal anlage tumors, often contain melanin, tend to arise from the cranial base, and invade the adjacent nasosinus structures or orbit. All of these tumors tend to have similar imaging appearances, consisting of a soft tissue mass with CT-isodensity to hyperdensity, T2-isointensity to hypointensity, prominent contrast enhancement, permeative bone destruction, and calcification. Schwannomas, neurofibromas, and plexiform neurofibromas rarely arise in the nasal cavity, paranasal sinuses, or nasopharynx.
Tumors of Cutaneous and Mucosal Epithelial Origin

Nasal papillomas are benign mucosal tumors that often extend into the maxillary, ethmoid, sphenoid, or frontal sinuses. Malignant transformation is extremely rare. CT and MRI often demonstrate a small or large polypoid nasal cavity mass with remodeling and ostiomeatal obstruction. Cylindric cell and inverted papillomas tend to be more aggressive. Complete surgical excision may be difficult, and recurrence is common. Squamous cell carcinoma and adenocarcinoma of the nasal cavity and sinuses are extremely rare in childhood. Imaging often demonstrates a sinus mass of homogeneous density and intensity with bone destruction. Contrast enhancement is uncommon. Necrosis and hemorrhage may occur, along with regional extension, nodal spread, and distant metastases.

Ear and Temporal Bone

Congenital cholesteatoma grows from ectopic epithelial rests (Fig. 10-68). Classically, there is no prior inflammation, trauma, or surgery. The most common site is the anterior middle ear cavity, although it may also arise in the external canal, petrous apex, or mastoid, or deep to an atresia plate (see Fig. 10-16). Usually there is conductive hearing loss and a white mass behind an intact tympanic membrane. The mass tends to be circumscribed, CT-isodense, T1-hypointense, and T2-hyperintense. Occasionally, it may be more extensive and may produce bony erosion.

Temporal bone involvement is uncommon and usually monostotic in fibrous dysplasia. Painless fibro-osseous expansion may be associated with external canal narrowing, hearing loss, or second ary cholesteatoma. CT shows expansion with “ground-glass” appearance, sclerosis, or lytic destruction. The differential diagnosis may include other fibro-osseous lesions, benign or malignant.

Exostosis is a common, benign bony hyperplasia of the external canal. It arises from the sutures of the tympanic ring, is usually localized, and is often bilateral. There may be hearing loss, pain, infection, or tinnitus. CT often shows nodular bony thickening with canal narrowing. Osteoma is an uncommon benign bony tumor that is usually unilaterial and more often arises in the outer bony canal.

Of the nerve sheath tumors in the region, neurofibromas (e.g., plexiform, NF-1) may involve the auricle and external canal. Acoustic or vestibular schwannoma is rare in childhood, suggests neurofibromatosis, and must be considered in retrocochlear hearing loss (see Chapter 8). CT may show only widening or shortening of the IAC. MRI is the study of choice, on which the tumor tends to be T2-hyperintense and gadolinium-enhancing. Schwannomas of other cranial nerves (e.g., VII, IX-XII) in this region are rare in children. Characteristically, there is an enhancing mass that expands the facial canal, jugular foramen, or hypoglossal canal.

Paragangliomas (glomus tumors) are also rare in childhood. They may be hereditary, familial, multicentric, or hormonally active. Paragangliomas are vascular, but slow-growing, tumors that may arise within the jugular bulb (glomus jugulare), middle ear cavity (glomus tympanicum), or the auricular branch of the vagus nerve (glomus vagale). Conductive hearing loss, pulsatile tinnitus, and a red retrotympanic mass are characteristic. The mass may be obscured by otitis media. Differentiation from carotid and jugular anomalies is necessary. CT may show an expanding and enhancing mass. MRI shows a T2-heterogenous mass (“salt-and-pepper” appearance), with multiple vascular flow voids and marked gadolinium enhancement. Angiography and therapeutic embolization are helpful for surgical management.

Rhabdomyosarcoma is a common malignancy in this region (Fig. 10-69). Extensive local involvement is frequent, along with intracranial invasion and metastases. CT and MRI show an enhancing soft tissue mass with bony destruction. Vascular complications include internal jugular vein invasion, compression, and thrombosis.

Langerhans cell histiocytosis occasionally involves the temporal bone and may be bilateral (Fig. 10-70). CT and MRI often show enhancing soft tissue masses with marginated bony destruction. Other skeletal lesions and intracranial involvement should be sought.

Metastasis

The most common metastatic tumors of the temporal bone are neuroblastoma and leukemia. Permeative, lytic bone destruction on CT is often seen. Differentiation from coalescent mastoiditis may be difficult. MRI better delineates intracranial involvement.

Neck, Oral Cavity, and Jaw

Benign “tumors” of the neck may be developmental, inflammatory, or neoplastic. Such lesions include cysts, ectopias, vascular anomalies, fibrosiritis colli, dermoid-epidermoid, teratoma, lipoma, and nerve sheath tumors. These are to be distinguished from lymphadenopathy, cellulitis, and abscess.

Primary malignant tumors of the pediatric head and neck vary with the age of the patient. Malignant teratoma is primarily congenital. Neuroblastoma usually arises in infants and young children. Rhabdomyosarcoma typically occurs in the preschool years. Other sarcomas and non-Hodgkin lymphoma occur over a broad age range but particularly in later childhood. Hodgkin disease, thyroid carcinoma, nasopharyngeal carcinoma, and salivary gland neoplasms most often occur in adolescence. These tumors may be asymptomatic with variable size and growth. Other symptoms and signs may be related to associated lymphadenopathy, paranasal sinus or ear involvement, aerodigestive compromise, or headache.

Figure 10-68. Left congenital cholesteatoma (arrow) on axial (A) and coronal (B) CT scans. C, cochlea; o, ossicles.
Fibromatosis coli (also known as congenital muscular torticollis) is a common benign condition of the neonate and young infant (Fig. 10-71). A firm, nontender, fibrous mass is usually palpated in the sternocleidomastoid muscle (SCM). Suggested causes include in utero deformation and birth trauma. There is venous hemorrhage evolving to fibrosis. SCM enlargement often occurs early, followed by muscle contracture and atrophy with torticollis. Ipsilateral hemifacial microsomia or plagiocephaly may also be seen. US shows a mass, or enlargement, of the SCM with variable echogenicity. CT shows an isodense SCM mass, hemorrhage, or calcification. MRI may show hemorrhage or mineralization along with enhancement. Treatment usually consists of physical therapy, but surgery may be required.

Dermoid (epidermoid) cysts are of ectodermal origin, usually occur as near-midline upper neck or scalp lesions, and may be asymptomatic (Figs. 10-72 and 10-73). They may also be associated with a dimple and dermal sinus and manifest as infection. US shows a circumscribed and thin-walled echogenic mass. CT and MRI demonstrate an encapsulated mass with fatty density or intensity. Particularly when occurring in the midline scalp, dermoid-epidermoid is to be distinguished from cephalocele (see Chapter 8) and vascular anomalies (e.g., venous malformation, hemangioma; Fig. 10-74).

Teratomas arise from pluripotential cells and usually manifest at birth as large neck masses causing respiratory or swallowing problems (Fig. 10-75). About one fifth are malignant. There is a higher incidence of polyhydramnios, stillbirth, and prematurity in infants with teratomas. Imaging shows a heterogeneous mass containing cystic areas, calcification, and variable amounts of fat.

Lipoma is a benign tumor composed of fat cells that tend to follow somatic growth (Fig. 10-76). On US, CT, and MRI, the mass has the same echogenicity, density, and intensity, respectively, as adipose tissue. The presence of other soft tissue characteristics, including enhancement, may require a differential diagnosis that includes teratoma, lipoblastoma, and liposarcoma.

Nerve sheath tumors (neurofibromas and schwannomas) arise from cranial or peripheral nerves in the neck. They may be sporadic or may occur as part of neurofibromatosis. Plexiform neurofibromas, which are pathognomonic of NF-1, consist of multiple nerve masses that incorporate adjacent soft tissues (Fig. 10-77).
**Figure 10-71.** Fibromatosis coli of the sternocleidomastoid muscle (M) on a longitudinal US image (A) as well as axial T2-weighted (B), sagittal gadolinium-enhanced T1-weighted (C), and coronal T2-weighted (D) MR images.

**Figure 10-72.** Anterior fontanelle dermoid (arrows) on sagittal T1-weighted (A) and coronal STIR (B) MR images.
The tumors are CT isodense to hypodense, T1-isointense to hypointense, and T2-isointense to hyperintense (“target sign”), and they enhance irregularly. Malignant degeneration occurs in a small percentage. Nerve sheath tumors are to be differentiated from neuroblastoma (Fig. 10-78).

Paragangliomas are exceedingly rare in this region in childhood.

Hodgkin lymphoma tends to involve contiguous nodal groups (Fig. 10-79). Non-Hodgkin lymphoma tends to be extranodal (e.g., adenotonsillar, nasal cavity, paranasal sinuses). Developmental or acquired immunodeficiency (e.g., PTLD; Fig. 10-80) predisposes to non-Hodgkin lymphoma. Asymptomatic lymphadenopathy is a common mode of presentation. Imaging findings include lymphadenopathy in several locations, usually with a dominant larger node or aggregate of nodes. Adenotonsillar involvement usually is bilateral and associated with airway obstruction. Necrosis and mineralization are uncommon except after treatment. These tumors are CT-isodense (relative to muscle), T1-isointense, T2-hyperintense, and variably enhancing. Local bone destruction may occur. Hypermetabolic activity on PET-CT correlates with tumor.

Rhabdomyosarcoma, typically the embryonal subtype, often originates in the head and neck (see Figs. 10-57 and 10-69). Other, less common sarcomas of childhood include fibrosarcoma, Ewing sarcoma, chondrosarcoma, osteosarcoma, malignant schwannoma, hemangiopericytoma, and Kaposi sarcoma.

**Figure 10-73.** Occipital dermoid (long arrows) and sinus (short arrows) with cerebellar dermoid (d) and enhancing abscess on sagittal T1-weighted (A) and gadolinium-enhanced T1-weighted (B) MR images.

**Figure 10-74.** Vertex parietal scalp hemangiomma (arrows) on sagittal T2-weighted (A), and gadolinium-enhanced T1-weighted (B) MR images, which show enhancement and vascular high-flow voids.
Thyroid adenoma and carcinoma are unusual in childhood. Carcinoma may be sporadic or may be associated with prior irradiation or multiple endocrine neoplasia type II. The papillary and follicular types are more common than the medullary type. A common presentation is an asymptomatic, firm, but mobile neck mass (Fig. 10-81). Rapid growth, fixation, aerodigestive symptoms, or cervical lymphadenopathy suggests malignancy. US may show an isoechoic to hypoechoic thyroid mass. Cystic masses can be aspirated. Solitary thyroid nodules are usually evaluated with \textsuperscript{99m}Tc or \textsuperscript{123}I scanning. Fine-needle aspiration for cytologic analysis, or open biopsy, is considered for nodules that lack, or show variable, radionuclide uptake.

Metastatic disease involving the head and neck may occur with neuroblastoma or leukemia. Primary neuroblastoma may rarely occur in the neck. Metastatic disease from a primary abdominal, thoracic, or pelvic neuroblastoma may also involve the skull, orbit, jaw, and neck nodes.

Salivary gland tumors of childhood most commonly involve the parotid gland. Hemangioma is the most common benign neoplasm (see Figs. 10-32 and 10-33), followed by the pleomorphic adenoma. Malignant tumors are more common in older children and adolescents. Mucoepidermoid carcinoma is the most common “low-grade” malignancy. Salivary gland tumors are often asymptomatic, solitary, firm, and slow-growing. Rapid growth, pain, facial nerve involvement, and cervical adenopathy suggest higher-grade malignancy. US may be used for localization and to differentiate solid from cystic masses. MRI is the best procedure for evaluating tumor character and extent. Most neoplasms (other than hemangioma) are T1-hypointense and T2-hyperintense with variable enhancement. T2 hypointensity suggests a highly cellular lesion, and local invasion often indicates malignancy.

Cysts and tumors of the jaw (maxilla and mandible) are categorized as odontogenic (dental origin) or nonodontogenic. Odontogenic cysts arise from tooth derivatives. The radicular cyst...
arises from the tooth apex, is circumscribed, and is surrounded by a thin rim of cortical bone. It is CT-hypodense and similar in appearance to periapical granuloma. The dentigerous cyst is a sharply defined unilocular or multilocular CT-hypodense cyst of the crown of an unerupted tooth (mandibular or maxillary; Fig. 10-82). The keratocyst is a unilocular or multilocular, keratin-containing, CT-hypodense cyst (usually mandibular). There may be cortical thinning and expansion. Multiple keratocysts are characteristic of the basal cell nevus syndrome.

Nonodontogenic cysts include fissural, hemorrhagic, and Stafne cysts. The fissural cyst occurs along bony fusion lines. It is usually a small, circumscribed, and corticated CT hypodensity. Hemorrhagic bone cysts tend to be mandibular, unilocular, and scalloped. Stafne cyst represents an anatomic variant (i.e., deep fossa of the submandibular gland) and is a well-defined, round or oval CT hypodensity near the mandibular angle.

Benign odontogenic tumors may be partially cystic; they include the ameloblastoma, calcifying epithelial odontogenic
tumors, and mixed epithelial odontogenic tumors (e.g., odontoma and cementoma). Ameloblastoma is the most common. It is benign but locally aggressive. It appears as a unilocular or multilocular lesion with distinct borders. There may be marginal sclerosis, expansion, a “soap-bubble” appearance, or cortical disruption with a soft tissue mass. CT often shows hypodensities interspersed with isodensities. MRI shows heterogeneous T1 and T2 intensities. Benign nonodontogenic tumors may be solid or partially cystic. These include the exostosis, osteoma, giant cell lesions, aneurysmal bone cyst, and fibro-osseous lesions.

Malignant jaw tumors may be of primary bone origin, may represent spread from an adjacent soft tissue tumor, or may be metastatic. Examples, respectively, include sarcoma, Langerhans cell histiocytosis, neuroblastoma, leukemia, and lymphoma.

**Suggested Readings**

**Texts**


**Monographs**