Practical Embryology of the Ear

The ear is composed of three interrelated but embryologically distinct regions: the external ear, the middle ear, and the inner ear. A basic knowledge of the developmental anatomy of these regions is essential to understanding congenital ear disorders, as well as normal anatomic variations that are frequently encountered in children with temporal bone pathology.

Development of the External Ear and Tympanic Membrane

The external ear consists of the auricle (pinna), the external auditory canal (EAC), and the external layer of the tympanic membrane and derives from the first and second branchial (pharyngeal) arches and first branchial cleft (or groove). The auricle begins developing from the mesoderm of the first and second branchial arches. During the 5th and 6th weeks of gestation, these two arches each give rise to three outgrowths (a total of six per side) which surround the portion of the first branchial cleft that goes on to form the EAC. These outgrowths, referred to as the auricular hillocks or the hillocks of His, enlarge and fuse to form the auricle, which is initially situated in the lower neck region, caudal to the mandible. Over the next several months, the auricle gradually ascends, ultimately attaining a normal adult position and configuration by 20 weeks of gestation. The auricle continues to grow after birth and reaches adult size by about 9 years of age (Kenna and Hirose 2003; Sadler 2010).

The EAC is a product of the dorsal portion of the first branchial cleft, which at 8 weeks of gestation deepens to form a funnel-shaped tube termed the “primary meatus.” The primary meatus is eventually surrounded by cartilage which ultimately forms the fibrocartilaginous portion of the EAC that makes up the outer third of the EAC in the mature ear (Kenna and Hirose 2003). At 9 weeks of gestation, the primary meatus further deepens and comes into contact with the epithelium of the primitive middle ear cavity (the tubotympanic recess) which derives from the first pharyngeal pouch. Proliferating ectodermal cells at the bottom of the primary meatus form an epidermal plug known as the meatal plate. The meatal plate begins to resorb during the 21st gestational week, with canalization proceeding from medial to lateral (Cole and Jahrsdoerfer 1990), and by the 28th week, only the innermost cells of the plug are left, forming the superficial layer of the tympanic membrane (Kenna and Hirose 2003; Sadler 2010). The inner layer of the tympanic membrane is formed from the endoderm of the tubotympanic recess. Separating the inner and outer layers is a middle fibrous layer which derives from first and second branchial arch mesenchyme that grows between the first branchial cleft and first pharyngeal pouch after the 9th fetal week (Kenna and Hirose 2003; Moore et al. 2013a).

The tube formed by the newly canalized meatal plug goes on to form the inner bony portion of the EAC, which at birth has not yet attained its full size nor, with the exception of the tympanic ring, become ossified. The EAC does not attain its adult size until roughly 9 years of age, while complete ossification of the EAC occurs by the second year of life (Kenna and Hirose 2003).
Development of the Middle Ear and Mastoid Cavity

The middle ear consists of the tympanic cavity, the ossicles, the eustachian tube, and various muscles, tendons, and ligaments. Development of the middle ear cavity begins in the 3rd fetal week, as expansion of the endoderm-lined first pharyngeal pouch forms the tubotympanic recess. At the 7th week of gestation, the mid-portion of the tubotympanic recess becomes narrowed, forming an isthmus that separates the tympanic cavity laterally and the eustachian tube medially (Kenna and Hirose 2003). The lateral tympanic end of the pouch subsequently divides into four sacs which progressively expand to form the middle ear proper, epitympanum, and portions of the mastoid. Expansion of the tympanic cavity proper is usually finished by the 30th week, while complete expansion of the epitympanum occurs by the 34th week. Pneumatization begins at approximately 30 weeks and is mostly complete at birth, at which time the middle ear cavity is approximately adult sized (Kenna and Hirose 2003).

The eustachian tube connects the middle ear cavity with the nasopharynx and represents the medial portion of the tubotympanic recess. At birth, the eustachian tube measures on average 18 mm in length – roughly half its adult length – and has a horizontal orientation. As a child grows, the eustachian tube gradually lengthens, ultimately reaching an average adult length of 37 mm (Ishijima et al. 2000), and its medial end angles progressively downward, eventually coming to rest by 6 years of age in the nasopharynx at the level of the inferior turbinate (Kenna and Hirose 2003).

The three middle ear ossicles (malleus, incus, and stapes) are derived primarily from the mesenchyme of the first and second branchial arches lying adjacent to the developing middle ear cavity. Specifically, a portion of the first branchial arch (Meckel’s cartilage) gives rise to the head of the malleus and the body and short process of the incus, while the second branchial arch mesenchyme (Reichert’s cartilage) produces the manubrium of the malleus; the long process of the incus; and the head, neck, crura, and lateral tympanic surface of the stapes footplate. The medial vestibular surface of the stapes footplate arises from the otic capsule (Kenna and Hirose 2003).

Ossification of the malleus and incus begins at 15 weeks gestation, while the stapes begins to ossify at 18 weeks gestation. Over time, the mesenchymal tissue surrounding the ossicles in the middle ear resorbs; however, the endodermal epithelium of the tympanic cavity remains, enveloping the ossicles and connecting them in a mesentery-like fashion to the walls of the tympanum. These epithelial connections...
go on to form the supporting ossicular ligaments. The ossicles are of adult size and shape at birth, but remnant mesenchymal tissue may remain for up to a year or even longer (Kenna and Hirose 2003; Sadler 2010).

The mastoid (or tympanic) antrum begins to form at approximately 18 weeks and at 21–22 weeks appears as a lateral extension of the epitympanum. Its lumen is generally well developed by the 34th week of gestation and is nearly of adult size at birth (Kenna and Hirose 2003; Moore et al. 2013a). The mastoid itself represents an expansion of the bone of tympanic plate and the tympanic antrum, which continue to grow during infancy and early childhood. Pneumatization of the mastoid begins at around the 34th week, and progressive invasion of the bone surrounding the antrum by the epithelium of the tympanic cavity results in the formation of multiple epithelial-lined sacs that progresses throughout infancy. At birth, the mastoid is mostly nonpneumatized, but by 2 years of age, mastoid air cells are mostly well developed. The mastoid process, the conical inferior projection of the mastoid bone, appears at approximately 1 year and is usually well developed by 3 years of age (Kenna and Hirose 2003; Moore et al. 2013a).

Development of the Inner Ear
The inner ear is made up of the membranous labyrinth and the surrounding bony labyrinth. The membranous labyrinth consists of the utricle, saccule, semicircular canals, endolymphatic sac and duct, and cochlear duct. With the exception of the endolymphatic sac and duct, these structures are of adult size and configuration at birth. The inner ears begin to develop toward the end of the third gestational week from thickenings of the surface ectoderm on either side of the developing rhombencephalon, referred to as the otic placodes. During the 4th week, the otic placodes invaginate into the underlying mesenchyme to form depressions (otic pits) which subsequently separate from the adjacent ectoderm to form the otic vesicles (otocysts). Each otocyst then divides into a dorsal utricular portion that gives rise to the utricle, semicircular canals, and endolymphatic duct and a ventral saccular portion that forms the saccule and cochlear duct (Sadler 2010). The utricle, saccule, and endolymphatic sac form during the 6th week of gestation and attain an adult configuration by 8 weeks of life (Kenna and Hirose 2003). The endolymphatic duct and sac form at 6 weeks of gestation and are among the few inner ear structures that continue to grow after birth. The endolymphatic sac rests in a fossa along the posterior wall of the petrous bone and functions to absorb endolymph and to equalize pressure between the CSF and endolymphatic systems.

The cochlea develops from the ventral saccular portion of the otocyst. During the 6th week of gestation, the saccule forms a tubular outgrowth (the cochlear duct) which penetrates the surrounding mesenchyme in a spiral fashion. The number of turns made by the cochlear duct increases with progressive development. One turn of the cochlea is present at 7 weeks of gestation, while the adult configuration of 2 1/2 to 2 3/4 turns is achieved by the 8th week of gestation. The organ of Corti arises in the wall of the cochlear duct and contains the sensory epithelium for hearing, including the hair cells. The cochlea remains connected to the rest of the saccule by a narrow tube referred to as the ductus reuniens.

The semicircular canals develop from the utricular portion of the membranous labyrinth during the 6th week of gestation, initially appearing as flattened discoid outpouchings. Over the subsequent 3 weeks, the central portions of these outpouchings become apposed to each other and then regress, resulting in the typical semicircular configuration. The superior canal is the first to develop, reaching maximal growth by week 19, followed by the posterior canal and then the lateral canal, which reaches maximal growth by week 22. Each canal has a dilated ampullary end and a nonampullary end, with the nonampullary portions of the posterior and superior canals fusing to form a common crus (Kenna and Hirose 2003; Sadler 2010).

By 7 weeks, the mesenchyme surrounding the otocyst begins to differentiate into cartilage, which eventually goes on to form the otic capsule. This cartilage undergoes vacuolization to form the perilymphatic spaces which surround the structures of the membranous labyrinth. The vestibule, which encloses the utricle, saccule, and part of the cochlear duct, begins to develop in the 8th week of gestation.
Shortly thereafter, in the 8th to 9th weeks, the cartilage around the cochlear duct begins to form two perilymphatic spaces, the scala vestibuli and scala tympani, which develop in close association with the cochlear duct. In its completed state, the bony cochlear canal is therefore divided into three separate compartments: the endolymph containing scala media (cochlear duct) and the surrounding perilymph-containing scala vestibuli and scala tympani. The lateral wall of the scala media is attached to the surrounding cartilage by a structure known as the spiral ligament, while the medial portion is connected to and supported by a cartilaginous process, referred to as the modiolus, which eventually ossifies to form the axis of the bony cochlea (Kenna and Hirose 2003; Sadler 2010).

Ossification of the otic capsule begins during the 15th week of gestation. Ossification begins in 14 centers and eventually results in formation of the petrous portion of the temporal bone. Fusion of the ossification centers is complete by the 23rd week of gestation. The adult otic capsule has a unique structure which is comprised of three layers of bone – inner and outer layers of perichondrial bone sandwiched around a central layer of intrachondral and endochondral bone (Kenna and Hirose 2003).

Development of the vestibulocochlear nerve (cranial nerve 8) parallels that of the membranous labyrinth. The nerve originates from the rest of cells which delaminate from the medial side of the otocyst by week 4 to give rise to the statoacoustic (vestibulocochlear) ganglion (Moore and Linthicum 2007). Like the otocyst, the vestibulocochlear ganglion also divides into two parts, which ultimately give rise to the cochlear and vestibular nerves. Dendrite bundles from each part run toward the otocyst by the middle of the 6th week, while medially directed axonal processes extend toward the brainstem, making contact by the 5th to 6th gestational week. The ganglion cells that form the cochlear division of cranial nerve 8 wind around the modiolus of the cochlea to form the spiral ganglion. Over the next several weeks, these primitive cochlear neurons extend axonal processes toward the developing organ of Corti, with these processes reaching the basal portions of the hair cells around the 9th to 10th week (Moore and Linthicum 2007; Bartel-Friedrich and Wulke 2007).

**Congenital Abnormalities of the Temporal Bone**

Congenital anomalies of the ear and temporal bone can present in various fashions, depending upon which parts of the ear are involved. These malformations may be obvious at birth, such as in children with abnormalities of the external ear (e.g., anotia or severe microtia); however, in patients whom pathology is very mild or limited to the middle or inner ear, these anomalies may not come to light until later in life. The overall incidence of ear malformations has been reported to be approximately 1:3,800 newborns, with outer ear malformations reported in 1:6,000–1:6,800 births (Bartel-Friedrich and Wulke 2007). The incidence of inner ear malformations is more difficult to estimate. It can be reasonably assumed that all children with congenital sensory hearing loss – which occurs with an incidence of 2–4 per 1,000 births (Smith et al. 2005) – would have identifiable abnormalities of the inner ears if their temporal bones were examined histologically. However, in the majority of cases, these abnormalities are only resolvable at a microscopic level and are not detectable with currently available imaging techniques. Therefore, the prevalence of inner ear anomalies identified with modern MRI or CT techniques in children with congenital sensorineural deafness has only been reported to be in the range of 20–30 % (Coticchia et al. 2006; McClay et al. 2008; Parry et al. 2005).

The location, extent, and severity of malformations one encounters by imaging depend upon which of the branchial arch or otocyst derivatives are affected and on the timing of developmental arrest. Disruption of ear development earlier in embryogenesis typically results in more severe and widespread malformations. While malformations involving all three parts of the ear do occur concurrently, it is actually more common to encounter outer/middle ear malformations without inner ear malformations and...
vice versa, a fact most likely related to the distinct embryogenesis of these regions (see above). Inner ear malformations are reported to occur in only 11–30% of individuals with outer and middle ear malformations (Swartz and Faerber 1985). On the other hand, malformations of the external ear usually occur in combination with middle ear anomalies, which is not surprising in light of their common embryologic origin from the first and second branchial arches. Therefore, although overlapping pathology is not particularly uncommon, from a practical standpoint, it is useful to divide the discussion of congenital ear malformations into two broad groups: (1) those predominantly involving the external and middle ear and (2) those involving the inner ear.

**External and Middle Ear Malformations**

**Microtia and Aural Atresia**

The term microtia is used to describe a spectrum of congenital malformations of the auricle ranging from mild structural abnormalities to complete absence of the external ear (anotia). Microtia is virtually always associated with stenosis or atresia of the external auditory canal, commonly referred to as congenital aural atresia (CAA), and is also frequently associated with anomalies of the middle ear. The prevalence of microtia has been reported to be in the range of 0.83–4.34 per 10,000 births, with males being more frequently affected than females, and there is an increased risk of microtia observed in children of Hispanic, Native American, Asian, or Pacific Islander descent compared to Caucasians and African-Americans (Kelley and Scholes 2007; Luquetti et al. 2012).

Between 70% and 93% of children with microtia have unilateral involvement, with the right ear being involved in approximately 60% of cases (Swartz and Faerber 1985; Luquetti et al. 2012). The majority of cases are sporadic, but approximately 14% are inherited. Additional extratemporal congenital anomalies are observed in roughly half of individuals with microtia, and bilateral microtia is more likely to be associated with birth defects outside of the ear than unilateral microtia (Kelley and Scholes 2007; Harris et al. 1996). Syndromes and anomaly spectrums associated with microtia include oculoauriculovertebral (OAV or Goldenhar) syndrome (Rollnick et al. 1987), Treacher Collins syndrome, Nager syndrome (Harris et al. 1996), Pfeiffer syndrome (Vallino-Napoli 1996), and Klippel-Feil syndrome (Miyamoto et al. 1983), among others. Several teratogens have also been implicated in the development of microtia, including thalidomide (Smithells and Newman 1992), retinoic acid (Lammer et al. 1985), and mycophenolate (Hoeltzenbein et al. 2012).

Both microtia and aural atresia result from anomalous development of the 1st and 2nd branchial arches and the intervening first branchial cleft. Arrested development of one or more of the six auricular hillocks, which first arise at approximately 6 gestational weeks (see above), results in auricular malformations. Failure of canalization of the meatal plug (which typically occurs between the 21st and 28th weeks of gestation) results in CAA. The severity of the malformation generally reflects the timing of the growth arrest, with earlier insults being likely to cause the more marked defects. Microtia and CAA typically present early in life, especially in severe cases, due to the visible anomaly.

Several classification systems have been proposed for both microtia and CAA. Among the most commonly used systems for categorizing malformations of the auricle is that of Weerda, which divides auricular defects into three grades, based on morphology and basic management principles (Weerda 1988). In first-degree dysplasia, only minor deformities are present, with most structures of a normal auricle being recognizable, and surgical reconstruction does not require use of additional skin or cartilage. Second-degree dysplasia includes ears in which some of the structures of a normal auricle remain recognizable, but reconstruction requires the use of some additional skin or cartilage. In third-degree dysplasia, none of the normal ear structures are recognizable, and total reconstruction using skin and large amounts of cartilage is required (Fig. 1).
In cases of microtia, the goal of imaging is not the characterization of the auricle, which is easily assessed by visual inspection, but rather to identify and characterize associated anomalies of the EAC, middle ear, and labyrinth. Generally speaking, the severity of the auricular malformation correlates with the severity of associated EAC and middle ear abnormalities, although exceptions occur. Approximately 70% of patients with mild (first- or second-degree) dysplasia of the auricle have EAC stenosis, while 10% demonstrate EAC atresia. In those with severe (third-degree) microtia, complete EAC atresia is present in approximately 80%, with EAC stenosis being observed in the remaining 20% (Mayer et al. 1997). Inner ear abnormalities are seen in between 11% and 30% of patients with middle and inner ear anomalies (Swartz and Faerber 1985; Mayer et al. 1997).

As is the case with microtia, a number of classification schemes have been proposed for malformations of the EAC (De la Cruz et al. 1985; Altmann 1955; Schuknecht 1989). In the widely used classification proposed by Schuknecht, CAA can be divided into four types based on the clinical evaluation, surgical findings, and type of repair needed (Schuknecht 1989). **Type A**, or high-grade meatal stenosis, is limited to the cartilaginous EAC. Left untreated, type A stenoses may result in cholesteatoma formation medial to area of narrowing, which may ultimately erode the bony canal or invade the middle ear. **Type B**, or partial

---

**Fig. 1** Microtia. (a) Photograph of the right ear demonstrating first-degree microtia. Minor deformities are present, with most structures of a normal auricle being recognizable. (b) Photograph of the left ear in a different child demonstrating second-degree microtia. Some of the structures of a normal auricle remain recognizable, but reconstruction would require the use of some additional skin or cartilage.

**Fig. 2** Type B congenital aural atresia with a secondary EAC cholesteatoma. Axial (a) and coronal. (b) CT images of the left temporal bone in a child with stenosis of the EAC involving both the bony and cartilaginous portions of the canal. There is a soft tissue medial to the stenosis scalloping the walls and floor of the EAC (arrow, b), compatible with a secondary cholesteatoma. The middle ear cavity is relatively normal.
atresia, involves narrowing of both the cartilaginous and bony EAC (Fig. 2), which may be filled with soft tissue (membranous atresia). Anomalies of the tympanic membrane and malleus are common in this form of CAA. The manubrium of the malleus is frequently short or curved, and the malleus may be fixed to the tympanic annulus or wall of the epitympanum. A bony septum may separate the middle ear cavity into a lateral compartment containing the malleus and incus and a medial compartment containing the stapes.

Type C, or total atresia, involves a completely atretic EAC with a well-developed and pneumatized tympanic cavity, while type D, or hypopneumatic total atresia, shares features of type C atresia, with the additional finding of markedly reduced pneumatization of the temporal bone. In both of these forms, there may be a partial or complete bony atretic plate (Fig. 3). The malleus and incus are usually fused and dysplastic, but the stapes is usually mobile. The course of the facial nerve is frequently abnormal, with the posterior genu and mastoid segment situated at or anterior to the level of the vestibule or round window coronally (Fig. 4). Normally, the posterior genu should be located posterior to these landmarks. In addition, the tympanic portion of the facial nerve may overlap the oval window. Characterization of the course of the facial nerve is critical in patients for whom surgical repair is being considered, given the potential for injury to the malpositioned nerve during canaloplasty. Type D CAA is also associated with anomalies of the inner ear.

Surgical correction is the treatment of choice for CAA; however, the decision of whether to repair the atretic canal depends on the likelihood of achieving serviceable hearing (Kelley and Scholes 2007). For cases of bilateral microtia and CAA, surgery is commonly delayed until the temporal bone is more fully

Fig. 3 Unilateral congenital aural atresia (CAA) in a 2-year-old child with oculoauriculovertebral syndrome. (a) Axial CT image through the midface and temporal bones demonstrates a normal right EAC and complete absence of the left EAC with a small opacified middle ear space (white arrow). (b) 3D surface-shaded lateral projection of the right face and temporal bone demonstrates a normal EAC (black arrow). (c) On the 3D reconstruction of the left face and temporal bone, no EAC is evident. In addition, there is hypoplasia of the mandibular ramus and zygomatic arch, and the glenoid fossa is shallow and poorly formed (compare with b).
pneumatized, usually at 5–6 years of age. Atresia repair typically follows microtia repair by 2 months to preserve the vascular supply to the skin and subcutaneous soft tissues. For unilateral CAA, surgery may be delayed until adulthood or not performed, depending on patient preference and hearing function in the contralateral ear. In 1992, Jahrsdoerfer and colleagues introduced a point-based grading system for CAA examining 9 CT parameters (Jahrsdoerfer et al. 1992) (Table 1). The involved ear is given a point for each parameter which is normal on CT, with two points given for presence of the stapes, resulting in a score ranging from 0 to 10 points. Higher scores are indicative of better surgical candidacy, with scores of 5 or less typically indicating poor surgical candidates. Furthermore, patients with scores of 7 have been shown on average to have significantly better hearing postoperatively than those with scores of 6 or lower (Shonka et al. 2008).

CT is the modality of choice for imaging patients with CAA and is used for identifying the type and extent of the abnormality and determining if it is surgically correctable. The findings in CAA vary with the severity of the anomaly. Types A and B are associated with stenosis and a relatively normal middle ear (Fig. 2). Types C and D have a thick irregular atresia plate and are often associated with ossicular

---

**Table 1** Jahrsdoerfer grading scale for congenital aural atresia (Jahrsdoerfer et al. 1992)

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Points awarded</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stapes present</td>
<td>2</td>
</tr>
<tr>
<td>Oval window open</td>
<td>1</td>
</tr>
<tr>
<td>Middle ear space</td>
<td>1</td>
</tr>
<tr>
<td>Facial nerve</td>
<td>1</td>
</tr>
<tr>
<td>Malleus/incus complex</td>
<td>1</td>
</tr>
<tr>
<td>Mastoid pneumatization</td>
<td>1</td>
</tr>
<tr>
<td>Incus-stapes connection</td>
<td>1</td>
</tr>
<tr>
<td>Round window</td>
<td>1</td>
</tr>
<tr>
<td>Appearance of external ear</td>
<td>1</td>
</tr>
<tr>
<td>Total points possible</td>
<td>10</td>
</tr>
</tbody>
</table>

Points are awarded for each parameter which is normal on CT.
anomalies and an abnormal course of the facial nerve. In complex forms of CAA (types C and D), the malleus and incus are often malformed, fused, or absent (Fig. 4). By definition, the tympanic membrane is absent in complete EAC atresia.

Patients with atresia or high-grade stenosis of the EAC are also at risk of developing a secondary cholesteatoma, which is estimated to occur in 4–7 % of atretic ears (Kelley and Scholes 2007). These cholesteatomas most likely develop as a result of obstruction of the normal egress of desquamated squamous epithelium lining the EAC (Cole and Jahrsdoerfer 1990; Schuknecht 1989). They are usually slow growing and are rarely diagnosed before the age of three; however, in patients 12 years and older, they are reported to be present in over 90 % of ears with an EAC caliber of 2 mm or less (Cole and Jahrsdoerfer 1990). Left untreated, these cholesteatomas can erode the bony canal or invade the middle ear (Schuknecht 1989). On imaging, they appear as soft tissue masses medial to the atretic or stenotic segment, which, when large, will erode the bony canal or ossicles (Mazita et al. 2011) (Fig. 2).

We prefer to defer evaluation with CT until 4–5 years of age to allow for maximal growth of the temporal bone.

**Ossicular Malformations**

Congenital ossicular malformations commonly present with conductive hearing loss. They occur with equal frequency in males and females and may be unilateral or bilateral. These anomalies include the absence or maldevelopment of any of the ossicles, and they are often associated with altered anatomy of other middle ear structures as well as with CAA. As is the case with CAA, ossicular malformations are best characterized with CT. MRI plays no significant role in their characterization unless concomitant inner ear or cranial nerve anomalies are suspected.

A widely used system for classifying isolated ossicular malformations is the Cremers classification, which divides congenital middle ear anomalies into four types: class I, isolated stapes ankylosis; class II, stapes ankylosis in combination with other congenital anomalies of the ossicular chain; class III, anomalies of the ossicular chain with a mobile stapes footplate; and class IV, aplasia or severe dysplasia of the oval window or round window (Teunissen and Cremers 1993).

Stapes ankylosis (Cremers class I and II anomalies) can occur as a result of fixation of either the stapes footplate or the suprastructure, with the former being much more common (Nandapalan and Tos 2000). Congenital footplate fixation is postulated to be due either to failure of differentiation of or to ossification of the annular ligament. In most cases of stapes footplate fixation, the suprastructure is of normal shape, but in 20 %, the suprastructure may be abnormal, usually taking the form of either a monopod or unicrurate type stapes (Nandapalan and Tos 2000; Teunissen et al. 1990) (Fig. 5). Unless there is an associated abnormality of the suprastructure or abnormal footplate thickening, congenital stapes footplate fixation may not be evident on CT (Swartz et al. 1986). The stapes footplate normally measures 0.25 mm in thickness and when visible on CT should only be evident as a thin white line traversing the oval window (Zeifer et al. 2000). Any thickening of the footplate should be viewed with suspicion in a patient with congenital conductive hearing loss.

Fixation of the stapes suprastructure can be due to ossification of the stapedius tendon (Fig. 6), fixation to the cochlear promontory, fixation to the tympanic part of the facial canal, or fixation of the stapes head to the posterior tympanic wall by a bony bar with a normal stapedius tendon (Nandapalan and Tos 2000; Wetmore and Gross 2011). Diagnosis requires close attention to the relationship of the suprastructure to the adjacent middle ear structures.

Class III defects consist primarily of malleoincudal fixations and incudostapedial disconnections. Malleoincudal abnormalities are probably most commonly seen in patients with CAA, in whom the malleus and incus are frequently fused and the neck of the malleus is attached to the lateral wall of the epitympanum or to the atretic plate (Fig. 4). In the absence of CAA, malleoincudal fixations are actually
the least commonly reported ossicular malformations and generally manifest as fibrous or bony bands fixing the ossicles to the epitympanum, likely reflecting incomplete pneumatization of the epitympanum or ossification of the ossicular suspensory ligaments (Fig. 7). These isolated malformations usually occur in a sporadic fashion, although cases of congenital familial malleus fixation have been reported (Miller et al. 2010). Less common is the so-called malleus bar, which refers to a bony bar that causes fixation of the handle of the malleus to the posterior tympanic wall (Kurosaki et al. 1998) (Fig. 8). This malformation can occur in isolation or in the setting of CAA (Carfrae et al. 2010).

Incudostapedial disconnections are the most common isolated congenital ossicular malformations. Patients with these malformations typically present with maximal conductive hearing loss due to large air gaps. They are characterized by an anomalous articulation between the long process of the incus and head of the stapes (Wehrs 1999). Most cases of congenital incudostapedial dissociation are due to incomplete...
formation of the long process of the incus (Fig. 9). There may be associated absence of the stapes capitulum and crura (Fig. 10), as these structures, like the long process, derive from Reichert’s cartilage. These findings may be unilateral or bilateral, with cases of inherited (either autosomal or X-linked dominant) congenital bilateral incus long process deficiency having been reported (Wehrs 1999). Congenital complete absence of the incus can also occur (Rahbar et al. 2002).

**Congenital Absence of the Oval Window**

In the normal temporal bone, the oval window niche is a rectangular recess which is easily identified along the medial tympanic wall, opening into the vestibule and housing the stapes footplate. Congenital absence of the oval window (CAOW) is a rare anomaly characterized by stenosis or atresia of the oval window (Cremers class IV anomaly). CAOW is thought to occur when the primitive stapes fails to fuse with the primitive vestibule, thus removing the normal stimulus for oval window development (Zeifer et al. 2000).
In virtually all cases of CAOW, stapes malformations are present, which is not surprising given the close anatomic and developmental relationship between the stapes and the oval window. In fact, Jahrsdoerfer reported finding a normal stapes in only 4% of ears with CAOW (Jahrsdoerfer 1977). In addition, roughly 20% of cases of CAOW are associated with CAA, and other ossicular abnormalities, usually involving the incus, are also common (Zeifer et al. 2000).

On CT, CAOW manifests either with concentric narrowing of the oval window or with complete obliteration of the oval window by a thick bony plate (Zeifer et al. 2000). The stapes is usually dysplastic and/or displaced inferiorly and posteriorly away from the obliterated oval window (Fig. 11), or it may be completely absent. Incus abnormalities, when present, usually involve the long process (Zeifer et al. 2000; de Alarcon et al. 2008). In 20% of cases, inner ear anomalies, usually manifesting as mild dilatation of the vestibule or vestibular aqueduct, are also evident (Zeifer et al. 2000).

Perhaps most important from a surgical standpoint is the association of CAOW with anomalies of the tympanic facial nerve, which occurs in roughly two thirds of cases. Like the stapes, the facial nerve develops from the 2nd branchial arch. Embryologically, the tympanic segment of the facial nerve is identifiable by the 6th week passing between the otocyst and the primitive stapes (Jahrsdoerfer 1988). Shortly thereafter, by week 8, after the stapes has contacted the otic capsule, a sulcus forms along the

---

Fig. 9 Incudostapedial disconnection due to congenital absence of the incus long process. Axial CT image through the left epitympanum (a) in a patient with left-sided conductive hearing loss demonstrates a normal malleoincudal articulation with the classic “ice cream cone” configuration, but a more inferior image at the level of the oval window (b) demonstrates absence of the incus long process which normally should articulate with the superstructure of the stapes (black arrowhead). (c) Axial right temporal bone CT image at a level comparable to (b) demonstrates a normal incus long process (white arrowhead) coursing toward the stapes (black arrowhead). The manubrium of the malleus is normal on each side (arrows, b and c).
lateral margin of the cartilaginous otic capsule, initiating formation of the horizontal facial nerve canal. If this groove is well formed, the facial nerve becomes locked into its normal anatomic position. One proposed mechanism for congenital absence of the oval window is that during the 5th and 6th weeks of gestation, the facial nerve becomes displaced and interposed between the otic capsule and stapes, thus preventing the necessary contact required to initiate oval window development (Zeifer et al. 2000).

On imaging, the facial nerve anomalies associated with CAOW include an inferiorly or medially located tympanic facial nerve course, which may pass over the oval window niche (Fig. 11), and lack of a well-defined bony facial nerve canal. Identifying an abnormal course of the facial nerve, particularly when it obstructs access to the oval window, is critical for assessing surgical candidacy and risk for facial nerve injury when an oval window drill-out is being considered.

**Congenital Absence of the Round Window**

Congenital absence of the round window (CARW) represents the other Cremers class IV anomaly. It is a rare cause of conductive hearing loss and has been reported both in syndromic and isolated cases. Associated syndromes include mandibulofacial dysostosis (Franceschetti syndrome) and endemic cretinism. CARW is also seen in association with ossicular anomalies, including stapes fixation, as well as with Mondini-type inner ear malformations. Round window absence can be unilateral or bilateral, and autosomal inherited cases are reported (Borrmann and Arnold 2007).

Embryologically, the round window develops in a connective tissue condensation between the scala tympani and the fossula of the cochlear fenestra. During otic capsule ossification, a cartilage ring located in the round window niche prevents ossification of the opening. Failure of this cartilage ring to develop results in round window absence (Martin et al. 2002). Conductive hearing loss due to round window obstruction can be explained by considering the labyrinth as a hydraulic system. The round window membrane functions to release the mechanical energy transmitted to the labyrinth from the stapes footplate. Because this fluid is noncompressible, rigid closure of the round window prevents movement of the footplate at the oval window, resulting in a conductive hearing loss.

As with most middle ear processes, CARW is best characterized with high-resolution CT and is diagnosed when there is nonvisualization of the normally air-filled round window niche, which is
Fig. 11 Oval window atresia. Axial (a) and coronal (b) CT images of the left temporal bone in a 3-year-old child with hearing loss demonstrate an atretic oval window covered by a thick bony plate (black arrows). The tympanic segment of the facial nerve (arrowheads) can also be seen coursing over the oval window. On the coronal image (b), the oval window niche is narrowed and the normal stapes superstructure is not evident. (c) Oblique axial MPR along the plane of the tympanic facial nerve (arrowhead) demonstrates an abnormally positioned stapes (white arrow) which courses posteriorly where it is fused with the posterior wall of the hypotympanum.

Fig. 12 Round window obstruction caused by a dehiscent jugular bulb. Axial (a) and coronal (b) CT images through the left temporal bone in a child with left-sided conductive hearing loss demonstrate a dehiscent jugular bulb (arrow) protruding into the hypotympanum and obstructing the round window niche (arrowhead). J jugular vein.
particularly well seen on coronal images just below the vestibule and posterior to the basal turn of the cochlea. Congenital round window obstruction can also occur as a result of a dehiscent or high-riding jugular bulb partially or completely occluding the round window niche (Weiss et al. 1997) (Fig. 12).

**Congenital Cholesteatoma**

Congenital cholesteatomas are defined as cholesteatomas that occur medial to an intact tympanic membrane in individuals without a history of otorrhea, tympanic membrane perforation, or previous otologic procedures. They are rare and account for only 2–5 % of middle ear cholesteatomas. Unlike the acquired form of the disease, congenital cholesteatoma often occurs in patients without a history of otitis media or eustachian tube dysfunction, although the coexistence of either of these entities is not grounds for excluding the diagnosis. They tend to be clinically occult until they become large enough to interfere with hearing, as patients usually present with conductive hearing loss or with an asymptomatic middle ear mass (Potsic et al. 2002; Kazahaya and Potsic 2004). The average age at presentation is approximately 5 years of age, and males are affected roughly 2.5 times more frequently than females (Potsic et al. 2002).

Although the origin of congenital cholesteatomas is controversial, most are thought to arise from remnant embryonic epithelial rests known to reside in the fetal tympanic cavity (Kazahaya and Potsic 2004). These rests are most frequently observed in the anterosuperior quadrant, which is where the majority of congenital cholesteatomas occur. Michaels et al. referred to epithelial rests occurring in this location as “epidermoid formations” (Michaels 1986). The epidermoid formation normally resorbs by the 33rd fetal week but may persist into the postpartum period, and it is hypothesized that most congenital cholesteatomas develop as a result of failure of the epidermoid formation to regress after birth (Karmody et al. 1998).

As mentioned above, the anterior superior quadrant of the middle ear is by far the most common site for congenital cholesteatomas, being involved roughly 80 % of patients, but because they are often clinically silent early on, roughly half of congenital cholesteatomas will involve two or more quadrants of the middle ear at the time of diagnosis, with roughly half involving the posterosuperior quadrant at the time of surgery (Potsic et al. 2002). Among cholesteatomas confined to 1 quadrant of the middle ear, 77 % are located in the anterosuperior quadrant, and 22 % are found in the posterosuperior quadrant. Mastoid extension is also seen roughly a quarter of the time.

While most congenital cholesteatomas originate in the middle ear, they rarely can also arise from the mastoids (Warren et al. 2007), the petrous portion of the temporal bone (Moffatt et al. 2008), or the EAC (Choi et al. 2011). The clinical presentation depends on the location of the lesion. Congenital mastoid cholesteatomas are probably the least commonly reported. Unlike congenital cholesteatomas of the middle ear, primary mastoid cholesteatomas are usually clinically silent for many years due to their location that may not be diagnosed until well into adulthood (Warren et al. 2007; Giannuzzi et al. 2011). Symptoms of congenital mastoid cholesteatomas usually only begin when the lesions become large enough to involve surrounding structures and can include otalgia, retroauricular swelling, neck pain, headache, hearing loss, or dizziness. At the time of diagnosis, congenital mastoid cholesteatomas are frequently found to erode the posterior wall of the EAC, descending facial nerve canal, sigmoid plate, jugular bulb, or the posterior fossa dural plate. They may also be associated with the development of a retroauricular abscess (Warren et al. 2007; Giannuzzi et al. 2011).

Petrous cholesteatomas are defined as cholesteatomas occurring medial to the otic capsule and can be classified as supralabyrinthine, infralabyrinthine, massive labyrinthine, apical, or combinations of these depending on their location and extent (Moffatt et al. 2008). The most frequently presenting complaint in patients with petrous cholesteatoma is hearing loss, which is usually due to labyrinthine invasion, followed by facial nerve palsy (Moffatt et al. 2008; Aubry et al. 2010). Less common complaints include vertigo, tinnitus, and otalgia. Petrous cholesteatomas may invade the labyrinth, fallopian canal, internal...
auditory canal (IAC), or cerebellopontine angle at the time of diagnosis, and surgical removal may in some cases require a middle cranial fossa or a transsphenoidal approach. Complete removal of petrous cholesteatomas may be difficult or even impossible due to adherence of cholesteatoma matrix to the internal carotid artery, jugular bulb, or dura (Aubry et al. 2010).

As mentioned above, external auditory canal cholesteatomas in children are most often associated with congenital aural stenosis and atresia. True congenital cholesteatomas occurring in the setting of a nonstenotic EAC are extraordinarily rare, with only a handful reported in the literature (Choi et al. 2011; Quantin et al. 2002). These patients present with a white mass arising along the floor of either the cartilaginous or bony EAC.

CT is the study of choice for evaluating patients suspected of harboring congenital cholesteatomas. They typically appear as sharply marginated soft tissue masses causing variable degrees of erosion of adjacent bone. Congenital cholesteatomas localized to the middle ear cavity may be impossible to differentiate from more common acquired cholesteatomas radiographically, but lesions located near the oval window are more suggestive of congenital cholesteatomas rather than the acquired form (Fig. 13).

When CT findings are equivocal, MRI can be useful in differentiating both acquired and congenital cholesteatoma from other entities in the middle ear. Both types appear as well-circumscribed masses which are of low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. They do not enhance with contrast. One key feature of cholesteatomas is that they demonstrate high signal intensity on diffusion-weighted imaging (DWI) (Warren et al. 2007), a finding which helps to distinguish cholesteatomas from most other congenital or acquired lesions in the middle ear and mastoid (see section on acquired cholesteatomas below).

Middle ear and petrous cholesteatomas may occasionally grow along the facial nerve. Identification of this spread pattern is important in order to assure complete resection. The status of the ossicles must be determined if an ossicular reconstruction is contemplated. Large congenital petrous cholesteatomas may extend into the middle cranial fossa and insinuate themselves between the dura and the floor of the middle cranial fossa. Identification of this spread pattern is important for adequate removal. Extension into the middle cranial fossa places the patient at higher risk for postoperative CSF leak. When identified, the surgeon may perform dural grafting to prevent this complication.
EAC cholesteatomas arising in the absence of CAA usually appear as soft tissue masses situated along the floor or posterior wall of the EAC with smooth erosion of the adjacent bone. Occasionally, small intralesional bony fragments may be evident (Heilbrun et al. 2003).

**First Branchial Cleft Anomalies**

Duplication anomalies of the first branchial cleft comprise fewer than 10% of branchial cleft abnormalities. They arise due to incomplete closure of the ectodermal portion of the first branchial cleft and may take the form of a fistula, a sinus tract, or a cyst (Triglia et al. 1998). Fistulas and sinuses are most common, each representing roughly half of first cleft remnants, with isolated cysts being the least common. These anomalies can be located anywhere from the floor of the external auditory canal to the submandibular region, in a roughly triangular region with its apex formed by the EAC medial to the intertragic notch (the superficial remnant of the external groove of the first cleft) and its base formed by a line between the tip of the chin and the middle of the hyoid bone. Most occur toward the apex of this triangle near the EAC or in the parotid region. This range of locations for first branchial cleft anomalies can be explained by the fact that the fetal auricle initially resides in a ventromedial position in the fetal neck caudal to the mandible before migrating upward and laterally to its final location.

Work classified first branchial cleft anomalies into two types based on their clinical and histologic features (Work 1972). Type I duplication anomalies are located medial to the concha with the cyst or sinus tract opening usually being situated inferior or posterior to the pinna or concha. These lesions are purely ectodermal and contain squamous epithelium but no adnexal skin structures or cartilage. Type II lesions pass through or adjacent to the parotid gland may lie medial to, lateral to, or between the branches of the facial nerve. A sinus tract may be evident and can end in the cartilaginous EAC or extend into the face or upper neck above the hyoid bone. Localization within the parotid gland can be explained by fact that the parotid arises as an outward evagination of the gut endothelium that envelops the facial nerve and occasionally the branchial remnant (Nofsinger et al. 1997). Type II cysts are of both ectodermal and mesodermal origin and, therefore, contain squamous epithelium with adnexal structures and/or cartilage.

Children with first branchial cleft anomalies generally present with a persistently draining or infected sinus and purulent otorrhea or with a mass in the canal, periauricular region, or upper neck (Nofsinger et al. 1997; Jakubikova et al. 2005). Most present in childhood, at a mean age of 2.4 years; however, in some cases they may not be diagnosed until adulthood and may initially go unrecognized or be mistaken for tumors or transient inflammatory lesions (Triglia et al. 1998).

Both CT and MRI can be useful adjuncts in characterizing first branchial cleft anomalies. CT demonstrates the relationship of the lesion with the EAC, the tympanic membrane, and middle ear cavity, while MRI allows better assessment of the extent of the lesion, particularly in the parotid area (Triglia et al. 1998). Unfortunately, while both modalities are excellent in detecting cysts, they perform less well for diagnosing sinuses and fistulae, and neither is particularly good for establishing the relationship of tract to the facial nerve preoperatively (Goff et al. 2012; Chan et al. 2012).

First branchial cleft cysts appear as oval or round cystic masses along the floor of the EAC, posterior to the auricle, or in a parotid or periparotid location. Generally speaking, cysts and sinuses within the ear canal or postauricular area are most likely type I lesions (Fig. 14), while lesions opening into the cheek, below the angle of the mandible, or upper neck represent type II anomalies (Fig. 15). That said, type II first branchial cleft anomalies can initially present as cysts within the ear canal (Nofsinger et al. 1997). Branchial cleft cysts are generally of fluid attenuation on CT, while on MRI they are usually of low to intermediate signal on T1-weighted images and high signal on T2-weighted images. The wall of the cyst can be of variable thickness and may enhance following intravenous contrast administration (Ibrahim et al. 2011).
**Fig. 14** Type 1 first branchial cleft cyst. (a) Coronal contrast-enhanced CT image of the head in a 12-month-old child with recurrent ear infections demonstrates a circumscribed, ovoid, cyst (arrow) along the floor of the lateral EAC. (b) Corresponding axial T2-weighted MR image demonstrates homogeneous high signal intensity within the lesion (arrow). No sinus tract was evident.

**Fig. 15** Type 2 first branchial cleft cyst. Axial (a–c) and sagittal (d) contrast-enhanced CT images through the neck in a child with a draining sinus in her right neck. The axial images at the level of the parotid (a) and mandibular angle (b) and the sagittal image (d) demonstrate a fluid-filled, peripherally enhancing sinus tract in the right neck (arrows) just below the EAC (asterisk, d). More inferiorly (c), the sinus is seen opening into a small defect in the skin of the right neck (arrowhead).
Treatment of first branchial cleft anomalies is a complete surgical extirpation of the cyst and/or tract, as recurrences may occur if the malformation is not removed along its entire course. Some recommend early surgical excision, ideally before infections occur, as inflammatory changes can distort surgical dissection planes (Goff et al. 2012). When preoperative knowledge of the course of the anomalous tract is necessary, CT fistulography can be performed to delineate the course of the fistula or sinus tract and aid in surgical planning (Whetstone et al. 2006) (Fig. 16).

**Congenital Vascular Anomalies**

Congenital vascular anomalies of the middle ear may be arterial or venous in nature. The most common vascular abnormalities described in the ear are the aberrant internal carotid artery (ICA), the persistent stapedial artery, and anomalies of the jugular bulb. These may be detected incidentally on otoscopic examination as vascular masses behind the tympanic membrane or present with nonspecific complaints including tinnitus, vertigo, aural fullness, or hearing loss (Windfuhr 2004). Although these anomalies represent developmental variants, their recognition is important preoperatively, as their presence increases the likelihood of significant hemorrhagic complications, particularly if the surgeon is unaware of them.

**Relevant Arterial Embryology**

The carotid arteries are derivatives of the primitive aortic arches, which accompany each branchial arch and connect the paired dorsal and ventral aortas. Specifically, the first aortic arch becomes the ipsilateral maxillary artery and portions of the external carotid artery, while the third aortic arch forms the common carotid artery and joins with the dorsal aorta to form the internal carotid artery (Moore et al. 2013b). The origin of the proximal external carotid artery is controversial, but it is believed to arise from either persistence of the ventral aorta or the surrounding mesenchyme (Som et al. 2003). The dorsal portion of the second aortic arch becomes the hyoid artery, a branch of the ICA which during the 5th week of gestation gives rise to the stapedial artery, around which the stapes crura later form (Hitier et al. 2013) (see above).

The stapedial artery divides into a superior (supraorbital) branch, which will become the future middle meningeal artery, and an inferior branch, which provides the infraorbital and inferior alveolar arteries. At around the same time, the ventral aorta gives rise to ventral pharyngeal arteries, which form the ventral portions of the pharyngeal arches, and it is the first ventral pharyngeal artery which is believed to give rise
Fig. 17 Embryology of the temporal bone arteries and anatomic variants. (a) Primitive internal and external carotid arterial anastomoses existing in the first trimester temporal bone. The hyoid artery (HA) is a branch of the ICA which during the 5th week of gestation gives rise to the stapedial artery (SA), around which the obturator foramen (ObF) forms. The middle meningeal artery (MMA) initially forms as a branch of the SA. The pharyngeal artery (PhA) arises from the ECA and eventually gives rise to the internal maxillary artery (IMaxA). The IMaxA anastomoses with the SA and the foramen spinosum (FSp) form around this anastomosis. The inferior tympanic artery (ITA) is a small branch of the ascending pharyngeal artery (APhA) which enters the middle ear through the inferior tympanic canaliculus (ITC) and anastomoses with the HA. (b) Normal arterial anatomy at birth. During the 10th week of gestation, the stapedial artery trunk disappears and its terminal branches becoming supplied by the IMaxA. The vestigial remnant of the HA becomes the caroticotympanic artery which anastomoses with the ITA. (c) Aberrant carotid artery. The aberrant carotid artery (red vessel) arises when there is agenesis of the proximal first embryonic segment of the ICA. The supply of the vessel is taken over by enlargement of the collateral anastomotic pathway.
to the external carotid artery and the primitive mandibular artery (the future internal maxillary artery). This mandibular artery anastomoses with the ventral portion of the stapedial artery, and the foramen spinosum forms around this anastomosis. During the 10th week of gestation, the stapedial artery trunk disappears, and its terminal branches become supplied by the external carotid system through the maxillary artery (Hitier et al. 2013). The vestigial remnant of the hyoid artery becomes the caroticotympanic artery, a small artery which passes through the middle ear and anastomoses with the inferior tympanic artery, a small branch of the ascending pharyngeal artery which enters the middle ear through the inferior tympanic canaliculus (Lasjaunias and Moret 1978) (Fig. 17).

**Aberrant Internal Carotid Artery** Lasjaunias and Moret described four variant arterial anomalies occurring in the middle ear based on angiography. These were (1) persistence of the stapedial artery, (2) an aberrant internal carotid artery without stapedial artery persistence, (3) an aberrant ICA with stapedial artery persistence, and (4) a pharyngo-tympano-petrosal artery (Lasjaunias and Moret 1978). Of these, the aberrant carotid artery, with or without stapedial artery persistence, is the most commonly reported congenital arterial variant in the middle ear, although the true prevalence of this entity is unknown. Embryologically, an aberrant ICA arises when there is agenesis of the proximal first embryonic segment of the ICA, with its flux being taken over by enlargement of a collateral anastomotic pathway existing between the inferior tympanic branch of the ascending pharyngeal artery and the caroticotympanic artery (a remnant of the embryologic hyoid artery that normally originates from the first ascending portion of the petrous ICA) (Lasjaunias and Moret 1978; Roll et al. 2003) (Fig. 17c). The result is an ICA which courses through the middle ear over the cochlear promontory before communicating with a normal horizontal petrous ICA anteriorly.

Among reported cases of aberrant ICA, females outnumber males by slightly greater than 2 to 1. In most cases, the aberrant ICA is a unilateral finding, although in 15% of cases it may be bilateral. A slight predilection for involvement of the right side over the left has been reported (Windfuhr 2004).

Hearing loss is the most commonly reported symptom, occurring in nearly half of patients found to have an aberrant ICA. Pulsatile tinnitus is the next most frequent symptom, occurring in 30%, and other less common signs and symptoms include serous otitis media, nonpulsatile tinnitus, otalgia, vertigo or dizziness, aural fullness, and headache (Windfuhr 2004). Roughly 5% of patients are asymptomatic (Windfuhr 2004). Iatrogenically induced bleeding caused by injury to an aberrant ICA during myringotomy or other middle ear surgeries is also a well-known risk, making recognition of the anomaly critical prior to instrumentation in the middle ear, and spontaneous bleeding of an aberrant ICA has also been reported (Sinnreich et al. 1984; Hunt and Andrews 2000).

CT is the diagnostic procedure of choice for distinguishing an aberrant ICA from other vascular retrotympanic masses, with the main differential considerations being glomus tympanicum and an exposed jugular bulb. An aberrant ICA classically appears as a tubular soft tissue structure arising in the hypotympanum from an enlarged inferior tympanic canaliculus, which courses anteriorly over the inferior aspect of the cochlear promontory before joining with the normal horizontal petrous ICA anteromedially through an opening in the carotid plate (Lo et al. 1985) (Fig. 18). The vertical segment of the petrous ICA canal will not be present. When contrast is administered, the aberrant vessel will show strong continuous intraluminal enhancement identical to that of other arteries. On MRA, an aberrant ICA is suggested when there is posterior, lateral, and superior positioning of the carotid genu (Davis and...
Harnsberger (1991). Frequently, the vessel will appear narrowed and irregular in contour compared to the contralateral ICA (Fig. 18).

Rarely, patients may present with a “duplicated” or “fenestrated” ICA, in which both a normally coursing petrous ICA segment and an aberrant ICA are present on the same side. In these situations, simultaneous development of both arteries creates an arterial fenestration, with both branches being of reduced caliber. As in cases of aberrant ICA, these variants will demonstrate enlargement of the inferior tympanic canaliculus, with a tubular soft tissue mass traversing the middle ear cavity before rejoining with the horizontal segment of the ICA more distally (Gartrell et al. 2012; Koenigsberg et al. 1995).

Some authors make a distinction between a truly aberrant ICA and a “lateralized” ICA (Glastonbury et al. 2012). The latter entity is a normal anatomic variant in which the genu of the petrous ICA is located posterolateral to its normal position (defined as a line drawn perpendicular to the midpoint of the basal turn of the cochlea), potentially protruding into the middle ear. The lateral bony wall overlying the carotid in this location is often thinned or dehiscent. Unlike the aberrant ICA, a lateralized ICA arises from a normal vertical petrous carotid segment and does not communicate with the inferior tympanic canaliculus. These differences are generally well depicted on CT, but differentiation can be more difficult on MRA as

Fig. 18 Aberrant carotid artery. (a) Axial unenhanced CT image through the right temporal bone demonstrates a tubular soft tissue structure (*asterisks*) which courses anteromedially over the inferior aspect of the cochlear promontory before joining with the normal horizontal petrous ICA. (b) Coronal CT image at the level of the oval window shows that this structure enters the hypotympanum through an enlarged inferior tympanic canaliculus (black arrow). (c) Contrast-enhanced axial CTA image demonstrates strong intraluminal enhancement within the aberrant carotid artery. (d) Anteroposterior MIP-reconstructed image from a 3D time-of-flight MRA in another patient demonstrates an aberrant right ICA (white arrow), which is laterally positioned, narrowed, and irregular in contour compared to the left ICA. The aberrant vessel looks like a backward number “7”
both will demonstrate a lateral position of the genu on the MRA. Clinically, however, the distinction is perhaps less important to make, as both entities can present in similar fashions, and they are both associated with an increased risk of iatrogenic injury during intratympanic surgery.

**Persistent Stapedial Artery** The stapedial artery is a fetal artery that connects branches of the primitive external carotid artery (ECA) to the ICA. Specifically, it arises from the hyoid artery near its origin from the proximal artery and extends superiorly through the mesenchymal primordium of the stapes (thus forming the obturator foramen of the stapes). After entering the cranium, it gives off an upper and a lower branch. The upper division supplies the middle meningeal artery and the orbit, while the lower division exits the cranial vault via foramen spinosum to give off mandibular and infraorbital branches, which anastomose with ventral pharyngeal branches of the ECA. Foramen spinosum itself results from the formation of the skull base around this anastomotic pathway between the primitive stapedial artery and the ECA (Hitier et al. 2013). The stapedial artery normally regresses during the 10th week, after which time its territory becomes supplied by the ECA and the ophthalmic artery (Silbergleit et al. 2000).

Persistence of the stapedial artery is extremely uncommon with a reported prevalence of 1:4,000 to 1:10,000 (Silbergleit et al. 2000). It can be associated with an aberrant ICA or duplicated ICA or may occur in the setting of a normally formed petrous ICA (Roll et al. 2003; Koenigsberg et al. 1995). When present, the persistent stapedial artery (PSA) gives rise to the middle meningeal artery, which is usually a branch of the maxillary artery. In these cases, the foramen spinosum is characteristically absent or hypoplastic.

A PSA may manifest with tinnitus or with conductive hearing loss due to associated stapes ankylosis (Jain et al. 2004; Pirodda et al. 1994). Alternatively, it may present as a pulsatile retrotympanic mass or be an incidental finding during middle ear surgery. Its presence may complicate or even preclude middle ear surgery, so preoperative identification can be essential to surgical planning and decision-making. In symptomatic cases or when the vessel crosses the operative field, surgical ligation of the PSA can generally be performed without complication (Hitier et al. 2013).

Four anatomic variants of PSA are described: (i) the hyoido-stapedial artery, (ii) the stapedial artery with aberrant carotid artery, (iii) the pharyngo-stapedial artery (PSA arising from the pharyngeal artery via the inferior tympanic artery), and (iv) the pharyngo-hyo-stapedial artery (PSA supplied from both the inferior tympanic hyoid arteries) (Hitier et al. 2013). Of these variants, the hyoido-stapedial artery and the stapedial artery with aberrant carotid artery are the most frequently reported types, while the latter two variations are exceedingly rare.

The hyoido-stapedial variant is the most commonly described form of PSA and represents persistence of the stapedial artery trunk (the hyoid artery) (Fig. 17d). Anatomically, these vessels arise from the proximal petrous ICA and enter the anteromedial hypotympanum contained in an osseous canal. They then course through Jacobsen’s canal for a short segment, exit the canal at the cochlear promontory, and proceed dorsally and cephalad through the obturator foramen of the stapes before entering the facial canal just posterior to the cochleariform process. Typically, they measure between 1.5 and 2.0 mm in diameter, although vessels as small as 0.4 mm have been reported (Silbergleit et al. 2000). The PSA then travels anteriorly in the facial canal, exiting just posterior to the geniculate ganglion where it becomes the middle meningeal artery (Hitier et al. 2013; Silbergleit et al. 2000). On high-resolution CT, the intratympanic course of the PSA can generally be followed (Fig. 19). Additional indirect signs of PSA include apparent enlargement of the tympanic segment of the facial nerve and nonvisualization of the ipsilateral foramen spinosum. It should be noted, however, that the absence of foramen spinosum does not necessarily equate to the presence of a PSA, as isolated agenesis of the foramen can be seen in up to 3 % of the population, particularly in cases in which the meningeal artery originates from the ophthalmic artery (Ginsberg...
et al. 1994). Nevertheless, if foramen spinosum is absent, a careful assessment of the middle ear should be made to rule out a PSA.

When associated with an aberrant ICA, the PSA arises from the intratympanic segment of the ICA (Fig. 17e). In these cases, the intratympanic portion of the PSA may not be directly evident on high-resolution CT, but its presence can be inferred indirectly by the absence of foramen spinosum and the enlargement of the anterior tympanic facial nerve canal. If necessary, MRA or cerebral angiography would be required to confirm the diagnosis and would demonstrate the PSA arising from the aberrant ICA and continuing intracranially as the middle meningeal artery (Silbergleit et al. 2000; Lau et al. 2004).

**Jugular Bulb Anomalies** The jugular bulb is a focal venous dilatation located at the confluence of the lateral dural venous sinuses in the jugular fossa and represents the junction between the sigmoid sinus and the internal jugular vein. It typically resides below and medial to the hypotympanum and inferior and posterior to the IAC, although its precise location within the temporal bone can vary. The jugular bulb does not actually form in humans until after 2 years of age, and it has been hypothesized that the bulb develops as a result of hemodynamic changes that only occur once an infant begins to assume an erect

![Fig. 19](image)

**Fig. 19** Persistent stapedial artery (PSA) – right temporal bone CT from a 7-year-old child with hearing loss. (a) Axial image through the hypotympanum demonstrates a PSA (arrow) coursing superiorly in the middle ear. Notice the absence of foramen spinosum in its expected location (black arrowhead) posterolateral to foramen ovale. (b) Axial image at the level of the mesotympanum shows the PSA (arrow) coursing along the cochlear promontory. (c) Axial image through the epitympanum. After passing through the obturator foramen, the PSA joins the canal for the tympanic facial nerve. Note the apparent enlargement of the facial nerve canal (white arrowhead). (d) Right anterior oblique reconstructed image demonstrates the PSA (arrow) coursing from the petrous internal carotid artery (ICA) through the hypotympanum.
position (Okudera et al. 1994; Friedmann et al. 2011). Specifically, erect posture from standing may create an ascending pulse wave from the heart that travels upward to strike the jugular sinus at the jugular foramen, causing gradual enlargement of the bulb during childhood that stabilizes sometime during adulthood.

The jugular bulbs are frequently asymmetric, with the right bulb being twice as likely to be larger than the left (Friedmann et al. 2011; Durgun et al. 1993). The predilection for right-sided jugular dominance may be due to nonsynchronous embryologic development of the venous sinuses and/or vena cava leading to asymmetric flow preferentially to the right. It has been alternatively suggested, based on the above described hemodynamic theory of jugular bulb development, that the increased prevalence of right-sided jugular bulb dominance may be due to the greater relative length of the left brachiocephalic vein, which could be better at dissipating upward forces from the venous pulsations of the heart, resulting in a smaller left jugular bulb (Friedmann et al. 2011). This asymmetry is generally not clinically significant and rarely presents a diagnostic problem; however, on MRI, asymmetric flow alterations or contrast enhancement in a prominent jugular bulb can occasionally mimic jugular thrombosis or a jugular foramen mass (Fig. 20). Unenhanced CT will demonstrate relative enlargement of one jugular foramen, with preservation of the cortex and jugular spine being the clue that one is dealing with normal asymmetry rather than a true jugular foramen mass.

In addition to normal jugular bulb asymmetry, several other jugular bulb variants occur in the pediatric population. While these anomalies may cause symptoms such as pulsatile tinnitus or hearing loss, they are usually asymptomatic or incidental findings on imaging (Atmaca et al. 2014). The most commonly described jugular bulb anomalies are the high-riding jugular bulb, the dehiscent jugular bulb, and the jugular bulb diverticulum. Together, these variants occur in roughly 10–15 % of individuals, but they are seen less frequently in children, being present in fewer 2 % of patients under the age of 10 and fewer than 5 % of individuals between the ages of 11 and 20. The prevalence of jugular bulb anomalies actually increases gradually during the first four decades before plateauing after around 50 years of age, suggesting that most are acquired and that anomalies in children are either congenital or develop very early in life (Friedmann et al. 2012a).

A high-riding jugular bulb is the most commonly encountered jugular bulb anomaly, with an overall prevalence of up to 15 %, but its prevalence in children is probably lower (Atmaca et al. 2014; Friedmann et al. 2012a; Kupfer et al. 2012). Although definitions vary, the jugular bulb is generally considered high
riding when its dome lies at or above the level of the floor of the IAC or reaches the level of the basal turn of the cochlea (Atmaca et al. 2014; Kupfer et al. 2012) (Fig. 21). A high-riding jugular bulb is usually an incidental finding, but it can be a cause of pulsatile tinnitus or hearing loss. They frequently may encroach upon or erode the vestibular aqueduct, but whether this finding is consequential is debatable as it is frequently found in asymptomatic individuals (Kupfer et al. 2012).

A dehiscent jugular bulb is considered when the bony sigmoid plate separating jugular bulb from the middle ear is absent, allowing protrusion of the bulb into the hypotympanum (Fig. 22). The prevalence of jugular bulb dehiscence detected on CT or histology is approximately 1–7 % of temporal bones, or approximately 13 % of patients, and it is frequently, but not always, described in the setting of a high-riding jugular bulb (Atmaca et al. 2014; Kuhn et al. 2012). This anomaly has alternatively been referred to as an “exposed jugular bulb,” “lateral jugular bulb diverticulum,” or simply “jugular diverticulum” (Bush et al. 2009). Unfortunately, there is a lack of uniformity in the literature regarding the usage of the term “jugular diverticulum,” which is often used interchangeably with jugular bulb dehiscence (Kuhn et al. 2012; Kutz and De la Cruz 2007; Shihada et al. 2008), potentially leading to some confusion in
diagnosis (see discussion of jugular bulb diverticulum below). In our opinion, the term “diverticulum” should be avoided when describing a dehiscent jugular bulb that protrudes into the middle ear. Patients with an exposed jugular bulb may present with a blue vascular retrotympanic mass lesion, which is usually situated in the posterior portion of the hypotympanum but may occasionally extend to the level of the mesotympanum. Left unrecognized, a dehiscent jugular bulb may place patients at risk for jugular injury and massive bleeding during myringotomy (Atmaca et al. 2014; Shihada et al. 2008). Patients can also present with tinnitus or conductive hearing loss. The latter symptom occurs when the protruding bulb contacts the tympanic membrane or ossicular chain or obstructs the round window niche (Weiss et al. 1997) (Fig. 12).

A jugular diverticulum, also referred to as “petrous jugular malposition,” is a focal outpouching of the jugular bulb that typically extends superiorly, medially, and posteriorly in the petrous bone (Bilgen et al. 2003; Pappas et al. 1993). Jugular diverticula are considered by some to be true congenital venous anomalies, but others question whether they are actually in fact progressive lesions capable of expanding to cause clinical symptoms (Bilgen et al. 2003). They are less common than the high-riding jugular bulbs and jugular dehiscence and occur with a reported prevalence of 1–3 % in patients undergoing temporal bone CT (Friedmann et al. 2012a; Bilgen et al. 2003). In spite of the common predilection for right-sided jugular dominance in most patients, jugular diverticula occur more frequently on the left side (Bilgen et al. 2003; El-Kashlan et al. 2000). The relationship between jugular diverticula and clinical symptoms is questionable, as the majority are asymptomatic, but given its proximity to the inner ear, it is believed that they may produce sensorineural hearing loss, tinnitus, or vertigo if they extend into inner ear structures such as the cochlea, vestibular aqueduct, posterior semicircular canal, or IAC.

On CT, a jugular diverticulum can be identified as a circumscribed, smoothly margined soft tissue mass contiguous with the jugular bulb which extends superiorly into the petrous bone (El-Kashlan et al. 2000) (Fig. 23). MR venography or contrast-enhanced MRI, preferably in the coronal plane, demonstrates a superiorly directed focal outpouching arising from the jugular bulb.

**Congenital Abnormalities of the Inner Ear**

Inner ear malformations occur as a result of an interruption of the development of the ear during the first trimester of pregnancy. Anomalous development of the membranous labyrinth leads to derangements in formation of the otic capsule and organ of Corti, which ultimately results in sensorineural hearing loss (SNHL) that is usually detected at or shortly after birth (Jackler 2010). The disturbance of inner ear
development may be idiopathic, associated with an inborn genetic error, or caused by exposure to a teratogenic agent during the period of inner ear organogenesis. Genetic errors may be autosomal or recessive and can be transmitted in an X-linked or autosomal fashion. Hearing loss in hereditary cases may be isolated or associated with various syndromes. Nonhereditary causes of SNHL include in utero exposure to agents including ototoxic drugs (e.g., aminoglycosides), chemical teratogens (e.g., thalidomide), in utero viral infections (particularly rubella and CMV), and radiation exposure. Furthermore, the risk of hearing loss may not be limited to first trimester exposures. Milder degrees of hearing loss have been noted in animals with aminoglycoside exposures during the human equivalent of the last two trimesters of pregnancy (Jackler 2010).

From a practical standpoint, developmental malformations of the inner ear can be divided into two broad categories: those with presumed intrinsic malformations limited to the membranous labyrinth, which appear “normal” by imaging, and those with malformations involving both the membranous and bony labyrinth, which are radiographically detectable (Jackler 2010). With higher-resolution CT and new developments in MRI, the distinction between these two groups is likely to become increasingly blurred. Nevertheless, most children with SNHL currently still demonstrate normal morphology on temporal bone imaging studies.

Up to 50 % of children with prelingual SNHL harbor mutations of the GJB2 gene, which encodes for the gap junction protein connexin 26 (Denoyelle et al. 1999). Most reports suggest that inner ear malformations are uncommon in children with GJB2 mutations, and many have suggested that routine imaging is not necessary in these individuals (Azaiez and Smith 2007; Greinwald and Hartnick 2002; Preciado et al. 2004). This topic is controversial, though, as Propst et al. reported that 72 % of subjects with biallelic GJB2 mutations demonstrated at least one temporal bone anomaly by CT imaging, with mild endolymphatic fossa enlargement and modiolar hypoplasia being the most common findings (Propst et al. 2006). These findings are usually quite subtle, however, and it is debatable whether identification of these very mild abnormalities ultimately alters patient management. That said, identification of more severe anomalies is important in surgical planning, as these malformations may make cochlear implant placement more challenging and also may increase the risk for complications including perilymph or CSF leak, postimplantation meningitis, and electrode misplacement, including placement into the IA-C. Furthermore, the course of the tympanic facial nerve is reported to be abnormal in approximately 15–32 % of patients with cochleovestibular anomalies, a finding which often influences surgical planning and occasionally precludes placement of the cochleostomy in an optimal position relative to the round window (Papsin 2005).

The following sections will focus primarily on conditions in which abnormalities of the inner ears are evident by imaging.

Malformations of the Membranous and Bony Labyrinth
With modern MRI and CT techniques, inner ear anomalies are identified only in approximately 20–30 % children with congenital SNHL (Coticchia et al. 2006; McClay et al. 2008; Parry et al. 2005). The majority of inner ear malformations are bilateral and symmetric, and roughly one-half of those with a unilateral anomalies also have some hearing loss in the “normal” ear, suggesting the presence of an abnormality limited to the membranous labyrinth on that side (Jackler et al. 1987). Most combined malformations of the membranous and bony labyrinth are probably due to arrest in the development of one or more components of the inner ear during the 4th to 8th weeks of gestation. The most convincing evidence for this theory arises from the fact that these malformations bear a strong resemblance to the appearance of the developing inner ear during this period in embryogenesis (Jackler 2010). In general, earlier developmental insults result in more severe deformities and worse hearing loss.
In 2002, Sennaroglu and Saatchi (Sennaroglu and Saatci 2002) proposed a classification for cochleovestibular malformations (Table 2), based in part on earlier work by Jackler and colleagues (Jackler et al. 1987), which categorized vestibulocochlear malformations into six types based on the theoretical timing of the causative in utero insults. In order of decreasing severity (or increasing age at developmental arrest), these malformations were labyrinthine aplasia (Michel anomaly), cochlear aplasia, common cavity deformity, cystic cochleovestibular malformation (incomplete partition type I or IP-I), cochleovestibular hypoplasia, and incomplete partition type II (IP-II).

**Labyrinthine Aplasia (Michel Anomaly)** First described in 1863, Michel anomaly (complete labyrinthine aplasia) is a rare malformation characterized by complete aplasia of the membranous labyrinth (Michel 1863). This anomaly is the most severe of the deformities involving the osseous and membranous labyrinth and is presumed to be caused by arrest of otic placode differentiation, which normally occurs...
during the 3rd week of gestation (Marsot-Dupuch et al. 1999). Patients present with complete SNHL and do not benefit from amplification or cochlear implantation. CT demonstrates a complete absence of the inner ear structures including the cochlea, vestibule, and semicircular canals (Fig. 24). Because the external and middle ears do not arise from the otic capsule, the EAC and middle ear cavity may be normal, but the absence of the stapes and an anomalous course of the facial nerve can be associated findings. In addition, in cases of Michel aplasia, the medial wall of the middle ear is typically flattened due to the absence of a normal cochlear promontory and lateral semicircular canal, and the IAC is usually small or atretic, findings which can help to distinguish the entity from labyrinthitis ossificans (see discussion on labyrinthitis ossificans below) (Marsot-Dupuch et al. 1999).

**Cochlear Aplasia** In cochlear aplasia, the cochlea is absent. The vestibule and semicircular canals are formed, but they may be normal, dilated, or hypoplastic. According to Jackler et al., this deformity results from an arrest in the development of the cochlear bud during the 5th week of gestation (Jackler et al. 1987); however, Sennaroglu and Saatci maintain that the arrest occurs earlier, before the 4th week (Sennaroglu and Saatci 2002). CT demonstrates an absent cochlea with a partially or completely formed vestibule and semicircular canals (Fig. 25). Because the cochlea is absent, the course of the labyrinthine facial nerve is more anterior than usual (Sennaroglu and Saatci 2002). As with complete labyrinthine aplasia, the main differential consideration for cochlear aplasia is labyrinthitis ossificans. In cochlear aplasia, the normal bulge of the cochlear promontory is absent.

**Common Cavity** The common cavity deformity is characterized as a malformed inner ear in which the vestibule and cochlea are confluent and completely devoid of differentiation. This anomaly results from arrested otic development between the 4th and 5th weeks of gestation prior to the separation of the otocyst into dorsal and ventral divisions. The common cavity is characterized by an ovoid or spherical smooth-walled cavity containing primordia of the membranous cavity. The overall neural population is sparse although some cells that resemble those of the organ of Corti may be scattered along the walls of the cavity. Patients typically present with profound SNHL, but good hearing has been observed in patients with this deformity after cochlear implantation. On CT, this dysplasia is characterized by replacement of
the vestibule and cochlea by a common cystic cavity, which is devoid of internal features (Fig. 26). The cavity can be variable in size but, on average, measures 10 mm in its largest dimension horizontally (Jackler 2010). Usually, the IAC enters the common cavity at its center. This finding can be helpful in distinguishing a common cavity deformity from cochlear aplasia with dysplastic semicircular canals. In the latter malformation, the cavity representing the dysplastic vestibule and SCCs is centered posterior to the IAC (see below) (Jackler 2010; Sennaroglu and Saatci 2002).

Incomplete Partition Type I In incomplete partition type I (IP-I or cystic cochleovestibular malformation), there is a cystic, dilated vestibule accompanied by a cystic, dilated cochlea. In these cases, the cochlea and vestibule are distinguishable, but they demonstrate no internal architecture. The modiolus is completely absent from the base to the apex, and the vestibule is grossly dilated. These malformations occur due to arrest of development at the 5th week (Sennaroglu and Saatci 2002). Unlike incomplete partition type II (IP-II or Mondini malformation), IP-I is not associated with vestibular aqueduct enlargement. On CT, IP-I malformations demonstrate some cochlear and vestibular separation, which distinguishes the malformation from the common cavity deformity, but both cavities are dilated and featureless and together form a characteristic “Fig. 8” or “snowman” contour (Fig. 27). In addition, the IAC is usually enlarged, and the cribriform area between the cochlea and IAC is often widened or defective.

Cochlear Hypoplasia Cochlear hypoplasia is believed to result from an insult at approximately 6 weeks of gestation. In this malformation, the cochlea is more differentiated than in IP-I, and there is clear separation of the cochlea and vestibule, but histologically, the modiolus is either malformed or absent. The cochlea is smaller than normal, and the vestibule may also be absent, hypoplastic, or enlarged (Jackler 2010; Sennaroglu and Saatci 2002). Vestibular aqueduct enlargement is not typically associated with these malformations (Sennaroglu and Saatci 2002). Patients with cochlear hypoplasia have variable hearing loss that is dependent on the degree of development of the membranous labyrinth. CT shows a small cochlear bud usually measuring between 1 and 3 mm (Fig. 28). In addition to potential vestibular anomalies, the semicircular canals are malformed in roughly 50 % of patients (Jackler 2010).
Incomplete Partition Type II (Mondini Malformation)  

Incomplete partition type II (IP-II or Mondini malformation) is the mildest and most common type of cochlear anomaly detected by imaging, accounting for approximately 50% of cochlear malformations (Jackler 2010). Some confusion surrounds the use of the term “Mondini” malformation, as it is often and incorrectly used to describe any malformation of the bony labyrinth. The term should be reserved for malformations characterized by an incomplete interscalar or osseous spiral lamina with fusion of the apical and middle turns, leading to a cochlea with only 1.5 turns (Jackler 2010; Sennaroglu and Saatci 2002). This malformation is caused by an arrest in labyrinthine development at the 7th week of gestation (Sennaroglu and Saatci 2002). The degree of development of the organ of Corti and other auditory neural elements is variable, and the degree of hearing

---

**Fig. 27** Type 1 incomplete partition anomaly (IP-1 or cystic cochleovestibular malformation). **(a)** Axial CT image of the left temporal bone in a 1-year-old child with bilateral sensorineural hearing loss shows the cochlea, vestibule, and semicircular canals to be dysplastic. There is some cochlear and vestibular separation, which distinguishes this malformation from the common cavity deformity, but both cavities are dilated and featureless and together form a characteristic “Fig. 8” or “snowman” contour. **(b)** Axial CISS MR image from the same patient again demonstrates separate but featureless cochlear and vestibular cavities bilaterally.

**Fig. 28** Cochlear hypoplasia. Axial CT image of the left temporal bone in a 7-year-old child with sensorineural hearing loss demonstrates a small, featureless cochlear bud (arrowhead) which is distinct from the relatively normal-appearing vestibule (arrow).
Fig. 29 Type 2 incomplete partition anomaly. Axial CT image through the left temporal bone in a 4-year-old child with sensorineural hearing loss demonstrates a small dysplastic cochlea (arrow), with a deficient modiolus. In these patients, the cochlea appears more fully formed than in cases of cochlear hypoplasia, but the cochlea only makes 1 1/2 to 1 3/4 turns rather than the normal 2 1/2 turns.

Fig. 30 Enlarged vestibular aqueduct. (a) Axial CT image through the right temporal bone in a child with sensorineural hearing loss demonstrates a widened, fan-shaped vestibular aqueduct, which is wider at its midpoint (double arrow) than the limbs of the nearby semicircular canals. (b) Axial CT image slightly inferior to (a) demonstrates a mildly dysplastic cochlea (arrowhead) with an incompletely formed modiolus, resulting in a fused appearance of the apical and midturns (IP-II or Mondini malformation).

loss depends on the extent of the underlying dysplasia. On CT, IP-II is characterized by a small dysplastic cochlea with a deficient modiolus or absent interscalar septum, resulting in a cochlea with only 1 1/2 to 1 3/4 turns and with only the basal turn appearing normal (Figs. 29 and 30). The vestibule and semicircular canals may also be deformed, though typically only mildly.

Enlarged Vestibular Aqueduct An extremely common, and probably characteristic, feature of IP-II malformations is enlargement of the vestibular aqueduct and endolymphatic sac. In fact, Sennaroglu and Saatchi reported that all patients with IP-II malformations had vestibular aqueduct enlargement.
(Sennaroglu and Saatci 2002), although we have observed cases of IP-II with a normal-appearing vestibular aqueduct on CT. Hearing loss occurring in the setting of this vestibular aqueduct enlargement is commonly referred to clinically as the “large vestibular aqueduct” (LVA) or “enlarged vestibular aqueduct” (EVA) syndrome (Valvassori and Clemis 1978). This anatomic anomaly is described in association with Pendred syndrome (see below), but is also frequently seen in children with nonsyndromic hearing loss (nonsyndromic enlargement of the vestibular aqueduct or NSEVA). It has also been reported in association with other syndromes including branchio-oto-renal (BOR) syndrome (Chen et al. 1995) and Waardenburg syndrome (Madden et al. 2003) and in association with distal renal tubular acidosis (Berrettini et al. 2001).

The vestibular aqueduct is a bony canal extending from the medial aspect of the vestibule to the posterior wall of the petrous bone and contains the endolymphatic duct and sac. The endolymphatic duct forms from the union of the utricular and saccular ducts and communicates directly with the membranous labyrinth and the endolymphatic sac. Traditionally, the endolymphatic duct has been depicted as a long thin tubular structure ending in a short, blunt, pouchlike endolymphatic sac. In reality, the endolymphatic duct is quite short – measuring only 2 mm in length – while the endolymphatic sac is a much larger and complex structure consisting of multiple interconnecting tubules, cisterns, and crypts (Lo et al. 1997). The proximal portion of the sac is located in the vertical segment of the vestibular aqueduct, and the distal extraosseous portion of the sac rests in a recess along the posterior wall of the petrous bone, between layers of dura. The endolymphatic sac serves two main functions: equalization of pressure between CSF and the endolymphatic system and endolymph absorption (Kenna and Hirose 2003).

EVA syndrome is more common in males, and bilateral involvement is more common than unilateral involvement by a ratio of 2:1. Affected children are usually born with normal or only mildly impaired hearing which gradually deteriorates through childhood and into adolescence (Sennaroglu and Saatci 2002). Vertigo and tinnitus are also common symptoms. In a small fraction of patients, the loss of hearing may be precipitated by certain events, with head trauma being the most widely reported. In addition, barotrauma, upper-respiratory infections, high fevers, noise trauma, and physical exertion have also been linked to hearing loss in patients with EVA syndrome (Gopen et al. 2011). Because of these associations, children found to have this malformation are usually instructed to avoid contact sports and activities associated with extreme barometric pressure changes (e.g., scuba diving).

The cause of hearing loss in patients with EVA syndrome remains a controversial topic, and many theories on the development of hearing loss have been proposed. These include (i) increased transmission of pressure shifts generated from the intracranial space through the vestibular aqueduct leading to inner ear damage; (ii) electrolyte imbalance due to endolymphatic sac dysfunction causing damaging the inner ear; and (iii) hyperosmolar fluid reflux into the inner ear causing osmotic damage to the neuroepithelium. Some feel that the enlarged vestibular aqueduct is a secondary epiphenomenon rather than an actual cause of hearing loss, with the true pathology only being detectable at the molecular level (Gopen et al. 2011).

On CT, the vestibular aqueduct is considered to be enlarged when it is greater than 1.5 mm in width (roughly the diameter of the simultaneously visualized posterior SCC) at the midpoint between the common crus and its external aperture (Jackler 2010; Swartz 2004) (Fig. 30). MRI additionally demonstrates enlargement of the endolymphatic sac (Fig. 31), and fluid levels may occasionally be evident within the sac. Endolymphatic sac enlargement may also be seen on MR without enlargement of the aqueduct evident by CT.

In their initial report of EVA syndrome, Valvassori and Clemis found that roughly 60 % of patients with enlarged vestibular aqueducts had associated malformations of the inner ear (Valvassori and Clemis 1978); however, with modern high-resolution CT and MRI techniques, the prevalence of other inner ear malformations detected with imaging is considerably higher. In fact, in one study, Lemmerling and colleagues found that some degree of modiolar deficiency, characteristic of IP-II anomalies, was present.
on CT in all ears with an enlarged vestibular aqueduct (Lemmerling et al. 1997). Another study examining MRI findings in EVA syndrome reported that 84 % of affected ears demonstrated additional inner ear anomalies, with cochlear anomalies being present in 76 % (Davidson et al. 1999). The most common abnormality, seen in 94 % of abnormal cochleae, was modiolar deficiency, with gross cochlear dysmorphism being observed occasionally but much less frequently. The most common vestibular anomaly in the study was simple enlargement of the vestibule, which was seen in a third of cases, while gross vestibular and semicircular canal dysplasias were present in 7 %.

Semicircular Canal Anomalies Malformations of the semicircular canals (SCC) frequently occur in association with other inner ear anomalies and can be seen in syndromes such as CHARGE, trisomy 13, and trisomy 18 (Koch et al. 2006). Patients with SCC anomalies present with vestibular dysfunction, while conductive hearing loss may occasionally occur due to congenital stapes fixation. These malformations most likely result from interruption of SCC development from the vestibular anlage, and most SCC malformations fall within a predictable spectrum based on the timing of the causative growth disturbance. As described earlier, the SCCs begin as disk-shaped evaginations arising from the vestibular appendage in the 6th gestational week. Over subsequent weeks, the outpouchings flatten into disk-like structures and the central portion of each disk is then resorbed and replaced by mesenchyme which results in the formation of characteristic semicircular ducts (Kenna and Hirose 2003). Failure of one of these disks to form results in the absence of the involved SCC, while incomplete absorption of the central portion of the disk results in a dysplastic or pocket-shaped SCC. The superior SCC is the first to form, reaching full development by the 19th week of gestation, followed by the posterior and then the lateral SCC which are developed by the 22nd week (Kenna and Hirose 2003; Sadler 2010). Therefore, superior and posterior SCC anomalies are almost always associated with anomalies of the lateral SCC, whereas abnormalities of the lateral SCC can occur in isolation (Jackler et al. 1987) (Fig. 32). The two main exceptions to this rule are in Waardenburg syndrome and Alagille syndrome, both of which may show the absence of the posterior SCC without involvement of other SCCs (see below) (Koch et al. 2006; Higashi et al. 1992), but we have also observed this finding in a child without phenotypic or chromosomal alterations of either of these syndromes.
Subtle abnormalities of the lateral SCC may also be indicated by a small or enlarged lateral SCC bony island (Fig. 33). Normally, the transverse diameter of this bony island measured on axial CT is between 2.6 mm and 4.8 mm, and diameters outside of this range should be considered suspicious in a child with inner ear symptomatology (Purcell et al. 2006). At times, it may be difficult to distinguish a common cavity malformation from cochlear aplasia with a dilated vestibule and malformed SCCs. As mentioned earlier, in a common cavity deformity, the IAC will be directed toward the center of the cavity, while in the latter malformation, the dilated vestibule will be centered posterior to the IAC (Figs. 25 and 26).

Cochlear Nerve Deficiency  Cochlear nerve deficiency (CND) refers to absence or reduction in caliber of the cochlear nerve and is present in 12–18 % of ears affected with SNHL (McClay et al. 2008; Parry et al. 2005). Although it is usually congenital, CND occasionally develops subsequent to birth due to long-standing hearing loss and atrophy of the nerve (Glastonbury et al. 2002). CND can be seen in isolation (with the vestibular nerve divisions present) or in conjunction with aplasia of vestibular nerve (complete absence of the eighth cranial nerve). While aplasia of the cochlear nerve is considered a contraindication for cochlear implants, the presence of a small hypoplastic nerve is not (Gray et al. 1998; Maxwell et al. 1999). It is therefore extremely important, when one has identified this condition in a child
being considered for implantation, to determine whether the nerve is completely absent or simply hypoplastic.

MRI is the most sensitive modality for diagnosing CND. On CISS or similar fluid-sensitive, high-resolution sequences (e.g., thin-section T2-weighted fast spin echo), the normal cochlear nerve is easily identifiable within the IAC. Oblique sagittal reconstructions oriented orthogonal to the long axis of the IAC are particularly useful for visualizing the cochlear nerve, the intracanalicular segment of the facial nerve (CN7), and the superior and inferior divisions of the vestibular nerve (Fig. 34). In this plane, the cochlear nerve is situated in the anterior inferior quadrant of the canal, CN7 is in the anterior superior quadrant, and the vestibular nerves are in the posterior half. Qualitatively, the cochlear nerve can be considered hypoplastic if its diameter is smaller than that of the adjacent facial nerve (Fig. 35). Commonly, the IAC is stenotic (diameter less than 4 mm) or even atretic, and it is theorized that this association occurs because canalization and development of the IAC depends upon the presence of vestibulocochlear nerve cells to form normally (Glastonbury et al. 2002; Walton et al. 2008).

CT is less sensitive than MRI for the detection of CND, as it only demonstrates secondary signs, such as absence or stenosis of the bony cochlear nerve canal (CNC) or IAC stenosis (Fig. 36). The CNC is normally between 1.4 and 3.0 mm in width, so any CNC measuring less than 1.4 mm should be viewed with suspicion in a child with SNHL (Stjernholm and Muren 2002). Children with narrow IACs on CT have been noted to perform worse following implantation than those with normal caliber IACs, presumably because the cochlear nerve is likely to be deficient when the IAC is narrow (Papsin 2005). It should be noted, however, that identifying a normal-sized CNC or IAC on CT does not rule out CNC, as up to 23 % of ears with CND may demonstrate normal width CNCs and 73 % can demonstrate normal-sized IACs (Huang et al. 2010; Adunka et al. 2007).

Syndromes and Inherited Disorders Associated with Ear Malformations
Roughly 60 % of cases of congenital hearing loss can be linked to a genetic cause, with approximately 30 % of these considered syndromic and the remaining 70 % being nonsyndromic (Lalwani and Castelein 1999). The term “syndromic” implies the presence of other distinctive clinical features in addition to hearing loss, and to date, over 300 syndromic forms of hearing loss have been described (Morton and Nance 2006). In many syndromes, hearing loss is an inconstant feature, but there are a number of well-
Fig. 34 Cochlear nerve deficiency. (a) Axial 3D CISS MR image through the temporal bones in a child with profound right-sided SNHL. The cochlear nerve is not seen on the right. Compare to the normal left cochlear nerve (white arrow). (b) Oblique sagittal image reconstruction through the left IAC in this patient demonstrates the normal four-nerve configuration with the facial nerve (arrowhead) located in the anterior superior quadrant, the cochlear nerve (arrow) in the anterior inferior quadrant, and the superior and inferior branches of the vestibular nerves located in the posterior half of the IAC. (c) On the right side, the cochlear nerve is not visible in the anterior inferior quadrant of the IAC (A anterior; P posterior).

Fig. 35 Cochlear nerve hypoplasia. (a) Oblique sagittal reconstructed 3D CISS MR image through the right IAC in a 7-year-old child with right-sided SNHL demonstrates a hypoplastic left cochlear nerve (arrow), which is smaller in diameter than the nearby facial nerve (arrowhead). (b) Oblique image through the contralateral IAC demonstrates a normal left-sized cochlear nerve, which normally has a diameter equal to or larger than that of the facial nerve.
characterized entities in which hearing loss is a frequent and/or major component. Although many of these syndromes and inherited forms of hearing loss do not demonstrate gross temporal bone anomalies by imaging, there are several, discussed below, in which ear malformations are a common and sometimes defining feature of the disease.

Pendred Syndrome Pendred syndrome is the most common form of syndromic hearing loss, accounting for up to 7.5% of cases of hereditary deafness (Fraser 1965). The syndrome is inherited as an autosomal recessive trait and is characterized by the combination of goiter and severe SNHL with vestibular aqueduct enlargement. The specific mutation causing the syndrome in most cases has been mapped to the SLC26A4 (previously called PDS) gene locus on chromosome 7q31 (Ito et al. 2011; Coyle et al. 1996). This gene codes for the protein pendrin, which is believed to function as a plasma membrane-bound chloride-iodine transporter (Scott et al. 1999).

Most patients with Pendred syndrome are euthyroid; however, many demonstrate varying degrees of hypothyroidism (Goldfeld et al. 2005). Goiter in individuals with Pendred syndrome usually manifests in mid-childhood and is due to a specific defect that prevents normal organification of iodine taken up by the gland by inhibiting its binding to thyroglobulin. Administration of perchlorate unmasks defects of iodine organification by provoking discharge of the unbound iodine from the gland. This fact is the basis for the perchlorate discharge test that is used to diagnose Pendred syndrome. In this test, potassium perchlorate is given to the patient orally after the thyroid is primed with radioiodide. In the presence of an organification defect, radioactivity in the thyroid will decrease dramatically after perchlorate administration, with a discharge of 10% or greater being considered abnormal (Reardon and Trembath 1996).

Studies incorporating molecular genetic testing of patients with hearing loss associated with nonsyndromic LVA anomalies – sometimes referred to as DFNB4 or nonsyndromic enlarged vestibular aqueduct (NSEVA) – have revealed that many of these cases are also associated with SLC26A4 mutations (Ito et al. 2011). In fact, SLC26A4 mutations are found in approximately 5–10% of children with NSEVA (Park et al. 2003) and in roughly 50% of patients with PDS or DFNB4 (Alasti et al. 1993). While the trait is thought to be inherited in an autosomal recessive fashion, Pryor et al. demonstrated a potential correlation between the clinical phenotype and the number of mutated alleles of SLC26A4 in patients with EVA (Pryor et al. 2005). In that study, Pendred syndrome was associated with the presence of two mutant alleles of SLC26A4, while NSEVA correlated with one or no mutant alleles. Furthermore,
unilateral EVA correlated with 0–1 mutant alleles, while bilateral EVA was tightly correlated with two mutant alleles.

Inner ear malformations are almost invariably present on CT among patients with Pendred syndrome and are characterized by IP-II type malformations – i.e., modiolar deficiency and vestibular enlargement, absence of the interscalar septum between the upper and middle cochlear turns, and enlargement of the vestibular aqueduct (Goldfeld et al. 2005) (Fig. 31). Enlargement of the vestibular aqueduct is now considered the most penetrant feature of the Pendred syndrome, and endolymphatic sac enlargement has been reported to be present on MRI in 100 % of patients with the syndrome (Phelps et al. 1998).

**Branchio-Oto-Renal Syndrome**

Branchio-oto-renal (BOR) syndrome is an autosomal dominant syndrome characterized by hearing loss, auricular malformations, branchial arch closure defects (preauricular pits and tags), and renal anomalies (Kochhar et al. 2007). Hearing impairment occurs in 70–93 % of individuals with BOR syndrome, and the syndrome has been reported to account for 2 % of children with profound deafness. The age of onset of hearing loss varies from early childhood to young adulthood, and hearing loss may be conductive, sensorineural, or mixed and may range from mild to profound.

In addition to hearing loss, common characteristics of the BOR phenotype include cup-shaped pinnae, preauricular pits, branchial cleft fistulae, and bilateral anomalies of the renal collecting system. Additional clinical features which have been reported in patients in the syndrome include lacrimal duct stenosis, a narrow face, palatal abnormalities, renal agenesis, anomalies of the bladder and ureters, and shoulder abnormalities (Kochhar et al. 2007).

BOR syndrome has an estimated incidence of 1:40,000. Mutations of two genes, EYA1 and SIX1, are known to cause the BOR phenotype. EYA1 mutations account for approximately 40 % of cases, while relatively little is known currently about SIX1 mutations and their prevalence (Kochhar et al. 2007). Based on genotype-phenotype analyses using coding sequence analysis of EYA1, Chang et al. proposed updated criteria in 2004 for the clinical diagnosis of BOR syndrome, with 4 major criteria (branchial anomalies, deafness, preauricular pits, and renal anomalies) and several additional minor criteria (Table 3). Using these guidelines, clinical diagnosis of BOR syndrome meritig genetic testing for EYA1 mutation requires the presence of 3 major criteria, 2 major and at least 2 minor criteria, or 1 major criterion and an affected first-degree relative meeting criteria for BOR (Chang et al. 2004).

In addition to demonstrating malformations of the auricle, patients with BOR syndrome also frequently demonstrate temporal bone abnormalities, which may involve any part of the ear. Inner ear anomalies are perhaps the most frequently seen imaging findings in BOR syndrome, with the most characteristic being

### Table 3  Updated diagnostic criteria for BOR syndrome (Chang et al. 2004)

<table>
<thead>
<tr>
<th>Major criteria</th>
<th>Minor criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Branchial anomalies</td>
<td>External ear anomalies</td>
</tr>
<tr>
<td>Deafness</td>
<td>Middle ear anomalies</td>
</tr>
<tr>
<td>Preauricular pits</td>
<td>Inner ear anomalies</td>
</tr>
<tr>
<td>Renal anomalies</td>
<td>Preauricular tags</td>
</tr>
<tr>
<td>Other: facial asymmetry, palate abnormalities</td>
<td></td>
</tr>
</tbody>
</table>

An affected individual must have at least three major criteria, two major criteria and at least two minor criteria, or one major criteria and an affected first-degree relative meeting criteria for BOR.
cochlear hypoplasia, particularly involving the apical turn (100%); deviation of the labyrinthine facial nerve canal medial to the cochlea (90%); and a funnel-shaped IAC with a large porus acusticus (86%) (Propst et al. 2005) (Fig. 37). Vestibular dysplasias, hypoplasia of the SCCs, enlargement of the vestibular aqueduct, and cochlear nerve deficiency can also be observed (Chen et al. 1995; Ceruti et al. 2002).

The most common and easily identifiable finding in the middle ear on temporal bone CT is a patulous Eustachian tube, which is seen in roughly 60% of BOR syndrome ears (Propst et al. 2005) (Fig. 38). Ossicular chain abnormalities are also common, and ossicles may be dysplastic, fused, fixed laterally, or malpositioned. Additional reported findings in the middle ear include a malformed and usually small middle ear cavity, a hypoplastic pyramidal eminence with the absence of the stapedius tendon, a tympanic facial nerve which overhangs the oval window, and the absence of the oval window (Chen et al. 1995; Propst et al. 2005; Ceruti et al. 2002). EAC stenosis and atresia can also been seen.
The acronym "CHARGE" was originally coined by Pagon et al. (Pagon et al. 1981) in 1981 and stands for Coloboma, Heart defects, Atresia of the choanae, Retarded growth and development, Genital hypoplasia, and Ear anomalies and/or deafness—reflecting what were believed to be the six cardinal clinical features of the syndrome. Since the initial description, the definition of CHARGE has evolved to accommodate several additional anomalies, including rhombencephalic dysfunction, cranial neuropathies, and dysfunction of the hypothalamic-hypophyseal axis. In 2005, Verloes (Verloes 2005) proposed an updated set of criteria, emphasizing the triad of coloboma, choanal atresia, and abnormal SCCs, and additionally suggested categories for partial and atypical forms of the syndrome (Table 4).

CHARGE syndrome is usually a sporadic autosomal dominant disorder, which in approximately two thirds of patients is associated with mutations of the CHD7 gene located on chromosome 8. Chorioretinal colobomas are present in 75–90% of cases, with bilateral involvement seen in 70–80% (Sanlaville and Verloes 2007) (Fig. 39). Choanal atresia, another of the major features of CHARGE, is observed in 35–65% of cases. Roughly 75% of patients with CHARGE have clinical signs or symptoms suggesting an abnormality of at least one cranial nerve. Absence or hypoplasia of the olfactory bulbs, resulting in anosmia or hyposmia, is possibly the most common cranial nerve anomaly, with one study observing this finding on MRI in 100% of patients with CHARGE (Pinto et al. 2005). Other frequently involved cranial nerves include CN8 (60%), CN7 (43%), and CN 9 and 10 (31%), although any cranial nerve can be involved (Byerly and Pauli 1993).

Hearing loss is observed in up to 90% of children with CHARGE and can be conductive, sensorineural, or mixed (Sanlaville and Verloes 2007). Inner ear anomalies are present in greater than 90% of patients. The characteristic inner ear anomalies in CHARGE are SCC aplasia and associated vestibular dysplasia, which are present in over 80% of patients with the syndrome (Holcomb et al. 2013; Morimoto et al. 2006) (Fig. 39). Narrowing or atresia of the cochlear aperture is common being present in over 50% of cases, and in ears exhibiting SNHL, cochlear nerve deficiency is evident in MRI in roughly 80% (Holcomb et al. 2013; Morimoto et al. 2006). Abnormalities of cochlear partitioning are very common, and vestibular aqueduct enlargement may also occasionally be seen (Lemmerling et al. 1997; Morimoto et al. 2006).

Outer ear anomalies are seen in virtually all patients with CHARGE, with the characteristic malformation being a misshaped pinna, which is small, low set, anteverted, and cup shaped. Middle ear anomalies are also frequently demonstrated on CT and include a small middle ear cavity, absence of the stapedius muscle, absence of the round and oval windows, dysplastic ossicles, ossicular fixation, and an

### Table 4  Updated diagnostic criteria for CHARGE syndrome (Verloes 2005)

<table>
<thead>
<tr>
<th>Major signs (the 3 Cs)</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Coloboma</td>
<td></td>
</tr>
<tr>
<td>Choanal atresia</td>
<td></td>
</tr>
<tr>
<td>Hypoplastic semicircular Canals</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Minor signs</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Rhombencephalic dysfunction (brainstem and cranial nerve, including SNHL)</td>
<td></td>
</tr>
<tr>
<td>Hypothalamo-hypophyseal dysfunction</td>
<td></td>
</tr>
<tr>
<td>Abnormal middle or external ear</td>
<td></td>
</tr>
<tr>
<td>Malformation of mediastinal organs (heart, esophagus)</td>
<td></td>
</tr>
<tr>
<td>Mental retardation</td>
<td></td>
</tr>
</tbody>
</table>

Typical CHARGE = 3 major signs OR 2 major signs + 2 minor signs
Partial/Incomplete CHARGE = 2 major signs + 1 minor sign
Atypical CHARGE = 2 major signs + 0 minor signs OR 1 major sign + 3 minor signs

**Charge** The acronym “CHARGE” was originally coined by Pagon et al. (Pagon et al. 1981) in 1981 and stands for Coloboma, Heart defects, Atresia of the choanae, Retarded growth and development, Genital hypoplasia, and Ear anomalies and/or deafness—reflecting what were believed to be the six cardinal clinical features of the syndrome. Since the initial description, the definition of CHARGE has evolved to accommodate several additional anomalies, including rhombencephalic dysfunction, cranial neuropathies, and dysfunction of the hypothalamic-hypophyseal axis. In 2005, Verloes (Verloes 2005) proposed an updated set of criteria, emphasizing the triad of coloboma, choanal atresia, and abnormal SCCs, and additionally suggested categories for partial and atypical forms of the syndrome (Table 4).

CHARGE syndrome is usually a sporadic autosomal dominant disorder, which in approximately two thirds of patients is associated with mutations of the CHD7 gene located on chromosome 8. Chorioretinal colobomas are present in 75–90 % of cases, with bilateral involvement seen in 70–80 % (Sanlaville and Verloes 2007) (Fig. 39). Choanal atresia, another of the major features of CHARGE, is observed in 35–65 % of cases. Roughly 75 % of patients with CHARGE have clinical signs or symptoms suggesting an abnormality of at least one cranial nerve. Absence or hypoplasia of the olfactory bulbs, resulting in anosmia or hyposmia, is possibly the most common cranial nerve anomaly, with one study observing this finding on MRI in 100 % of patients with CHARGE (Pinto et al. 2005). Other frequently involved cranial nerves include CN8 (60 %), CN7 (43 %), and CN 9 and 10 (31 %), although any cranial nerve can be involved (Byerly and Pauli 1993).

Hearing loss is observed in up to 90 % of children with CHARGE and can be conductive, sensorineural, or mixed (Sanlaville and Verloes 2007). Inner ear anomalies are present in greater than 90 % of patients. The characteristic inner ear anomalies in CHARGE are SCC aplasia and associated vestibular dysplasia, which are present in over 80 % of patients with the syndrome (Holcomb et al. 2013; Morimoto et al. 2006) (Fig. 39). Narrowing or atresia of the cochlear aperture is common being present in over 50 % of cases, and in ears exhibiting SNHL, cochlear nerve deficiency is evident in MRI in roughly 80 % (Holcomb et al. 2013; Morimoto et al. 2006). Abnormalities of cochlear partitioning are very common, and vestibular aqueduct enlargement may also occasionally be seen (Lemmerling et al. 1997; Morimoto et al. 2006).

Outer ear anomalies are seen in virtually all patients with CHARGE, with the characteristic malformation being a misshaped pinna, which is small, low set, anteverted, and cup shaped. Middle ear anomalies are also frequently demonstrated on CT and include a small middle ear cavity, absence of the stapedius muscle, absence of the round and oval windows, dysplastic ossicles, ossicular fixation, and an
abnormal course of the tympanic facial nerve, which may course over the oval window niche (Morimoto et al. 2006; Lemmerling et al. 1998; Morgan et al. 1993).

Venous abnormalities of the temporal bone are also a common feature of CHARGE syndrome. The most common venous anomalies described are large emissary veins, which may be associated with hypoplasia of the ipsilateral sigmoid sinus or jugular bulb. These veins may traverse the mastoid portion of the temporal bone and may complicate or alter middle ear and mastoid surgery (Friedmann et al. 2012b). Therefore, it is important to look for and identify these venous anomalies in CHARGE patients for whom cochlear implantation or other otologic surgeries are being contemplated.

Oculoauriculovertebral Syndrome Oculoauriculovertebral syndrome (OAVS) covers a heterogeneous and complex spectrum of disorders which include conditions such as hemifacial microsomia and Goldenhar syndrome (Passos-Bueno et al. 2009). The syndrome is characterized primarily by ocular and aural anomalies, and most patients have hemifacial microsomia. Eye anomalies include epibulbar dermoids and upper eyelid colobomas, and these are often associated with macrostomia or facial clefts. Spinal anomalies include scoliosis, vertebral body fusion, vertebral segmentation anomalies (hemivertebrae, butterfly vertebrae, and supplementary vertebrae), and fusion anomalies of the skull base and atlas (Anderson and David 2005). In addition to craniofacial and vertebral anomalies, cardiac, renal, and central nervous system defects are also observed in patients with OAVS (Passos-Bueno et al. 2009).
et al. 2009; Johnson et al. 2011). There is no agreement on the minimal diagnostic criteria, but isolated microtia or hemifacial microsomia with mild ear anomalies (preauricular tags) probably represent the mildest manifestations of the disease.

The prevalence of OAVS is estimated to be roughly 1/20,000. Males with hemifacial microsomia are affected nearly twice as frequently as females (Senggen et al. 2011). The etiology of the phenotypic changes of OAVS remains unknown, but it is most likely that both genetic and nongenetic factors play a role in the pathogenesis of the disease. Most cases are sporadic, but familial cases with autosomal dominance inheritance are seen in 2–10% of cases, with wide clinical variability observed within families. Most chromosomal alterations identified appear to be patient specific, but involvement of chromosomes 5, 18, 22, and X has been reported in multiple instances. OAVS has also been described in infants born to diabetic mothers and women living in high altitude regions and with in utero exposure to thalidomide (Passos-Bueno et al. 2009).

On CT, patients with OAVS frequently show asymmetric hypoplasia of the maxilla and mandible (Fig. 40). The contralateral side of the face may be normally developed, but bilateral anomalies are seen in up to 30% of patients (Johnson et al. 2011; Senggen et al. 2011). Variable degrees of TMJ malformation can occur, and the degree of TMJ dysplasia does not appear to correlate with the degree of mandibular dysplasia (Johnson et al. 2011).

Ear deformities range from isolated preauricular tags to atresia of the EAC (Fig. 3), with associated anomalies of the middle ear and facial nerve (see above). In addition, inner ear anomalies have been reported to be present in 36% of patients with OAVS (Bisdas et al. 2005). The most commonly reported inner ear findings are dysplasias of the SCCs and vestibules, hypoplasia of the cochlea, and abnormalities of the IACs (stenosis or enlargement).

Treacher Collins Syndrome Treacher Collins syndrome (TCS), also referred to as mandibulofacial dysostosis, is an autosomal dominant craniofacial disorder of the first and second branchial arches with clinical features that include down-slanting palpebral fissures with lower eyelid coloboma; malar and maxillary hypoplasia; microtia and other ear malformations, with resultant conductive hearing loss; and, in severe cases, cleft palate (Passos-Bueno et al. 2009). Hypoplasia of the facial bones, particularly the
mandibular ramus and condyle and zygomatic complex, occurs in the greater 75 % of patients. The maxilla is also usually hypoplastic, but may occasionally appear overprojecting.

The birth prevalence of the disorder is estimated at 1:50,000, and the majority of cases of TCS occur in a sporadic fashion, with only 40 % seen in patients with a family history of the disease (Dixon 1995). Mutations in the TCOF1 gene, located on chromosome 5, are implicated in most cases of TCS, with approximately 120 pathogenic mutations of the gene currently described. TCOF1 encodes for a nucleolar phosphoprotein, known as treacle, which is involved in ribosomal DNA gene transcription and in processing of pre-ribosomal RNA. Deficiency of the treacle protein leads to insufficient ribosome biogenesis, which is associated with decreased cell proliferation, increased neuroepithelial apoptosis, and a reduced number of cranial neural crest cells migrating into the core of the first and second branchial arches (Passos-Bueno et al. 2009).

On imaging, craniofacial defects in TCS are usually bilateral and symmetric. They include hypoplastic or aplastic zygomatic arches, choanal shortening, micrognathia, and maxillary narrowing or overprojection (Johnson et al. 2011). Temporal bone anomalies in TCS include microtia, CAA, and associated middle ear dysplasias, including hypoplasia of the tympanic cavity, ossicular malformations and fixation, and abnormalities in the course of the facial nerve (Takegoshi et al. 2000; van Vierzen et al. 1995) (Fig. 41). Abnormalities of the lateral SCC and vestibule have also been reported to occur rarely (Takegoshi et al. 2000).

Surgical techniques are available to address most of the anomalies of TCS. Surgical correction of the zygoma, orbits, and mandible are usually delayed until patients are 4–10 years of age. Auricular repair is often delayed until after 6 years of age to allow time for adequate development of the costal cartilage, which is used for reconstruction (Johnson et al. 2011).

**Waardenburg Syndrome** Waardenburg syndrome (WS) is an autosomal dominant syndrome characterized by the constellation of hypertelorism with a prominent broad nasal root (dystopia canthorum); eyebrow hyperplasia and synophris; pigmented disturbances including heterochromia iridis, white forelock, leukoderma, and white eyelashes; and SNHL. The abnormalities seen in WS are believed to result from abnormal development of neural crest-derived melanocytes, which are normally found in the skin, inner ear, glia, peripheral and enteric neurons, and craniofacial skeletal tissue. SNHL in WS is thought to be caused by the absence of melanocyte-derived intermediate cells of the stria vascularis, which induces endolymphatic collapse and secondary agenesis of the organ of Corti (Elmaleh-Berges
et al. 2013). Several types (I–IV) and additional subtypes have been identified since the syndrome was first described in 1951 (Waardenburg 1951), with type I (WS1) and type II (WS2) occurring most frequently and types III (WS3 or Klein-Waardenburg syndrome) and IV (WS4 or Shah-Waardenburg syndrome) being extremely rare. To date, six different genes (PAX3, MITF, EDN3, EDNRB, SOX10, and SNAI2) have been implicated as being responsible for the varying phenotypes of the syndrome (Cullen et al. 2006; Pingault et al. 2010).

WS1 is the classic phenotype first described by Waardenburg. It is an autosomal dominant disorder characterized by dystopia canthorum, pigmentary anomalies, and hearing loss. Most cases of WS1 are attributable to mutations of the PAX3 gene located on the distal long arm of chromosome 2 (Pingault et al. 2010). WS3 shares the features of WS1, with the addition of flexion contractures and muscle hypoplasia of the upper limbs, and is also linked to PAX3 mutations.

WS2 differs from WS1 in that dystopia canthorum is not present. In reality, WS2 likely covers a heterogeneous collection of auditory-pigmentary syndromes that do not fit in other categories (Read and Newton 1997). In WS2, MITF and SOX10 mutations are each involved in roughly 15 % of cases, while mutations of SNAI2, EDNRB, and possibly EDN3 account for small percentages (Pingault et al. 2010).

WS4 is characterized by an association with Hirschsprung’s disease. About 50 % are due to SOX10 mutations, with another 20–30 % caused by EDN3 or EDNRB mutations. Patients with SOX10 mutations also frequently exhibit severe neurologic abnormalities resulting from impaired myelination of the central and peripheral nervous systems in addition to the characteristic findings of WS, a condition referred to as peripheral demyelinating neuropathy, central dysmyelinating leukodystrophy, Waardenburg syndrome, and Hirschsprung disease (PCWH) (Elmaleh-Berges et al. 2013).

Hearing loss is the most common feature of WS, occurring in approximately 60 % of children with WS1 and 90 % of children with WS2. Temporal bone anomalies on imaging are reported in up to 50 % of WS patients but are uncommon in those with WS1, suggesting that most detectable abnormalities occur in patients with other subtypes (Madden et al. 2003; Cullen et al. 2006; Oysu et al. 2001). When present, inner ear anomalies are usually bilateral and symmetric. The most common findings in WS are SCC anomalies (Fig. 42). In particular, aplasia or hypoplasia of the posterior SCC, which is seen in roughly 26 % or patients with WS, is characteristic of the disease (Fig. 43). A recent retrospective analysis of 15 patients with WS and inner ear anomalies associated with SOX10 mutations (comprised of patients

![Fig. 42 Waardenburg syndrome. Axial CT image in a 21-month-old child with Waardenburg syndrome demonstrates a dysplastic lateral and posterior semicircular canals](image-url)
with WS2, WS4, and PCWH) demonstrated vestibular enlargement and semicircular canal abnormalities in all of the patients with 67% demonstrating posterior SCC agenesis, 53% demonstrating superior SCC agenesis, and 33% lateral SCC agenesis (Elmaleh-Berges et al. 2013).

Other inner ear anomalies observed in WS include vestibular aqueduct enlargement, widening of the upper vestibule, IAC hypoplasia, and decreased modiolus size (Madden et al. 2003; Higashi et al. 1992). In their series of patients with WS associated with SOX10 mutations, Elmaleh-Berges and colleagues noted cochlear nerve agenesis in 21% and cochlear abnormalities—generally consisting of a small cochlea or flattening of the midturn and apex of the cochlea with preservation of normal cochlear partitioning—in 93% (Elmaleh-Berges et al. 2013). White matter signal abnormalities and agenesis of the olfactory bulbs, lacrimal glands, and parotid glands were also common findings in the series.

Alagille Syndrome The absence of the posterior semicircular canal with a normal lateral semicircular canal has also been reported in patients with Alagille syndrome (arteriohepatic dysplasia), a genetic syndrome caused by mutations or deletion of the JAG1 gene located on chromosome 20p12. The syndrome is characterized by chronic cholestasis, posterior embryotoxon (congenital opacity at the margin of the cornea), butterfly vertebral anomalies, cardiovascular malformations, and characteristic facies (Koch et al. 2006). Additional temporal bone histopathologic findings reported in Alagille syndrome include mild cochlear hypoplasia, absence of the cochlear aqueduct, and partial defects of the superior SCC (Okuno et al. 1990).

X-Linked Deafness with Stapes Gusher X-linked deafness with stapes gusher (also known as X-linked deafness type 3, DFN3, X-linked stapes gusher syndrome, or Nance deafness) is a sex-linked recessive disorder caused by a loss-of-function mutation of the POU3F4 gene at the Xq21 region (the DFN3 locus). DFN3 is one of the four X-linked loci implicated in congenital deafness (the others being DF2, DF4, and DF6). Mutations at this locus are identified in 50% of families with X-linked hearing loss (Petersen et al. 2008). Deficiency of POU3F4 has been shown to cause defects in both otic fibrocytes and the stria vascularis in the cochlear lateral wall in mouse models (Song et al. 2011).

Symptomatic patients with this mutation are typically male and present with hearing loss at birth which rapidly progresses to severe deafness within the first decade of life. The hearing loss is typically mixed, but some individuals demonstrate SNHL without a conductive component. Vestibular problems are also common. Female carriers of the gene mutation may have normal hearing or only mild-to-moderate...
hearing loss (Song et al. 2010). In patients with the DFN3 mutation, there is a communication between the subarachnoid and perilymphatic spaces due to deficiency of the lamina cribrosa separating the IAC from the basal turn of the cochlea. This communication causes elevated perilymphatic pressure and fixation of the stapes footplate which is the cause of conductive hearing loss (Papadaki et al. 1998). In this setting, manipulation of the stapes footplate during surgery can result in a gush of perilymph through the oval window (“stapes gusher”), and therefore, stapedectomy is generally contraindicated in these patients.

Characteristic imaging findings on both CT and MRI in children with X-linked deafness with stapes gusher include enlarged, bulbous IACs; a widened cochlear aperture with deficiency of the lamina cribrosa, resulting in a wide communication between the basal turn of the cochlea and the IAC; cochlear hypoplasia with modiolar deficiency; and widening of the bony canal for the labyrinthine segment of the facial nerve (Kumar et al. 2003) (Fig. 44). Vestibular aqueduct dilatation has also been reported (Talbot and Wilson 1994).

**Imaging in Cochlear Implantation**

Cochlear implantation is a therapeutic option for children and adults with severe-to-profound SNHL, which has been approved for use in the United States since the early 1980s (Miyamoto and Kirk 2003). As the majority of cases of SNHL result from degeneration of the hair cells in the organ of Corti, cochlear
implants essentially bypass nonfunctioning hair cells by converting mechanical sound into electrical signals that directly stimulate spiral ganglion cells.

Modern cochlear implants are multichannel, multielectrode systems that are designed to take advantage of the tonotopic organization of the cochlea, by providing complex sound analysis and stimulating different locations in the cochlea. The devices consist of both internal and external components. The internal component consists of a surgically implanted receiver/stimulator – usually placed in a subperiosteal pocket behind the ear – which is attached by a wire to an electrode array placed within the cochlea (Fig. 45). The electrode array is ideally inserted into the scala tympani through the round window or through an anteroinferior cochleostomy, with exposure to the round window niche typically achieved through a canal wall-up mastoidectomy with a facial recess approach. The external components of the device include a microphone and speech processor worn over the ear and an attached transmitter – usually held in position by a magnet – which transmits the processed sound signals across the skin to the internal receiver/stimulator coil.

Current FDA guidelines allow cochlear implantation of children as young as 1 year of age. Implantation is indicated in children aged 12–24 months with profound bilateral hearing loss (greater than 95 dB) who have shown a lack of auditory skill development and minimal hearing aid benefit. For children with severe-to-profound hearing loss (71–95 dB), cochlear implantation is indicated after the age of 24 months, if they show a lack of auditory skills development and little benefit from conventional amplification. Although cochlear implants are only approved in the United States for children over the age of 12 months, a number of infants younger than this are implanted every year. If children are identified with profound hearing loss at an early age and have completed an adequate trial of hearing aid use before the age of 1 year without demonstrating an appreciable benefit, clinicians may see little need to delay cochlear implantation (Miyamoto and Kirk 2003).

In our practice, most pediatric cochlear implant candidates undergo imaging evaluation with both CT and MRI. Temporal bone CT is particularly useful for surgical planning as it nicely demonstrates the degree of pneumatization of the mastoid air cells, anomalies of the middle ear space, cochlear ossification,
and the presence of vascular anomalies, such as aberrant blood vessels or a dehiscent jugular bulb which may affect the surgical approach. Special attention should be paid to the patency of the round window, course of the facial nerve, size of the facial recess, patency of the cochlea and cochlear aperture, and presence of the internal auditory canal.

Congenital malformations of the cochlea are also well demonstrated on CT and are generally not – with the exception of Michel aplasia – contraindications for CI. Identification of inner ear anomalies is important in surgical planning, however, as they may make implant electrode placement more challenging and also increase the risk for complications such as perilymph or CSF leak, postimplantation meningitis, and potential electrode misplacement, including placement into the IAC (Papsin 2005; Buchman et al. 2004). In general, children with relatively mild inner ear malformations (e.g., vestibular aqueduct enlargement, IP-2, partial SCC dysplasias) tend to perform better on tests of speech perception following cochlear implantation than children with more severe malformations (e.g., common cavity malformation and cochlear hypoplasia) or syndromes such as CHARGE syndrome, but it is important to keep in mind that even the presence of a severe malformation does not preclude implantation provided that both the cochlea and cochlear nerve are present.

Furthermore, facial nerve course is aberrant in approximately 15–32% of patients with cochleovestibular anomalies, a finding which influences surgical planning and occasionally precludes placement of the cochleostomy in the optimal position in relation to the round window (Papsin 2005; Buchman et al. 2004). Two specific anomalies of the facial nerve that place it at risk are lateral and anterior displacement of the vertical portion of the facial nerve and a facial nerve that courses over the promontory over or anterior to the round window (Miyamoto and Kirk 2003).

Fluid in the middle ear cavity should always be noted as this may indicate otitis media and delay surgery until appropriately treated. Active middle ear infection must be eradicated prior to surgery in order to prevent infection of the implanted device. Intracochlear bony overgrowth results in obliteration of the membranous labyrinth and may be detected by CT. Although labyrinthitis ossificans is not a specific contraindication for CI, advanced cases may limit the insertion of the electrode or require drilling of the basal turn or in select cases use of a split electrode device (Miyamoto and Kirk 2003). Furthermore, ossification can occur after electrode placement.

The primary utility of MRI in pediatric cochlear implant candidates is to establish the presence of the cochlear nerve, as complete cochlear nerve deficiency is a contraindication to implantation (Gray et al. 1998; Maxwell et al. 1999). In addition, soft tissue obliteration of the inner ear structures without ossification (labyrinthine fibrosis) is not detected by CT. In these cases, thin-section fast spin-echo T2-weighted images or CISS images will demonstrate loss of the normally hyperintense intralabyrinthine signal, which indicates replacement of the normal perilymph and endolymph. MRI is also superior to CT for detecting associated brain anomalies, such as can be seen in children with hearing loss due to congenital CMV infection.

For postoperative evaluation or surveillance of implanted ears, plain radiographs and CT generally suffice. Our surgeons routinely obtain portable transorbital radiographs in the operating suite immediately after implantation, which generally suffice to document the position and configuration of the electrode array (Fig. 46). Alternatively, radiographs obtained using the Chausse III projection, a variant of the Stenvers view obtained with the head rotated 30° away from the side of interest and the X-ray source angled 15° cephalad, produce an orientation roughly along the axis of the cochlea and provide a clear depiction of deeply inserted intracochlear electrode arrays with less electrode overlap with the cochleostomy position projected beneath the vestibule (Czerny et al. 1997). CT is also excellent for demonstrating electrode course (Fig. 46). Complications of cochlear implantation include misplacement of the electrode, which may become kinked in the cochlea (Fig. 47) or penetrate into the IAC (Fig. 48), electrode extrusion or migration, perilymph or CSF leak, meningitis, facial nerve stimulation,
Fig. 46 Normal cochlear implants. (a) Intraoperative transorbital radiograph obtained immediately following right-sided cochlear implantation in an 8-year-old child. The patient had undergone left-sided cochlear implantation in the past. Note the normal spiral curvature of the intracochlear electrode arrays (arrows). (b) Modified right anterior oblique (Stenver) reconstructed CT image from the same patient demonstrates a normal intracochlear course of the electrode.

Fig. 47 Malpositioned cochlear implant. (a) Transorbital radiograph in a child with a failed left cochlear implant demonstrates an abnormal configuration of the electrode array. Instead of making a normal smooth clockwise rotation (compare with the previous figure), the electrode takes an abrupt upward turn at the cochleostomy site (arrowhead). (b) 3D reconstructed CT image shown in a left anterior projection again demonstrates the abnormal configuration of the electrode array (shown in blue) which courses upward upon entering the cochlea (arrowhead) and winds opposite of its normal direction in the mid cochlear turn before making a hairpin turn (arrow) and winding back in the normal direction.
cholesteatoma formation, flap necrosis and infection, and otitis media (Ikeya et al. 2013; Tarkan et al. 2013). Labyrinthitis ossificans can also develop after implantation.

MRI can be safely performed in patients with select cochlear implants under certain conditions, but we generally reserve MRI for cases in which a strong medical indication is present and no equivalent alternative imaging options exist (Teissl et al. 1998). Many modern implants feature a removable internal magnet, which must be surgically removed prior to MRI and which can be replaced after MRI has been completed. Failure to remove the internal magnet prior to MRI can result in magnet displacement and local skin reactions (Broomfield et al. 2013). As of this writing, only a handful of cochlear implants (Med-El Pulsar, Med-El Sonata, and Med-El Concert) have been approved in the United States by the Food and Drug Administration recently for MRI at field strengths up to 1.5 T without magnet removal.

**Primarily Acquired Diseases of the Temporal Bone in Children**

**Otitis Media and Other Infections**

**Acute Otitis Media**

Otitis media is primarily a disease of childhood and is the most common illness leading to pediatric doctor’s visits. The term itself simply refers to inflammation of the middle ear without reference to cause or pathogenesis. As such, in the acute setting, otitis media is generally subdivided into either acute (or suppurative) otitis media or otitis media with effusion (also referred to as serous or nonsuppurative otitis media), each of which has distinct diagnostic criteria and therapeutic implications. Acute otitis media (AOM) is diagnosed when there is rapid onset of the signs and symptoms of a middle ear infection, which include otalgia, otorrhea, fever, irritability, anorexia, vomiting, or diarrhea. Otoscopic evaluation in cases of AOM reveals a full or bulging, opaque, and immobile tympanic membrane. Otitis media with effusion (OME), on the other hand, is relatively asymptomatic, and otoscopy frequently reveals a retracted or convex tympanic membrane with decreased mobility. In OME, the membrane is often translucent, and air-fluid levels and/or bubbles may be seen behind it (Bluestone and Klein 2003a).
The pathophysiology of both AOM and OME is multifactorial and includes anatomic and physiologic factors, infectious considerations, and environmental and host factors, but eustachian tube dysfunction generally plays an important role in their development in children. The eustachian tube has three main physiologic functions: (i) pressure regulation and ventilation, (ii) protection from reflux of secretion from the nasopharynx, and (iii) drainage of secretions from the middle ear. In infants and children, the eustachian tube is shorter, wider, and more horizontally oriented than it is in adults, all features that likely contribute to the relatively high rate of otitis media in early childhood. In fact, by the age of 7 years, when the eustachian tube has assumed a more adult configuration, the prevalence of otitis media is actually quite low (Casselbrant and Mandel 2010).

Streptococcus pneumonia, Haemophilus influenzae, and Moraxella catarrhalis are the most common bacterial pathogens cultured in AOM, together accounting for slightly over 70 % of cases (Bluestone et al. 1992). In addition, there is strong evidence that viruses play an important role in the development of AOM, which is usually preceded by a viral infection of the upper-respiratory tract mucosa that, in turn, triggers a cascade of events ultimately leading to AOM as a complication (Heikkinen and Chonmaitree 2003). Furthermore, a number of viruses, including respiratory syncytial virus, influenza virus, adenovirus, parainfluenza virus, and rhinovirus, have been identified in middle ear effusions using polymerase chain reaction techniques. While many cases of AOM can be treated conservatively with observation, severe cases (defined as cases with moderate to severe otalgia, high fevers, or a toxic appearing child) and cases in children under 6 months should be treated with antibiotics (Casselbrant and Mandel 2010).

Management of OME is a more controversial subject. While most episodes of OME resolve within 3 months without active therapy, persistent or recurrent OME is not uncommon, with roughly 5–10 % of episodes lasting for a year or longer (American Academy of Family Physicians et al. 2004). Even in those patients with persistent OME, watchful waiting may be appropriate as long as the patients are otherwise asymptomatic and not at risk for long-term damage to hearing or development. Surgical intervention may be indicated in certain situations, however, including (i) OME which persists for 4 months or longer with persistent hearing loss or other signs and symptoms, (ii) persistent or recurrent OME in children with risk factors for developmental difficulties (e.g., autism spectrum disorders or syndromes including cognitive, speech, or language delays), and (iii) structural damage to the tympanic membrane or middle ear (American Academy of Family Physicians et al. 2004).

In general, imaging is not indicated for uncomplicated cases of AOM or OME and should be reserved for severe cases in which complications are suspected. The complications of AOM can be intra- or extratemporal. Intratemporal complications include mastoiditis, petrous apicitis and osteomyelitis, subperiosteal abscess, facial nerve involvement causing facial paralysis, external otitis, and labyrinthitis. Extratemporal complications include meningitis, intracranial abscess/empyema, venous sinus thrombosis, and otitic hydrocephalus. These are discussed in more detail below.

**Mastoiditis**

Mastoiditis refers to inflammation of the mastoid air cells and can be acute, subacute, or chronic. Acute mastoiditis is the most common suppurative complication of AOM and, in the preantibiotic era, had an incidence of 5–10 % in patients with AOM (Bluestone 2000). Since the widespread adoption of antimicrobial therapy, the incidence of acute mastoiditis has dramatically declined. Among the most commonly cultured organisms from the ears of children with acute mastoiditis are S. pneumonia, S. pyogenes, and anaerobes (Bluestone 2000; Quesnel et al. 2010).

Acute mastoiditis generally progresses in the following stages: (i) acute mastoiditis without peristitis/osteitis, (ii) acute mastoiditis with periostitis, and finally (iii) acute mastoid osteitis with or without subperiosteal abscess (Bluestone 2000). The first stage – mastoiditis without periostitis or osteitis – is not a true complication but rather a natural extension of otitis media. Since the middle ear and mastoid air
system communicate freely through the aditus ad antrum, inflammation can easily spread from one compartment to the other. In fact, if CT were obtained on all patients with an episode of AOM, most, if not all, would have evidence of mastoid involvement, manifesting as mastoid air cell opacification without bone erosion. MRI is not generally performed for uncomplicated cases of acute mastoiditis without periostitis or osteitis, and this stage is frequently an incidental finding on MRI performed for other reasons, typically appearing as a middle ear and mastoid effusion with occasional thickening and enhancement of the mastoid mucosa (Fig. 49). Most cases of acute mastoiditis without periostitis resolve in concert with resolution of middle ear disease.

Acute mastoiditis with periostitis develops when infection in the mastoid spreads to the periosteum covering the mastoid process, usually through venous channels such as the mastoid emissary vein, and is characterized clinically by postauricular erythema, swelling, and tenderness. CT is the imaging method of choice in this stage and is usually performed to rule out osteitis, coalescence of the mastoids, or subperiosteal abscess formation. The imaging findings at this stage are similar to the earlier stage with the possible addition of swelling of the postauricular soft tissues, but there should be no evidence of bone erosion or abscess formation at this stage.

When infection progresses to involve the bone, it is referred to as acute mastoiditis with osteitis. This stage is also commonly referred to as acute coalescent mastoiditis because the bony trabeculae of the mastoid, which normally separate individual mastoid air cells, become eroded leading to formation of an intramastoid empyema. On CT, there is mastoid opacification with loss of mastoid trabeculations and, if the infection persists for long enough, coalescence of the mastoid air cells into a single mastoid cavity (Fig. 50) (Antonelli et al. 1999). Subperiosteal abscess may also develop at this stage. These abscesses result from extension of infection through vascular channels or via frank erosion of bone. Whenever mastoid osteitis is suspected by imaging, careful inspection along the mastoid walls should be performed for evidence of erosion of the mastoid cortex or for the presence of a loculated extramastoid fluid collection. Contrast-enhanced CT and MRI are more sensitive for detection of abscess than a noncontrast CT and will demonstrate a crescentic, peripherally enhancing fluid situated adjacent to the mastoid cortex (Fig. 51).
Once the infection has broken out of the mastoid, it may spread in several directions. Most characteristic is abscess formation along the lateral surface of the mastoid process, due to the relatively thin trabecular bone in this region, resulting in a postauricular subperiosteal abscess. Alternatively, the infection may spread anteriorly into the cells in the zygomatic root, resulting in an abscess in the anterior and superior portion of the pinna and preauricular area that can spread into the malar region. When the abscess extends through the mastoid tip into or beneath the sternocleidomastoid muscle, it is referred to as a Bezold abscess (Fig. 52). Because aeration of the mastoid tip is a predisposing factor for their development, Bezold abscesses generally occur in older children and adolescents, and they are rarely seen in infants and young children, in whom mastoid pneumatization is not yet complete (Vazquez et al. 2003). Less common locations for abscess development are posterior to the occipital bone (Citelli

**Fig. 50** Acute mastoiditis with osteitis. Axial unenhanced CT image of the left temporal bone in a young child demonstrates complete mastoid opacification with erosion of mastoid trabeculae and early coalescence. There is erosion of the lateral mastoid cortex (arrowhead). Note the overlying soft tissue swelling (no abscess was present in this child)

**Fig. 51** Postauricular abscess due to mastoiditis in an 8-year-old child who presented with swelling behind the right ear. (a) Axial contrast-enhanced CT viewed with bone windows demonstrates mastoid opacification with erosion of the lateral mastoid cortex (arrow) and loss of normal mastoid septations. (b) The corresponding image reconstructed with a soft tissue algorithm demonstrates a rim-enhancing fluid collection (asterisk) overlying the mastoid, compatible with a subperiosteal abscess

Once the infection has broken out of the mastoid, it may spread in several directions. Most characteristic is abscess formation along the lateral surface of the mastoid process, due to the relatively thin trabecular bone in this region, resulting in a postauricular subperiosteal abscess. Alternatively, the infection may spread anteriorly into the cells in the zygomatic root, resulting in an abscess in the anterior and superior portion of the pinna and preauricular area that can spread into the malar region. When the abscess extends through the mastoid tip into or beneath the sternocleidomastoid muscle, it is referred to as a Bezold abscess (Fig. 52). Because aeration of the mastoid tip is a predisposing factor for their development, Bezold abscesses generally occur in older children and adolescents, and they are rarely seen in infants and young children, in whom mastoid pneumatization is not yet complete (Vazquez et al. 2003). Less common locations for abscess development are posterior to the occipital bone (Citelli
Uncomplicated acute mastoiditis can be treated conservatively with antibiotics and myringotomy. Surgery is indicated for patients who fail to respond to conservative options or when there is mastoiditis with osteitis, subperiosteal abscess, or other associated complications. Therefore, differentiation between acute mastoiditis without osteitis and acute mastoiditis with osteitis or subperiosteal abscess is critical, as most cases of the former respond to antibiotics and tympanocentesis, while the latter usually requires treatment with mastoidectomy (Bluestone and Klein 2003b).

Subacute mastoiditis represents a later stage of otitis media and mastoiditis that develops when an acute infection fails to totally resolve within 10–14 days. It is characterized by signs and symptoms which are similar to those of AOM but milder and persistent. Development of this stage is attributable to failure of antibiotics to eradicate the initial infection, and children with subacute mastoiditis should undergo tympanocentesis for middle ear drainage and to identify the causative organism(s), so that culture-directed antibiotics can be initiated (Bluestone and Klein 2003b). Occasionally, patients may develop a latent suppurative process in the mastoid after treatment of AOM, in which the middle ear may appear free of disease on examination. These patients with “masked” mastoiditis fail to demonstrate the classic signs and symptoms of otitis media and may not actually present for care until after intracranial complications.
have developed (Holt and Gates 1983). It is likely that obstruction of mastoid drainage at the aditus ad antrum contributes to development of this entity, and mastoidectomy is indicated to relieve obstruction. Diagnosis of latent mastoiditis is typically made with CT, which demonstrates typical findings of acute mastoiditis; however, the middle ear cavity may appear normal. Intracranial complications are generally best characterized with MRI (see below).

**Petrous Apicitis**

Petrous apicitis (also referred to as apical petrositis) is a rare suppurative complication that occurs when infection from the middle ear and mastoid extends medially into a pneumatized petrous apex. Roughly a third of individuals have pneumatized petrous bones, but pneumatization usually does not occur until 3 years, making petrous apicitis an uncommon occurrence below this age (Bluestone and Klein 2003b; Razek and Huang 2012). Like acute mastoiditis, most cases of petrositis are self-limited and improve in concert with resolution of the precipitating middle ear and mastoid infection. However, the infection may, on occasion, fail to resolve due to impaired drainage caused by mucosal swelling or granulation. This can result in acute osteomyelitis, with bony resorption and formation of a purulent abscess, a situation analogous to the development of coalescing mastoiditis. Patients progressing to this stage of petrous apicitis usually present with an acute febrile illness and some or all of the symptoms of the classic Gradenigo’s triad (ear pain with aural discharge, sixth nerve palsy, and facial pain). Occasionally, the maxillary and mandibular divisions of the trigeminal nerve are involved, causing pain in the teeth and jaw. Potential complications of petrous apicitis include meningitis, cerebral abscess formation, and venous sinus thrombosis (Koral and Dowling 2006).

CT findings in acute petrous apicitis include opacification of petrous air cells in the early stage of the disease and bony lysis with resorption of petrous septations or erosion of cortical bone (Koral and Dowling 2006). The MRI findings of petrous apicitis are a high signal intensity lesion on T2-weighted images within a pneumatized anterior petrous apex, which is of low signal on T1-weighted images and demonstrates peripheral contrast enhancement (Koral and Dowling 2006; Dave et al. 1997). There may also be associated enhancement of the adjacent dura and cranial nerves and of Meckel’s cave due to meningitis. Abscesses will demonstrate ring enhancement and restricted diffusion on DWI (Ibrahim et al. 2010) (Fig. 53). Gallium SPECT imaging is useful for evaluating response to therapy (Lee et al. 2005).

**Fig. 53** Petrous apicitis. (a) Axial gadolinium-enhanced fat-suppressed T1-weighted MR image through the temporal bones in a patient with fever, headache, and double vision demonstrates a peripherally enhancing fluid collection (arrow) in the right petrous apex with marked surrounding abnormal enhancement. There is also diffuse mucosal enhancement throughout the right mastoids and middle ear, compatible with otomastoiditis. (b) The corresponding DWI image shows the fluid in the collection to have restricted diffusion (arrowhead), consistent with abscess...
Chronic Otitis Media

Some confusion surrounds the term chronic otitis media, which is frequently used synonymously with chronic suppurative otitis media (CSOM), but which is occasionally confused with chronic otitis media with effusion. The latter term generally refers to OME which persists beyond 3 months but does not involve a perforation of the tympanic membrane (Bluestone and Klein 2003b). This form of “chronic otitis media” is not generally considered a complication of OME. On the other hand, CSOM is a stage of middle ear disease in which there is chronic inflammation of the middle ear and mastoid associated with a defect in tympanic membrane, usually due to a chronic perforation or a tympanostomy tube (Bluestone and Klein 2003b).

The pathogenesis of CSOM is multifactorial. Factors associated with its development include anatomic factors such as eustachian tube dysfunction, immature or impaired immune function, method of feeding (bottle vs. breast), presence of upper-respiratory infection or allergy, familial predisposition, and various environmental (e.g., exposure to second hand smoke) and social factors. The prevalence of CSOM is reported to be particularly high (7–46 %) in certain population groups such as Alaskan, Canadian, and Greenland Inuits and Australian Aborigines (Bluestone and Klein 2003b). A higher than usual prevalence has also been reported among certain Native Americans such as members of the Apache and Navajo tribes, in certain South Pacific Islanders, and in some African populations.

CSOM is usually preceded by an episode of AOM with perforation or for which a tympanostomy tube was placed; however, it may develop as a consequence of chronic otitis media with effusion. Diagnosis is typically made when there is persistent drainage through a central perforation of the tympanic membrane lasting for at least 2–3 months. Tympanic membrane defects usually involve most of the pars tensa. Conductive hearing loss is a common presenting complaint, and chronic hearing loss is the most common long-term sequela of CSOM. If the level of hearing loss is greater than 20–30 dB, ossicular involvement should be considered, with potential causes of hearing loss including ossicular erosion and fixation due to fibrous or granulation tissue. Erosion of the ossicular chain occurs in approximately 20 % of cases of CSOM without cholesteatoma (Dornelles et al. 2005). Mixed hearing loss can also occur, with the sensorineural component usually being due to development of a serous labyrinthitis (see below). Cholesteatoma is a common complication when medical treatment fails, and progression to any number of suppurative complications including labyrinthitis, facial paralysis, or intracranial abscess can also occur (see below).

Chronic mastoiditis is usually seen in the setting of chronic suppurative otitis media and chronic tympanic membrane perforations, but may rarely occur in the absence of otitis media. In the latter situation, children may present with a fever of unknown origin or with chronic or recurrent otalgia or tenderness over the mastoid process. Similar to children with subacute mastoiditis, patients with chronic mastoiditis may progress to develop intracranial or other intratemporal complications.

CT should be considered for patients with CSOM and chronic mastoiditis if intensive medical treatment fails, if there is early recurrence after completion of therapy, or if cholesteatoma is suspected. The typical CT imaging findings are an opacified, sclerotic, or underdeveloped mastoid with a tympanic perforation. Key features to look for include discontinuity of the ossicular chain or other bone defects. In addition, other unusual causes of a chronic draining ear, such as neoplasms and Langerhans cell histiocytosis, should be excluded (Bluestone and Klein 2003b).

Acquired Cholesteatoma

Acquired cholesteatomas are much more common than congenital cholesteatomas and are almost always the result of chronic middle ear disease (otitis media or eustachian tube dysfunction) or of implantation of squamous epithelium into the middle ear or mastoid as a result of trauma or surgery (see above for a discussion of congenital cholesteatomas). They can be divided into primary (those occurring due to
tympanic retractions) and secondary types (those forming secondary to tympanic membrane perforations). Both types are characterized by the presence of keratinizing stratified squamous epithelium similar to that of the skin of the EAC and accumulation of desquamated epithelium or keratin within the middle ear or other pneumatized portions of the temporal bone, typically forming a squamous epithelium-lined cyst-like structure containing laminated keratin debris (Bluestone and Klein 2003b). Morbidity from middle ear cholesteatoma is primarily due to its propensity to cause bone resorption, which may be secondary to either pressure erosion from mass enlargement or, in part, the activity of various enzymes such as collagenase which are produced in the cholesteatoma or elaborated by osteoclasts in their immediate environment (Abramson and Huang 1977; Abramson 1969; Chole and Sudhoff 2010).

Several explanations for the pathogenesis of acquired cholesteatomas have been proposed, including (i) the invagination (retraction pocket) theory, (ii) the basal cell proliferation theory, (iii) the epithelial invasion theory, and (iv) the squamous metaplasia theory (Chole and Sudhoff 2010; Sudhoff and Tos 2000). The invagination theory is perhaps the most widely accepted and proposes that negative middle ear pressure resulting from eustachian tube dysfunction causes retraction of the tympanic membrane. The retraction usually occurs in the pars flaccida or in the posterior superior portion of the pars tensa, which tend to be the thinnest and morphologically weakest portions of the tympanic membrane (Lim 1970; Ars 1981). Inflammation causes adhesions to form between the medial surface of the retracted tympanic membrane and surrounding structures, including the ossicles, resulting in retraction pocket formation. Finally, desquamated keratin debris from the lining of the retraction pocket becomes trapped, leading to the formation of a primary cholesteatoma.

The basal cell proliferation theory suggests that inflammation causes hyperplasia of basal epithelial cell cones in the tympanic membrane which invade the basement membrane into the subepithelial tissues as columns of proliferating epithelium (Sudhoff et al. 1996). Sudhoff and Tos (Sudhoff and Tos 2000) proposed that the pathogenesis of most primary acquired cholesteatomas may actually be best explained by a combination of the invagination and basal proliferation theories.

In the epithelial invasion theory, keratinizing squamous epithelium from the outer skin of the tympanic membrane is thought to migrate into the middle ear through a perforation. This theory may explain the pathogenesis of less common secondary cholesteatomas which develop at the site of a central TM perforation (Bluestone and Klein 2003b; Chole and Sudhoff 2010). Furthermore, cholesteatomas can also arise secondary to postsurgical (iatrogenic) or posttraumatic implantation of epidermal elements into the middle ear cavity (McKennan and Chole 1989).

The final theory, squamous metaplasia, posits that chronic inflammation could cause metaplastic transformation of the normal pluripotent squamous or cuboidal epithelial cells of the middle ear cavity into keratinizing epithelium (Sade et al. 1983). Although some experimental data have demonstrated that certain stresses can lead to the formation of keratinizing epithelium in the middle ear, there is no direct evidence that cholesteatomas arise by this mechanism from the middle ear mucosa (Chole and Sudhoff 2010), and as a result, the squamous metaplasia theory is probably the least accepted explanation for the pathogenesis of acquired cholesteatomas.

The incidence of cholesteatoma in children is reported to be 4.5–15 per 100,000, with males being more frequently affected than females (Bluestone and Klein 2003b; Swartz 2009). The likelihood of cholesteatoma development is even higher in children with a cleft palate, among whom nearly 10 % develop aural cholesteatomas, likely as a consequence of functional obstruction of the eustachian tube due to impaired tubal opening mechanisms.

Signs and symptoms of ear disease in a child may be absent or mild, and therefore, acquired cholesteatoma may go undiagnosed for years. Adult patients often present with progressive conductive hearing loss and otorrhea; however, children rarely complain of hearing loss, especially if involvement is unilateral, and they are often unaware of more subtle symptoms associated with the disease, like otorrhea.
The presence of severe symptoms such as facial paralysis, vertigo, vomiting, fever, or headache should raise the concern for a suppurative complication. Diagnosis of acquired cholesteatoma is primarily based on otoscopy, which frequently shows a pearly white mass associated with tympanic membrane defect, usually situated in the posterior superior quadrant (Bluestone and Klein 2003b) (Fig. 54).

Because most cholesteatomas are diagnosed by otoscopy, the main role of imaging in the preoperative setting is to evaluate the extent of the disease and to assess for bone erosion. On CT, the diagnosis of cholesteatoma is confirmed by presence of an expansile, sharply margined soft tissue mass in the middle ear cavity eroding bone (Barath et al. 2011). The presence of an intratympanic mass that does not erode bone is a nonspecific finding and could represent, in addition to cholesteatoma, granulation tissue, fibrous tissue, secretions, or even a neoplasm. Furthermore, the presence of bone erosion is not entirely specific either, as it can be seen as a consequence of chronic otitis media without cholesteatoma, although it is seen more frequently when cholesteatoma is present (Swartz 2009).

![Fig. 54 Pars tensa cholesteatoma. Otoscopic view of the left tympanic membrane in a patient with an acquired posterosuperior pars tensa cholesteatoma. The cholesteatoma (asterisk) demonstrates a characteristic pearly white appearance (A anterior, P posterior, arrow – manubrium of the malleus) (Photo provided courtesy of Dr. Oliver Adunka, University of North Carolina, Chapel Hill, NC)](image_url)

![Fig. 55 Pars flaccida cholesteatoma. Axial (a) and coronal (b) CT images through the right temporal bone in a 5-year-old child with otorrhea and hearing loss demonstrate a soft tissue mass filling the epitympanum, including Prussak’s space (asterisk, b), and eroding the short process of the incus (arrowhead). The scutum is also mildly blunted in (arrow, b)](image_url)
The vast majority of acquired cholesteatomas arise from retraction pockets of the pars flaccida or the posterosuperior quadrant portion of the pars tensa (Lau and Tos 1989). Pars flaccida cholesteatomas typically originate within the Prussak’s space (Fig. 55), a small space situated between the pars flaccida and the neck of the malleus, which is bounded inferiorly by the lateral process of the malleus and superiorly by the lateral malleolar ligament. Cholesteatoma originating in this location tends to grow posteriorly into the lateral epitympanum (attic cholesteatoma) and from there can extend further posteriorly through the aditus ad antrum into the mastoid antrum. As they expand, pars flaccida cholesteatomas tend to displace the ossicular chain medially. Rarely, they may develop anterior to the head of the malleus and extend into the anterior epitympanic (supratubal) recess. When this occurs, facial nerve involvement at the level of the geniculate ganglion may occur (Chu and Jackler 1988).

Pars tensa cholesteatomas are less common and are usually result of posterosuperior retraction pockets. These cholesteatomas, which are also referred to as sinus cholesteatomas, tend to spread first to the posterior recesses of the middle ear, beginning with the more laterally positioned facial recess before spreading medially to the sinus tympani (Swartz 2009). From the posterior tympanum, these cholesteatomas may extend along the prominence of the facial nerve, medial to the incus body, into the posterior epitympanum, from which they may then spread to the antrum (Sudhoff and Tos 2007). Because they tend to extend medial to the ossicular chain, sinus cholesteatomas generally displace the ossicles laterally as they grow (Kikuchi et al. 1993) (Fig. 56).

At the time of diagnosis, nearly all cholesteatomas are associated with ossicular erosions at surgical or histologic evaluation. The most common sites of erosion are the long process of the incus (Fig. 57) followed by the incus body (Fig. 55) and the head of the malleus (Swartz 2009). Incus erosion may be present in up to 90 % of cases, with malleus and stapes erosion seen in roughly 30 % (Dornelles et al. 2005). Erosion of scutum is typically a late finding seen with advanced disease (Figs. 55 and 57).

For all cases of known or suspected cholesteatoma, each of the following should be closely looked for on CT: (i) ossicular chain erosion, (ii) scutum erosion, (iii) dehiscence of the tegmen tympani, (iv) lateral semicircular canal dehiscence, (v) dehiscence of the facial nerve canal, and (vi) sinus tympani involvement. When ossicular chain erosion is evident, an effort should be made to determine which ossicles are involved, as this will determine what type of reconstruction or prosthesis is necessary if ossiculoplasty is...
being considered. In particular, the status of the stapes should be carefully assessed, as involvement of the stapes superstructure will determine whether partial or total replacement prosthesis is indicated.

Large cholesteatomas in the epitympanum can also erode through the tegmen tympani (Fig. 58). When tegmen dehiscence is identified, it is critical to exclude the development of a temporal lobe encephalocele, which may be difficult or impossible to differentiate from cholesteatoma tissue in the epitympanum on CT. In these instances, MRI should be performed, and we find that high-resolution coronal or sagittal T2-weighted sequences (e.g., CISS or FIESTA) are particularly well suited for demonstrating herniation of brain or dura through the tegmen defect.

**Fig. 57** Cholesteatoma eroding the incus long process and scutum. Coronal CT image of the left temporal bone in a 4-year-old child with a pars flaccida cholesteatoma demonstrates complete erosion of the incus long process, which normally articulates with the stapes superstructure (arrow). The scutum is also blunted.

**Fig. 58** Cholesteatoma eroding the tegmen tympani. Coronal CT image of the right temporal bone at the level of the oval window in an 11-year-old child with a large epitympanic cholesteatoma shows a large defect of the tegmen tympani, which has been eroded by the cholesteatoma. In addition, the incus long process, which is normally visible at this level, has also been eroded, and the scutum appears slightly blunted.
Another potential complication resulting from medial extension of cholesteatoma is that of a labyrinthine fistula, which occurs in roughly 10% of ears with cholesteatoma and is usually due to erosion of the lateral semicircular canal (Moody and Lambert 2007) (Fig. 59). The bony coverings of the semicircular canals and oval window must be carefully evaluated, particularly in patients with SNHL or progressive vertigo. It is particularly important to be aware of semicircular canal erosion preoperatively because it may preclude complete cholesteatoma removal. Surgeons may initially elect to leave cholesteatoma matrix which is adherent to the dehisced SCC rather than risk potential perilymph leak (Lambert 2010).

Cholesteatomas may also dehisce and spread along the facial nerve either along its tympanic segment or, in the case of anterior epitympanic cholesteatomas, in the region of the geniculate ganglion, putting the nerve at risk for injury during surgery. Facial nerve dehiscence is identified in close to 20% of ears operated on for cholesteatoma, with the tympanic segment being the most commonly involved segment (usually just above the oval window), followed by the geniculate ganglion and the mastoid and labyrinthine segments (Chu and Jackler 1988; Moody and Lambert 2007; Ikeda et al. 2006). In spite of the relatively high frequency of facial dehiscence, preoperative facial paralysis secondary to cholesteatoma is uncommon, occurring in only 1.1% of patients with acquired cholesteatoma (Swartz 1984).

Finally, for cholesteatomas involving the posterior tympanum, it is important to determine if there is medial extension of the mass into the sinus tympani. This area cannot be visualized during surgery via the standard facial recess approach. Therefore, if the surgeon is not aware of the presence of disease within the sinus tympani preoperatively, he or she may inadvertently leave cholesteatoma behind in this location. Furthermore, the presence of sinus tympani disease predicts failure in controlling disease in a significant number of children (Stern and Fazekas-May 1992).

The treatment for acquired cholesteatoma is surgical debridement with either canal wall-up or canal wall-down mastoidectomy and tympanic membrane reconstruction. Ossicular reconstruction may be necessary in advanced cases depending upon integrity of the ossicular chain. In general, recurrence rates are higher with canal wall-up procedures (Mutlu et al. 1995). Furthermore, some authors believe that cholesteatomas behave more aggressively in children than they do in adults, and it is generally accepted that recurrence rates after surgery are higher in children (Edelstein et al. 1989; Sie 1996). Because of this, many surgeons choose to perform planned second-look procedures roughly 6 months after canal wall-up mastoidectomy to clear out residual or recurrent disease and, if necessary, to reconstruct the tympanic membrane and ossicular chain (Mutlu et al. 1995; Darrouzet et al. 2000; Ho and Kveton 2003).
MRI has been shown to be of some benefit for differentiating cholesteatoma from other entities that can reside in the middle ear, such as granulation or scar tissue, cholesterol granuloma, retained secretions, and other debris, which may all appear identical on CT imaging. MRI can therefore be particularly useful for ruling out cholesteatoma in the postoperative setting, potentially obviating the need for a second-look procedure. On standard noncontrast pulse sequences, the imaging features of cholesteatomas are nonspecific, with most being hypo- to isointense on T1-weighted images and hyperintense on T2-weighted images relative to brain tissue, characteristics that are similar to other processes in the middle ear. Administration of contrast media can help to distinguish cholesteatoma from granulation tissue, as the latter enhances on T1-weighted images following contrast administration while cholesteatoma does not (Martin et al. 1990) (Fig. 60); however, a number of investigators have demonstrated a poor correlation between conventional post-contrast T1-weighted MR images and intraoperative findings for differentiation of cholesteatoma and other tissues, particularly in cases of smaller cholesteatoma pearls (Kimitsuki et al. 2001; Vanden Abeele et al. 1999).

Williams, Ayache, and colleagues (Ayache et al. 2005; Williams et al. 2003) have suggested that performing delayed (30–45 min) post-contrast T1-weighted sequences may improve the accuracy of MRI for detecting residual cholesteatoma postoperatively, based on the observation that scar tissue that develops in the middle ear and mastoid cavity postoperatively may not demonstrate appreciable enhancement on immediate post-contrast imaging and may therefore produce false-positive results. Because scar

---

**Fig. 60** Epitympanic cholesteatoma. Axial T2-weighted (a), unenhanced T1-weighted (b), and gadolinium-enhanced T1-weighted (c) images through the left temporal bone demonstrate a lesion that is of predominantly low T1-weighted signal intensity and high T2-weighted signal, with thin peripheral enhancement (arrows). (d) Echo-planar DWI image demonstrates increased signal within the lesion, confirming the diagnosis of cholesteatoma.
tissue is poorly vascularized and often enhances in a delayed fashion, enhancement may only become evident on images obtained after a significant delay following the contrast injection. Cholesteatomas, being avascular, do not enhance on either immediate or delayed post-contrast images. In their report, these authors reported a sensitivity of 90% and a specificity of 100% for cholesteatoma recurrence using the delayed contrast-enhanced technique (Ayache et al. 2005); however, more recent studies have shown less robust results, with sensitivities ranging from 57% to 90% and specificities between 55% and 68% (De Foer et al. 2010; Venail et al. 2008).

Diffusion-weighted imaging (DWI) is a promising technique for diagnosing cholesteatomas, which typically demonstrate high signal intensity on DWI images (Fig. 60) due partly to restricted water diffusion but predominantly to T2 shine-through (Vercruysse et al. 2006). Echo-planar (EPI) DWI sequences, traditionally used intracranially for stroke evaluation, can be helpful for detecting cholesteatoma in the surgically altered middle ear, but suffer from susceptibility effects at the air/bone interfaces in the skull base which may hide or partially obscure small mural cholesteatomas. More recent attention has been devoted to novel non-EPI single-shot turbo spin-echo DWI sequences (e.g., HASTE DWI, PROPELLER DWI, and BLADE DWI), which allow the use of higher imaging matrices and which show fewer susceptibility artifacts, thus potentially improving visibility of smaller cholesteatomas and

---

**Fig. 61** HASTE DWI versus echo-planar (EPI) DWI for cholesteatoma detection. (a) Axial T2-weighted MR image of the left temporal bone in a patient who had undergone mastoidectomy in the past demonstrates nonspecific hyperintense soft tissue along the roof of the mastoidectomy bed (arrow). On the EPI DWI image (b), the area is obscured by curvilinear artifacts caused by magnetic susceptibility effects at the bone-soft tissue interface. Note the similar appearance on the normal right side. However, on the HASTE DWI image, the hyperintense cholesteatoma is well visualized (arrow).
mural cholesteatomas (De Foer et al. 2008; De Foer et al. 2007; Lehmann et al. 2009; Schwartz et al. 2011) (Fig. 61). In 2011, Jindal et al. (Jindal et al. 2011) performed an analysis of published studies on the use of EPI and non-EPI DWI for postoperative cholesteatoma detection and found a pooled sensitivity and specificity for non-EPI DWI of 91.4 % and 95.8 %, respectively, compared with 70.6 % and 87.3 % for EPI DWI. Despite the apparent advantages of non-EPI, however, the sequences still do not appear to be reliably able to detect cholesteatomas smaller than 3 mm (Li et al. 2013).

Cholesterol Granuloma

Cholesterol granuloma (CG) is a lesion characterized by an inflammatory reaction to byproducts of extravascular hemoglobin degradation resulting in formation of an expanding cyst in the temporal bone. CGs preferentially involve in the petrous apex, but can also occur in the middle ear cavity and mastoid. CGs rarely develop in the pediatric patients, among whom they most commonly occur in adolescents and teens; however, they have been reported to occur in children as young as 6 months of age (Miura et al. 2002; Sanna et al. 2009). In one series of 17 adult and pediatric petrous apex CGs, 18 % occurred in adolescents as young as 14 years of age (Brodkey et al. 1996). Another series of petrous apex cholesteatomas included patients as young as 12 years (Sanna et al. 2009).

Histologically, these lesions are composed of a large number of cholesterol crystals engulfed by multinucleated foreign-body giant cells, which are surrounded by a thick capsule of fibrous granulation tissue mixed with histiocytes, round cell infiltration, macrophages, and numerous accompanying capillary-sized blood vessels. Old and recent hemorrhage may also be evident (Rinaldo et al. 2005).

The pathogenesis of CG formation remains controversial, but two main theories are most widely accepted. Originally, it was thought that CGs arise as a consequence of occlusion of pneumatized air cells, resulting in negative pressure and subsequent extravasation of intravascular fluid into the mucosa of the air cells which, over a prolonged period of time, eventually leads to mucosal breakdown and hemorrhage into the air cells. The degrading blood products subsequently incite an inflammatory granulomatous-type reaction, and repetition of this cycle leads to formation of an expansile mass which may eventually erode surrounding bone (DiNardo et al. 2003). This “negative pressure” theory has recently been challenged by an alternative “exposed marrow” hypothesis, which states that marrow-filled cavities are eroded as air cells develop, causing a predilection for subacute hemorrhage in the air cells, and the subsequent inflammatory reaction to the degrading blood products leads to cyst development, with expansion of the cyst causing further re-hemorrhage from the marrow cavities (Jackler and Cho 2003).

CGs of the middle ear and mastoid are probably distinct entities from those occurring in the petrous apex. The latter type of CG, typically seen in older patients, usually only occurs when there is extensive pneumatization of the temporal bone. However, children with persistent eustachian tube dysfunction and middle ear effusion commonly have early arrest of pneumatization and, therefore, do not develop apical air cells (Sanna et al. 2009). In addition, because petrous apex pneumatization does not occur until after 3 years of age, petrous apex CGs are virtually unheard of in very young children, in whom CGs most commonly occur in the mastoid and epitympanum (Bluestone and Klein 2003b; Miura et al. 2002). Although CGs are commonly associated with long-standing chronic otitis media (occurring in 12–21 % of patients with chronic OME), they occasionally arise in patients without a previous history of infection (Rinaldo et al. 2005; da Costa et al. 1992).

Hearing loss is the most common presenting symptom in patients with petrous apex CGs, followed by vertigo, headaches, tinnitus, and facial nerve dysfunction. Rarely, patients may develop trigeminal neuralgia, diplopia, or visual symptoms (Terao et al. 2001). These symptoms most likely are due to the enlarging cyst impinging upon nearby cranial nerves or the otic capsule. Not infrequently, petrous apex CGs may be detected incidentally on imaging studies performed for unrelated reasons (Sanna et al. 2009). When they occur in the middle ear, otoscopic examination may reveal a blue eardrum, which probably is
due to reflection of light from the granuloma in the middle ear. In these cases, the condition must be
differentiated from a dehiscent high jugular bulb (see above) (Bluestone and Klein 2003b). Large middle
ear or mastoid CGs can erode into the EAC, inner ear, or middle cranial fossa (Jang and Cho 2009; Martin
et al. 2012; Murugasu et al. 2004; Nikolaidis et al. 2010).

Asymptomatic petrous apex CGs can be followed, while symptomatic lesions are treated with surgical
drainage or cyst resection (Schmalfuss 2009). Middle ear and mastoid cholesterol granulomas can initially
be treated with myringotomy; however, if this approach fails, the granuloma should be surgically removed
(Bluestone and Klein 2003b).

On CT, CGs of the middle ear cavity appear as nonspecific, nonenhancing soft tissue masses causing
variable degrees of bone erosion and as such may be indistinguishable from cholesteatoma. MRI allows
differentiation of the entities, as CGs demonstrate homogeneously high signal on both T1- and T2-weighted sequences (Fig. 62) surrounded by a low signal rim, while cholesteatomas are typically iso-
to hypointense relative to the brain (Martin et al. 1989; Morioka et al. 1995). The characteristic T1

---

**Fig. 62** Middle ear cholesterol granuloma. Axial unenhanced T1-weighted (a) and T2-weighted (b) MR images through the epitympanum demonstrate a lesion in the posterior epitympanum (arrow) which is of high signal intensity on both pulse sequences, compatible with a cholesterol granuloma.

**Fig. 63** Petrous apex cholesterol granuloma. (a) Axial noncontrast CT through the right temporal bone in a 12-year-old child demonstrates a well-circumscribed, ovoid, lucent lesion with sclerotic margins in the petrous apex (black arrow). (b) At a slightly inferior level, note the pneumatized but fluid-filled petrous apex air cells (arrowhead).
shortening displayed by CGs is thought to be due to the paramagnetic effects of hemoglobin breakdown products derived from microhemorrhages around cholesterol crystals within the cyst (Martin et al. 1989).

CGs of the petrous apex characteristically appear as a well-marginated, expansile mass centered in the petrous apex which remodels and erodes the bone on CT (Fig. 63). They can be differentiated from the more common petrous apex effusion by destruction of the normal septations between air cells. As in the middle ear, CGs of the petrous apex demonstrate high signal on T1- and T2-weighted sequences, with no internal enhancement evident following contrast administration (Fig. 64). They may show a thin rim of peripheral enhancement, however (Griffin et al. 1987). Occasionally, fatty marrow in an asymmetrically nonpneumatized petrous apex (a normal anatomic variant) may be mistaken for a petrous apex CG. Fat-suppressed MR sequences help to differentiate the two processes. On these pulse sequences, the signal from normal marrow fat drops out, while CGs remain hyperintense (Razek and Huang 2012) (Fig. 64).

**Labyrinthitis**

The term labyrinthitis refers to infection or inflammation of the inner ear involving the cochlear or vestibular apparatus. The process typically begins in the perilymph-containing spaces and then causes secondary changes of the endolymphatic spaces. Several methods of categorizing labyrinthitis have been proposed. These include grouping the disease based on the mechanism or route of spread (tymanogenic, meningogenic, hematogenic, or posttraumatic), by the causative agent (viral, bacterial, autoimmune, luetic, or toxic), by the type of tissue or fluid replacing the labyrinthine space (serous, suppurative, hemorrhagic, fibrous, ossific), or by the chronicity of the process (acute, subacute, chronic) (Bluestone and Klein 2003b; Swartz and Mukherji 2009).
Serous (or toxic) labyrinthitis is defined as a sterile inflammatory response within the labyrinth and can be due to various factors, including bacterial toxins or other inflammatory mediators, contamination of perilymph due to surgical procedures or perilymph fistula, neoplastic influences, inner ear hemorrhage, and viral infections (Nadol 2010). Viral infections are probably the most common cause of serous labyrinthitis, with mumps virus, measles virus, and varicella-zoster virus being among the most frequently reported causes of viral labyrinthitis (Davis 2010).

Acute suppurative labyrinthitis typically develops as a consequence of otitis media or bacterial meningitis. When labyrinthitis develops as a result of an infection spreading into the inner ear from the middle ear, it is referred to as tympanogenic (or otogenic) suppurative labyrinthitis (Swartz and Mukherji 2009; Nadol 2010). Passage of pathogens into the inner ear is usually through the oval window or round window, with spread through a cholesteatomatous lateral semicircular canal fistula being a less common route of entry. The process should be considered in patients with acute or chronic middle ear infections who develop sudden or rapidly progressive hearing loss or severe vertigo (Bluestone and Klein 2003b). Patients with underlying Mondini malformations, a previous history of stapes footplate fracture, or a fistula between the middle and inner ear is at increased risk for suppurative labyrinthitis.

Bacterial infection spreading to the inner ear from infected CSF or the meninges is referred to as meningogenic suppurative labyrinthitis (Nadol 2010). Meningitis is the most common cause of acquired childhood deafness, accounting for roughly 6% of all hearing loss in children. It is estimated that anywhere from 5 to 35% of patients with bacterial meningitis develop permanent sensorineural hearing loss (Davis 2010; Dodge et al. 1984; Fortnum and Davis 1993). S. pneumoniae is the bacterial species most often associated with postmeningitic SNHL (Wellman et al. 2003). Spread of bacteria into the inner ear in cases of meningogenic labyrinthitis is usually via the fundus of the IAC – typically through the lamina cribrosa or cochlear nerve foramen – or via the cochlear aqueduct (Swartz et al. 1985). Due to its route of transmission to the inner ear, meningogenic labyrinthitis is more likely to be bilateral, while the tympanogenic form tends to be unilateral.

Symptoms of labyrinthitis depend on the severity of the inflammatory process. Serous or toxic labyrinthitis can present with sudden, progressive, or fluctuating hearing loss and/or vertigo; however, in children (especially infants), the presence of vertigo may not be obvious clinically (Bluestone and Klein 2003b; Swartz and Mukherji 2009). Symptoms of suppurative labyrinthitis, on the other hand, are more fulminant and usually manifest as sudden or rapidly progressive hearing loss or severe vertigo. Patients may be afebrile, and when fever is present, meningitis should be suspected. Autoimmune causes of labyrinthitis are also recognized causes of labyrinthitis that produce progressive SNHL and vestibular defects. One such autoimmune disorder is Cogan syndrome, which consists of progressive bilateral SNHL and interstitial keratitis of the eyes. Although Cogan syndrome most frequently affects young adults, children may also develop the disease (Vasileiadis et al. 2012). Treatment of labyrinthitis is directed at the underlying cause. Mastoidectomy may be indicated for patients with underlying cholesteatoma or acute mastoid osteitis (Bluestone and Klein 2003b). Meningogenic suppurative labyrinthitis improves after treatment of the underlying meningitis.

From an imaging standpoint, it may be most practical to classify labyrinthitis as acute, subacute, or chronic labyrinthitis. CT and MRI are complementary in evaluating patients with suspected labyrinthitis. CT allows for evaluation of middle and inner ear structures, is better at demonstrating bony defects, and defines extent of underlying mucosal thickening or cholesteatoma. MRI detects abnormal signal or enhancement of the membranous labyrinth confirming an inflammatory process.

There are no characteristic CT findings in uncomplicated acute serous or suppurative labyrinthitis; however, CT may be helpful in identifying congenital or acquired defects, such as cochlear dysplasias or findings suggestive of a perilymph fistula. MRI is usually also normal, but patients with acute/subacute labyrinthitis may occasionally demonstrate faint enhancement of the labyrinthine structures on contrast-
enhanced MRI, which can persist for as long as 6 months after symptoms have resolved (Seltzer and Mark 1991; Papadopoulos et al. 1995; Downie et al. 1994) (Fig. 65). The labyrinthine enhancement is presumed to be due to accumulation of gadolinium in the labyrinthine membranes caused by breakdown of labyrinthine vasculature or leakage of gadolinium into the labyrinthine fluid due to breakdown of the blood-perilymph barrier (Seltzer and Mark 1991; Vignaud et al. 1995). In cases of hemorrhagic labyrinthitis, T1-weighted MR images may show increased signal intensity in the structures of the inner ear (Lowe and Vezina 1997).

Long-standing inflammation leads to replacement of the membranous and bony labyrinth by fibrous (fibrosing labyrinthitis) or osseous tissue (labyrinthitis ossificans) over a period of months to years. This process is a nonspecific end-stage healing process that may be seen after an episode of either serous or suppurative labyrinthitis. It is theorized that these changes develop due to abnormal proliferation of undifferentiated mesenchymal cells which eventually differentiate into fibroblasts and later osteoblasts (Swartz and Mukherji 2009).

In the fibrous stage of chronic labyrinthitis, which can occur by as early as 2 weeks, granulation tissue consisting of hypertrophic fibroblasts and neovascularity causes membranous fibrosis (Hegarty et al. 2002). The CT appearance of the inner ear in this stage remains normal. Therefore, MRI can be quite useful in establishing the diagnosis, as it will demonstrate partial or complete loss of normal high signal intensity perilymph within the inner ear on T2-weighted images (Fig. 66). We use CISS images for...
this purpose, as they provide high fluid contrast and excellent spatial resolution. Labyrinthine enhancement with gadolinium is also commonly present at this stage due to the presence of angiogenesis (Casselman et al. 1993a).

In the ossific stage of chronic labyrinthitis, intracochlear bone formation is well characterized by CT and may be localized or diffused. Normal labyrinthine structures are replaced by bone, with ossification generally beginning in the scala tympani in the basal turn of the cochlea early on and progressing toward the apex (Axon et al. 1998; Green et al. 1991) (Fig. 67). When there is complete ossification, differentiation from congenital cochlear or labyrinthine aplasia may be difficult, as the labyrinthine structures may become similar in density to the surrounding otic capsule bone (Fig. 68) (see section on congenital inner ear anomalies above). Helpful in distinguishing the two entities is the tendency for labyrinthine aplasia to demonstrate a small inner ear with associated flattening of the medial middle ear wall and a hypoplastic or atretic IAC (Marsot-Dupuch et al. 1999).

MRI findings are similar to those observed in fibrosing labyrinthitis. Abnormal enhancement of vestibule or cochlea may persist if there are mixed fibrous and ossific changes. The distinction between fibrous and osseous obstruction of the membranous labyrinth is particularly important in candidates for

Fig. 67 Early labyrinthitis ossificans. Axial left temporal bone CT image in a 5-year-old child with left-sided SNHL demonstrates faint calcification of the basal turn of the cochlea (arrow), with preservation of the mid and apical turns.
cochlear implantation, as significant ossification of the cochlea can make implantation more difficult, if not impossible, and often results in poorer functional results (Waltzman et al. 1995).

Ramsay-Hunt Syndrome

Ramsay-Hunt syndrome (herpes zoster oticus) is a disease caused by the reactivation of latent varicella-zoster virus residing within the geniculate ganglion, with subsequent spread of the inflammatory process to the seventh and eighth cranial nerves. Patients may present with ipsilateral facial paralysis, tinnitus, hearing loss, hyperacusis, vertigo, dysgeusia, decreased tearing, or ear pain. Characteristic vesicles are frequently seen in the external auditory canal (Fig. 69), on the pinna, or, less often, on the anterior pillar of the fauces. In some patients, cranial nerves V, IX, and X are also involved.

Although Ramsay-Hunt syndrome is the most common cause of atraumatic facial paralysis in adults, it is relatively rare in children, and the incidence appears to increase with age. Furuta et al. found that the syndrome presents in 53% of children with facial paralysis between the ages of 6 and 15 years, but only in 9% of those under 5 years (Furuta et al. 2005). Compared with adults, children with Ramsay-Hunt syndrome tend to experience milder degrees of facial palsy and are less likely to develop other associated cranial neuropathies. Furthermore, the appearance of vesicles tends to be delayed in children with the disease (Hato et al. 2000).

MRI usually shows contrast enhancement of the seventh and eighth nerve trunks within the distal internal auditory canal and along the labyrinthine segment, as well as enhancement of the cochlea, vestibule, and parts of the semicircular canals (Kuo et al. 1995; Sartoretti-Schefer et al. 1994). Intense enhancement of the geniculate ganglion and the tympanic and mastoid facial nerve segments may also be observed (Sartoretti-Schefer et al. 1994) (Fig. 69). In addition, focal T2 hyperintensities of the brainstem, most often within the trigeminal nuclear complex, have been reported (Haanpaa et al. 1998), as well as enhancing, ischemic lesions in the region of the pontine facial nerve nucleus (Sartoretti-Schefer et al. 1999). The latter finding has been considered the result of a primary neuritis of the intrameatal nerve trunks of the seventh nerve, with secondary anterograde and retrograde spread of the inflammation to the intratemporal nerve segment and to the brainstem (Sartoretti-Schefer et al. 1999).

Intracranial Complications of Acute Otitis Media and Mastoiditis

Intracranial complications of mastoiditis include meningitis, intracranial abscesses and empyemas, venous sinus thrombosis, and otitic hydrocephalus (Bluestone 2000; Go et al. 2000). Prior to the antibiotic era, intracranial complications developed in 6% of patients with mastoiditis, and nearly 80% of these cases were fatal. The widespread adoption of antibiotics has fortunately led to a significant decline in the
mortality and hospitalizations due to mastoiditis; however, among children hospitalized or requiring surgery for mastoiditis, intracranial involvement by the disease is still fairly common, with an incidence of up to 16% (Zevallos et al. 2009).

Fever and otalgia are the most common symptoms in this subset of patients, but these complaints are nonspecific and are frequently seen in cases of mastoiditis without central nervous system involvement. On the other hand, headache or alterations in mental status are uncommon complaints in uncomplicated mastoiditis and, when present, should be considered signs of intracranial involvement (Go et al. 2000).

Sigmoid sinus thrombosis and epidural abscess are probably the most frequently diagnosed intracranial complication of mastoiditis, but meningitis, subdural empyema, and otitic hydrocephalus are not uncommon (Bluestone 2000; Go et al. 2000; Zevallos et al. 2009).

**Venous Sinus Thrombosis** Thrombus within a venous sinus may be evident on unenhanced CT, appearing as high density material within the occluded sinus, but this finding is only evident in approximately 25% of cases of sinus thrombosis (Leach et al. 2006). On contrast-enhanced CT or MRI, an intraluminal filling defect will usually be evident within the occluded sigmoid sinus (Fig. 70). Thrombus is also suggested on spin-echo MRI sequences by the absence of a normal flow void in the occluded venous structure. Acute thrombus (0–5 days) is predominantly isointense on T1-weighted images and hypointense on T2-weighted images, while subacute thrombus (6–15 days) becomes increasingly hyperintense on both T1- and T2-weighted images. Sequences susceptible to blood may also be useful for detecting thrombosed sinuses and veins, which will appear markedly hypointense on SWI and T2* images. Phase contrast or time-of-flight MR venography in patients with CVT demonstrates absence of normal venous flow-related signal (Fig. 70). In some cases, however, time-of-flight MRV may appear falsely negative due to high signal from thrombus being mistaken as normal flow. In these cases, contrast-enhanced CT venography or MRV usually clearly depicts the filling defect.

**Epidural Abscess** Epidural abscesses are typically due to contiguous spread of infection from the mastoids. These collections reside in the potential space between the dura and the inner table of the skull. The thick, inelastic dura acts initially as a barrier to protect the underlying brain from concomitant infections.
involvement. As a result, epidural abscesses may present with a prolonged and insidious clinical course (usually over the course of several weeks to months) (Karampekios and Hesselink 2005). Early on, patients may complain only of fever or headache; other neurological symptoms do not develop until the infection has breached the dura into the subdural space, after which point meningitis, cerebritis, or subdural empyema may develop.

MRI is the most sensitive imaging modality for detecting epidural abscesses. They classically have a biconvex shape and most frequently occur in the posterior fossa posterior to the sigmoid plate and above the sigmoid sinus, where they are referred to as perisinus abscesses (Minks et al. 2013). They may resemble CSF on T2-weighted imaging, but they are typically of higher signal intensity than CSF on T1-weighted and FLAIR images. On contrast-enhanced T1-weighted imaging, epidural abscesses demonstrate a peripheral enhancement (Fig. 71). Unless the process has breached the dura or there is concomitant cortical venous thrombosis, the adjacent brain parenchyma is often normal in appearance. On diffusion-weighted imaging, epidural abscesses may demonstrate reduced diffusion.

**Hydrocephalus** Otitic hydrocephalus refers to the development of increased intracranial pressure in the setting of middle ear and mastoid infection. Presenting signs and symptoms include headache, vomiting, abducens palsies, and papilledema. When lumbar puncture is performed, the opening pressure is elevated,
but CSF findings are otherwise normal (Sennaroglu et al. 1996). The use of the term “hydrocephalus” in this case is perhaps a misnomer, as actual ventricular enlargement is uncommon. The pathogenesis of otitic hydrocephalus is controversial. Most authors ascribe the increase in intracranial pressure to thrombosis of the venous sinuses resulting in impaired venous drainage, while others – citing cases in which sinus thrombosis is absent – believe that the process is idiopathic. Still others have suggested that the process is caused by a vasomotor reflex phenomenon originating from the thrombosed epithelium of the vein rather than by mechanical obstruction of the vein (Sennaroglu et al. 1996; Teichgraeber et al. 1982). MRI should be performed in cases of otitic hydrocephalus, primarily to assess for possible venous sinus obstruction or other intracranial complications.

**Temporal Bone Trauma and Associated Findings**

Among patients presenting to trauma centers with closed head injuries, roughly 10–20 % have temporal bone fractures evident clinically or by CT (Nageris et al. 1995). Temporal bone fractures comprise 22 % of all skull fractures, and 8–29 % occur bilaterally (Brodie 2010; Cannon and Jahrsdoerfer 1983). Historically, temporal bone fractures have been classified based on their course relative to the long axis of the petrous pyramid as longitudinal or transverse fractures. Longitudinal fractures course parallel to the long axis of the petrous pyramid, whereas transverse fractures are oriented perpendicular to this axis (Cannon

---

**Fig. 72**  Longitudinal temporal bone fracture. (a) Axial CT image of the right temporal bone in a 16-year-old who sustained a head injury. There is a fracture line (arrow) oriented parallel to the axis of the petrous bone which extends through the mastoid into the epitympanum. (b) Image slightly inferior to (a) again demonstrates the fracture (arrow) traversing the mastoids and extending through the posterior and anterior (black arrowhead) walls of the EAC. (c) Image located superior to (a) demonstrates that the fracture line (arrowhead) extends into the squamosa of the temporal bone.
and Jahrsdoerfer 1983). Using this classification system, longitudinal fractures account for roughly 80% of temporal bone fractures (Parisier et al. 2003).

Longitudinal fractures classically result from a blow to the temporoparietal region and typically involve squamosal portion of the temporal bone and the posterosuperior wall of the EAC, with extension through the tegmen mastoideum and tegmen tympani. The fracture line typically courses along the long axis of the petrous bone with the vector extending through the middle ear cavity (Fig. 72), the region of the incudomalleolar joint, and the anterior genu of the facial nerve. Longitudinal fractures are often associated with ossicular fractures or subluxations. In addition, the facial nerve is injured in roughly 25% of patients with these fractures, with the injury usually occurring along the tympanic portion of the nerve between the geniculate ganglion and its posterior genu (Parisier et al. 2003). Classic findings in patients with longitudinal fractures include hemotympanum, perforation of the tympanic membrane, and blood in the EAC.

Transverse fractures usually occur as a result of severe trauma to the frontal or occipital regions, often resulting in loss of consciousness. The anteroposterior direction of the force results in a fracture that begins in the region of the foramen magnum or jugular foramen and courses perpendicular to the long axis of the temporal bone. These fractures may extend to the region of the IAC or across the cochlea and vestibule (Fig. 73), so this type of fracture often results in sensorineural hearing loss and vertigo. Furthermore, approximately 50% of patients with transverse fractures suffer from facial nerve injury resulting in paralysis (Parisier et al. 2003). Bleeding from the EAC, hemotympanum, and perforation of the tympanic membrane are less commonly associated with transverse fractures than longitudinal fractures.

In reality, most temporal bone fractures are more accurately described as oblique or complex in nature (Aguilar et al. 1987; Williams et al. 1992) (Fig. 74). Because of this, it has been suggested that it is more useful to classify fractures based on whether they involve or spare the otic capsule (Fig. 73), as otic capsule-disrupting fractures almost always result in sensorineural hearing loss and are also associated with a much higher incidence of facial nerve paralysis compared to fractures sparing the otic capsule (30–50% vs. 6–14%) (Brodie and Thompson 1997; Dahiya et al. 1999). Based on this method of classification, only 2.5–5.8% of temporal bone fractures involve the otic capsule.
CT imaging is indicated for evaluation of suspected temporal bone fractures and provides information regarding the course of the fracture and the structures involved, including the IAC, otic capsule, facial nerve canal, and ossicular chain. The majority of temporal bone fractures are associated with ipsilateral opacification of the mastoid air cells, and patients who present with signs of a basilar skull fracture and an opacified mastoid without an obvious fracture by CT should be assumed to have an occult temporal bone fracture. Reversible causes of facial nerve palsy, such as hematomas or bone fragments impinging on the facial nerve, should be identified as emergent decompressive surgery, or intravenous steroid administration may be warranted in certain instances in order to preserve facial nerve function. Overall, facial

**Fig. 74** Oblique temporal bone fracture with malleoincudal dislocation. Axial left temporal bone CT from a 9-year-old child who suffered head trauma demonstrates an obliquely oriented fracture line (arrowhead) traversing the mastoids and extending into the epitympanum. The malleoincudal joint has lost its normal ice cream cone configuration, with the scoop of ice cream, representing the malleus, being medially subluxed relative to the cone, representing the incus body and short process.

**Fig. 75** Oblique temporal bone fracture resulting in a perilymph fistula. Axial right temporal bone CT image in a patient with an oblique temporal bone fracture (arrow) demonstrates foci of gas (arrowheads) within the cochlea and vestibule, indicating the presence of a traumatic perilymphatic fistula.
paralysis occurs in 5–33 % of children with temporal bone fractures (Parisier et al. 2003). In addition, carotid artery dissection may result from extension of the fracture into the carotid canal; therefore, if fracture is seen extending into the petrous carotid canal, dedicated imaging of the carotid artery with either CTA or MRA should be performed to rule out injury to the vessel.

In all cases, the status of the tegmen tympani and tegmen mastoideum should also be determined, as fracture extension through these areas places patients at higher risk for recurrent meningitis and CSF leak (Brodie and Thompson 1997). Such patients may benefit from exploratory surgery and dural grafting to prevent complications. Delayed or fluctuating SNHL and persistent vertigo suggest a posttraumatic perilymph fistula (see below), and air visualized within the vestibule (pneumolabyrinth) is evidence of direct communication between the middle ear/mastoid cavity and the labyrinth (Fig. 75). In children, extensive longitudinal fractures may completely separate the petrous apex from the other portions of the temporal bone. This is termed a “floating cochlea” and is associated with acute conductive hearing loss and paralysis of the abducens and facial nerves (Merwin et al. 1989).

A variety of ossicular abnormalities, ranging from mild subluxation to complete fractures, can result from temporal bone fractures (Meriot et al. 1997). Subluxation of the incudostapedial joint is the most commonly reported posttraumatic derangement, occurring in roughly 80 % of cases, although subtle cases may be difficult to detect by CT. Therefore, incudostapedial subluxation is only identified in slightly over 50 % of cases on CT. Disruption of the incudomalleolar joint is visualized more frequently on CT (seen in 60 % of radiographically evident ossicular injuries) and is diagnosed when there is derangement of the normal “ice cream cone” configuration or subtle widening of the joint on axial images through the epitympanum (Fig. 74). Complete dislocation of the incus, in which both the incudostapedial and malleoincudal joints are dislocated, occurs in approximately 30 % of cases (Fig. 76). Stapediovestibular dislocation, evident as displacement of the stapes footplate laterally away from the oval window or medially into the vestibule, is rare and accounts for only 3 % of ossicular injuries. This, in addition to fractures of the stapes footplate, can be associated with perilymphatic fistulas (Mafee et al. 1984; Nishiike et al. 2008). Fractures of the ossicles are also relatively uncommon. The stapes is the most likely ossicle to be fractured (6 %), usually with involvement of the crura. Fractures of the incus occur in 4 % of cases and most often affect the long or lenticular process, while malleus fractures only occur in 1 % of cases (Meriot et al. 1997).
Perilymphatic Fistula

A perilymphatic or labyrinthine fistula is an anomalous communication between the inner and middle ear cavities resulting in leakage of fluid into the middle ear (Weber and Reilly 2003). Perilymphatic fistulae are felt to be an important cause of sensorineural hearing loss in children and may be congenital or acquired.

Congenital forms are associated with developmental dysplasias of the cochlea, vestibule, and semicircular canals, as well as with anomalies of the stapes, oval window, and round window (Weissman et al. 1994). Most congenital fistulae occur at the oval window and/or round window, with the former being the more common site (Weber and Reilly 2003). The prevalence of congenital fistulae in children with unexplained hearing loss is about 6 % (Reilly 1989).

Acquired fistulae may occur spontaneously or arise as complications of surgery (usually stapedectomy), infection, trauma (including barotrauma), or tumor. In one surgical series, 5.6 % of stapedectomy failures were attributed to perilymphatic fistulae. The fistulae were usually due to incomplete graft coverage of the stapedectomy defect, but also occurred as a result of dislocation of the prosthesis into the vestibule (Scheid et al. 2001; Vincent et al. 2010). As mentioned previously, roughly 10 % of patients with acquired cholesteatomas develop perilymphatic fistulae, usually as a consequence of lateral semicircular canal erosion (Moody and Lambert 2007). Less commonly, cholesteatoma may breach the inner ear directly through the oval or round window. Trauma resulting in fractures through the otic capsule or stapediovestibular dislocations may also cause leakage of perilymph into the middle ear.

The diagnosis of perilymphatic fistula should be suspected in a patient who presents with sudden sensorineural hearing loss in the setting of recent exertion, head trauma, or barotrauma. Patients may alternatively complain of fluctuating hearing loss, tinnitus, ataxia, or episodic vertigo. Fistula formation should also be suspected in patients who develop labyrinthitis, sensorineural hearing loss, or recurrent episodes of meningitis following otitis media. The treatment of idiopathic perilymphatic fistulae or those associated with congenital ear dysplasia is initially conservative, allowing for spontaneous recovery, while treatment of acquired fistulas is directed to the underlying cause. For a perilymphatic fistula which persists after conservative management, exploratory tympanotomy with grafting of the fistula site (usually the oval and/or round window) is recommended (Weber and Reilly 2003).

CT is the modality of choice for imaging patients suspected of having a perilymphatic fistula. In patients with idiopathic fistulae, imaging is often normal, but in some instances small fluid collections adjacent to the oval or round window may be evident. In these cases, the diagnosis should be suggested in patients with the proper clinical findings and unexplained middle ear effusions. In addition, the presence of pneumolabyrinth is strongly suggestive of underlying labyrinthine fistula (Scheid et al. 2001; Nishizaki et al. 1998) (Fig. 75). The primary role of imaging is to identify patients with underlying predisposing conditions for developing a perilymphatic fistula and to exclude other causes for the patient’s symptomatology. Imaging studies should be closely evaluated for malformations involving the cochlea, vestibule, and semicircular canals, and abnormalities of the oval window and stapes should also be searched for, as these are commonly found intraoperatively in patients with congenital fistulae. That said, only a minority of stapes and oval window malformations can be identified on CT (Weissman et al. 1994; Weber et al. 1993).

The bony covering over the lateral semicircular canal and the margins of the oval and round windows must be specifically evaluated in patients with middle ear cholesteatoma, as erosion of these bony coverings may portend development of a labyrinthine fistula. In the poststapedectomy patient, the position of the stapes prosthesis should be assessed as medial subluxation of the prosthesis into the vestibule may cause a perilymph fistula (Scheid et al. 2001).
Pediatric Temporal Bone Tumors and Tumor-Like Conditions

Tumors arising in the temporal bone are uncommon, comprising just 1.5% of head and neck tumors in children (Cunningham et al. 1987). They often present insidiously and with nonspecific symptoms, but children most frequently present with signs and symptoms of acute or chronic otitis media or external otitis (Van De Graff and Cass 2003). A number of different histologic tumor types have been reported in the temporal bone in children; however, most are extremely rare. Certain tumors that are relatively common in adults, such as paragangliomas and squamous cell cancers, are exceedingly uncommon in pediatric-aged patients and are therefore not covered in detail here. Among the more common tumors to affect the ear in children are rhabdomyosarcoma and Langerhans cell histiocytosis, which are reviewed below.

Rhabdomyosarcoma

Rhabdomyosarcoma (RMS) is the most common soft tissue malignancy of childhood, accounting for greater than 60% of pediatric soft tissue sarcomas (Sbeity et al. 2007). Although the tumor commonly affects the head and neck region, involvement of the temporal bone occurs in only 8–15% of cases with head and neck disease, while the masticator space and orbit are the most commonly affected sites. Nonetheless, RMS is still the most common malignancy of the pediatric temporal bone, accounting for roughly 80% of lateral skull base tumors in children (Sbeity et al. 2007; Castillo and Pillsbury 1993; Schwartz et al. 1980).

Several theories have been proposed concerning the cellular origins of RMS, including derivation from totipotential mesenchymal cells, from immature myoblastic tissue, or from muscles within the middle ear (Sbeity et al. 2007). The tumors are characterized microscopically by small, anaplastic, round, and spindle-shaped cells exhibiting hyperchromatic nuclei and granular acidophilic cytoplasm. Four histological subtypes of RMS are traditionally described: (i) embryonal, (ii) botryoid (considered by some to be a subtype of the embryonal variety), (iii) alveolar, and (iv) pleomorphic (Parham 2001). Embryonal RMS is by far the most common subtype in the head and neck, including the temporal bone, and is more commonly found in younger children (Robson 2010). The alveolar type is seen in older children, most commonly occurs in the extremities, and carries a worse prognosis than embryonal sarcomas (Robson 2010; Stein-Wexler 2009). The rare pleomorphic type of RMS occurs almost exclusively in adults (Parham 2001).

RMS has a bimodal age distribution with the first peak occurring in the first decade of life, and the second peak occurring during adolescence (Robson 2010). They may arise primarily or develop secondary to previous irradiation. While most cases occur sporadically, some children have been identified with mutations of the p53 tumor suppressor gene which may lead to a hereditary predisposition to cancer (Robson 2010). In addition, the embryonal subtype has been associated with loss of heterozygosity at chromosome 11p (Parham 2001; Robson 2010).

RMS of the temporal bone usually present insidiously with symptoms which mimic those of chronic otitis media but which are unresponsive to treatment. Occasionally, they may present with retroauricular masses. Additional clinical findings may include a polyp in the EAC, discharge or bloody otorrhea, pain, or focal neurologic deficits including cranial nerve palsies (Van De Graff and Cass 2003; Sbeity et al. 2007). Roughly 30% of patients have neurologic deficits at the time of diagnosis and facial nerve palsy is often present (Schwartz et al. 1980; Castillo et al. 1998). These tumors are often extensive and may involve multiple sites including the intracranial compartment. Tumor spread may be via contiguous destruction of tissue, including bone, and by perineural extension along nerve trunks. Tumors confined to the middle ear usually spread via invasion of the fallopian canal with infiltration of the facial nerve and extension into the IAC (Myers et al. 1968). Other routes of spread include extension into the middle cranial fossa through the tegmen; spread into the vestibular and cochlear aqueducts following invasion...
into the labyrinth; inferior extension to involve the jugular bulb, styloid process, and carotid artery; and direct extension anteriorly into the nasopharynx and infratemporal fossa (Wiatrak and Pensak 1989).

Approximately 15% of patients with temporal bone RMS have distant metastasis at the time of diagnosis, with the lungs being the most common site of hematogenous spread, while regional lymph node metastasis occurs in 5–20% of patients (Crist et al. 1990). Treatment of temporal bone RMS involves multimodality therapy with combined radiotherapy and chemotherapy, with or without surgery, and reported 5-year survival rates range from 41% to 81% (Sbeity et al. 2007; Durve et al. 2004; Raney et al. 1983). The presence of meningeal involvement is considered the worst prognostic indicator in head and neck RMS (Latack et al. 1987).

The imaging findings of temporal bone RMS are nonspecific and show an aggressive soft tissue mass causing bone destruction (Fig. 77). MRI is preferred over CT in evaluating the extent of tumor spread, particularly when dural involvement and intracranial spread are suspected. On MRI, the lesions are heterogeneous but may demonstrate iso- to hypointense signal relative to brain on T2-weighted images suggestive of hypercellularity (Robson 2010). On T1-weighted images, the lesions are usually iso- to slightly hyperintense relative to muscle and enhanced following contrast administration. Central necrosis may also be evident (Castillo and Pillsbury 1993). Coronal or sagittal contrast-enhanced T1-weighted images are particularly useful for evaluating intracranial involvement (Fig. 77). On DWI, RMS may show reduced diffusion, likely due to the cellular nature of the tumors (Lope et al. 2010).
Langerhans Cell Histiocytosis

Langerhans cell histiocytosis (LCH – previously referred to as histiocytosis X) refers to a group of diseases involving various organ systems, characterized by an abnormal clonal proliferation of Langerhans cells. In normal circumstances, Langerhans cells act as tissue macrophages in the dermis and also interact with lymphocytes in T-cell-dependent areas of lymph nodes (Saliba et al. 2008). LCH includes three overlapping entities formerly known as eosinophilic granuloma, Letterer-Siwe disease, and Hand-Schuller-Christian disease.

The etiology of LCH remains a mystery, but the observation that all forms of the disease are monoclonal (with the exception of the adult pulmonary form) suggests that it may be a neoplastic process. Alternatively, it has been hypothesized that LCH is a reactive disorder which is caused by environmental or other triggers leading to an aberrant reaction between Langerhans cells and T lymphocytes (Weitzman and Egeler 2008).

Pathologic diagnosis of LCH requires electron microscopic identification of Birbeck granules – laminar rod-shaped organelles which are present in the nuclear cytoplasm of Langerhans cells – or positive immunohistochemical staining with S-100 and/or CD1a (Saliba et al. 2008; Satter and High 2008). The lesions of LCH are granulomatous lesions containing pathologic Langerhans cells as well as normal inflammatory cells such as T cells, eosinophils, and macrophages. In addition, the lesions contain multinucleated giant cells which have been shown to be osteoclastic and produce cytokines causing bone lysis (da Costa et al. 2005).

The clinical course of LCH ranges from relatively benign to extremely aggressive and lethal, and single or multisystem involvement can occur. Bone is the most commonly involved organ in children with single-organ disease, and these patients present with unifocal or multifocal lytic bone lesions. Although the skull is the most common sight of involvement, any bone may be involved, and LCH is the most common cause of vertebra plana in children (Van De Graff and Cass 2003; Weitzman and Egeler 2008).

Multisystem disease, which occurs in roughly 50 % of patients with LCH, can involve any organ system and can be classified as high risk or low risk depending on the organs affected (Weitzman and Egeler 2008; Satter and High 2008). High-risk LCH makes up roughly 10 % of cases and includes disease in which two or more organs are involved including at least one “high-risk” organ (liver, spleen, lung, or hematopoietic system). Multisystem disease with involvement of any of these high-risk organ systems is associated with higher likelihood of relapse and mortality compared to low-risk and single-organ disease.

Roughly two-thirds of patients with LCH present before 10 years of age (Bonafe et al. 1994). Temporal bone involvement occurs in 15–61 %, and bilateral involvement is seen in approximately a third of cases (Van De Graff and Cass 2003; Saliba et al. 2008). Up to 25 % may present initially with otologic complaints. As with other tumors of the temporal bones, the clinical signs and symptoms of LCH are often nonspecific, and this may lead to a delay in diagnosis. The most common presenting symptoms are otitis media, otitis externa, or a temporal bone mass. Patients may also complain of otorrhea, otalgia, postauricular swelling, an aural polyp, or conductive sensorineural hearing loss. Labyrinthine and cranial nerve involvement are unusual although there have been isolated reports of patients presenting with facial nerve deficits, vertigo, or sensorineural hearing loss (Van De Graff and Cass 2003).

For localized osseous lesions, vigorous therapy is not generally indicated, and surgical curettage or low-dose radiation therapy often suffices. Treatment of multisystem disease is controversial, with high dose steroids, single-agent chemotherapy, and multi-agent chemotherapy each having their proponents (Satter and High 2008). Studies have shown that failure to respond to chemotherapy at 6 weeks is the single most important predictor of poor survival among patients with the disease (Weitzman and Egeler 2008).

On CT, LCH of the temporal bone manifests as a destructive lesion, usually involving the mastoid, with the squamous portion and the middle ear being less commonly affected sites. Petrous apex involvement is
relatively uncommon. Smaller structures such as the ossicles and otic capsule may be eroded; however, involvement of these structures is not seen as frequently as one might expect given the extensive destruction usually seen (Fernandez-Latorre et al. 2000) (Fig. 78). The presence of additional lytic calvarial lesions with well-defined beveled edges is highly suggestive of the diagnosis. On MRI, the lesions of LCH demonstrate high signal intensity on T2-weighted images and variable signal on T1-weighted images. The lesions enhance strongly with gadolinium administration, which helps distinguish LCH from cholesteatoma (Fig. 79). The lesions may be single or multiple and may occur bilaterally. Following appropriate treatment, the soft tissue mass resolves, and this is followed by reossification and remodeling of the affected bone.

Other Pediatric Temporal Bone Tumors
A variety of other tumors and tumor-like lesions have been reported to occur in the temporal bone in children, although these are for the most part exceedingly rare (Van De Graff and Cass 2003).
**Paraganglioma** Paragangliomas are the most common neoplasms of the middle ear in adults, with a peak incidence in the fourth decade of life, but are uncommon in the pediatric age group (Jacobs and Potsic 1994). The incidence of paragangliomas is increased in patients with von Hippel-Lindau disease and multiple endocrine neoplasia type II, but the tumors also occur sporadically in children, with isolated glomus tumors being described in children as young as 6 months of age (Jacobs and Potsic 1994; Boedeker et al. 2009). Compared to adult paragangliomas, glomus tumors in children tend to behave more aggressively, are more likely to secrete vasoactive substances, and are more likely to be multifocal. In addition, glomus tumors tend to be more difficult to diagnose clinically, as children are not as easily able to verbalize complaints, and the tumors, which classically appear as reddish retrotympanic masses on otoscopy, are often masked by serous otitis media (Jacobs and Potsic 1994; Magliulo et al. 1996).

In the temporal bone, paragangliomas typically arise from paraganglia in the middle ear along Jacobson’s nerve (glomus tympanicum) or in the jugular foramen along Arnold’s nerve (glomus jugulare). Their imaging features are similar to those of paragangliomas occurring in adults. Glomus tympanicum tumors appear as soft tissue masses situated along the cochlear promontory on CT, while glomus jugulare tumors are soft tissue masses that classically produce permeative bony erosion around the jugular foramen on CT. On MRI, these tumors, particularly when large, may demonstrate a characteristic “salt-and-pepper” appearance on T1-weighted images. The hypointense “pepper” represents high-velocity flow voids of feeding arterial branches in the tumor (also evident on T2-weighted images), while the less commonly seen hyperintense “salt” represents underlying foci of hemorrhage. Paragangliomas enhance avidly following contrast administration, which helps distinguish them from cholesteatomas in the middle ear (Razek and Huang 2012).

**Endolymphatic Sac Tumor** Endolymphatic sac tumors, which are low grade hypervascular tumors arising from the epithelium of the endolymphatic sac or duct, have been reported in a small number of children, both sporadically and in association with von Hippel-Lindau disease (Kupferman et al. 2004). On imaging, these tumors appear as destructive masses arising along the posterior wall of the petrous temporal bone. They often demonstrate speckled internal calcifications on CT. On MRI, most show foci of increased signal intensity on T1-weighted imaging – presumably due to previous hemorrhage – and enhancement is typical following gadolinium administration (Mukherji et al. 1997) (Fig. 80).

**Metastases** Most primary pediatric malignancies can metastasize to the temporal bone, with leukemia and neuroblastoma being the ones most likely to do so (Robson et al. 1998). In patients with leukemia, temporal bone involvement occurs in 16–35 % (Van De Graff and Cass 2003) and may present clinically in a variety of fashions, including auricular or EAC skin lesions, tympanic membrane thickening or redness, middle ear effusions, otitis media, hearing loss, mastoiditis, or facial nerve paralysis. On physical examination, there may be ulceration, hemorrhage, and diffuse mucosal thickening of the EAC and middle ear cavity. Perineural infiltration of the 7th and 8th cranial nerves also occurs, causing facial paralysis or hearing loss. MRI is the imaging modality of choice for evaluation of suspected cranial nerve involvement and demonstrates abnormal enhancement and thickening of the involved nerve along its cisternal or peripheral segments (Fig. 81). Rarely, leukemia may manifest as a focal destructive soft tissue mass in the temporal bone (referred to as a granulocytic sarcoma). In these cases, the radiographic findings are indistinguishable from other metastases and malignant temporal tumors and consist of an aggressive soft tissue mass causing lytic bone destruction (Kaufman et al. 1993).

**Exostoses (Surfer’s Ear)** Exostoses are benign bone outgrowths that occur along walls of the EAC, which primarily develop in individuals with repeated exposure to cold water submersion, such as surfers, swimmers, divers, kayakers, and sailors. In fact, the condition has been termed “surfer’s ear” due to its
high incidence among the surfing population. Although the actual pathogenesis of EAC exostoses is not known, repeated exposure of the ear to cold water is believed to play a role in their development, and these lesions seldom present before 10 years of age (Kroon et al. 2002; Nakanishi et al. 2011). Radiographically, exostoses present as smoothly bordered, broad-based bone overgrowths in the bony portion of the external auditory canal. They are usually multiple and bilateral (Fig. 82). The cortex of these lesions is continuous with the cortex of the adjacent EAC, and they are not associated with a soft tissue mass or bone destruction.

**Osteoma** Exostoses should be distinguished from rarer osteomas, which are benign bony neoplasms that can affect a wide range of age groups starting in the second decade of life. Osteomas are usually unilateral and classically appear as solitary, pedunculated, and calcified masses originating from the tympanosquamous or tympanomastoid suture line and extending into the EAC (Carbone and Nelson 2012). They may also occur within the middle ear cavity, where they may cause conductive hearing loss if they encroach upon the ossicles or tympanic membrane (Cho et al. 2005) (Fig. 83). Both osteomas and exostoses are usually discovered incidentally, but when they become large enough, they may cause symptoms such as recurrent cerumen impaction, otitis externa, or hearing loss.
Other Tumors  Ewing’s sarcoma is the second most primary bone malignancy in children, but only scattered cases have been reported to arise in the temporal bone, where the imaging features of these tumors are nonspecific (Kadar et al. 2010). Other temporal bone tumors more typically seen in adults, such as squamous cell carcinoma, adenoid cystic carcinoma, neurofibromas, and schwannomas, have rarely been reported in children (Van De Graff and Cass 2003). The latter two tumor types may be seen in patients with neurofibromatosis types 1 and 2 (Fig. 84), respectively, and should be considered in patients who present with retrocochlear hearing loss.

Fig. 81  Leukemia involving the temporal bone. Axial gadolinium-enhanced T1-weighted image in a child with leukemia demonstrates abnormal enhancement within both IACs (arrowheads) and along the geniculate and tympanic segments of both facial nerves (white arrows). There is also abnormal enhancement within both Meckel’s caves (black arrows), compatible with diffuse leukemic involvement of the cranial nerves.

Fig. 82  EAC exostoses (surfer’s ear). Axial CT image of the left temporal bone in a patient with a history of repeated cold water exposure demonstrates smoothly bordered, broad-based bone overgrowths along the anterior and posterior walls of the bony EAC (arrows) which are continuous with the underlying cortex.
Fibrous Dysplasia

Fibrous dysplasia (FD) is a disease characterized by slow, progressive replacement of normal bone by an abnormal proliferation of isomorphic fibrous tissue mixed with poorly formed, haphazardly arranged woven bone (Megerian et al. 1995). The disease is due to disordered osteoblastic activity which disrupts formation of mature lamellar bone, resulting in bone which is expanded, distorted, and structurally weakened (Swartz and Mukherji 2009). FD may involve a single bone (monostotic FD; 70–75 % of cases) or multiple bones (polyostotic FD). The polyostotic variety of FD may occur as a component of McCune-Albright syndrome, a genetic disorder consisting of the triad of polyostotic FD, precocious puberty, and cutaneous pigmentation anomalies (cafe-au-lait spots). In order of decreasing frequency, FD most commonly affects the ribs, femur, tibia, maxilla, calvarium, and humerus. Temporal bone
involvement is fairly uncommon and is usually monostotic, occurring in approximately 18% of cases in which there is craniofacial involvement (Megerian et al. 1995; Morrissey et al. 1997).

FD of the temporal bone is slightly more common in males than in females and usually presents in the first and second decades of life, although diagnosis of FD has been reported in children as young as 9 months of age (Megerian et al. 1995; Younus and Haleem 1987). The most commonly reported clinical manifestations of FD of the temporal bone are progressive stenosis of the external auditory canal and conductive hearing loss. EAC cholesteatomas develop in up to 40% of patients secondary to progressive EAC stenosis. Patients may also present with progressive painless swelling in the squamous or mastoid portions of the temporal bone, otorrhea, or trismus (likely due to temporomandibular joint involvement). Approximately 14% of individuals with temporal bone FD may demonstrate SNHL, most likely caused by involvement of the otic capsule or the IAC (Megerian et al. 1995). Facial nerve paralysis or paresis due to encroachment on the facial nerve canal occurs in approximately 9%. Rarely, craniofacial FD may undergo malignant transformation (estimated at 0.4% of cases), but to our knowledge, no reported cases have been associated with temporal bone FD (Morrissey et al. 1997).

Treatment of FD is based on the severity of the symptoms. Surgical intervention is limited to biopsy and for the relief of functional deficits or to address cosmetic deformity. Surgery may be performed to maintain a patent EAC, for removal of cholesteatoma, preservation of vestibular and cochlear function, or facial nerve decompression. Radiation therapy is generally not indicated due to a theoretically increased risk of malignant degeneration.

CT is the imaging modality of choice for evaluating patients with temporal bone FD and typically demonstrates enlargement of all or part of the bone, including the mastoid and petrous portions. Three classic radiographic patterns of FD (pagetoid, sclerotic, and cystic) were first described on plain radiographs by Fries in the 1950s and are now commonly used to describe the CT features of the disease.

Fig. 85 Fibrous dysplasia. (a, b) Axial CT images through the temporal bones in a 13-year-old with fibrous dysplasia demonstrate diffuse expansion of the left temporal and sphenoid bones, which demonstrate a characteristic “ground glass” appearance. Note the encroachment on the epitympanum (arrow, a) and narrowing of the left EAC (b).
The pagetoid pattern is the most common (56%) and is characterized by the classic “ground glass” pattern due to a mixture of dense and radiolucent areas of fibrosis (Fig. 85). The sclerotic form of FD (23%) is homogeneously dense, while the cystic form (21%) is characterized by a spherical or ovoid lucency surrounded by a dense bony shell.

On CT, one should be attentive to the presence and degree of EAC stenosis (Fig. 85); the presence of a soft tissue mass (potentially reflecting cholesteatoma); encroachment on the IAC, labyrinth, and facial nerve canal; and bony overgrowth into the temporomandibular fossa (Brown et al. 1995). Although it was previously thought that fibrous dysplasia spares the inner ear structures, involvement of the otic capsule, including the cochlea, vestibule, and vestibular aqueduct, can occur and likely contributes to SNHL in some patients with FD (Megerian et al. 1995; Brown et al. 1995). Middle ear involvement is uncommon and usually results from long-standing EAC stenosis with secondary cholesteatoma, although ossicular impingement due to direct middle ear encroachment can occur (Fig. 85).

The MRI characteristics of temporal bone FD are nonspecific and can be easily mistaken for tumor. The majority of the lesion is usually of low to intermediate signal intensity on all spin-echo pulse sequences (Fig. 86), but regions of high signal on both T1- and T2-weighted images can be seen. Enhancement following intravenous contrast administration is variable but is usually present. The presence of high signal areas or intense contrast enhancement may indicate increased clinical and pathologic activity (Casselman et al. 1993b).

**Otosclerosis**

Although otosclerosis most commonly presents in early adulthood, patients with the disease occasionally present with hearing loss in childhood or adolescence (Millman et al. 1996). Otosclerosis is a disorder affecting the bony labyrinth which is characterized by resorption of the normally dense middle
endochondral layer of bone of the otic capsule which is replaced by disorganized foci of Haversian bone. In the early or active phase of the disease (sometimes referred to as otospongiosis), there is formation of spongy, vascular, demineralized new bone, which is less dense than normal surrounding bone. Over time, these spongy foci become less vascular and calcify, resulting in sclerosis. This phase is referred to the inactive or otosclerotic phase (Virk et al. 2013; Sakai et al. 2011).

Otosclerosis has an autosomal dominant inheritance pattern with variable degrees of penetrance and expression. The disease has a prevalence of 0.3–0.4 %, and it affects females roughly twice as frequently as males (Virk et al. 2013; Sakai et al. 2011; Smith et al. 2003). Although numerous theories about the etiology of otosclerosis have been postulated, the true nature of the origin of the disease is not known. In addition to possible genetic factors, proposed explanations include endocrine factors, autoimmunity, and viral infections (Menger and Tange 2003). A number of potential candidate genes have been implicated, but current data are limited making it impossible to point to a single responsible gene (Bittermann et al. 2014). Defects in the COL1A1 gene, one of the genes associated with osteogenesis imperfecta (see below), have been reported to be associated with clinical otosclerosis (McKenna et al. 1998); however, this association is controversial, as some studies have reported no association, while others suggest a gender-related association between COL1A1 and otosclerosis (Kalafallah et al. 2011; Rodríguez et al. 2004). Infection with the measles virus has been proposed as a contributing factor in sporadic cases of otosclerosis, and electron microscopic and immunohistochemical studies have shown the presence of measles virus and proteins in active otosclerotic foci (Arnold and Friedmann 1987; McKenna and Mills 1989; McKenna et al. 1986). Furthermore, Niedemeyer et al. found a decrease in the incidence of otosclerosis in younger patients in Germany which appeared to coincide with the widespread introduction of measles vaccination, suggesting that measles infection may play a role in disease development in some patients (Niedermeyer et al. 2001).

Two forms of otosclerosis are typically described: fenestral and retrofenestral (or cochlear) types. The fenestral type of otosclerosis is characterized by lesions involving only the oval window region, usually along the anterior margin of the oval window niche (the location of the fissula ante fenestram) (Linthicum 1993). In the retrofenestral type of otosclerosis, there is demineralization of the cochlear capsule in addition to the typical oval window changes seen in fenestral otosclerosis. Oval window involvement is almost always evident, even in fenestral disease (Sakai et al. 2011).

Most patients with otosclerosis present during the third to fourth decades of life (Salomone et al. 2008); however, roughly 15 % of cases of clinical otosclerosis occur in patients younger than 18 years, and cases have been described in children younger than 6 years of age (Salomone et al. 2008; Robinson 1983). Hearing loss due to otosclerosis is typically conductive in nature, has an insidious onset, and is slowly progressive. Conductive hearing loss in otosclerosis is almost always secondary to fixation of the stapes footplate at the anterior oval window margin due to overgrowth of otosclerotic bone. Interestingly, subclinical otosclerosis detected by histologic temporal bone analysis is not uncommon, with foci of disease being evident in the temporal bones of 0.6 % of children younger than 5 years, 4 % of individuals between 5 and 19 years, and in 10 % of adults; however, less than 10 % of those with histologic findings of otosclerosis develop hearing loss as a result of stapes footplate fixation (Smith et al. 2003).

Sensorineural and mixed hearing loss occur less commonly (approximately 10 %) and are associated with the retrofenestral form of the disease (Robinson 1983; De la Cruz et al. 1999). In 70–80 % of patients, hearing loss is bilateral, but it is usually asymmetric (Smith et al. 2003; Menger and Tange 2003). The etiology of sensorineural hearing loss in otosclerosis is unclear, but proposed explanations include diffusion of cytotoxic enzymes into the perilymph, expansion of the otosclerotic focus into the endosteal layer of the otic capsule, changes in the spiral ligament and stria vascularis, and vascular shunts (Smith et al. 2003; Mafee et al. 1985a).
The diagnosis of otosclerosis is suggested by history, physical examination, audiologic evaluation, and imaging. Patients often complain of a hollow quality in their hearing with improved hearing in background noise. Tinnitus and vestibular symptoms may be present; however, these symptoms rarely appear prior to 9 years of age (Smith et al. 2003). On otoscopic exam, the cochlear promontory may have a faint pinkish hue (Schwartz’s sign), presumably reflecting the vascularity of the process. This finding is highly suggestive of otosclerosis but is only evident in 10% of cases (Nourollahian and Irani 2013). Audiograms demonstrate evidence of conductive hearing loss, and there may be a decrease in bone conduction at 2,000 Hz (a finding known as Carhart’s notch), which is also suggestive of the disease (Smith et al. 2003).

**Fig. 87** Fenestral otosclerosis. Axial CT image of the right temporal bone in a 13-year-old female with conductive hearing loss demonstrates a subtle focus of demineralized bone along the anterior margin of the oval window (*arrow*), consistent with spongiotic changes of otosclerosis.

**Fig. 88** Retrofenestral otosclerosis. Axial CT image of the left temporal bone in a young adult patient with progressive mixed hearing loss demonstrates subtle demineralization adjacent to the apex of the cochlea (*arrowhead*), compatible with early retrofenestral otosclerosis. Note as well the focus of fenestral otospongiotic bone along the anterior margin of the oval window (*arrow*).
Imaging can play a useful role in the diagnosis of otosclerosis, but may not be necessary in patients presenting conductive hearing loss with characteristic clinical and audiometric signs and disease limited to the oval window. High-resolution CT has been shown to have sensitivity and specificity of greater than 90% for diagnosing otosclerosis (Virk et al. 2013). In the fenestral form of the disease, CT will demonstrate a subtle focus of bone demineralization along the anterior margin of the oval window, representing abnormal spongiotic bone (Fig. 87). As the disease progresses, these lesions enlarge, protruding slightly into the middle ear cavity, and they may impinge upon or even obliterate the oval window. In the inactive sclerotic phase, the lesions may remineralize and become indistinguishable from the normal otic capsule, but subtle thickening of the stapes footplate may be evident (Mafee et al. 1985b; Sakai et al. 2000). On contrast-enhanced MRI, a small enhancing focus may be evident in the oval window niche during the active phase of the disease (Ziyeh et al. 1997).

In virtually all cases of cochlear otosclerosis, there will be evidence of fenestral involvement on CT. Cochlear otosclerosis will additionally demonstrate demineralization of the bone around the cochlea (Fig. 88), occasionally producing a lucent halo around the cochlea (the so-called “double ring” sign). The otospongiotic bone along the basal turn of the cochlea may also give the appearance of an extra or “fourth” cochlear turn (Mafee et al. 1985a). On MRI, the characteristic finding in cochlear otosclerosis is pericochlear enhancement, reflecting contrast pooling in active, vascular otospongiotic foci. Pericochlear otospongiotic lesions may demonstrate soft tissue signal on unenhanced T1-weighted images and high signal on T2-weighted images; however, frequently the otic capsule will appear normal on these unenhanced sequences (Sakai et al. 2000; Ziyeh et al. 1997).

Round window involvement has been reported to be present in 13% of patients with clinical otosclerosis and be seen in both fenestral and cochlear otosclerosis, although it is more common in cases of the latter (Mansour et al. 2011). Isolated round window involvement is rare. On CT, round window involvement by otosclerosis is evident by the presence of a demineralized plaque along the inferior bony edge of the round window, which may cause apparent thickening of the round window membrane, or even obliterate the round window recess. In the active phase of disease, these lesions may demonstrate enhancement on post-contrast T1-weighted MR images.

The standard treatment for conductive hearing loss due to footplate fixation from otosclerosis is stapedectomy with placement of a stapes replacement prosthesis. In adult patients with SNHL, sodium fluoride is recommended to prevent progression of hearing loss; however, in children the efficacy of sodium fluoride therapy is not established (Smith et al. 2003). Cochlear implantation can be considered; however, some authors believe that patients with extensive retrofenestral otosclerosis are at increased risk for facial nerve stimulation from the cochlear implant, presumably due to increased conductivity of otospongiotic bone (Merkus et al. 2011; Rotteveel et al. 2004).

**Osteogenesis Imperfecta**

Osteogenesis imperfecta (OI) is a connective tissue disorder characterized by development of brittle bones which are susceptible to fracture, leading to progressive bone deformity and growth deficiency (Forlino et al. 2011; Swinnen et al. 2013). The disease, which occurs with an incidence of approximately 1 in 15,000–20,000 births, is usually caused by an autosomal dominant mutation of either the COL1A1 or COL1A2 gene. These genes code for elements of type I collagen, which is an important constituent of bone, blood vessels, skin, joints, and tendons.

Four major types of OI have classically been described (Sillence et al. 1979). In these classic forms of the disease, which are inherited in an autosomal dominant fashion, a common bone defect is believed to occur downstream of the various specific collagen mutations associated with each subtype. This defect alters bone cell function and remodeling mechanisms which normally maintain bone homeostasis (Forlino et al. 2011). Type I OI (formerly osteogenesis imperfecta tarda) is the most common and mildest
form of the disease. This form of the disease has variable penetrance and is characterized by the triad of blue sclerae, multiple fractures occurring in childhood, and hearing loss. The fractures usually begin with ambulation and decrease with puberty. Type II OI (formerly osteogenesis imperfecta congenita) is lethal in the perinatal period due to extreme bone fragility which affects the respiratory tract and causes insufficient skull ossification. Affected newborns usually have short, bowed long bones with crumpling from in utero fractures, blue/gray sclerae, and a large soft cranium. Types III and IV OI have clinical manifestations of intermediate severity between those of types I and II. Type III OI (progressive deforming OI) is the most severe, nonlethal form of the disease. Patients with this type of OI may sustain hundreds of fractures, resulting in extremely short stature, progressive bone deformities, and scoliosis; most have platybasia or basilar invagination. Type IV OI is a moderately severe disease displaying a range of phenotypes that overlap types I and III. These patients may not demonstrate the blue sclerae that are typically observed in the other forms of OI.

Recently, a number of additional types of OI (types V–XI) have been described, with many of these being inherited in an autosomal recessive fashion. These types are usually caused by defects in genes whose products interact with type I collagen. Recessive forms of OI can have variable clinical severity ranging from moderate to lethal disease (Forlino et al. 2011).

Regardless of the underlying phenotype, approximately 50 % of patients with OI develop conductive hearing loss, usually between the second and fourth decades of life, with approximately 20 % developing hearing loss by age 5 (Smith et al. 2003; Swinnen et al. 2013). Patients may present with conductive, sensorineural, or mixed hearing loss. Conductive hearing loss usually presents by the second to third decade of life and is caused by structural changes in the ossicles. The incus and stapes become thin and fragile and are susceptible to pathologic fractures, resorption, and subluxation. The stapes is the most frequently affected ossicle in OI, and in addition to fractures, patients can develop marked thickening of the footplate causing mild ankylosis at the oval window. Bony changes of the otic capsule can also encroach on the stapes footplate, causing a picture of conductive hearing loss similar to what is observed in patients with otosclerosis (Swinnen et al. 2013; Armstrong 1984). In fact, it has been suggested that there may be a relationship between otosclerosis and mild OI, both of which may be associated with mutations of the COL1A1 gene (McKenna et al. 1998). However, the two diseases are histologically different, with OI involving all three layers of the otic capsule (endosteum, endochondral layer, and periosteum), while otosclerosis is limited to the endochondral layer (Nager 1988; Berger et al. 1985).

Sensorineural hearing loss may be caused by encroachment on the cochlea by reparative tissue or by hemorrhages or microfractures involving the otic capsule (Shapiro et al. 1982). The treatment of hearing
loss in OI is mainly symptomatic and may include amplification or stapedectomy (Forlino et al. 2011). Patients presenting with SNHL may be considered for cochlear implantation.

CT is the imaging modality of choice for imaging patients with otologic manifestations of OI. OI results in diffuse demineralization of the otic capsule that may extend to the petrous apex. The demineralization seen in OI is similar to that seen in otosclerosis. In fact, in nearly 80% of cases of OI, fenestral changes are present manifesting as hypodense bony plaques situated along the anterior margin of the oval window (Swinnen et al. 2013). The stapes footplate is often thickened and hypodense, and ossicular dislocations or fractures may be evident. In addition, the oval and round windows may be obliterated by proliferating abnormal bone. Retrofenestral lucencies, appearing as lucency surrounding the labyrinthine structures, are present in slightly over 60% of cases. These findings have been described as a “double ring sign” or “fourth” cochlear turn (Fig. 89). A hypodense focus anteroinferior to the fundus of the IAC can be seen in nearly 50% of cases, and demineralization can also be seen around the semicircular canals and about the facial nerve canal.

MRI with contrast may demonstrate enhancement in the fundus of the IAC or of the bone surrounding the cochlea, vestibule, or semicircular canals, which may indicate the presence of active otospongiotic lesions (Swinnen et al. 2013; Heimert et al. 2002).

Osteopetrosis

Osteopetrosis is a rare hereditary sclerosing bone dysplasia characterized by a progressive increase in bone mass and density which leads to formation of dense, brittle bone. The disease has an estimated incidence of approximately 1:250,000, with two major forms of the disease recognized based on their mode of inheritance (Sobacchi et al. 2013). Autosomal dominant osteopetrosis (ADO, formerly Albers-Schönberg disease) is typically a more benign, adult-onset form of the disease and will not be discussed here. Autosomal recessive osteopetrosis (ARO), also known as malignant infantile osteopetrosis, is a more severe form of the disease, which presents shortly after birth and may lead to death if left untreated. Mutations to seven different genes have been identified as causing ARO, with phenotypic manifestations and severity varying depending upon the specific gene defect. Mutations in TCIRG1 and CLCN7 together account for approximately 70% of cases of ARO, with the former gene being involved in 50–55% and the latter in roughly 10–15%. These genes encode proteins that are critically involved in osteoclast function, and mutations in these genes lead to an osteoclast-rich form of osteopetrosis in which osteoclasts are abundant but nonfunctional. Rarer gene mutations affecting osteoclast differentiation have also been described. In these subtypes, osteoclasts are absent or reduced in number (Sobacchi et al. 2013; Ihde et al. 2011).

Regardless of the gene mutation involved, the common end result is decreased osteoclast activity leading to inability to resorb bone or calcified cartilage. Osteoblast activity is therefore unchecked, leading to excessive deposition of new, immature osteoid and formation of thick sclerotic fragile bones that are prone to fracture. Furthermore, in the more severe malignant recessive forms of osteopetrosis, obliteration of the marrow containing spaces can occur, leading to bone marrow failure. As a result, patients may suffer from anemia, thrombocytopenia, and recurrent infections, and they frequently develop compensatory extramedullary hematopoiesis and splenomegaly. Neurologic changes are also common and are most likely the consequence of inadequate formation of the cranial foramina and increased intracranial pressure, which may lead to hydrocephalus, Chiari I malformation, cranial nerve compression, visual loss, or respiratory and eating difficulties (Sobacchi et al. 2013). Most patients with malignant osteopetrosis present in infancy with severe anemia or early visual problems secondary to optic nerve compression (Dozier et al. 2005).

Otologic manifestations of childhood osteopetrosis include both conductive and sensorineural hearing loss, otitis media, vertigo, and facial nerve palsy (Dozier et al. 2005). Roughly a quarter of patients with
ARO demonstrate hearing loss during the first year of life, and the majority of children will eventually develop hearing deficits. In those children with hearing loss, 100% have a conductive component, while 26% will have an additional sensorineural component. Conductive hearing loss may be secondary to ossicular involvement and ankylosis or overgrowth of the bony margins of the epitympanum. EAC stenosis also occurs due to bony overgrowth. Sensorineural hearing loss, when present, is thought to be due to cochlear nerve compression caused by bone encroachment on the IAC (Stocks et al. 1998; Steward 2003).

Currently, the treatment of choice for ARO is hematopoietic stem cell transplantation, which can reverse many of the bone manifestations of the disease; however, a subgroup of patients develop progressive neurodegenerative disease which cannot be reversed by stem cell transplantation (Castellano Chiodo et al. 2007). Treatment of hearing loss is typically supportive, but surgery may be required to treat complications such as optic or facial nerve compression and EAC stenosis (Dozier et al. 2005).

CT is the imaging modality of choice for imaging patients suspected of having osteopetrosis, although patients with the milder forms of disease studied early in life may have few imaging findings. Characteristic findings include diffuse thickening and sclerosis of the calvarium and temporal bones with poor pneumatization of the middle ear (Fig. 90). The mastoid air cells are also poorly pneumatized, and the ossicles are often sclerotic and may be enlarged. The subarachnoid fossae are typically enlarged, which may persist into later childhood, resulting in a fetal appearance of the bone that persists past infancy (Castellano Chiodo et al. 2007; Elster et al. 1992). Narrowing of the EAC, eustachian tube, IAC, and petrous carotid canal are also observed (Dozier et al. 2005). Other diseases to consider in the differential for diffuse sclerosis of the temporal bone in a child include sclerotic fibrous dysplasia, hyperparathyroidism, progressive diaphyseal dysplasia (Camurati-Engelmann-Ribbing syndrome), metaphyseal dysplasia (Pyle’s disease), cranio-metaphyseal dysplasia, craniodiaphyseal dysplasia, juvenile Paget’s disease, pyknody sostosis, and osteopathia striata (Voorhoeve’s syndrome) (Sakai et al. 2011). On MRI, findings in osteopetrosis include generalized bone thickening and sclerosis with obliteration of the normal marrow containing medullary spaces in the calvarium and skull base.
References


Murugasu E, Yong TT, Yoon CP. Invasive middle ear cholesterol granuloma involving the basal turn of the cochlea with profound sensorineural hearing loss. Otol Neurotol. 2004;25:231–5.


