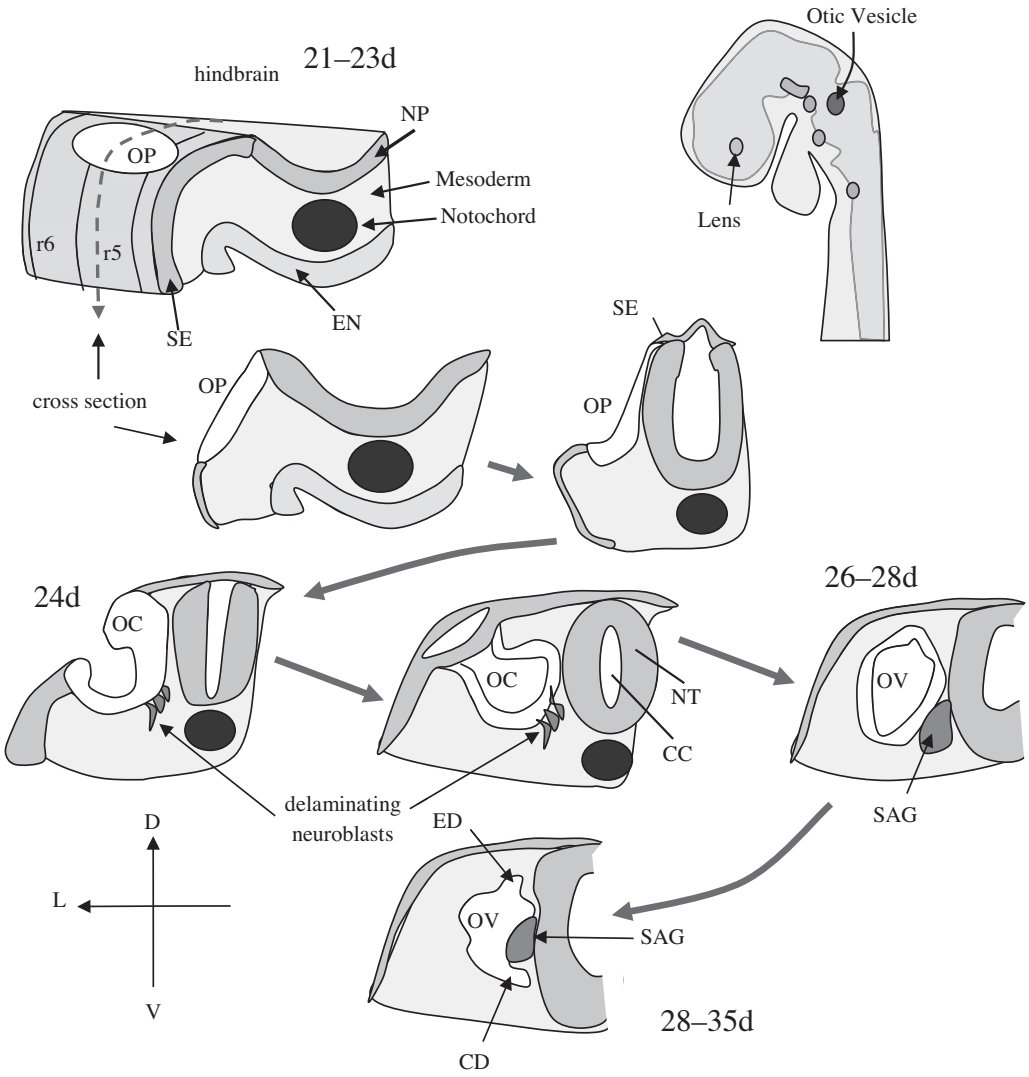


Figure 2–2. Formation of the otic vesicle (OV) from the otic placode (OP) over a period from approximately 3 to 4 weeks in the human embryo (follow arrows: 21–23 to 28–35 days post fertilization. [E8.5 to just beyond E10.5 in mouse]). d = days post fertilization. ED = endolymphatic duct; EN = endoderm; CC = central canal; CD = cochlear duct; D = dorsal; L = lateral; Mesoderm = mesenchyme presumptive mesoderm; NP = neural plate; NT = neural tube; OC = otic cup; OP = otic placode; OV = otic vesicle; r5-r6 = rhombomeres 5 and 6; SAG = statoacoustic ganglion; SE = surface ectoderm (epidermis); V = ventral. From *Genetics, Embryology, and Development of Auditory and Vestibular Systems* (2011) (p. 158) by Sherri M. Jones and Timothy A. Jones.

FORMATION OF THE OTIC PLACODE AND OTIC VESICLE

The inner ear is formed from primordial ectodermal cells near neuromeres (rhombomeres) 5 and 6 in the embryonic hindbrain. These cells form a flat, thickened patch of ectoderm called the otic placode during week 3 (see Figure 2–2). The otic placode appears morphologically as the first step in the formation of the inner ear. A variety of genetic signals define a region of tissue committed to form the otic placode, and subsequent molecular signaling leads to the separation of the otic placode from epidermal tissue (reviewed by Ohyama, Groves, & Martin [2007]). According to Groves (2005), the cells ultimately forming the placode are not simply

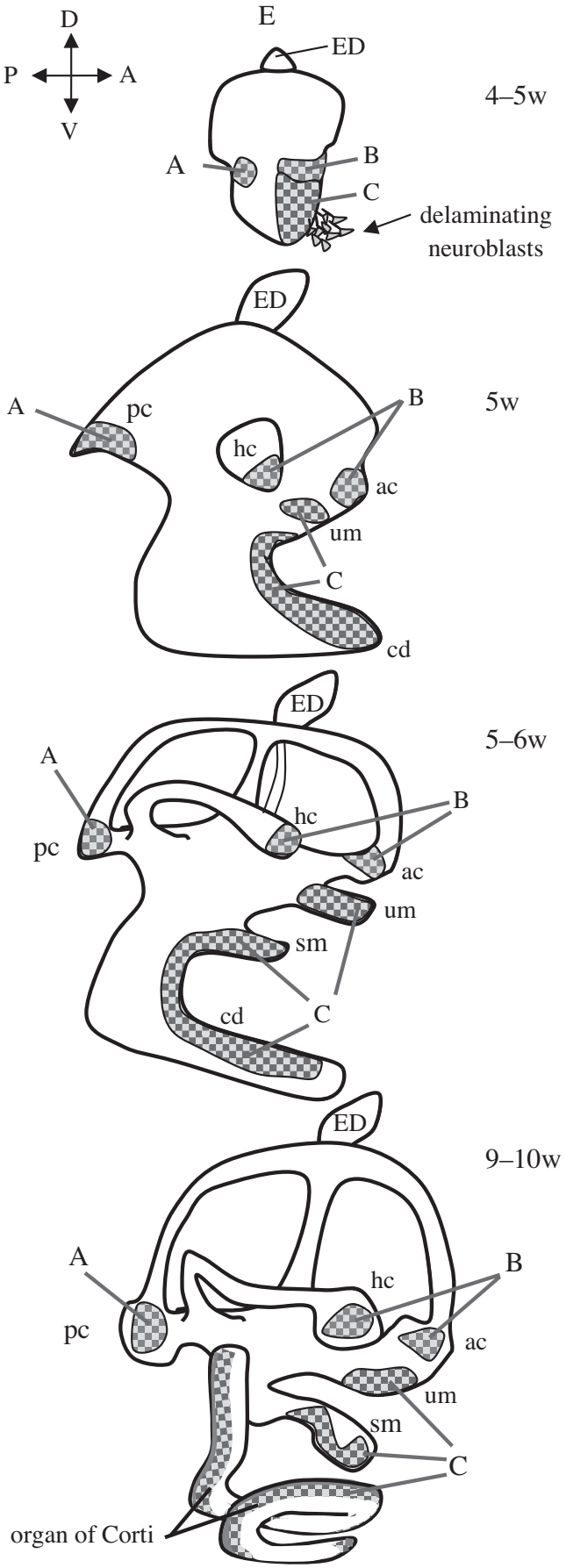


Figure 2–3. Lateral views of locations for important markers of prosensory and sensory domains during development. Regions forming the most dorsal sensory epithelia (cristae of semicircular canals) are distinguished early by the expression of particular genes (labels A and B), whereas the more ventral regions are marked with a different combination of signals (labeled C). The ventral prosensory domains elaborate the sensory epithelia of the utricular macula (um), saccular macula (sm) and cochlear duct (cd) in that order. ac = anterior crista. hc = horizontal (lateral) crista. pc = posterior crista. Schematics are based on data from the mouse. Corresponding ages in human are given (weeks post fertilization). Adapted from Bok, Chang, and Wu, 2007 with permission from *International Journal of Developmental Biology, 51*, 526.

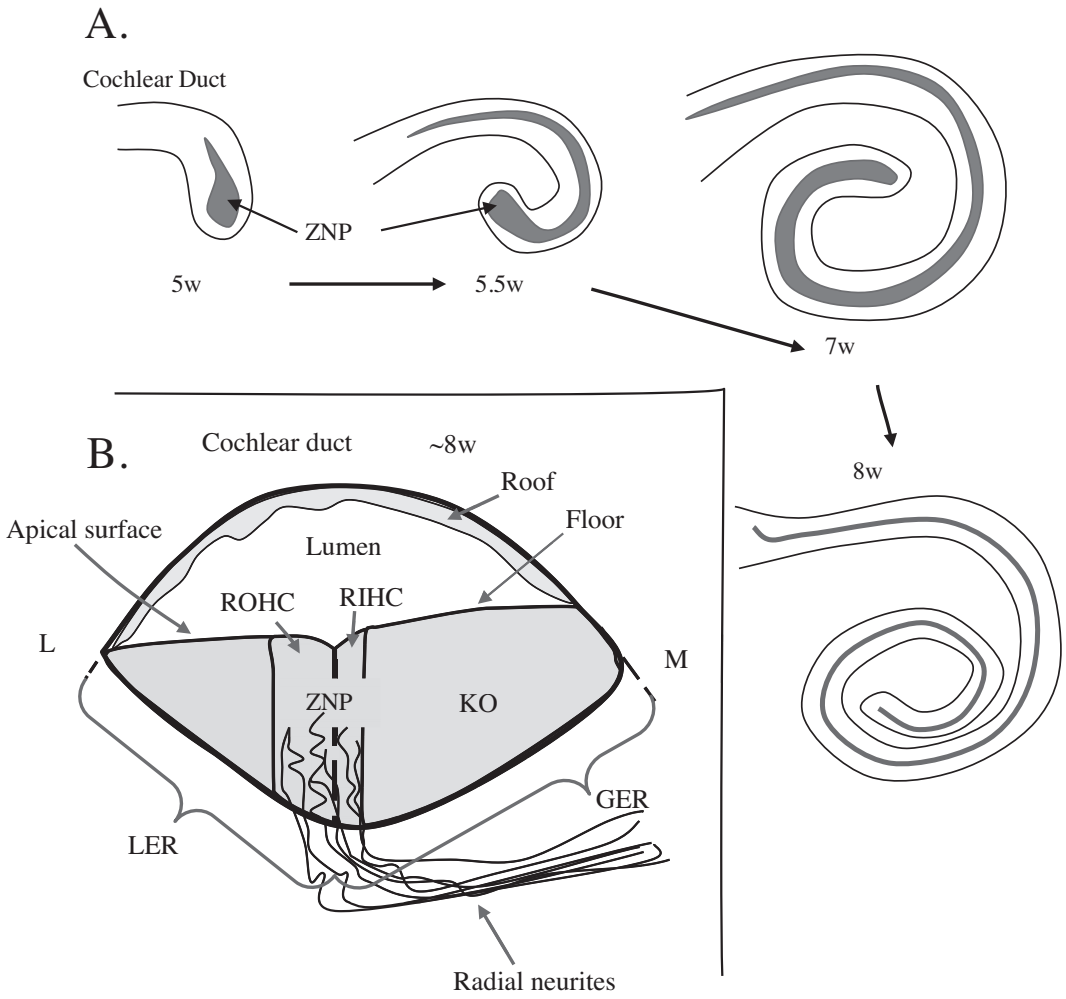


Figure 2–4. A. Formation of the zone of non-proliferation (ZNP, marked gray) in the cochlear duct at four periods of development, based on studies in the mouse model (e.g., Chen et al., 2002, Lee et al., 2006). Corresponding estimated ages for the human are shown. The ZNP appears first in the apex and then sweeps towards the base over the next week (48 hours in mouse). The formation of ZNP is completed by approximately week 6 (E14 mouse, Chen et al., 2002, Lee et al., 2006), however, the cochlea continues to elongate and coil until weeks 9 to 10 (P0, mouse). During this latter period of elongation, the sensory epithelium thins and narrows. **B.** Schematic of the cochlear duct circa week 6 (E14 mouse). According to Lim and Rueda (1992), the greater epithelial ridge (GER) incorporates the region of inner hair cells, whereas the lesser epithelial ridge (LER) incorporates the region where outer hair cells will form. Together they form the floor of the early cochlear duct, also called the epithelial ridge. The zone of non-proliferation identifies the epithelial region of prospective inner and outer hair cells. Kolliker's organ (KO) has been defined a number of ways. Here, Kolliker's organ includes only that portion of the GER that does not include prospective sensory hair cells. RIHC = region of inner hair cells. ROHC = region of outer hair cells. From *Genetics, Embryology, and Development of Auditory and Vestibular Systems* (2011) (pp. 164, 166) by Sherri M. Jones and Timothy A. Jones.

2006). Mutations in, or knock outs of, core PCP genes (or related signaling pathways) prevent elongation and result in a shortened cochlea. The extent to which vestibular sensors depend on the convergent extension process is not clear. However, like the cochlea, the vestibular epithelia start out as a layered partition of

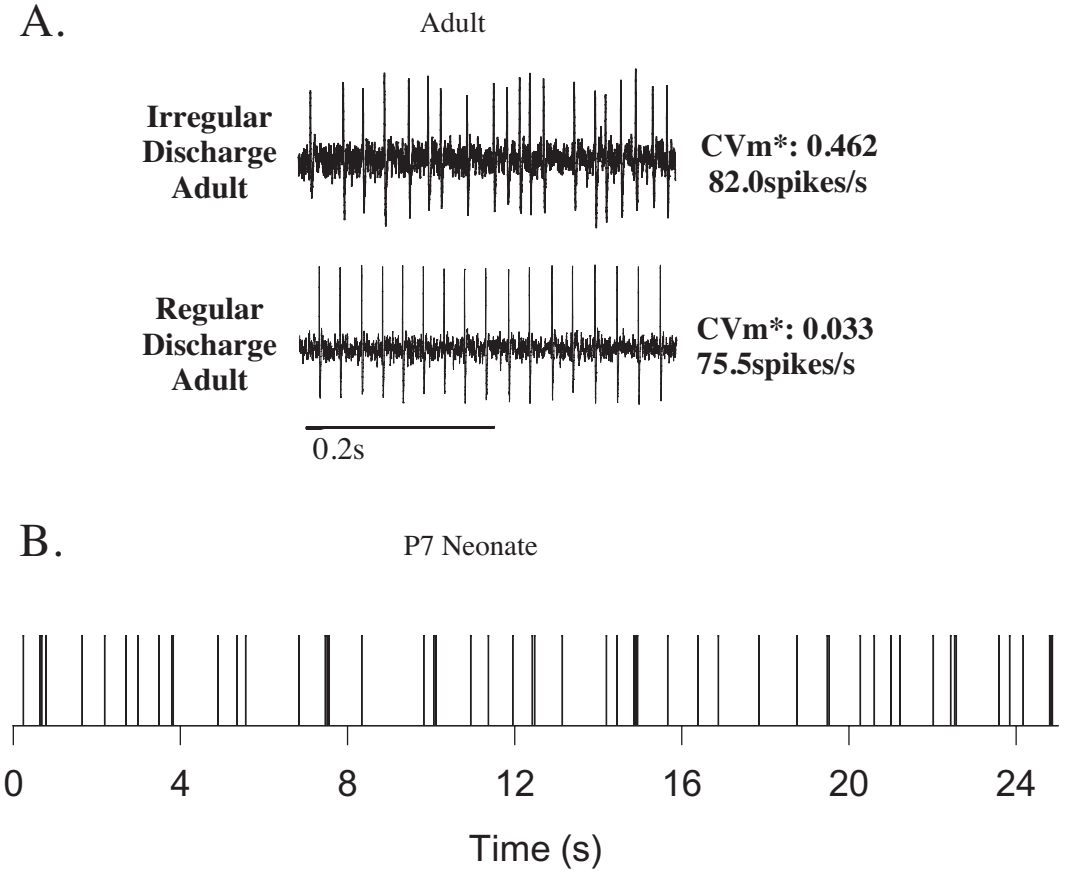


Figure 2–7. Spontaneous discharge activity of vestibular primary afferent neurons in mice. **A.** Adult mice: Each voltage spike represents an individual action potential. No stimulus is presented. These cells were chosen for illustration because they have similar high discharge rates. Two types of activity patterns are recognized. Irregular discharge (top tracing) is characterized by irregular spacing between spikes, and a high CV. A regular spike discharge pattern tends to show regular spacing between spikes and a low CV. Modified from Jones, Jones, et al. (2008). **B.** Spontaneous spike train of primary afferent neuron recorded from the superior vestibular nerve in the neonatal mouse at P7 (corresponding human age: 10 to 12 weeks). Note the time scale difference in B. This record was made over a period of approximately 24 seconds. Each vertical "spike" represents the onset time of the neural spike discharge during this portion of the recording. Discharge rate is slow and an irregular firing pattern (with high CV) typical for neonatal vestibular neurons is apparent. Unpublished data. CVm* = indicates that CV values were normalized for spontaneous rate based on mouse data. From *Genetics, Embryology, and Development of Auditory and Vestibular Systems* (2011) (p. 219) by Sherri M. Jones and Timothy A. Jones.

from vestibular sensors plays an important role in nervous system function.

Spontaneous vestibular discharge patterns are of two types in mammals and birds: regular and irregular. Just how regular or irregular the neural discharge is depends in part on the nature of the dendritic synaptic termination and on the nature of the membrane channels resident in the neuron (Eatock, Xue, & Kalluri, 2008; Iwasaki, Chihara, Komuta, Ito, & Sahara, 2008; Kalluri, Xue, & Eatock, 2010). The regularity can vary widely and this variety can be quantified using a single number called the coefficient of variation (CV = standard deviation/mean spike interval). The value of the CV normally varies between 0 and 1. The CV approaches 1.0 as the discharge pattern becomes more irregular (more stochastic, to be precise), whereas the CV approaches 0.0 as the discharge becomes more regular. Figure 2–7 illustrates the discharge patterns of reg-

ular and irregular vestibular afferents in mature (Figure 2–7A) and developing (Figure 2–7B, P7 neonate) mice. Discharge rates in mature mice range from less than 10 to over 140 spikes/sec, with most between 55 and 110 spikes/sec (e.g., Jones, Jones, et al., 2008). The recordings of Figure 2–7B were made in vivo from primary afferent neurons of the superior vestibular nerve (Jones & Jones, 2011). The discharge pattern of the neonate reflects a relatively low discharge rate (~8 spikes/sec) with irregular discharge timing (CV = 0.66).

Recording the activity patterns of individual vestibular primary afferent neurons in intact animals (i.e., *in vivo*) is challenging. Some investigators have instead explored the use of vestibular explant preparations. In this case, the labyrinth with the ganglia are removed and maintained in a physiological solution. Vestibular primary afferents recorded in a mouse inner ear explant preparation are also spontaneously active (Desmadryl, Raymond, & Sans, 1986). Afferent discharge patterns in explants have been measured on different postnatal days. Mean spontaneous discharge rates were low initially (day of birth, P0: 5 to 10 spikes/sec) and all neurons displayed irregular activity. Remarkably, regular discharge patterns were found at P1 and older. Beginning between P6 and P8, discharge rates increase dramatically (>80 spikes/sec), and the proportion of regular fibers increase as well. In a similar preparation in the chicken, Galicia, Cotes, and Galindo (2010) reported irregular spontaneous discharge rates on the order of 40 spikes/sec and CVs above 1.0 in recordings as early as five days before birth (E15). Many of the patterns found in the in vitro preparation were similar to those reported for in vivo studies (discussed below). In vitro studies provide many practical advantages while at the same time raising the question of whether neurons behave the same when studied in their natural environment, that is, in vivo.

Spontaneous discharge patterns of horizontal canal neurons have also been recorded in vivo in the neonatal rat from age P1 to P20 (Curthoys, 1983). Discharge rates were low at the youngest ages (<10 spikes/sec, P1 to P3) and all neurons exhibited irregular spontaneous discharge patterns. The first regular fibers were seen on P4. Discharge rates and the proportion of regular fibers increased substantially after P10 (30 to 40 spikes/sec). Nonetheless, rates for regular cells were still somewhat below the adult values at P20. Romand and Dauzat (1982) reported similar findings in the cat except that regular spontaneous discharge patterns were seen as early as P1. An example of an irregularly discharging macular primary afferent at P7 is shown in Figure 2–7B (mouse in vivo recording). In vivo recordings in the chick embryo at E19 showed that regular discharge patterns were present, and on average the rate for embryos was 22 spikes/sec versus 60 spikes/sec for post-hatch animals (Jones & Jones, 2000b). Note that discharge rates were somewhat lower in the in vivo recordings compared with explants. This may reflect the effects of anesthesia used in in vivo preparations or conditions associated with in vivo preparations. In summary, in the rodent at birth, spontaneous activity is immature, showing low discharge rates and being dominated by irregular firing patterns. Maturation of discharge patterns progresses over a period of weeks.

One role of the horizontal canal is to detect head turning and to activate vestibular neurons to send a signal to the brainstem that produces eye movements that compensate for head motion and maintain gaze on a stationary visual target. This is known as the vestibulo-ocular reflex (VOR). One question to ask is: When are primary afferents capable of delivering a compensatory signal comparable to that of the mature system? Curthoys (1983) evaluated this question by measuring the response of horizontal canal primary afferent neurons to head rotation in rats at ages from P1 to P20. At P1 neural responses were sluggish and highly variable. By P6 to P8, the neural response gain approached that of the adult. This suggests that, for some stimuli, the neuroepithelium at P8 is capable of generating signals comparable to those of an adult. These findings are consistent with results noted above showing that morphological and electrophysiological features of hair cell function (e.g., calyx, membrane channels, vestibular efferents) emerge during the first two to three postnatal weeks in rodents (weeks 13 to 23 in humans). Indeed, VOR gains in juvenile mice (P21 to P26) are slightly, but significantly, lower than those in mature animals at 3 to 4 months old (Faulstich et al., 2004). Central improvements likely mediate the final maturation changes. Similar maturational changes are reported for children (Casselbrant et al., 2010).

The basic structural elements of the macula are in place in the chick before hatch (by E16). The question is, when does macular function emerge? Little information is available regarding macular functional development in any species. Recordings of macular vestibular evoked potentials (VsEPs) have been made in embryos and hatchling chicks (E18 to P22; Jones & Jones, 2000a). Responses were obtained as early as E19 (1 to 2 days before hatch). Therefore, the onset of macular function occurs at least by E19 in the chick and likely earlier. Macular response thresholds decreased rapidly to approach adult values within days of hatch. Other response characteristics also matured systematically over similar periods (response latencies shortened and amplitudes increased). These findings, and the fact

that chicks are able to walk and run within hours of hatching, show that in the chick the vestibular periphery matures early and is functional even before hatch.

DEVELOPMENTAL MILESTONES OF VESTIBULAR FUNCTION AND BALANCE BEHAVIORS IN THE HUMAN

By the twelfth to fourteenth weeks of gestation, the human vestibular epithelium appears almost mature and contains types I and II hair cells, myelinated calyx-bearing and bouton-terminated afferent neurons, as well as efferent axon terminals (Dechesne, 1992). Ten weeks later (week 24) they are considered adult-like, and at birth, some 10 to 16 weeks later (weeks 36 to 40), the periphery is considered mature. Although the vestibular periphery may be mature at birth in the human, this is not readily apparent from human behavior (unlike the case in the chick above). The human neonate requires years (see below) to acquire a mature vestibular-linked behavioral repertoire. It is clear that central myelination and circuit refinements must take place in order to fully equip the otherwise largely immature cortical, cerebellar, extrapyramidal, and brainstem descending motor control systems for the support of mature behavioral motor programs and postures. Behaviors such as walking and running as well as mature posturing must develop after birth in the human (e.g., sitting up, rolling over, even holding one's head up; see Table 2–2; Eviatar & Eviatar, 1978; Frankenburg & Dodds, 1967). Table 2–2 summarizes well-known developmental milestones for motor skills in the human infant. Central immaturities therefore can obscure or mask true vestibular functional capability as well as deficits in neonates and children. Moreover, failure to recognize vestibular deficits may, in the absence of intervention, put individuals at risk for developing abnormal postural, dynamic motor control and/or motor effector systems (e.g., De Kegel et al., 2012; Fife et al., 2000; Shall, 2009; Van Cleave & Shall, 2006)

A functional vestibular periphery is evident in the human neonate. Full-term normal infants typically demonstrate vestibular mediated ocular compensation induced by head rotation (Cyr et al., 1985; Donat et al., 1980; Eviatar & Eviatar, 1978; Eviatar et al., 1974, 1979; Ornitz et al., 1979; Staller, 1986; Tibbling, 1969; Wiener-Vacher et al., 1996). Head rotation elicits a bilateral response from the vestibular periphery. VOR measurements at the youngest ages generally show tonic conjugate deviation of the eyes in a direction opposite that of rotation, and may include occasional rapid saccades. The slow compensatory movement is dependent on the vestibular periphery, whereas saccades depend on brainstem circuitry (see Chapters 3 and 4). A reduced number or frequency of saccades reflects the immature state of brainstem neural circuits. The presence of saccades in the neonate is in part dependent on frequency of rotation. Saccades may be absent in very young or premature infants but are found consistently in normal 1- to 3-month-old babies.

Caloric testing in the neonate evaluates each ear separately and has often provided evidence of central immaturity, especially in premature infants (Eviatar et al., 1974, 1979; Donat et al., 1980). VOR responses to calorics are variable and may be absent in infants less than 6 months. By 3 to 6 months a consistent VOR is generally present (Cyr et al., 1985, Eviatar et al., 1974, 1979; Donat et al., 1980). The absence of a VOR at

Table 2–2. Familiar Developmental Milestones for Motor Skills in the Human Infant*

Motor Skill	25% (mo)	50% (mo)	75% (mo)	90% (mo)
Lifts head (prone)				0.7
Lifts head 90 degrees (prone)	1.3	2.2	2.6	3.2
Chest up, arm support (prone)	2.0	3.0	3.5	4.3
Sits (head steady)	4.8	5.5	6.5	7.8
Rolls over	2.3	2.8	3.8	4.7
Stands holding on	5.0	5.8	8.5	10.0
Walks well	11.3	12.1	13.5	14.3

*Selected data from Frankenberg and Dodds (1967). Age (months) at which the respective fraction (25%, 50%, 75%, and 90%) of infants successfully completed motor tasks listed.

10 months or older is considered abnormal (Fife et al., 2000). Most abnormal responses (e.g., disconjugate eye movements, tonic ocular deviation, absent saccades) were found in premature or very young normal term babies (Donat et al., 1980; Tibbling, 1969), but such abnormalities generally reflected central immaturities which disappeared with maturation. Thus, improvements in vestibular responses largely represent central maturational processes which occur over several years.

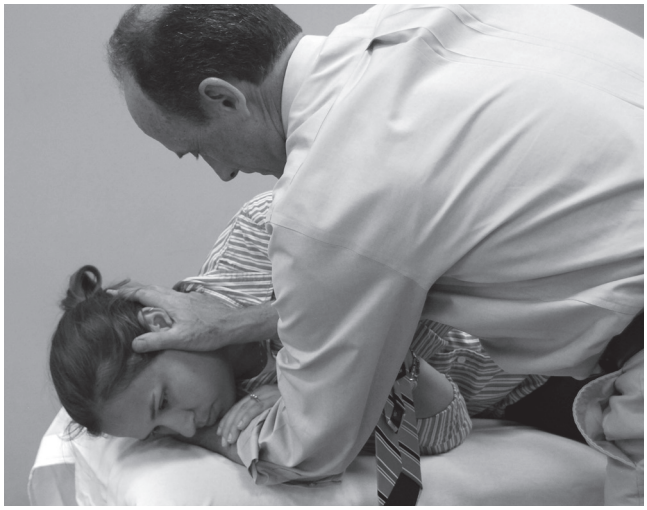
Vestibular reflex testing and posturography also have been used to assess the time course of maturation in children and demonstrate that the process begins very early in the neonate and continues into adolescence. There is considerable variation in the VOR findings for some parameters in growing children. Results depend on the particular metric used to characterize eye movements and the range of subject ages represented. Measurements can also be affected by the subjects' state of alertness. The ability to arouse and maintain alertness throughout testing is itself a function of age and this can influence results.

Using Barany rotary chair testing, a number of investigators have reported a decrease in velocity of the VOR slow component with maturation (Ornitz et al., 1979; Tibbling, 1969; Wiener-Vacher et al., 1996). Maturational changes appeared to be larger during the first year after birth and change more slowly thereafter into adolescence. Although these results outline a temporal profile, it is difficult to relate the findings to specific components of the vestibular ocular reflex.

VOR gain provides better insight regarding the effectiveness of vestibular reflex compensation for head motion. VOR gain reportedly increased linearly in a group of 120 children studied longitudinally over the period from 3 to 9 years of age (e.g., mean gains, 3 years: 0.58 to 9 years: 0.82, for 0.5 Hz rotations; Casselbrant et al., 2010), whereas thereafter from young adolescence to the adult, gain reportedly may decrease slightly at some frequencies (Herman et al., 1982; Valente, 2007). The findings of Charpiot et al. (2010) also indicated a decreasing gain for a slightly older group of children (6 to 12 years old). Others found little difference in VOR gain over ages studied (7 to 12 years old; Horak et al., 1988; 3 months to 6 years: Cyr et al., 1985). However, the range of ages and frequencies studied were somewhat different and sample sizes were smaller in the latter cases, which may in part account for differences in findings. At any given age the variance for VOR gain in humans is quite large (e.g., Peterka et al., 1990a, 1990b). Nonetheless, there is support for the hypothesis that a slow systematic maturation of VOR gain (increasing mean) occurs in the young child through preadolescence and small reductions in mean gain from the second to third decade of life. In addition to that cited above, evidence for this comes from a large cross-sectional study of 261 individuals, ages 7 to 81 years, which indicates a similar temporal profile where VOR gains appear highest for ages 7 to 20 years and then decrease somewhat and remain relatively stable over the ages from 30 to 81 years (Peterka et al., 1990a, 1990b). Changes in VOR gain are small relative to the variance in any case. Controversy persists in the literature regarding maturation over ages from approximately 10 to 30 years. In the future, it will be important to use standardized methods and metrics including standard frequencies and rotational velocities to minimize variability across studies. It would be helpful to see large longitudinal studies providing normative data for VOR in children and adults through the third decade of life. Such studies, although difficult to achieve, would serve to clarify the profile of change in the VOR over the human lifetime.

Computerized dynamic posturography (CDP) includes a sensory organization test (SOT), which provides a measure of how effectively sensory information is used by an individual to maintain balance and stability under dynamic postural challenge (see Chapter 15). SOT examines how one uses visual, somatosensory, and vestibular information to maintain balance and quantifies, among other things, the amount of sway exhibited. Several investigators have used the SOT in children between the ages of 3 and 15 years to characterize changes in balance skills with maturation and to develop normative data on sensory weighting strategies (e.g., Casselbrant et al., 2010; Charpiot, Tringali, Ionescu, Vital-Durand, & Ferber-Viart, 2010; Hirabayashi & Iwasaki, 1995; Peterka & Black, 1990; Peterson et al., 2006; Valente, 2007). The evidence suggests a steady improvement on vestibular tasks that continues into late childhood and by some reports into adolescent years when scores approached those of adults. In general, skills in the use of vestibular sensory input were the last to mature relative to use of somatosensory and visual cues for maintaining balance.

In summary, whereas the peripheral vestibular apparatus is essentially mature at birth, central maturation continues into adolescence. The VOR can be elicited in the neonate and it matures substantially over the first three years. Improvement generally continues into adolescence when adult balance skills are achieved. Although progress in our understanding has been made, it should be clear that we are only meagerly informed about the complex multisensory process of maturation in human postural balance and vestibular reflex systems. Our understanding of human vestibular development would benefit substantially from



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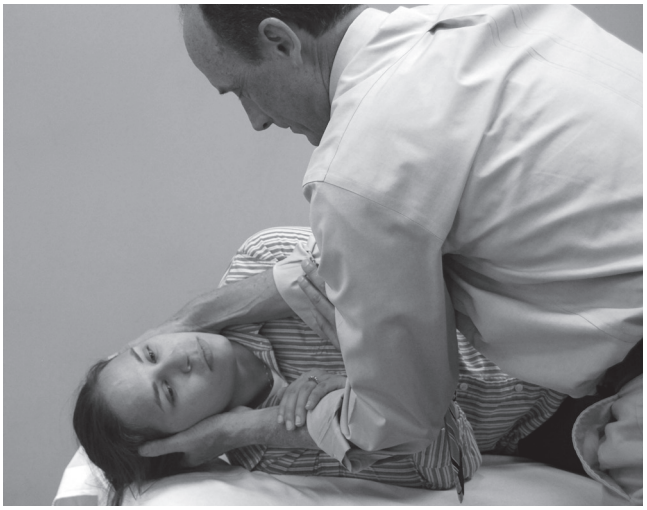

D

E

Figure 11–6. *continued* **C.** The clinician, along with assistance from the patient, rapidly moves the patient from left side lying to right side lying, maintaining the rightward neck rotation throughout the movement. There is a sudden deceleration of the patient's head in the clinician's hands as the patient's shoulder and the clinician's forearm hit the treatment table. The patient should experience vertigo at this point. If not, the clinician can rotate the patient's neck 90 degrees to the left (**D**) and then rapidly back down towards the treatment table (**E**). The original description of the treatment called for the patient to remain in this position for 5 minutes after the cessation of the vertigo and nystagmus. In clinical practice today, the patient typically remains in this position for 2 minutes after the vertigo and nystagmus stop. The clinician will then guide the patient back to a seated position, maintaining the cervical rotation until the patient is upright. Note that this treatment may be performed with the clinician standing behind the patient.

side, Mandalà and colleagues (2012) reported an 87% success rate in the treatment arm at 24 hours compared with a 0% success rate in the sham arm. Published clinical practice guidelines (Bhattacharyya et al., 2017; Fife et al., 2008) support the use of the Semont maneuver for the treatment of posterior SCC BPPV, but due to the paucity of studies comparing the Semont maneuver with CRP, the practice guidelines did not make recommendations regarding the comparative effectiveness of two PRMs.

Treatment Considerations

Since the posttreatment restrictions are not required, clinicians may retest and repeat treatment as needed within one treatment session. There is marked variability in the literature in terms of the number of maneuvers performed in a treatment session, from one per session (Lynn et al., 1995), to repeated maneuvers (maximum of five) until there is resolution of the nystagmus in the Dix–Hallpike test (Froehling et al., 2000). Due to this variability, the clinical practice guideline (Bhattacharyya et al., 2017) makes no recommendations as to the number of maneuvers performed in a treatment session. Based on the literature, repeat testing and treatment within a given session are not contraindicated. The clinician, however, should be mindful of the patient's overall symptoms as repeated provocation can lead to nausea and emesis.

Complications from PMRs are rare and mild. No serious complications were reported in the RCTs. Mild complications of nausea, vomiting, fainting, and canal

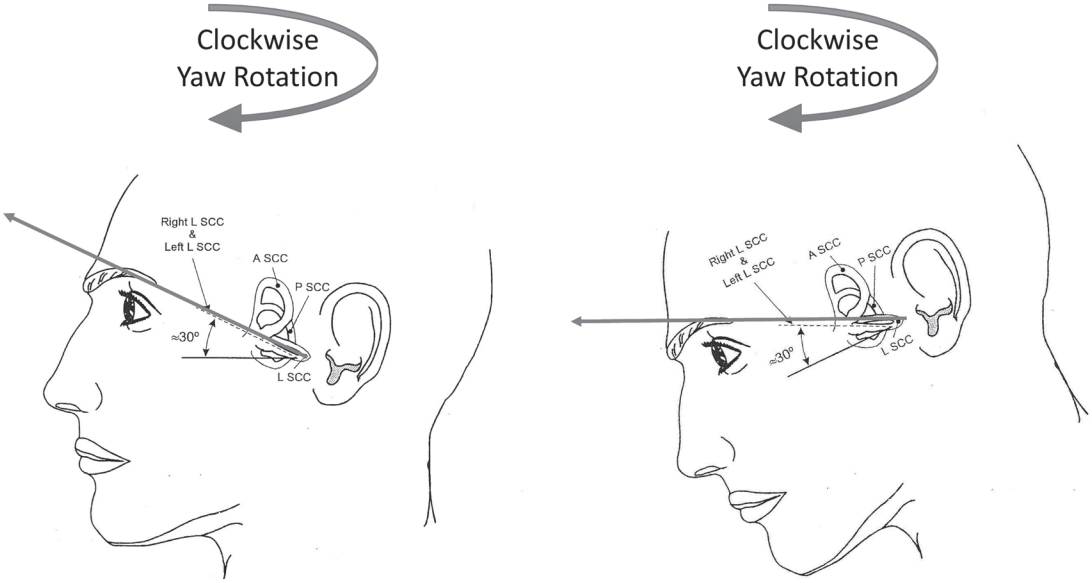

Figure 13–3. Rotation around the earth vertical axis is performed with the head tilted downward by 30° so as to align the horizontal semicircular canal in the optimal plane of (yaw) rotation (arrow), thus maximizing excitation and inhibition in accordance with Ewald's law. Adapted and reprinted with permission from Canalis and Lambert (2000). *The Ear: Comprehensive Otology* (p. 120). Philadelphia: Lippincott Williams & Wilkins.

offered an alternative reason that the disparity between the observed rotational and caloric responses might be due to the fundamental differences between the stimuli (physiologic versus non-physiologic).

PATIENT POPULATIONS IDEAL FOR ROTATIONAL ASSESSMENT

We have just discussed the various clinical indicators for when to perform rotational testing. However, there are a number of specific patient populations for whom rotational testing should be considered an absolute necessity. The first clinical population for whom rotational testing is a necessity comprises those who experience caloric irrigations that reveal bilateral vestibular areflexia. Rotational assessment is required in this situation in order to investigate whether there is a true and complete absence of vestibular reactivity. As previously mentioned, nearly all vestibular lesions first impact the lower stimulus frequencies (i.e., the caloric stimulus). This is analogous to the manner in which hearing loss nearly always impacts the higher frequencies first. Similar to the preservation of low-to-mid frequency hearing sensitivity in presbycusis, there is often preservation of residual vestibular function for higher frequency stimuli in the presence of vestibular pathology. Such residual vestibular function for higher frequencies can only be objectively quantified through rotational assessment. This is critical as the presence of residual vestibular function is essential if physical therapy and vestibular rehabilitation efforts are to be successful. Alternatively, if vestibular areflexia is determined for the entire frequency response, this also becomes critical information as vestibular substitution therapy is then recommended. Use of rotational testing can also confirm the validity of the bilateral absence of the caloric response. That is, if the caloric response is truly absent, then an abnormal VOR response is highly predicted at the lower end of the rotational test frequencies (Jacobson, McCaslin, Grantham, & Shepard, 2016). However, if the VOR response during rotational assessment is entirely within the normal limits for the lowest test frequencies for a patient with caloric areflexia, then a technical error in the delivery or recording of your caloric response should be considered a possible contributing factor to this finding.

The second clinical population for whom rotational testing is necessary consists of those for whom there is some question about the status of vestibular compensation. For case histories where persistent dizziness, vertigo, or imbalance is reported, a differential diagnosis should include an uncompensated vestibular

lesion. Although most peripheral vestibular lesions effectively compensate within one to two weeks, there may be select cases where compensation is incomplete or slow to respond. In such cases, rotational assessment is one of the only clinical tools that can effectively investigate this question. The use of SHA (in conjunction with velocity step testing, which is discussed in Appendix IV) can often highlight problems integral to the compensation process. Moreover, the use of rotational testing (due to its precisely controlled stimulus) makes it an excellent tool for monitoring central compensation progress over time, which is the next clinical population for whom rotation testing is highly recommended.

As mentioned, the third clinical population that would highly benefit from rotational testing are those patients who require monitoring of vestibular degenerative diseases, toxicity, pre- and postoperative status, compensation status, or rehabilitation progress. One of the greatest strengths of rotational testing is its ability to continuously deliver a precise stimulus over repeated clinical visits. Because of this, rotational testing offers a tremendous advantage when monitoring vestibular physiology over an extended period of time, even years. Although there is some variability in how various clinicians may interpret the results, or even some variability with how the test is administered (i.e., differences in mental tasking procedures), the stimulus delivery will remain exactly the same over time provided the chair is regularly maintained and calibrated. This is a significant advantage over other assessment tools such as caloric irrigations, which have a number of inherent factors that contribute to a much higher rate of variability (McCaslin, 2012). This is a distinct advantage when monitoring vestibular function during administration of potentially vestibulotoxic medications, whether for medical treatment or labyrinthine ablation. Moreover, the monitoring of vestibular function to show compensation, rehabilitation, or disease progression can be critical to the effective management of your patient.

A fourth clinical population for whom rotational testing is often a necessity consists of pediatric patients. The often noxious nature of the caloric test, concomitant with the wearing of vision-denied video goggles, often precludes the successful administration of rotational testing to young patients. Testing VOR function in very young patients is often limited to a binary yes/no decision when determining the presence or absence of a vestibular response. In these cases, it is sometimes best to simply identify the presence or absence of a VOR response during chair rotations while the young patient is seated on a parent's or caregiver's lap. If the child can tolerate the video goggles, this is even better. However, very young patients often have a low tolerance for tight-fitting goggles, or many goggles are not designed for very young pediatric faces. Therefore, it is often best to simply observe the child's eyes to determine the presence of a nystagmus response that beats appropriately to the rotations of the chair. This can effectively be accomplished while the patient is seated on a parent's or caregiver's lap with the video monitoring system focused on the child's face and the parent securely holding the child's face in the straightforward position. In this way, it is possible to confirm the presence or absence of a VOR, even in an infant. Although laterality of a vestibular lesion or general vestibular weakness cannot be determined from simple VOR observation, this approach can be effective when determining complete absence of a vestibular reactivity for a patient who is late to walk or has a congenital malformation such as bilateral enlarged vestibular aqueduct (EVA) or Mondini malformation.

THE ROTATIONAL CHAIR

Unlike audiometers, the number of options for purchasing a rotational chair is very limited. Currently, there are only a few manufacturers producing "turnkey" commercially available equipment dedicated to clinical rotational testing. The two most common manufacturers in the United States are Neuro Kinetics, Inc. (NKI) and Micromedical, Inc. Figure 13–4 shows two images of the Neurotologic Test Center Suite (NOTC) produced by Neuro Kinetics, Inc. Each image comprises a lightproof enclosure and torque-driven rotational chair. The primary difference between the two images is that the NOTC on the left is a standard rotational chair, while the image on the right is a chair that is capable of producing angular rotations with the chair positioned slightly off vertical axis. This latter testing is known as Off-Vertical-Axis-Rotation (OVAR), and requires an upgraded torque motor that sits higher off the floor and thus necessitates an eight-foot diameter booth in lieu of the six-foot diameter booth used with the standard NOTC chair. Figure 13–5 shows two images of the System 2000 produced by Micromedical, Inc. Both images show the same rotational chair. The difference between the images is that since the videooculography goggles are fully enclosed, the testing can be performed without the need for a lightproof enclosure. This can offer a distinct advantage in test environments that may not be able to support a six-foot or even eight-foot diameter enclosure. Moreover, the boothless nature of this rotational chair makes it

test to carry out—the patient is just asked to look at the spot. So language difficulties or cognitive deficits have minimal impact. SHIMP is even easier to carry out at the bedside than HIMP, and the corrective saccade is much easier for the clinician to see.

NEW DEVELOPMENTS

1. The ability of vHIT testing to probe the function of all canals has revealed unexpected outcomes. One is that a rather surprising number of patients have BVL of most (or all) semicircular canals. Prior to vHIT the extent of vertical canal loss could not be measured in a clinic. With vHIT it now can be. And this pattern of reduced or absent function of all canals (Ward, Agrawal, Hoffman, Carey, & Della Santina, 2013) gives new understanding of patient complaints.
2. Complementing that result is the equally unexpected result of the bilateral sparing of anterior canal function, when other canals show loss (Tarnutzer, Bockisch, Buffone, & Weber, 2017). Figure 14–19 shows data for a patient with bilateral loss of horizontal canal function, but bilateral sparing of both anterior and posterior canals (Akdal et al., 2016).
3. As mentioned above, the use of vHIT to quantify the progress of the loss of canal function during intratympanic gentamicin is now widespread.
4. The dissociation of vHIT and caloric test results may be an indicator of endolymphatic hydrops.

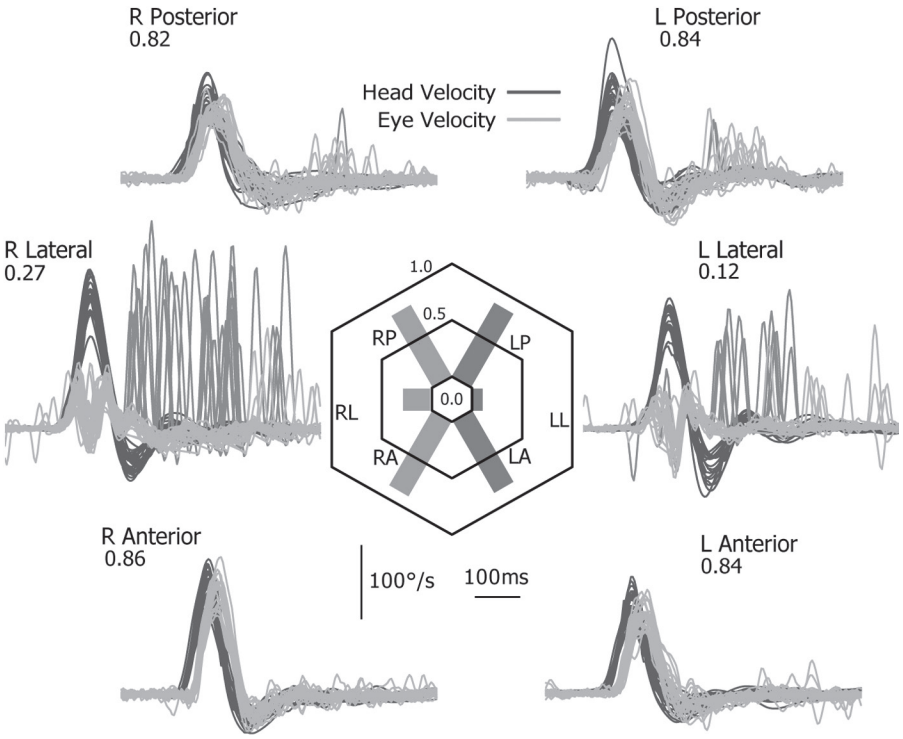

Figure 14–19. Results of testing all canals in a patient with BVL. Here there is loss of function in both horizontal canals, but sparing of both anterior canals and of both posterior canals. Video head impulse measures of patients with BVL often reveal relative sparing of anterior semicircular canal function. This clinical pattern may possibly indicate gentamicin vestibulotoxicity or Ménière's disease, or in Wernicke's encephalopathy as in this patient, but also occurs in BVL of unknown origin. Reprinted from *Journal of the Neurological Sciences*, Vol. 365, Akdal, G., MacDougall, H. G., Chen, L., Tanrıverdizade, T., Yiğitaslan, O., and Halmagyi, G. M., Selective impairment of horizontal vestibulo-ocular reflexes in acute Wernicke's encephalopathy, pp. 167–168, © 2016, with permission from Elsevier.

CONCLUSION

Vestibular testing is being revolutionized by vHIT, and when the results for vHIT are combined with the results of the new ocular and cervical vestibular evoked myogenic potential (oVEMP and cVEMP) tests it is possible to measure the function of all vestibular sense organs (Curthoys, 2012).

Video HIT is very simple and does not require specialized conditions—testing is done with the patients sitting in an ordinary chair in a normally lit room. Video HIT is undemanding and can be used on patients as young as 3 years. Video HIT is a fast, simple way of quickly, safely, and acceptably answering the question: Which side is affected? Is the canal function of each ear in the normal range? Video HIT allows repeated testing even within a few minutes. It gives an absolute level of canal function. It is very well tolerated by patients and it allows measurement of the function of all semicircular canals. The vHIT test is noninvasive, safe, simple, and quick (less than 10 min to test both sides), and very acceptable even to dizzy and nauseous patients. The analysis software provides objective, quantitative results in real time. In sharp contrast to caloric stimuli, with vHIT the magnitude of the stimulus at each instant in time is known and can be related directly to the response at that instant. Video HIT is portable—it can be used in the clinic or at the bedside or the emergency room, or even in the patient's home. Video HIT can be carried out even during acute attacks of vertigo (e.g., in a patient during a Menière's attack or an attack of vestibular neuritis [Manzari et al., 2011]).

VIDEOS ASSOCIATED WITH THIS CHAPTER

Video 14–1. How the horizontal head impulse test is carried out at Royal Prince Alfred Hospital Sydney for HIMPs and SHIMPs.

Video 14–2. How the standard LARP impulses are carried out at Royal Prince Alfred Hospital Sydney.

Video 14–3. How the standard RALP impulses are carried out at Royal Prince Alfred Hospital Sydney.

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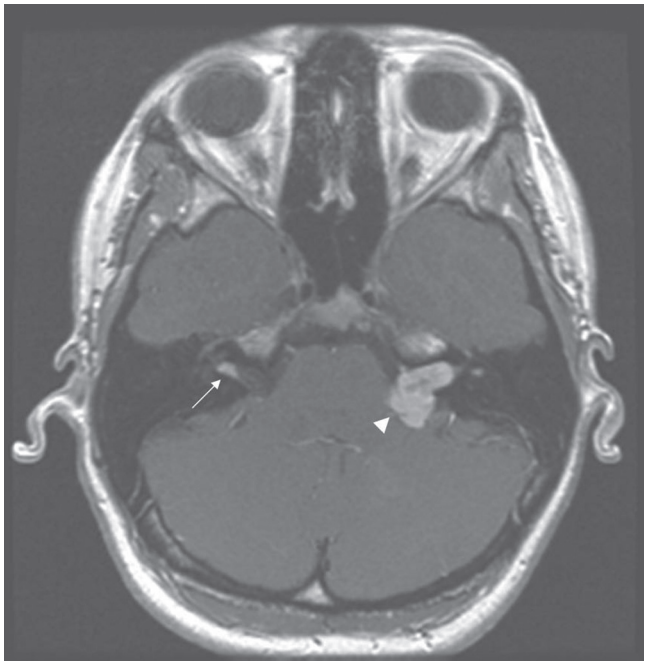

Figure 21–8. T1 Axial MRI with gadolinium-NF2 patient with a right internal auditory canal lesion (white arrow), and a left cerebellopontine angle lesion with extension into left internal auditory canal (white arrowhead).

Preservation of Residual Hearing

There is mounting evidence that hearing preserved by conservation surgery will usually continue to deteriorate, especially in Ménière's disease. In this case, the riskier intracranial procedure required for a vestibular neurectomy may provide no incremental benefit to the patient (Eisenman, Speers, & Telian, 2001; Tewary, Riley, & Kerr, 1998). This realization, coupled with the increased use of intratympanic gentamicin treatment, has led to a dramatic reduction of intracranial procedures for vertigo. Regardless, the patient's perception of useful hearing is the most important factor when determining whether or not to sacrifice residual hearing. Some patients claim subjective benefit from amplification of hearing ordinarily considered useless by audiologic standards. On the other hand, there are patients who have clinically measurable hearing that should be useful, but report no benefit from amplification and/or refuse to use a hearing aid. Thus, no specific hearing threshold or speech discrimination score should be used as a strict criterion to clarify this decision, and each case must be considered on an individual basis.

Labyrinthectomy

Labyrinthectomy procedures are reserved for patients who have minimal, if any, residual hearing. When the contralateral ear is stable and the patient is convinced that the involved ear is functionally useless, one may recommend a labyrinthectomy. This can be accomplished chemically with intratympanic aminoglycosides (discussed elsewhere in this volume) or surgically using a transcanal or transmastoid labyrinthectomy.

Also known as the oval window labyrinthectomy, the transcanal labyrinthectomy procedure is performed through the external auditory canal (Pulec, 1974; Schuknecht, 1991b). A tympanomeatal flap is elevated and the stapes is removed from the oval window, giving the surgeon access to the labyrinth. Some surgeons remove bone between the oval window and the round window to improve visualization of the vestibule. The saccule and the utricle are then removed. Removal of the horizontal and superior canal ampullae is not possible with this approach, but the surgeon may choose to section the singular nerve or remove the posterior canal ampulla. Control of vertigo may be augmented by placing absorbable packing material soaked with an aminoglycoside antibiotic into the vestibule. This procedure reliably eliminates episodic vertigo, even though removal of the sensory neuroepithelium in its entirety is not achieved. A threefold increase in postoperative disequilibrium compared with the transmastoid approach (63% versus 23%, respectively) has been documented (Langman & Lindeman, 1998).

The transmastoid labyrinthectomy, considered by the senior author to be the "gold standard" for surgical relief of vertigo, is accomplished through a post-auricular incision. A complete mastoidectomy is performed and the surgeon identifies the horizontal semicircular canal in the mastoid antrum. When the labyrinthine bone has been drilled away appropriately, the surgeon will be able to visualize and remove all three ampullae and both otolithic maculae (Figure 21–9). The wound is then irrigated and closed. As one would expect, this procedure results in highly reliable relief of vertigo (Kemink, Telian, Graham, & Joynt,1989; Schwaber, Pensak, & Reiber, 1995).

Vestibular Neurectomy

If a decision has been made to ablate vestibular function but hearing preservation is desired, a vestibular nerve section may be performed. This operation is more risky and slightly less reliable than labyrinthec-

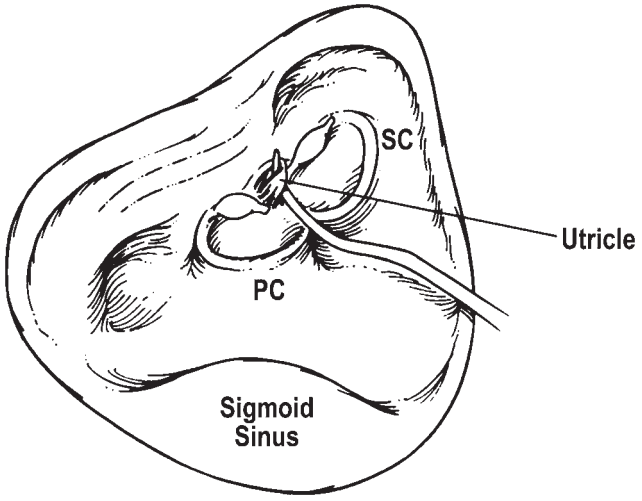

Figure 21–9. Surgical view of left temporal bone following transmastoid labyrinthectomy. Semicircular canals and vestibule are open. From Chapter 144, Surgery for vestibular disorders, by Steven A. Telian. in C. W. Cummings (Ed.), *Otolaryngology Head and Neck Surgery*, 4th ed., p. 3301. Copyright 2005, Elsevier. Reproduced with permission.

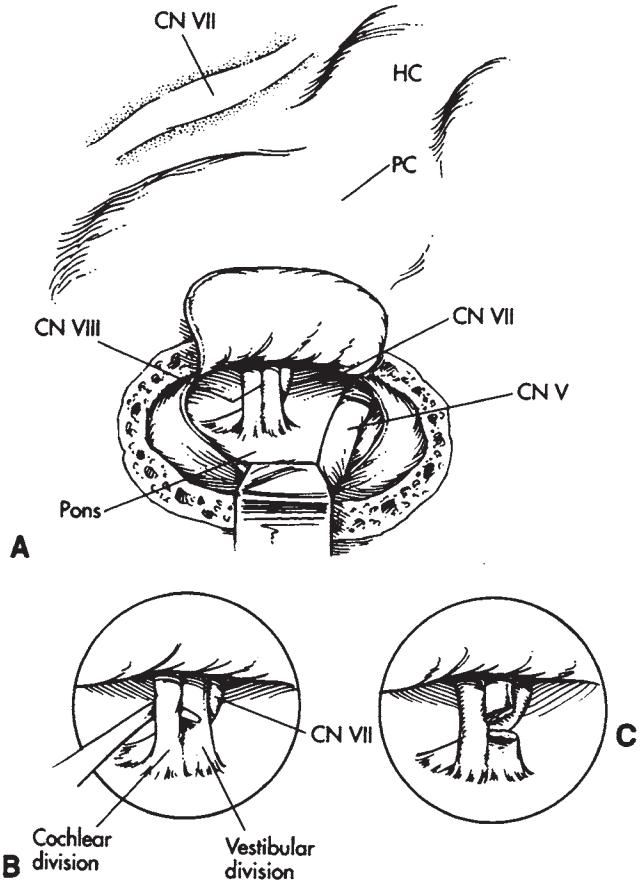

Figure 21–10. Left retrolabyrinthine vestibular nerve section. **A.** Depiction of the surgeon's view into the cerebellopontine angle through the craniotomy. **B.** Higher power magnification showing the separation of the cochlear and vestibular portions of the eighth cranial nerve. under the same magnification, **C.** Depicts the separation of the sectioned vestibular nerve. From Chapter 144, Surgery for vestibular disorders, by Steven A. Telian. in C. W. Cummings (Ed.), *Otolaryngology Head and Neck Surgery*, 4th ed., p. 3305. Copyright 2005, Elsevier. Reproduced with permission.

tomy procedures, but it does offer relief from episodic vertigo in the vast majority of cases. Reported rates of vertigo control range from 80% to 95%. The three most widely used approaches are the retrolabyrinthine approach, the retrosigmoid approach, and the middle fossa approach.

The retrolabyrinthine approach involves a complete mastoidectomy, with decompression of the posterior fossa dura and the sigmoid sinus. The bone anterior to the sinus is removed until the labyrinth is outlined and the ELS is exposed. The dura and the ELS are incised anterior to the sigmoid sinus, opening the cerebellopontine angle. The eighth nerve complex is identified and the vestibular portion of the eighth cranial nerve is divided, taking care to avoid injury to the facial nerve, the auditory fibers, or the nervus intermedius (Figure 21–10). The major disadvantage of this approach is the uncertainty about the plane of dissection between the cochlear and vestibular nerve bundles. This may result in an incomplete transection of the vestibular nerve fibers or inadvertent sectioning of some fraction of the auditory fibers. This is a reliable approach with a low incidence of hearing loss and facial nerve injury (House, Hitselberger, McElveen, & Brackmann, 1984; Kemink & Hoff, 1986; Silverstein & Norrell, 1980).

The retrosigmoid approach may allow for more selective sectioning of the vestibular nerve than the retrolabyrinthine approach. Because this procedure involves less temporal bone dissection, thus improving the speed of the procedure, it is the preferred approach by many neurotologists today. However, patient positioning and the surgeon's proximity to the nerve are less favorable than with the retrolabyrinthine approach. There is also an increased need for retraction of the cerebellum. This operation is also known to produce a higher incidence of postoperative headaches, especially if intradural drilling is performed to achieve more lateral exposure of the eighth nerve