7.3 The Brain Stem Auditory System

Upon entering the brain stem, the central processes of the spiral ganglion cells bifurcate and distribute to the cells of the dorsal and ventral cochlear nuclei (Sect. 7.3.1). The organization of the terminations was first described by Lorente de Nó (1933), based on his Golgi studies in a 4-day-old cat. In squirrel monkeys, fibres from the basal turn of the cochlea project to dorsal regions of the ventral cochlear nucleus, whereas apical fibres project to ventral regions (Moskowitz and Liu 1972). The primary cochlear nuclei contribute bilateral ascending projections to the superior olivary complex and to the lateral lemniscus (Sect. 7.3.2). The majority of the lateral lemniscal fibres ascend directly to the inferior colliculus (Sect. 7.3.3). Ascending projections from the inferior colliculus form the brachium of the inferior colliculus and reach the MGB (Sect. 7.4.1), which via the acoustic radiation (Sect. 7.4.2) projects to the auditory cortex (Sect. 7.4.3).

7.3.1 The Cochlear Nuclei: Diversification of Cochlear Input

The human cochlear nuclei consist of a large ventral nucleus and a smaller dorsal nucleus (Moore and Osen 1979; Terr and Edgerton 1985; Adams 1986). The **dorsal cochlear nucleus** contains a large variety of cell types, and is situated dorsolateral to the inferior cerebellar peduncle. The **ventral cochlear nucleus** contains many different cell types and has anteroventral, ventral and posteroventral subnuclei, which borders are not well defined, however. The cochlear nuclei receive a rich **blood supply** from multiple sources, including branches of the anterior and posterior inferior cerebellar arteries (Oas and Baloh 1992).

The **secondary auditory projections** from the cochlear nuclei to the superior olivary complex and the inferior colliculus take various routes (Fig. 7.8). Ipsilaterally, a major projection from both ventral and dorsal cochlear nuclei reaches the superior olivary complex (Sect. 7.3.2). Contralaterally, there are three major ascending cochlear projections (Strominger 1973; Strominger et al. 1977): (1) the largest originates in the ventral part of the ventral cochlear nucleus and forms the **trapezoid body**; its axons may proceed directly to the contralateral lemniscus or terminate in the superior olivary complex; (2) fibres from the dorsal part of the ventral cochlear nucleus form the **intermediate acoustic stria**; they contribute to the lateral lemniscus; and (3) a contralateral projection from the dorsal cochlear nucleus, forming the **dorsal acoustic stria**. The dorsal and intermediate acoustic striae and the trapezoid body converge to form the **lateral lemniscus**. The auditory nuclei do not only serve as relay nuclei in the ascending auditory projection, but also as reflex centres. Efferents from the cochlear nuclei enter the reticular formation, where they contact neurons of the

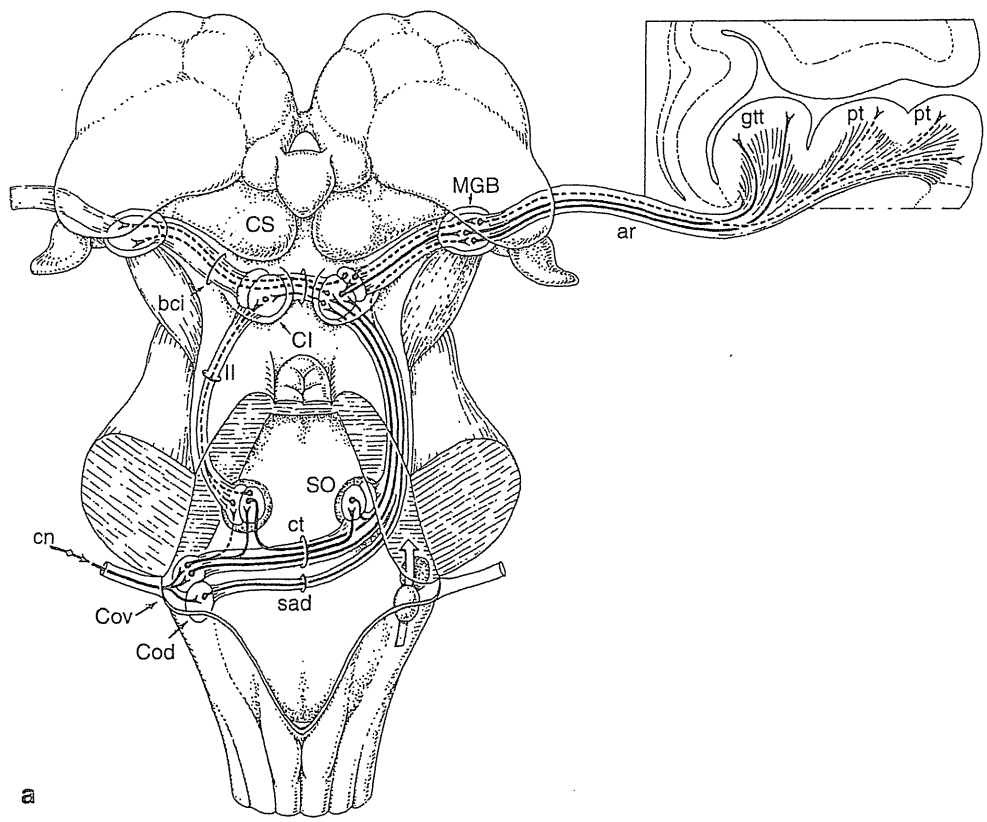

Fig. 7.8 (a) Overview of the auditory projections in the human brain (after ten Donkelaar et al. 2007); (**b–d**) the position of the cochlear nuclei (in *red*), the lateral lemniscus (in *light red*) and the colliculus inferior (in *red*) in horizontal sections of the brain stem (after Duvernoy 1995). *Abbreviations*: *ar* acoustic radiation; *bci* brachium of colliculus inferior; *CI* colliculus inferior; *cn* cochlear nerve; *Cod*, *Cov* dorsal and ventral cochlear nuclei; *CS* colliculus superior; *ct* corpus trapezoideum; *gtt* gyrus temporalis transversus (Heschl's gyrus); *ll* lateral lemniscus; *MGB* medial geniculate body; *pt* planum temporale; *sad* stria acoustica dorsalis; *SO* superior olive

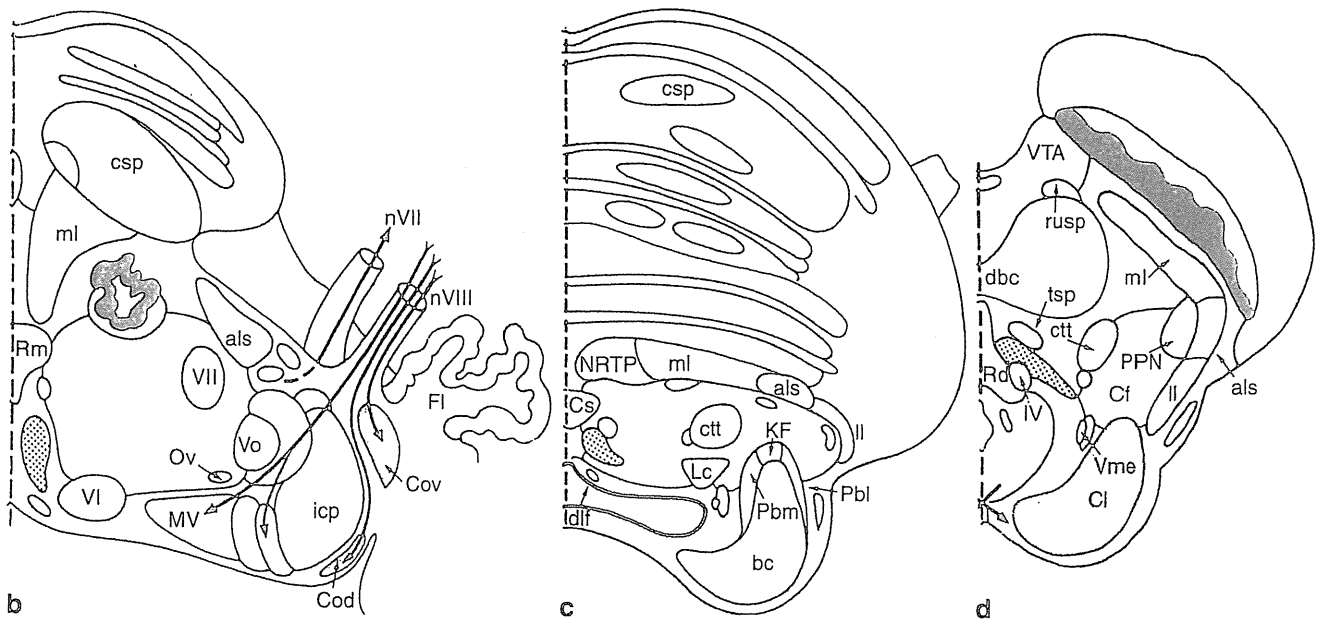

Fig. 7.8 (continued)

ascending reticular activating system (see Chap. 5), and give rise to the **auditory-evoked startle reflex**.

7.3.2 The Superior Olivary Complex: Recreation of Auditory Space

The superior olivary complex is the first site for **binaural convergence**. In primates, the cochlear nuclei project to the superior olivary complex on both sides of the brain stem (Strominger 1973; Strominger et al. 1977). The **superior olivary complex** is located in the caudal pons, lateral to the medial lemniscus and dorsal to the spinothalamic tract. The complex contains the medial superior olivary nucleus, the lateral superior olivary nucleus and the nucleus of the trapezoid body. The latter nucleus is indistinct in apes and vestigial in humans (Moore 2000). The superior olivary complex is important for the **localization of sounds** (Moore and Linthicum 2004). A sound is localized by two means depending on its frequency: (1) low-frequency sounds activate the two ears at somewhat different times (interaural time differences); (2) high-frequency sounds activate the two ears with somewhat different intensities (interaural intensity differences). Neurons in the medial superior olivary nucleus are tuned to low-frequency stimuli and are sensitive to interaural time differences. The projection from the ventral cochlear nucleus is thought to contribute to this sensitivity. In contrast, neurons in the lateral superior olivary nucleus are tuned to high-frequency stimuli and are sensitive to interaural intensity differences. The lateral superior olivary nucleus receives a monosynaptic excitatory connection from the ipsilateral ventral cochlear nucleus and a disynaptic inhibitory connection from the contralateral ventral cochlear nucleus via the nucleus of the trapezoid body. Since the dorsal cochlear nucleus does not innervate the superior olivary complex, it is believed not to play a role in the localization of sounds.

Behavioural studies in cats have implicated the superior olivary complex in the **recreation of auditory space**. Cats with lesions above the level of the superior olivary complex, in the lateral lemniscus, the inferior colliculus, the MGB or the auditory cortex, are unable to locate a sound source in the spatial field contralateral to the lesion, whereas cats with lesions below the superior olivary complex have more diffuse deficits (Casseday and Neff 1975; Thompson and Masterton 1978; Jenkins and Masterton 1982). A comparable deficit has been observed in human subjects with extensive midline pontine lesions that eliminated crossed input to the superior olivary complex on both sides (Griffiths et al. 1997a; Furst et al. 2000; see *Clinical case 7.2*). These animal and human studies suggest that the auditory spatial field is recreated in the brain stem by transformations occurring at the level of the superior olivary complex.

7.3.3 The Upper Brain Stem: Integration of Ascending Auditory Pathway

The **lateral lemniscus** is clearly visible in the rostral pons and the midbrain. Most of its fibres terminate in the inferior colliculus. Many of these fibres send a collateral branch to the nuclei of the lateral lemniscus, which innervate the inferior colliculus and also directly the MGB. In most mammalian

species, the lateral lemniscus contains sizable ventral, intermediate and dorsal lemniscal nuclei (Moore 1987). In humans, only the dorsal lemniscal nucleus is well developed (Geniec and Morest 1971; Moore 1987). It gives rise to Probst's commissure to the contralateral inferior colliculus.

The **inferior colliculus** is composed of three nuclei: central, external and pericentral. The **central nucleus** is the principal nucleus of the inferior colliculus and receives input from: (1) the direct pathway from the dorsal and ventral cochlear nuclei; (2) projections arising from the ipsilateral and contralateral superior olivary complex and (3) fibres from the dorsal nucleus of the lateral lemniscus. These projections all pass via the lateral lemniscus. The central nucleus is laminated (Geniec and Morest 1971) and processes the physical characteristics of sounds for auditory perception. In this nucleus, neurons in a single layer are maximally sensitive to similar tonal frequencies. The function of the other two nuclei of the inferior colliculus is not entirely clear. Lesion studies in cats suggest that the **external** and **pericentral nuclei** play a role in acousticomotor function such as the orientation of the head and body to auditory stimuli. The inferior colliculus projects to the MGB via the brachium of the inferior colliculus, which is macroscopically visible on the lateral surface of the midbrain. The inferior colliculi are interconnected via the commissure of the inferior colliculi.

7.3.4 Brain Stem Topography: Generation of Evoked Potentials

Waves I and II of the ABR are generated by the cochlear nerve. The subsequent waves III–VI are generated within the brain stem (see Fig. 7.5). Intrasurgical recordings made from the surface of the human brain stem and dipole studies suggest that wave III is generated by a volley of action potentials in axons emerging from the cochlear nuclei in the ventral acoustic stria (Stockard et al. 1978, 1986; Moller and Jannetta 1982; Scherg and von Cramon 1985). Waves IV and V are generated further rostrally in the brain stem: wave IV most likely at the level of the superior olivary complex contralateral to the stimulated ear, presumably by the bend in the axonal pathway occurring at that point, and wave V by synaptic activity in the inferior colliculus (Moller and Jannetta 1982; Moore et al. 1996).

Clinical Case 7.2 Impaired Sound Localization Following a Midline Pontine Lesion

In a 45-year-old female patient with an extensive **midline pontine lesion**, eliminating crossed input to the superior olivary complex on both sides, Griffiths et al. (1997a, b) observed that the patient had no difficulty in detecting frequency and amplitude modulation and no general deficit in detection of auditory temporal information, but she was *unable* to determine by sound alone the location and direction of motion of objects in the environment, such as ringing telephones and passing trains. Furst and co-workers analyzed sound localization in patients with multiple sclerosis and brain stem infarcts (Furst et al. 2000; 1995; Aharonson et al. 1998). Levine and Häusler (2001) reported another case (see **Case report**).

Case report: An 80-year-old male presented with sudden onset of vertigo and vomiting. On examination, he was found to have a left gaze palsy, dysphagia, dysarthria, and a right hemiplegia that included only the lower face. He had no auditory complaints, and his bedside hearing evaluation was unremarkable. MRI showed a left trapezoid body infarct, the location of which is indicated in Fig. 7.9a. A year later, he was evaluated with a battery of hearing tests. Despite an age-appropriate audiogram and normal BAERs, all fusion tests were abnormal for the three stimuli used (clicks, low-pass noise and high-pass noise) and for interaural time or level disparities (Fig. 7.9b). Just noticeable differences were highly abnormal, and regardless of the size or type of interaural disparity, the patient indicated that everything sounded as though it were coming from or near the centre of his head (Fig. 7.9c). Unlike normal subjects, nothing was heard coming from the far right or left.

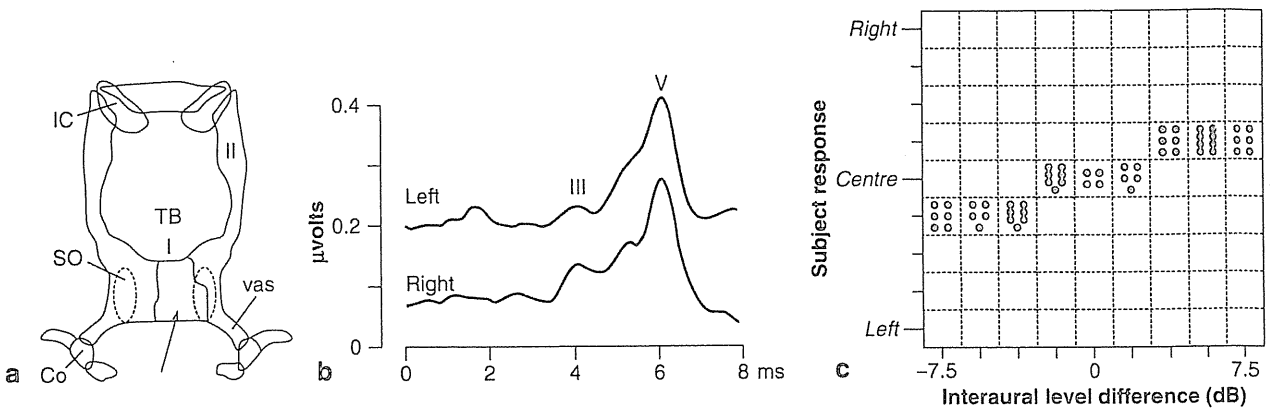

Fig. 7.9 Impaired sound localization in a patient with a lower pontine lesion (*arrow* in **a**) involving the trapezoid body; (**b**) brain stem auditory evoked responses; (**c**) sound lateralization (after Levine and Häusler 2001; see text for explanation). *Abbreviations*: *Co* cochlear nuclei; *IC* inferior colliculus; *ll* lateral lemniscus; *TB* trapezoid body; *vas* ventral acoustic stria

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This case is based on a case report by Levine and Häusler (2001).

7.4 The Forebrain Auditory System

For decades, the dominant species for research on the auditory forebrain has been the cat, but the focus has now clearly shifted to non-human primates. Although the subcortical auditory systems of monkeys and cats are largely similar, there are important differences in cortical organization (Kaas and Hackett 2000).

7.4.1 The Auditory Thalamus

The **medial geniculate body (MGB)** or **nucleus** is clearly visible on the inferior surface of the inferior thalamus. The MGB contains several divisions, the principal auditory relay nucleus is the **ventral** or **principal medial geniculate nucleus** (Winer 1984). The ventral division of the MGB is laminated. It receives the major ascending auditory projection from the also laminated central nucleus of the inferior colliculus. For both nuclei, lamination is a structural correlate of precise tonotopic organization. In contrast, the dorsal and medial divisions of the MGB are not laminated and receive much less dense input from the inferior colliculus. The ventral medial geniculate nucleus projects via the auditory radiation to the tonotopically organized primary auditory cortex. The dorsal and medial subnuclei project to higher-order auditory cortical areas in the planum temporale, areas that do not have such a precise tonotopic organization as the primary auditory cortex.

7.4.2 The Acoustic Radiation

In 1882, Constantin von Monakow first described the origin of the acoustic radiation from the MGB in rabbit experiments. The classic studies in the human brain located the proximal part of the acoustic radiation just caudal to the thalamus, where it originates from the MGB, then passes through the sublenticular, posterior part of the internal capsule to curve around the inferior sulcus of the insula before reaching Heschl's gyrus

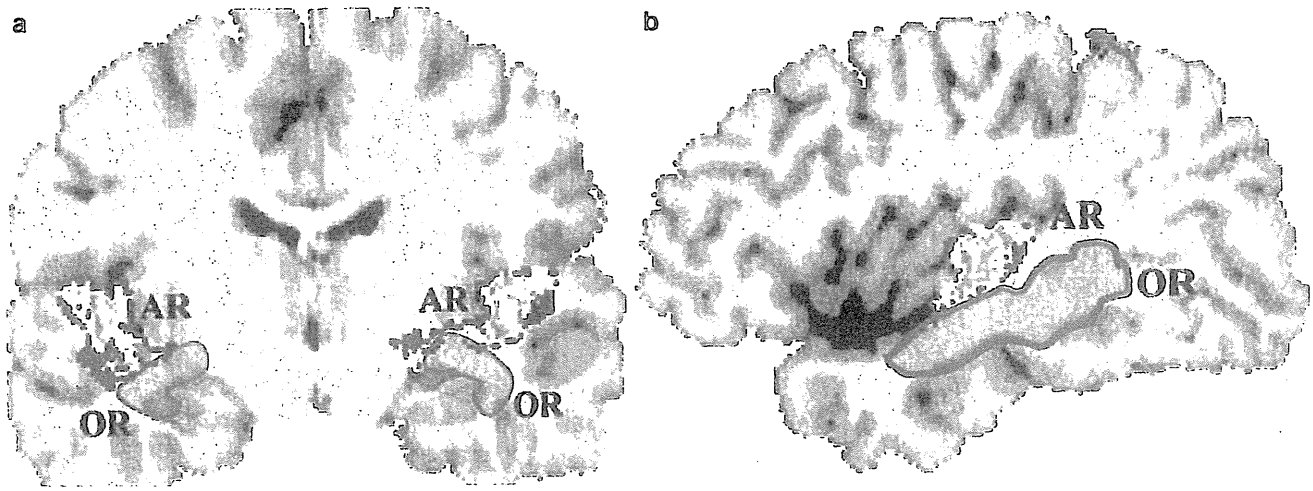

Fig. 7.10 The acoustic and optic radiations in coronal (**a**) and sagittal (**b**) probabilistic maps (after Rademacher et al. 2002). *Abbreviations*: *AR* acoustic radiation; *OR* optic radiation

(Dejerine 1895; Flechsig 1920; Pfeifer 1920). In a more recent study, Rademacher et al. (2002) showed the stereotaxic localization, intersubject variability and interhemispheric differences of the human acoustic radiation (Fig. 7.10). They showed that the location of the acoustic radiation varies considerably between individuals and hemispheres.

7.4.3 The Auditory Cortex: Sequential Levels of Auditory Processing

The **primary auditory cortex** (A1) is located on the transverse temporal or Heschl's gyrus in the temporal lobe of the cerebral cortex and corresponds to area 41. It is surrounded by secondary auditory areas (A2): caudally the caudomedial area, also known as the planum temporale, and rostrally, the rostral area. Geschwind and Levitsky (1968) demonstrated that the **planum temporale** is larger on the left side in the majority of the postmortem brains they examined. **Asymmetry** of the planum temporale may form the substrate for left hemispheric dominance for language-related auditory processes (Geschwind and Galaburda 1985; Dorsaint-Pierre et al. 2006) and is correlated with handedness (Steinmetz et al. 1989, 1991).

Heschl's gyrus is located largely within the lateral sulcus (von Economo and Horn 1930; Fig. 7.11). The transverse temporal gyrus is often partially duplicated into a double, or occasionally triple convexity (Pfeifer 1920; Steinmetz et al. 1989; Penhune et al. 1996; Leonard et al. 1998; Morosan et al. 2001). The cytoarchitecture of the human auditory cortex has been described by Brodmann (1908, 1909), von Economo and Koskinas (1925), Galaburda and Sanides (1980) and, more recently by Hackett et al. (2001) and Hackett

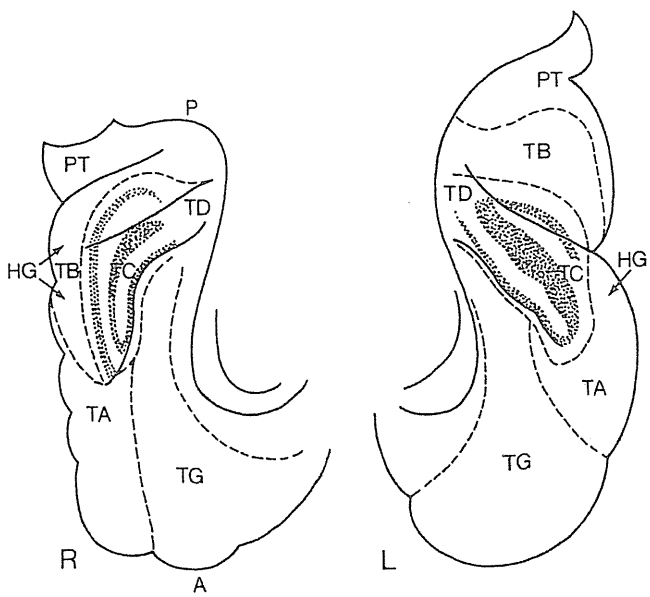

Fig. 7.11 The human auditory cortex. The primary auditory cortex is composed of two fields, TD and TC. On the right side (*R*), these occupy a double transverse temporal gyrus (Heschl's gyrus); on the left side (*L*), they correspond to a single Heschl's gyrus and a part of the more caudally situated planum temporale (*PT*). TD and TC are composed of markedly granular subareas (*dotted* in *red*) and less granular areas. Note the distinct right–left asymmetries with a larger planum temporale on the left side. *Abbreviations*: *A* anterior; *HG* Heschl's gyrus; *P* posterior; *TA* superior temporal area; *TB* magnocellular supratemporal area; *TC* transverse supratemporal area; *TD* intercalate supratemporal area; *TG* temporopolar area (after Brodal 1981)

and Kaas (2004) and Morosan et al. (2001) and Rademacher et al. (2001a, b). The primary auditory cortex was designated area 41 by Brodmann, TC by von Economo and Koskinas and

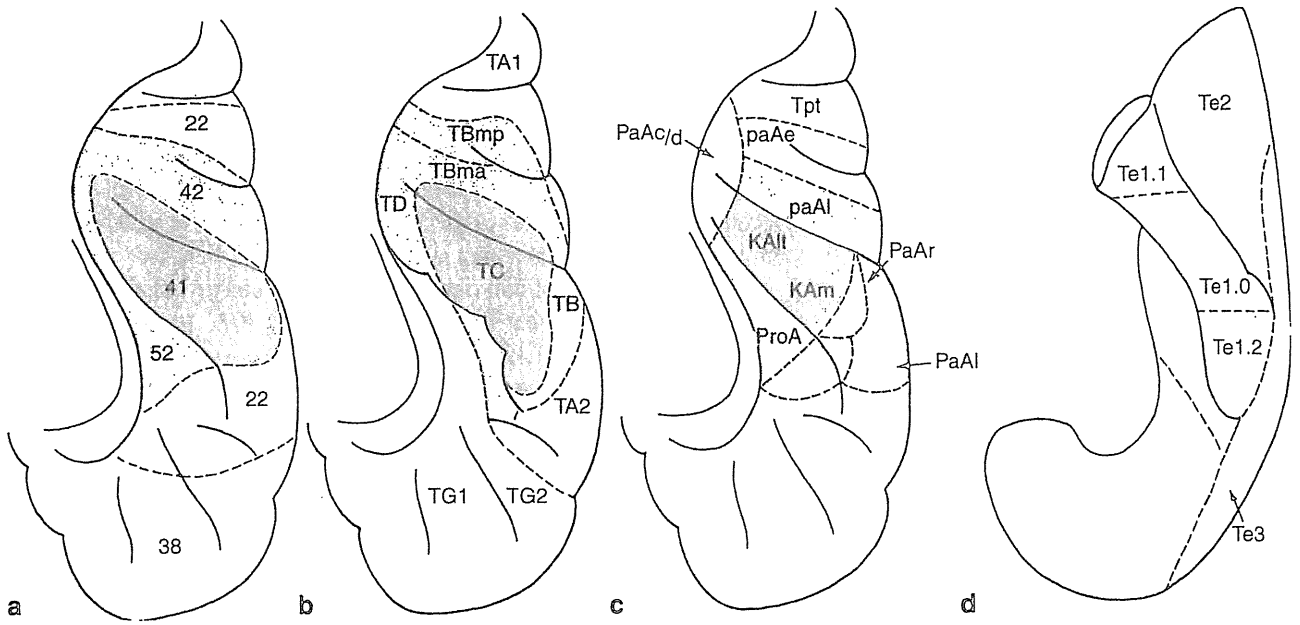

Fig. 7.12 Regional parcellation of the right human superior temporal cortex (rostral is below) according to (**a**) Brodmann, (**b**) von Economo and Koskinas, (**c**) Galaburda and Sanides and (**d**) Morosan and co-workers (after Hackett and Kaas 2004 and Morosan et al. 2001). In (**a–c**), core areas are shown in *red*, belt areas in *medium red* and parabelt areas in *light red*. *Abbreviations*: *KAlt*, *KAm* lateral and medial auditory koniocortex; *PaAc/d*, *paAe*, *PaAl/paAl*, *PaAr* caudal/dorsal, external, lateral and rostral auditory parakoniocortex; *ProA* proauditory cortex; *TA1*, *TA2*, *TB*, *TBma*, *TBmp*, *TC*, *TD* subdivisions by von Economo and Koskinas; *Te2*, *Te1.0*, *Te1.1*, *Te1.2*, *Te3* subdivisions by Morosan and co-workers; *TG1*, *TG2* temporopolar subdivisions of von Economo and Koskinas; *Tpt* temporoparietal area; *22–52* Brodmann areas

KAm and KAlt (medial and lateral auditory **koniocortex**) by Galaburda and Sanides (Fig. 7.12a–c). KAm is the most medial and the most granular area, whereas the more lateral KAlt is less granular. Morosan et al. (2001) suggested three areas with well-developed layers IV, Te1.1, Te1.0 and Te1.2, to represent the primary auditory cortex (Fig. 7.12d). There is considerable variability in size of the auditory koniocortex and its extent does not coincide with gyral or sulcal anatomy (Galaburda and Sanides 1980; Rademacher et al. 1993, 2001a, b; Hackett et al. 2001; Morosan et al. 2001). The human auditory koniocortex (area 41/TC/KA/Te) is homologous to the core area of the monkey auditory cortex. Based on parvalbumin staining, Wallace et al. (2002) suggested that Heschl's gyrus contains two core fields, partially surrounded by at least six belt fields that lie mostly on the superior temporal gyrus. In an fMRI study, Wessinger et al. (2001) showed that pure tones primarily activate the core and that more complex sounds activate belt areas.

The **primate auditory core area** is located in the centre of the superior temporal plane (Hackett et al. 2001; Fig. 7.13). In primates, a centrally located core region containing two or three subdivisions including the primary auditory area (A1), a surrounding belt of cortex with some seven divisions, and a lateral parabelt region comprised of at least two fields, have been described. In monkeys, the **core region** can be identified

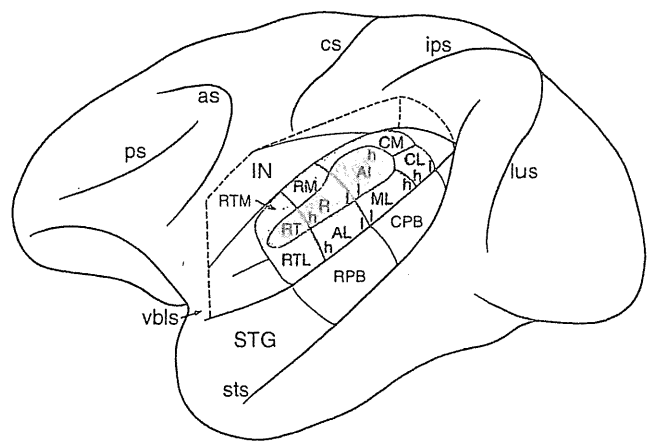

Fig. 7.13 Auditory and auditory-related cortices in macaque monkeys. Core areas (A1, R, RT) are shown in *red*, belt areas (CL, CM, AL, RM, RTL, RTM) in *medium red* and parabelt areas (CPB, RPB) in *light red*. Major sulci have been opened to show the extent of auditory-related cortex. *Abbreviations*: *A1* primary auditory area; *AL* anterolateral area; *as* arcuate sulcus; *CL*, *CM* caudolateral and caudomedial areas; *CPB* caudal parabelt area; *cs* central sulcus; *h*, *l* high and low frequencies; *IN* insula; *ips* intraparietal sulcus; *lus* lunate sulcus; *PL* posterolateral area; *ps* principal sulcus; *R* rostral area; *RM* rostromedial area; *RPB* rostral parabelt area; *RT* rostrotemporal primary auditory cortex; *RTL*, *RTM* lateral and medial rostrotemporal areas; *STG* superior temporal gyrus; *sts* superior temporal sulcus; *vbls* ventral bank of lateral sulcus (after Hackett et al. 2001)

on the basis of *specific* anatomical and physiological features. The region shows dense immunostaining for parvalbumin in layer IV, surrounded by a more lightly stained belt, which is flanked by a very sparsely stained parabelt (Jones et al. 1995; Kosaki et al. 1997). Parvalbumin staining also marks the human core auditory cortex in humans (Nakahara et al. 2000; Wallace et al. 2002; Chiry et al. 2003). In macaque, chimpanzee and human brains, Hackett et al. (2001) identified the auditory core from serial sets of adjacent sections processed for cytoarchitecture, myeloarchitecture, acetylcholinesterase and cytochrome oxidase. The position of the core region with respect to major sulci and gyri in the superior temporal region varied most in chimpanzee and human brains.

In monkeys, most neurons of the ventral division of the MGB project to the core cortex (Mesulam and Pandya 1973; Burton and Jones 1976; Luethke et al. 1989; Rauschecker et al. 1997). These **thalamocortical projections** terminate in layers IV and lower III in regular patches of higher density label, separated by areas of less dense labelling (Pandya and Rosene 1993; Hashikawa et al. 1995). In contrast, the medial and dorsal divisions of the MGB project to the core area diffusely. It seems likely that the human primary auditory cortex also receives dense thalamic input. This input explains the cochleotopic organization shown in this area by functional imaging, including magnetoencephalography (MEG) (Elberling et al. 1982; Hari et al. 1989; Pantev et al. 1995; Lutkenhoner and Steinstrater 1998), PET (Lauter et al. 1985; Ottaviani et al. 1997; Lockwood et al. 1999), fMRI (Wessinger et al. 1997; Scheich et al. 1998; Di Salle et al. 2001) and microelectrode mapping studies in epilepsy patients (Howard et al. 1996).

The human auditory koniocortex is surrounded rostrally, laterally and caudally by an area of **parakoniocortex** (Fig. 7.12). This region covers the lateral part of the transverse temporal gyrus, and extends rostrally and caudally over the superior temporal plane. The auditory parakoniocortex has been called area 42 by Brodmann and TB by von Economo and Koskinas. Galaburda and Sanides (1980) distinguished three regions: (1) a rostral auditory parakoniocortex (PaAr) on the rostral aspect of the superior temporal plane; (2) a lateral, internal auditory parakoniocortex (PaAl), lateral to A1; and (3) a caudal auditory parakoniocortex (PaAc), covering the caudal portion of the superior temporal plane and extending around the insula to the parietal operculum. In its turn, the parakoniocortex is surrounded by an extensive area of auditory cortex that covers the remaining of the superior temporal plane and the lateral surface of the superior temporal gyrus, except for its rostral pole. This region was described as area 22 by Brodmann (1909), as TA by von Economo and Koskinas (1925) and as external auditory parakoniocortex (PaAe) by Galaburda and Sanides (1980).

In primates (Fig. 7.13), anatomical and physiological studies defined a **belt area** surrounding the core rostrally, laterally and caudally (Pandya and Sanides 1973; Galaburda and Pandya 1983; Morel and Kaas 1992; Morel et al. 1993; Hackett et al. 1998a). The area rostral and lateral to the belt is nowadays known as the **parabelt** (Morel et al. 1993; Hackett et al. 1998a). Both belt and parabelt areas differ from the core area in their pattern of thalamic input. The macaque belt area receives projections from only the medial and dorsal divisions of the MGB (Rauschecker et al. 1997), whereas the parabelt area is also innervated by these two divisions of the MGB but, moreover, by the medial division of the pulvinar (Trojanowski and Jacobson 1975; Burton and Jones 1976; Hackett et al. 1998b).

Ablation of the core of macaque auditory cortex eliminates responses to auditory stimuli in the adjacent belt region (Rauschecker et al. 1997), suggesting that input from the medial and dorsal geniculate nuclei is not sufficient to support auditory processing in the absence of direct projections from the ventral geniculate nucleus. Instead, information processing to the secondary auditory cortical areas appears to depend on transcortical projections that pass successively from core to belt to parabelt cortex (Jones and Powell 1970; Seltzer and Pandya 1978; FitzPatrick and Imig 1980; Luethke et al. 1989; Morel and Kaas 1992; Morel et al. 1993; Hackett et al. 1998a). Tardif and Clarke (2001) studied the intrinsic connectivity of human auditory areas with anterograde and retrograde labelling of the carbocyanine dye DiI. With DTI, the tracts connecting the Heschl's gyri via the corpus callosum have been studied (Hofer and Frahm 2006; Westerhausen et al. 2009). These interhemispheric connections are located more rostrally within the posterior callosal third than those connecting the posterior parts of both superior temporal gyri.

The idea of a two-stream, **what/where** organization of sensory cortex originated in the visual system (see Chap. 8). In rhesus monkeys, such a dichotomy has also been demonstrated for the cortical auditory projections (Rauschecker and Tian 2000). The **'where' (dorsal) pathway** is thought to link A1 via the caudomedial belt with the frontal eye field and parietal targets (Romanski et al. 1999; Fig. 7.14) that are implicated in *spatial processing*. The **'what' (ventral) pathway** is thought to represent a *pattern information* stream that originates in the anterior core and belt areas and influences targets within the temporal lobe. A similar **two-stream organization** may exist in the human auditory cortex (Griffiths et al. 2000; Alain et al. 2001; Maeder et al. 2001; Wessinger et al. 2001; Clarke et al. 2002). The right insula is activated by a moving sound image (Griffiths et al. 1994) and, conversely, a patient with a right hemispheric stroke causing atrophy of the right insula was unable to detect sound source movement by either phase or loudness cues (Griffiths et al. 1996). Subjects listening to stimulus movement stimulated by changes in binaural timing show maximal activity in the inferior parietal area, particularly on the right side (Griffiths et al. 1998; Weeks et al. 1999). These findings suggest: (1) that there is a transcortical passage of information from

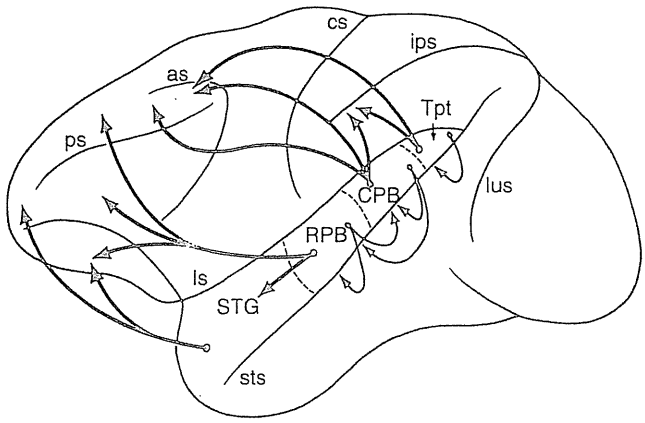

Fig. 7.14 Topography of auditory-related projections. Caudal (*CPB*) and rostral (*RPB*) subdivisions of the parabelt and the superior temporal gyrus (*STG*) project topographically to segregated regions of superior temporal, posterior parietal and prefrontal cortices. *Abbreviations*: *as* arcuate sulcus; *cs* central sulcus; *ips* intraparietal sulcus; *ls* lateral sulcus; *lus* lunate sulcus; *ps* principal sulcus; *sts* superior temporal sulcus; *Tpt* temporoparietal area (after. Hackett and Kaas 2004)

auditory koniocortex mediocaudalwards across the insula into the parietal lobe, during processing of information on sound source position and motion (see Hackett and Kaas 2004); and (2) a dominant functional role of the right hemisphere in sound localization.

In **functional imaging studies**, simple auditory tasks such as passive listening to white noise bursts, tones or consonant-vowel speech syllables, activate restricted areas within the lateral fissure on the superior temporal plane (Zatorre et al. 1992, 1994; Binder et al. 1994, 1997, 2000; Zatorre and Binder 2000). The extent of the activation varies from subject to subject and may spread rostralwards and caudalwards on the superior temporal plane. The area of activation is generally within and around the transverse temporal gyrus. With exposure of subjects to more complex stimuli such as passive listening to tone patterns, single words, pseudowords or narrative text, activity is not only present in the cortex of the superior temporal gyrus, but now foci of activation appear on the lateral aspect of the superior temporal gyrus in area 22/TA/PaAc (Binder et al. 1994). The human primary auditory cortex is functionally organized in a **tonotopic manner**. In a combined fMRI and DTI study, Upadhyay et al. (2007) showed that the connectivity pattern in the human primary auditory cortex is similar to that described in tonotopic mapping studies on macaque monkeys (Morel et al. 1993) and cats (Lee et al. 2004; Lee and Winer 2005).

In general, activity is **bilaterally equal**. With complex stimuli, language in particular, the question arises whether there is a right–left asymmetry in the response. Since handedness influences hemipheric lateralization, imaging studies of speech processing are normally restricted to neurologically normal right-handers. In them, there is a tendency for greater activation of the left hemisphere during tasks that depend on word meaning. Left lateralization of speech characterizes both males and females (Frost et al. 1999). The functional significance of greater left hemispheric activity is implied by imaging studies of stroke patients after infarctions of the left perisylvian area (Weiller et al. 1995; Heiss et al. 1997; Mummery et al. 1999). Subjects who showed good recovery of speech perception had increasing activation of the left temporal cortex surrounding the infarct. Some indications for an **opposite asymmetry** in processing **musical stimuli** come from cases of pathology:

1. A patient with a **right** thalamic tumour experienced ***distorted perception*** of ***music*** but not of voices (Roeser and Daly 1974).
2. Cortical activity has been demonstrated in the right superior temporal lobe during ***musical hallucinations*** (Kasai et al. 1999).
3. A case of ***amusia***, a form of auditory agnosia, was seen after an infarct involving the right insula (Griffiths et al. 1997b).

7.4.4 Auditory Disorders Related to Stroke

Disorders of auditory perception may follow strokes in the territory of the internal carotid arteries or of the vertebrobasilar system (Levine and Häusler 2001; Lechevalier et al. 2007; Kaga 2009), and appear as:

- ***Auditory agnosia***, the impossibility of recognizing environmental sounds, words and music, which the patient, however, is said to hear
- ***Pure word deafness***, the impossibility to understand spoken language to repeat or to write under dictation in the absence of other signs of aphasia
- ***Cortical deafness***, the feeling of being deaf contrasting with the integrity of the tonal audiogram
- ***Amusia***, auditory agnosia specific for music

The central disorders of auditory perception may result from lesions of either the right, the left or both cerebral hemispheres, usually involving parietotemporal cortical areas. ***Cortical deafness*** is characterized by bilateral abolition of the middle and late latencies of auditory potentials, caused by bilateral lesions of the primary auditory cortices. Such patients have the feeling of being deaf to all types of auditory stimuli, but often say they are *not* deaf, rather that they do not understand what is said to them. The term ***subcortical deafness*** is used to indicate an auditory disorder clinically identical to cortical deafness, but due to lesions in subcortical areas of the brain. It was first described by Le Gros Clark and Russell (1938). The ischaemic lesions involved the two external capsules and extended sufficiently downwards to interrupt the acoustic radiations, while sparing the auditory cortices. Recent cases were reported by Woods (1996), Levine and Häusler (2001) and Kaga et al. (2005; see ***Clinical case 7.3***).

Since the pioneering studies of Ferrier (1875) and Henschen (1920), there has been a longstanding debate as to whether bilateral destruction of either the primary auditory cortex or the acoustic radiation results in ***auditory agnosia***. In macaque monkeys, bilateral lesions of the primary auditory cortex apparently do not cause permanent deafness (Heffner and Heffner 1990). Less recovery of function in the human brain, compatible with the clinical diagnosis of auditory agnosia, may or may not have been caused by the inclusion of the surrounding auditory association areas (Lechevalier et al. 2007).

Tanaka et al. (1991) differentiated three clinical syndromes of auditory agnosia: (1) ***disconnection syndromes***, destroying the acoustic radiation and causing auditory agnosia (*prephonemic deficit*); (2) ***cortical lesions*** of the left superior temporal lobe may result in pure word deafness (*linguistic deficit*) and (3) unilateral or bilateral ***temporoparietal*** or ***subcortical lesions*** have been documented in patients with non-verbal auditory agnosia (*deficit* to *environmental sounds*). Lesions occurring peripherally to the MGB (*prethalamic*) may cause hearing loss and those bilaterally located centrally to the MGB (*postthalamic*) may result in auditory agnosia. Small lesions of the MGB may be related to auditory hallucinations (Fukutake and Hattori 1998). ***Pure word deafness*** may be the result of left or bilateral temporal lesions, possibly due to disconnection as suggested by Liepmann and Storch (1902). Recent cases were reported by Kaga et al. (2000; see ***Clinical case 7.4***) and Levine and Häusler (2001).

Disorders of ***music perception*** following cerebral damage can be divided into two categories (Lechevalier et al. 2007):

1. ***Multimode perceptive disorders*** affecting more or selectively musical sounds, but with verbal and environmental sound difficulties
2. A ***pure amusia***, where only music perception is affected (for ***congenital amusia*** see Ayotte et al. 2000)

In both monkeys and humans, neurons in **core areas** respond strongly to narrow-band sounds such as tones, whereas neurons in **belt areas** respond better to more complex sounds such as noise (Wessinger et al. 2001; Rauschecker and Tian 2004; Tian and Rauschecker 2004; Bendor and Wang 2006). Within the core areas, two mirror symmetric **tonotopic maps** sharing a low-frequency border have been identified, corresponding to A1 and the rostral field R (Formisano et al. 2003; Bendor and Wang 2006). In monkeys, a third core area (RT) has been found that lies rostral to R (Kaas and Hackett 2000; Hackett and Kaas 2004). Kaas and Hackett postulated that each core area is connected to medial and lateral neighbouring belt areas (see Fig. 7.14), with additional belt areas located on the rostral and caudal ends of the core. Three of these lateral belt areas (caudal-lateral, middle-lateral and antero-lateral) have been mapped electrophysiologically and possess similar mirror tonotopic maps to those of their adjacent core (Rauschecker and Tian 2004; Tian and Rauschecker 2004). In an fMRI study, Patterson et al. (2002) identified a specific region in the lateral part of Heschl's gyrus that was preferentially activated by temporally regular sounds with a **pitch**. They determined that only lateral Heschl's gyrus, a non-primary auditory region rostrolateral to the primary auditory cortex, responded to the temporal regularity of pitch of the acoustic stimuli. Other imaging studies (Penagos et al. 2004; Schneider et al. 2005) have confirmed these findings.

Musical perception is not a uniform competence in the general population. Some patients will have had musical training, others not. Peretz (2001) estimated that 5–10% of individuals are completely unable to distinguish the pitches of two notes of music or to memorize the smallest musical tone. Geschwind and Galaburda (1985) suggested that rightward deviation from the usual pattern of cerebral asymmetry may be associated with increased giftedness for talents for which the right hemisphere is assumed to be important. With MR morphometry, Schlaug et al. (1995) presented evidence for structural brain asymmetry in musicians. Musicians with **perfect pitch** revealed stronger leftward asymmetry of the planum temporale than non-musicians or musicians without perfect pitch. This suggests that outstanding musical ability is associated with increased leftward asymmetry of the cortex subserving music-related functions.

Neuropsychological studies in epileptic patients who underwent a unilateral temporal cortectomy have contributed to our knowledge of the localization of musical functions (Liégeois-Chauvel et al. 1998). A right temporal cortectomy was found to disturb melodic perception as well as the perception of pitch intervals, whereas a left-sided lobectomy did not disturb perception of the intervals. These data underline the key role of the superior temporal gyrus in discrimination of melodies. Cortectomy of the posterior part of T1, including the planum temporale, the lateral part of Heschl's gyrus and Brodmann area 22, is more striking for the processing of pitch and variations of rhythm than cortectomy of the rostral part of T1. Disorders of the perception of rhythm and metre (recognition of a cadence of march or waltz) can be dissociated. The right and left rostral parts of T1 would be implicated in the processing of metre. Griffiths et al. (1997b) reported a patient with lesions of the middle and posterior temporal areas and the insula of the right hemisphere. The patient complained of not being able to appreciate music. Neuropsychological testing showed a deficit of musical perception without disturbance of the perception of noises, environmental sound and speech sounds. His ability to detect continuous changes of sound frequency was preserved. However, a disturbance in the analysis of rapid sequences of notes seemed to be the basis of his musical perception deficit. Neuroimaging studies have revealed that rhythm perception

activates area 44 and that detection of pitch changes relies on the left cuneus and precuneus (Platel et al. 1997, 2003).

Auditory hallucinations are observed in brain stem (Ross et al. 1975; Cambier et al. 1987; Fisher and Tapia 1987; Griffiths 2000) and temporal lobe (Lechevalier et al. 1985) strokes. Cambier et al. (1987) reported five purely auditory observations of ***hallucinosis*** (hallucinations, regarded by the patient as abnormal), four of which were attributed to paramedian strokes of the pons and one to an infarct of the dorsolateral mesencephalon. Auditory hallucinations following temporal lobe lesions are unusual and have specific characteristics (Liepmann and Storch 1902; Lechevalier et al. 1985; Augustin et al. 2001; Evers and Ellger 2004; Sacks 2007; see ***Clinical case 7.5***).

Clinical Case 7.3 Auditory Agnosia Caused by Bilateral Lesions Restricted to the Auditory Radiations

Bilateral lesions of the auditory radiations are rare (Tanaka et al. 1991; Woods 1996; Kaga et al. 2005; see **Case report**).

Case report: Kaga et al. (2005) reported a patient with auditory agnosia due to bilateral lesions of the auditory radiations. A 43-year-old male patient experienced mild left temporal hemiplegia due to a right putaminal haemorrhage. He recovered completely but hypertension persisted. When he was 53 years old, he had a left putaminal haemorrhage and went into a coma. After recovering from the coma and the right hemiplegia, he could hear but could not discriminate speech sounds. Brain CT and MRI demonstrated small bilateral lesions restricted to the auditory radiations (Fig. 7.15a, b). Pure-tone audiograms recorded 1 and 4 years after the second haemorrhage are shown in Fig. 7.15c, d. MEG demonstrated the disappearance of middle latency responses and AEP studies showed a very small Pa peak. In contrast, a positron emission tomographic study showed a marked bilateral increase in blood flow in the auditory cortex in response to both click and monosyllable stimuli. This may be due to activation of the auditory cortex via non-specific pathways.

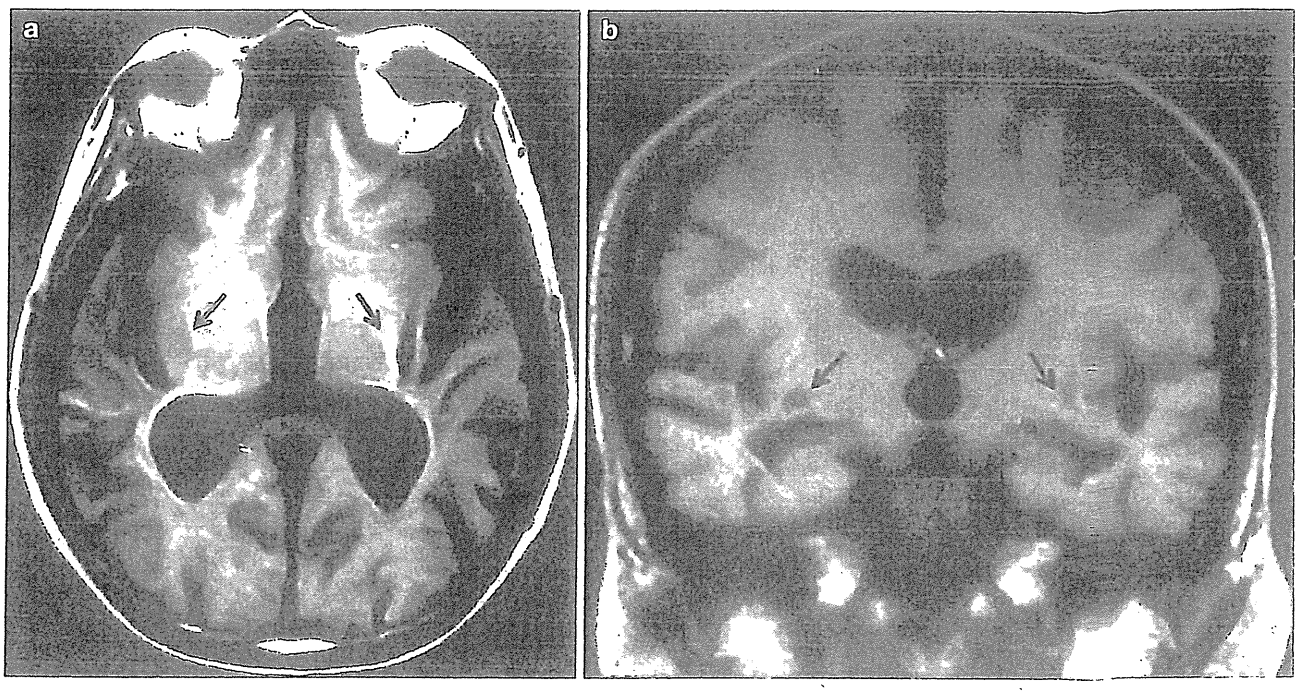

Fig. 7.15 Auditory agnosia caused by bilateral lesions restricted to the auditory radiations. In the axial (**a**) and coronal (**b**) MRIs, the auditory radiations are bilaterally damaged (*arrows*) by small brain infarcts. Pure-tone audiograms recorded one (**c**) and four (**d**) years after the second haemorrhage (from Kaga et al. 2005)

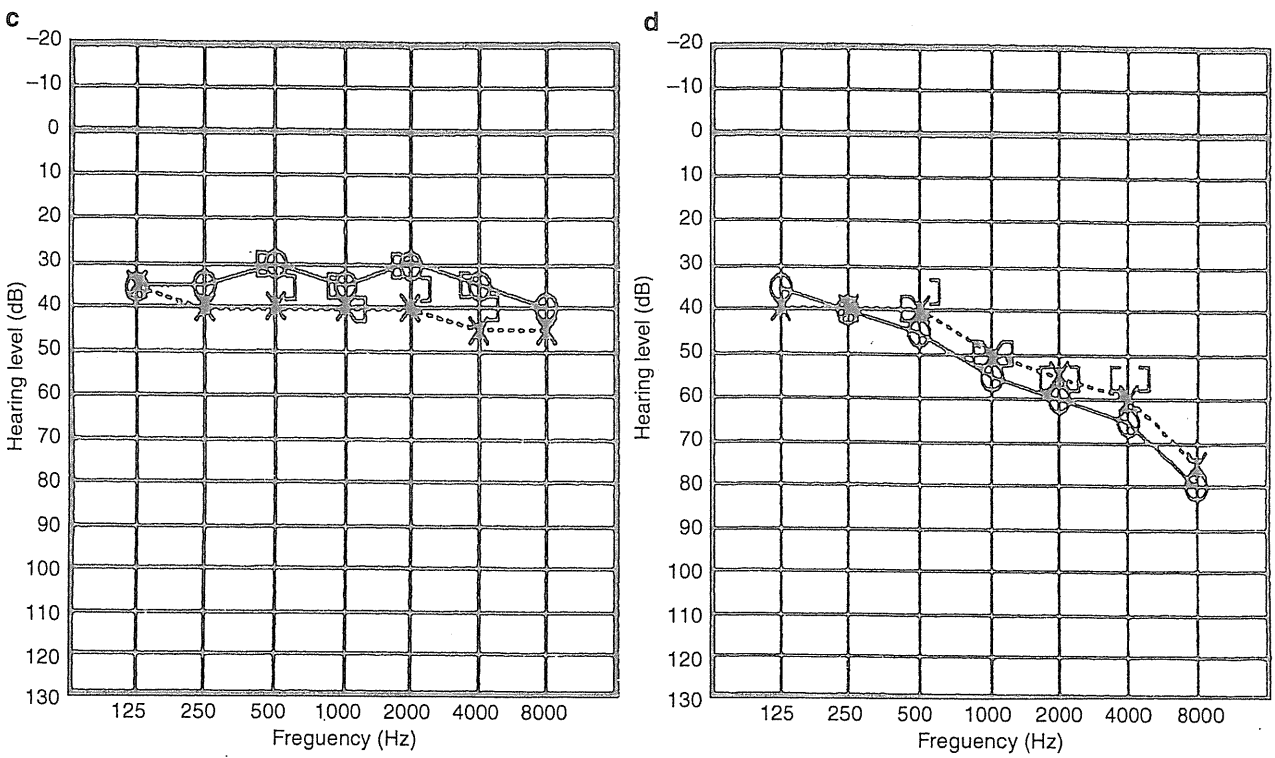

Fig. 7.15 (continued)

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Clinical Case 7.4 Neuropathology of Auditory Agnosia Following Bilateral Temporal Lobe Infarction

Severe auditory deficits due to bilateral lesions of the primary auditory cortex or the auditory radiations is very rare. The resulting hearing problem is referred to as ***auditory agnosia*** or ***cortical deafness***. Kaga et al. (2000) reported a patient who came to autopsy (see **Case report**).

Case report: Kaga's case of auditory agnosia due to bilateral lesions of the auditory cortex was first diagnosed in 1975 when the patient was 37 years old. He was admitted to hospital for examination following his second cerebrovascular accident. MRI of the lesions on admission is shown in Fig. 7.16a, b. A comprehensive follow-up examination of auditory function was periodically conducted until his sudden death 15 years later. His brain was studied neuropathologically. Initial pure-tone audiometry revealed moderate sensorineural hearing loss in the right ear and mild sensorineural hearing loss in the left ear. Repeated pure-tone audiometry revealed that bilaterally thresholds became progressively poorer over time. Speech audiometry of both ears consistently revealed that the patient was unable to discriminate any monosyllabic words. In general, speech and hearing tests demonstrated that he could not comprehend spoken words but could comprehend written commands and gestures. Neuropathological examination of the brain revealed a total defect and neuronal loss of the superior temporal gyrus, including Heschl's gyrus, and total gliosis of the MGB (MGB; Fig. 7.16c, d). In the right hemisphere, subcortical necrosis, gliosis in the centre of the superior temporal gyrus and partial gliosis of the MGB were found (Fig. 7.16c, e). These data support the clinical observations of imperception of speech sounds, music and environmental sounds, which may be due to progressive degeneration of both MGBs.

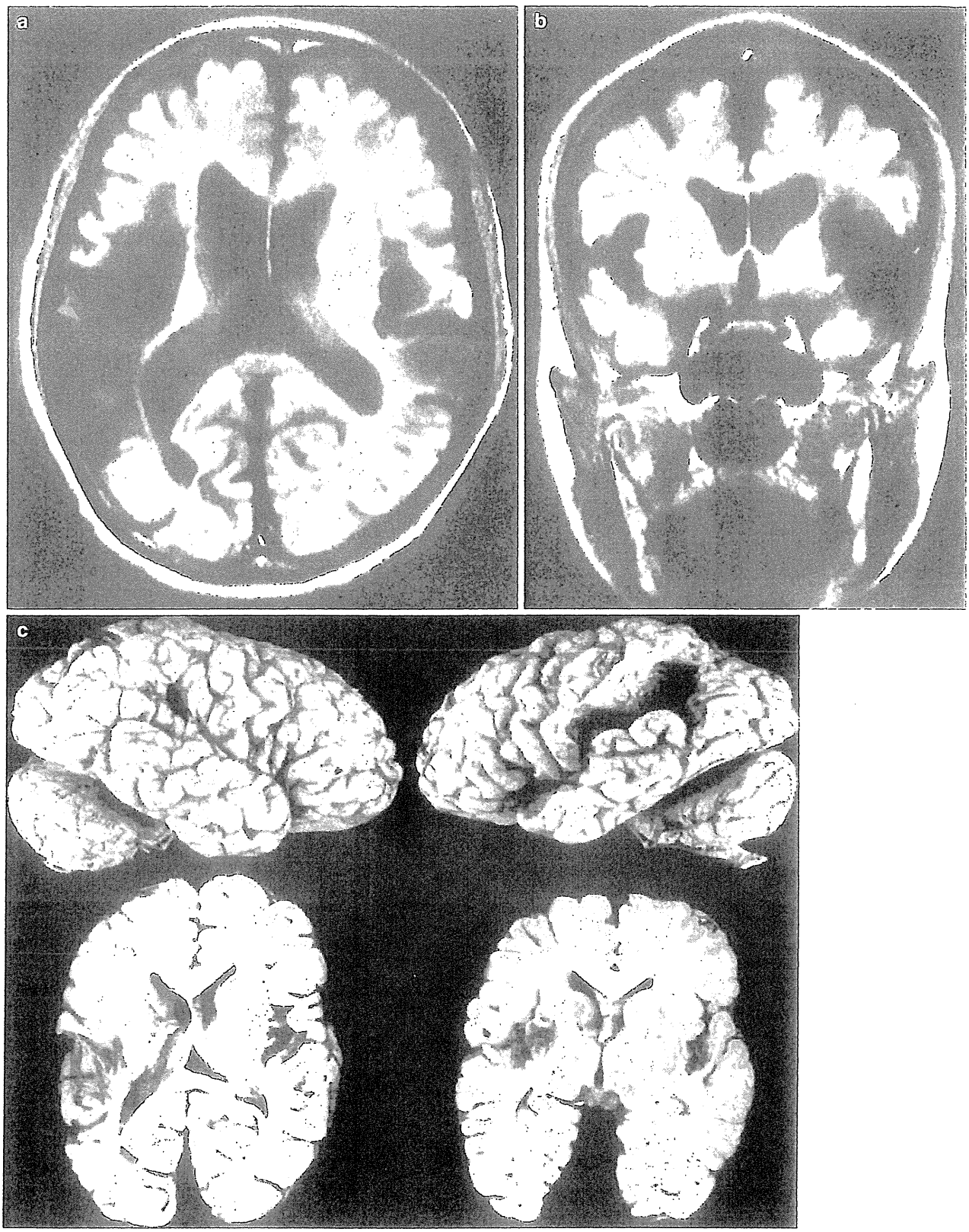

Fig. 7.16 Auditory agnosia following bilateral temporal lobe infarction. (**a**, **b**) Axial (here the left side is on the left) and coronal (here the left side is on the right) MRIs showing a large infarct in the left hemisphere and a small infarct in the right hemisphere including the auditory cortex. (**c**) Lateral views of the brain and two horizontal sections in which the auditory cortex is present. In the right hemisphere, a small infarct is present in the upper part of the lateral sulcus, whereas in the left hemisphere extensive infarction can be seen in Broca's area, the superior temporal gyrus and the supramarginal gyrus. (**d**, **e**) HE-stained sections of the medial geniculate body (MGB). In the left MGB, neurons have been completely replaced by glial cells (**d**), whereas in the right MGB (**e**) there is partial neuronal preservation (from Kaga et al. 2000)

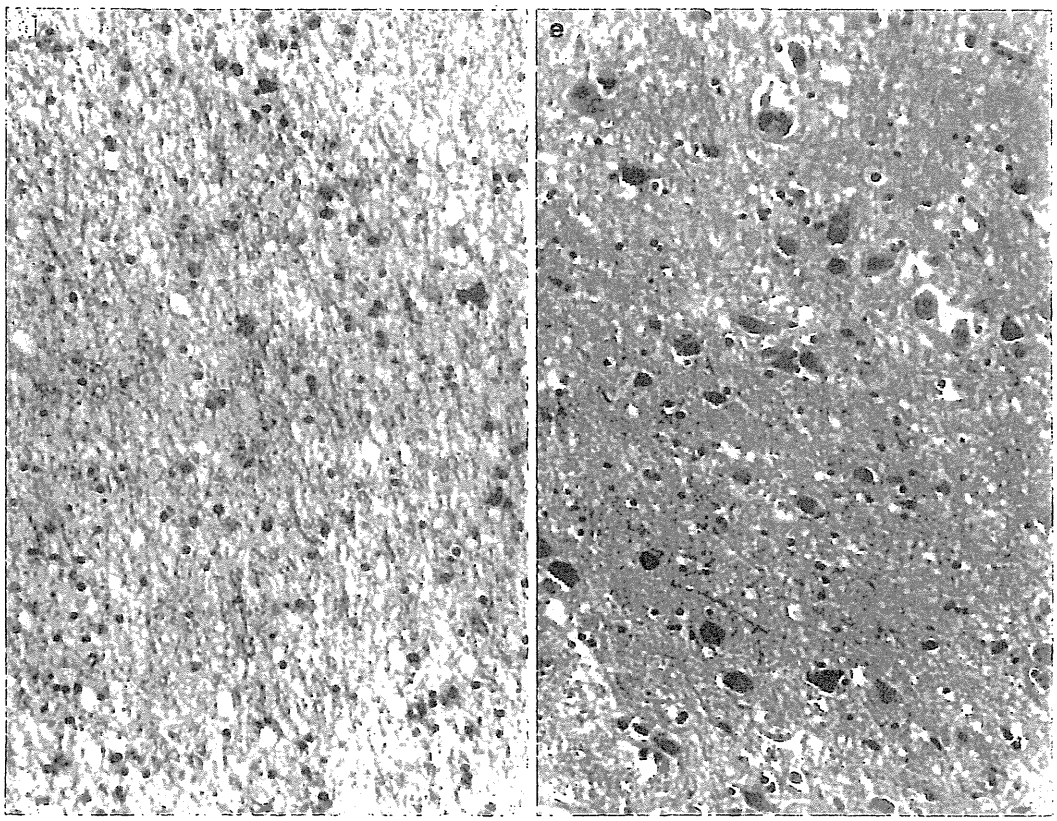

Fig. 7.16 (continued)

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Clinical Case 7.5 Auditory Hallucinations Following a Metastasis in Heschl's Gyrus

Case report: A 64-year-old patient presented with word-finding difficulties. He suffered from coronary sclerosis with exercise-induced angina pectoris but he had no previous neurological complaints. On neurological examination, there were no focal signs but his speech was non-fluent with word-finding difficulties and suboptimal comprehension. On hospital admission, he repeatedly complained of ***auditory hallucinations***, consisting of incomprehensible words and sounds. On MRI, a contrast-enhancing lesion was found in the left gyrus of Heschl (Fig. 7.17) that appeared to be part of a more lobular contrast-enhancing in the left parietotemporal region with surrounding oedema. The auditory hallucinations disappeared on treatment with dexamethasone. A biopsy showed that the tumour was a gemistocytary astrocytoma for which he was treated with radiotherapy and temozolamide.

This case was kindly provided by Peter van Domburg (Department of Neurology, Orbis Medical Centre, Sittard, The Netherlands).

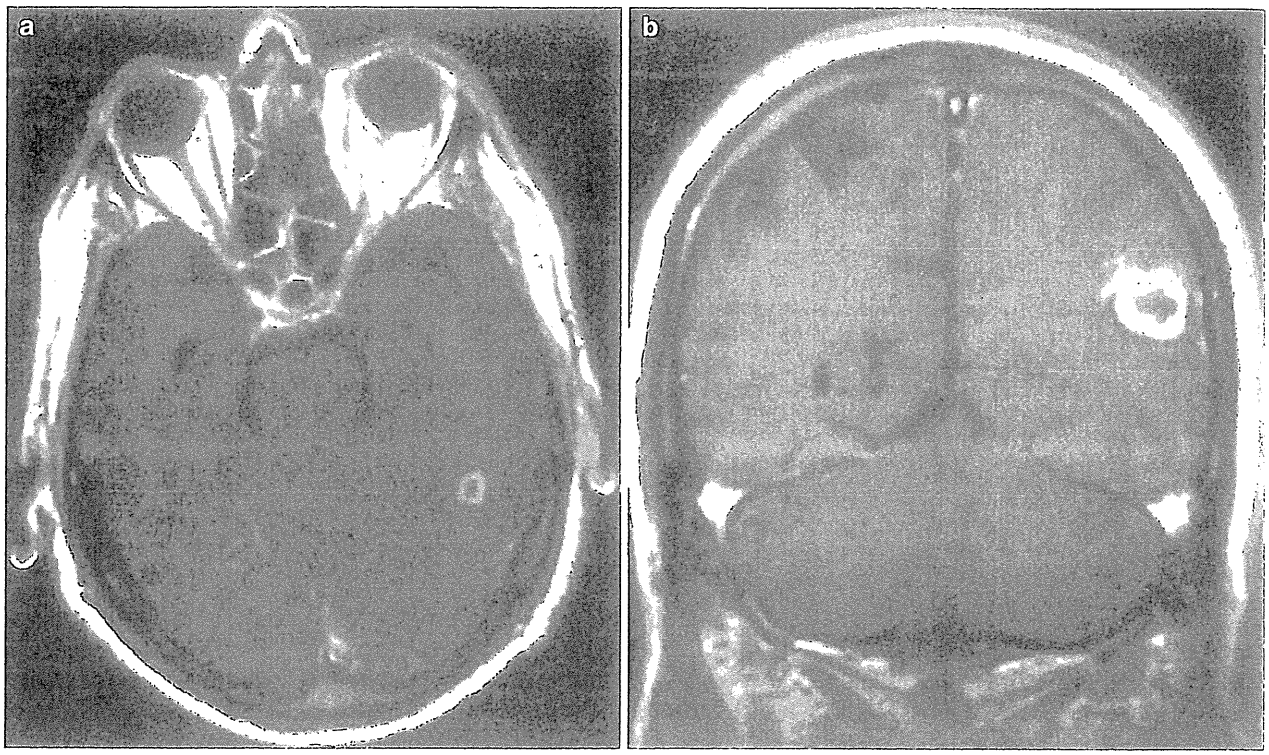

Fig. 7.17 T1-contrast MRIs of a metastasis in the left gyrus of Heschl that caused auditory hallucinations (courtesy Peter van Domburg, Sittard)

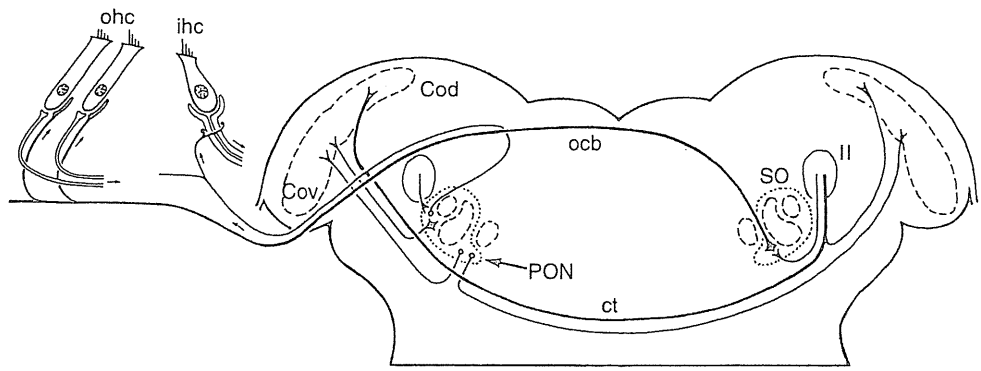

Fig. 7.18 Efferent control of the cochlea. *Abbreviations*: *Cod*, *Cov* dorsal and ventral cochlear nuclei; *ct* corpus trapezoideum; *ihc* inner hair cells; *ll* lateral lemniscus; *ocb* olivocochlear bundle; *ohc* outer hair cells; *PON* periolivary nuclei; *SO* superior olivary complex (after Nieuwenhuys 1984)

7.5 The Descending Auditory System

Parallel with the pathways from the organ of Corti to the auditory cortex, there is an uninterrupted chain of neurons conducting impulses in the opposite, descending direction. The final link in this **descending auditory system** is formed by the **olivocochlear bundle** of **Rasmussen**, which originates in the **peri-olivary nuclei** around the superior olivary nucleus (Fig. 7.18). Most of the fibres of the olivocerebellar bundle decussate in the tegmentum. They enter the vestibular nerve and join the cochlear nerve via the vestibulocochlear anastomosis (Schuknecht 1993) to terminate in the inner and outer hair cells of the organ of Corti. The human olivocochlear system has been identified with acetylcholinesterase histochemistry (Schuknecht et al. 1959) and choline acetyltransferase immunohistochemistry (Moore et al. 1999; Moore and Linthicum 2004).

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知っておきたい用語の解説

新生児聴覚スクリーニング

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Ⅰ．背　景

先天性難聴児は500～1,000人の出生に対し1人の割合で生まれる。これは世界共通である。先天性疾患の中で最も頻度が高い。いかにして産科入院中に電気生理学的，あるいは他覚的に難聴を発見するかを目的として技術が2つ開発された。1つはThorntonらによる自動ABR（Automated auditory brainstem response, AABR）で，もう1つは英国のKempが発見したOAE（Otoacoustic emission）耳音響放射検査である[1]。米国のItanoはAABRを用いて新生児聴覚スクリーニングに取り組み，精密聴力検査による真の難聴児について補聴器装用下の言語発達の追跡研究を行った[2]。すなわち，生後6カ月前より難聴が発見され補聴下の教育をしたグループと，生後6カ月以降に難聴が発見され，補聴下の教育を受けたグループに分けた。両グループを3歳になった時点での言語力を評価したところ，難聴の軽重にかかわらず，6カ月前のグループの方が6カ月以後のグループに比し有意に高い言語力を獲得することを1998年に報告した。この報告は世界各国に強い影響を与えたが，わが国もその一つである。

Ⅱ．目　的

出来る限り新生児期の聴覚スクリーニングを全国に普及させ，難聴の疑い例を次のステップである難聴の精密聴力検査に進める。このようにして真の難聴児を早期発見し，生後6カ月以内に補聴を早期に行い，成人した時に一般社会で共存共生して生きていけるだけの聴覚と言語力を身につけ聴いて話せるようにする。

Ⅲ．スクリーニングの方法（産科・新生児科）

AABRとOAEの2つの方法が使用されている。いずれの検査機器も外国製品で300～400万円もする。

1．AABR

35～40dBをスクリーニングのレベルに設定されている。ABRの波形そのものは出ない。結果だけがpassあるいはreferとして出る。したがって軽～中等度の難聴もreferとしてスクリーニングされる（図1）[3]。

2．耳音響放射

聴覚検査用のOAEにはTOAE（Transient otoacoustic emission）とDPOAE（Distortion product otoacoustic emission）の2つがある。15～20 dB前後がスクリーニング

〒152-8902　東京都目黒区東が丘2-5-1

音の大きさ（デシベル：dB）	聴力レベル
～20dB 以内	正常聴力
20～50	軽度難聴
50～70	中等度難聴
70～90	高度難聴
90dB 以上	重度難聴

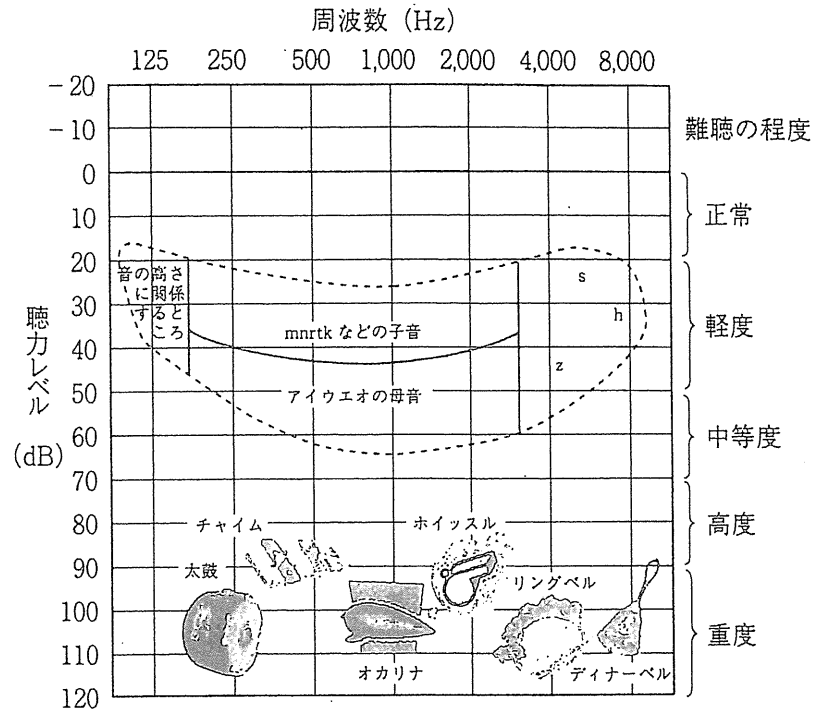

図1　難聴の重さの分類と新生児聴覚スクリーニングレベル（35～40dB）

レベルとなる。結果は反応あり，あるいは反応なしとして表現される。したがって中耳に滲出液があると反応は出現しない。

3．注意すべき点

AABR と OAE の両検査とも，もし refer であっても軽・中等度難聴か高度難聴か重度難聴か全く区別ができない。

Ⅳ．精密聴力検査（耳鼻咽喉科）

スクリーニングで refer とされた新生児は，生後1カ月前後で耳鼻咽喉科の外来で，① ABR，DPOAE，②行動反応聴力検査，③小児神経耳科的に立直り反射や原始反射の検査を行って総合的に診断する。

1．ABR

ABR は閾値だけでなく，強刺激時の ABR の波形も参考にする。ABR 強刺激で無反応であっても潜時 0～2 msec の間に，蝸牛マイクロフォン電位（CM，Cochlear microphonics），加重電位（-SP，summating potential）の有無もチェックする。強刺激の波形が Wave Ⅰを含め波形全体の潜時が延長していると伝音難聴成分が含まれることが多い。閾値が中等度の場合，Latency intensity curve を描き，伝音性か感音性か判断する。Wave Ⅰと Wave Ⅴの波間潜時が著しく延長している時には脳幹の未熟性あるいは脳幹障害を疑う。ABR が無反応であっても新生児期の蝸牛や脳幹の未成熟のために難聴がないこともあり，次に記載する行動反応聴力検査と比較して診断する。たとえ ABR が無反応でも残存聴力はほとんどの例で存在する[4]。

2．行動反応聴力検査（Behavioral Audiometry）

防音室で行う検査で，スピーカーより各周波数ごとに音圧を変えて音刺激を与え，驚愕反射（目を開ける，目を閉じる），定位反射（ふりむき反射）などを見る。このほかにネオメーターやインファントオージオメーターのように限られた周波数の音圧を変え反応を観察するものや，鈴や太鼓などを併用して音刺激に対しての反応の有無をチェックする。小生は伝声管（通称，ベートーベンの補聴器，ラッパ補聴器，トランペット型補聴器などと呼ばれる）を用いて新生児の反応を観察する（図2）。これは大いにすすめられる方