第6章 耳介の先天異常と小耳症

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キーワード

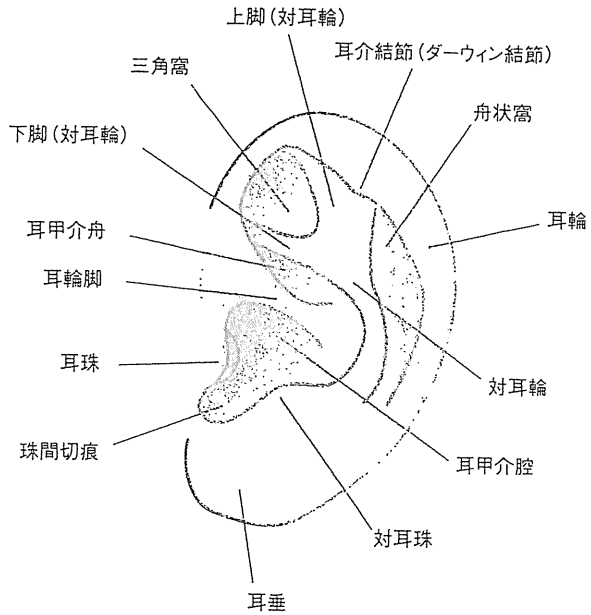
形成外科と耳鼻咽喉科共同のアプローチ
小耳症外耳道閉鎖症
耳介形態の再建
外耳道形成
副耳
耳瘻孔
耳垂裂
折れ耳
constricted ear
埋没耳
立ち耳
スタール耳
無耳症

はじめに

耳介は複雑な形態を示すが(図1)、その各部は胎生期の第一鰓弓と第二鰓弓、両者の間に存在する第一鰓溝から発生する。発生期の何らかの異常により各種の耳介形態異常が現れ、形成外科的治療の適応となる。

本稿では各種の耳介先天異常について述べるとともに、最も高度な変形を示す小耳症外耳道閉鎖症に対する形成外科と耳鼻咽喉科共同のアプローチについて紹介する。

小耳症外耳道閉鎖症に対しては、従来から形成外科では耳介形態の再建、耳鼻咽喉科では外耳道形成が別々に行われていた。先に外耳道形成のみが行われた場合、瘢痕形成のため繊細な耳介形態を再建することは困難であった。一方、耳介形態のみが再建されても耳介の位置が適切でないと、後に再建部位に対して外耳道形成を行うことができない。われわれは両科共同によるアプローチによってこれらの問題点を解決し、形態再建と機能再建の両立を目指している。



【図1】耳介各部位の名称

1 各種の耳介先天異常

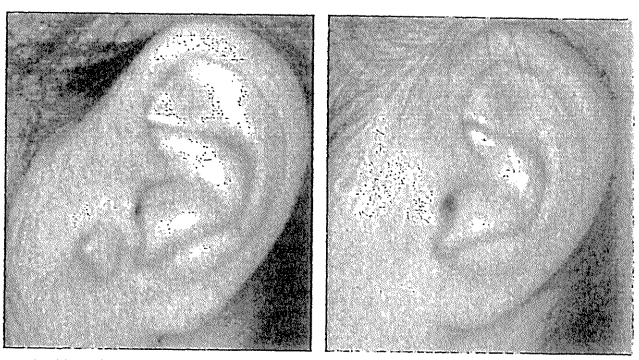
1 軽度の異常

副耳

通常耳珠の前方に存在する皮膚の隆起で、軟骨成分を含む場合が多い。茎が細い場合は糸で結紮するのみで脱落する場合もあるが、隆起が残存して見えることもあるので、切除縫合が確実である(図2)。

耳瘻孔

耳瘻孔には耳輪脚の前方に皮膚開口部を持つ耳前瘻孔と、耳介部分に開口する耳介瘻孔がある(図2)。深さはまちまちであるが、底部で軟骨に達するものが多い。時によって感染を起こす場合があるので根治切除の適応になるが、軟骨の一部を含めて完全切除しないと再発することがある。

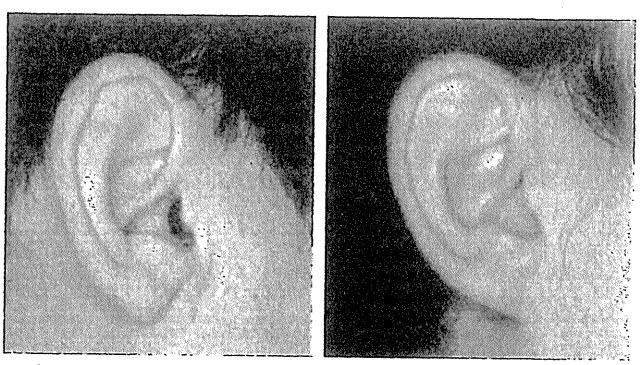


A:耳輪前方の耳前瘻孔、耳輪脚部分の耳介瘻孔、耳珠前方の副耳を合併した症例　B:切除術後4ヵ月の状態

【図2】副耳と耳瘻孔の合併例

耳垂裂

耳垂部分に切痕を生じているもので、切痕の向きや大きさはさまざまである。また、裂が高度のものは耳垂欠損と呼ばれる状態となる。単純に裂を閉じるだけでは瘢痕拘縮によりnotchを生じ、裂が残ったような形状になるので、局所皮弁術を応用した各種の方法が行われる。右の症例はW形成術を応用した方法(山田法)が用いられている(図3)。



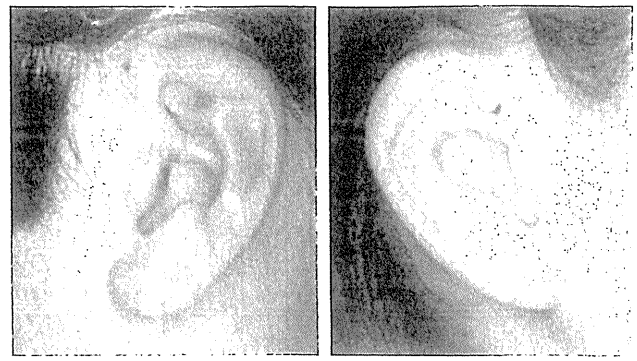
A:典型的な耳垂裂の症例、術前の状態　B:形成術を行って6ヵ月後の状態

【図3】耳垂裂

折れ耳とconstricted ear

折れ耳は、耳介の上方部が折れ曲がっている変形で、耳介自体の大きさが正常なものをいう(図4A)。折れ耳に近い変形を呈し、耳輪の長さが短縮している場合、constricted earと呼ばれる(図4B)。

いずれも対耳輪上脚部分の形成が不十分であり、この部位の軟骨形成を行う必要がある。またconstricted earは短縮した耳輪部分を広げて耳甲介部からの軟骨移植を必要とする場合がある。Constricted earが高度になったものが小耳症の耳甲介型であると考えられる。



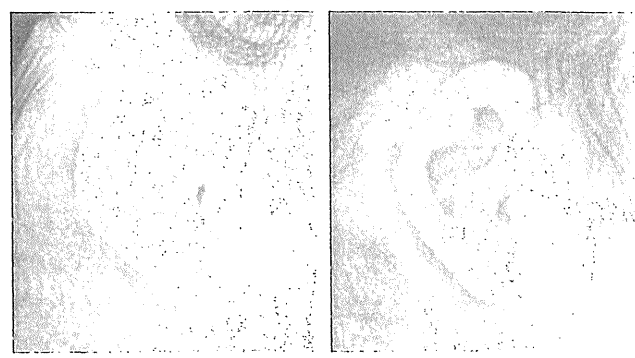
A：折れ耳　B：constricted ear

【図4】折れ耳とconstricted ear

2 中等度の異常

埋没耳

耳介軟骨の上部が側頭部皮下に埋没した状態になっているものを指す。用手的に引き出せるが手を離すと埋没状態に戻るため、マスクやめがねの装着に支障を来す。乳幼児期には非観血的矯正を行うことによって軽快する場合もあるが、多くは軽度の耳介軟骨の変形を伴うため、手術で耳介後面の筋の切離と軟骨形成を行う(図5)。

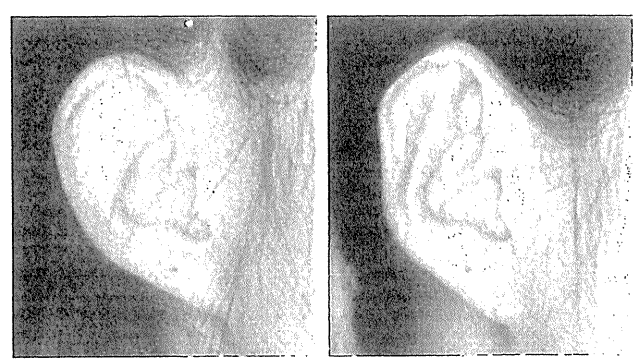


A：術前の状態　B：術後1年の状態、マスクの装着が可能となっている

【図5】埋没耳

立ち耳

耳介が側頭部から異常に聳立した状態で、多くは対耳輪の湾曲が不十分なため、あるいは耳甲介後壁が異常に高いために生じる。特に西洋ではコミカルな醜形として嫌われ、いじめの対象ともなるため手術がさかんに行われる。耳甲介腔の軟骨切除や対耳輪の軟骨形成を行う(図6)。

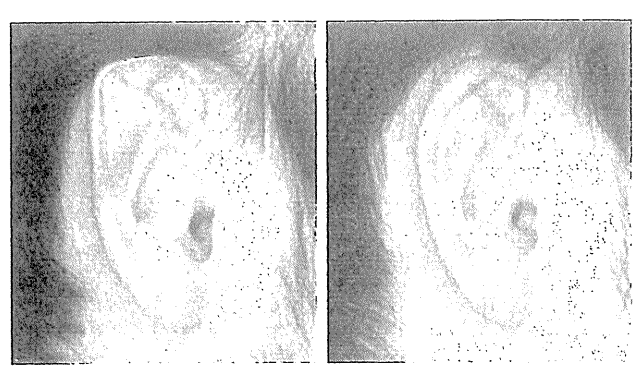


A：術前の状態　B：対耳輪の軟骨形成を行い、術後10ヵ月の状態

【図6】立ち耳

スタール耳

対耳輪が上脚と下脚のみでなく上方ないし後方へ第3脚として分枝して変形した耳介形態となっているものを指す。対輪第3脚(third crus of antihelix)とも呼ばれる。乳幼児期には矯正治療も行われるが、手術においては軟骨の形成を要し、さまざまな術式の報告がある(図7)。



A：術前の状態　B：軟骨形成術後6ヵ月の状態

【図7】スタール耳

3 高度の異常

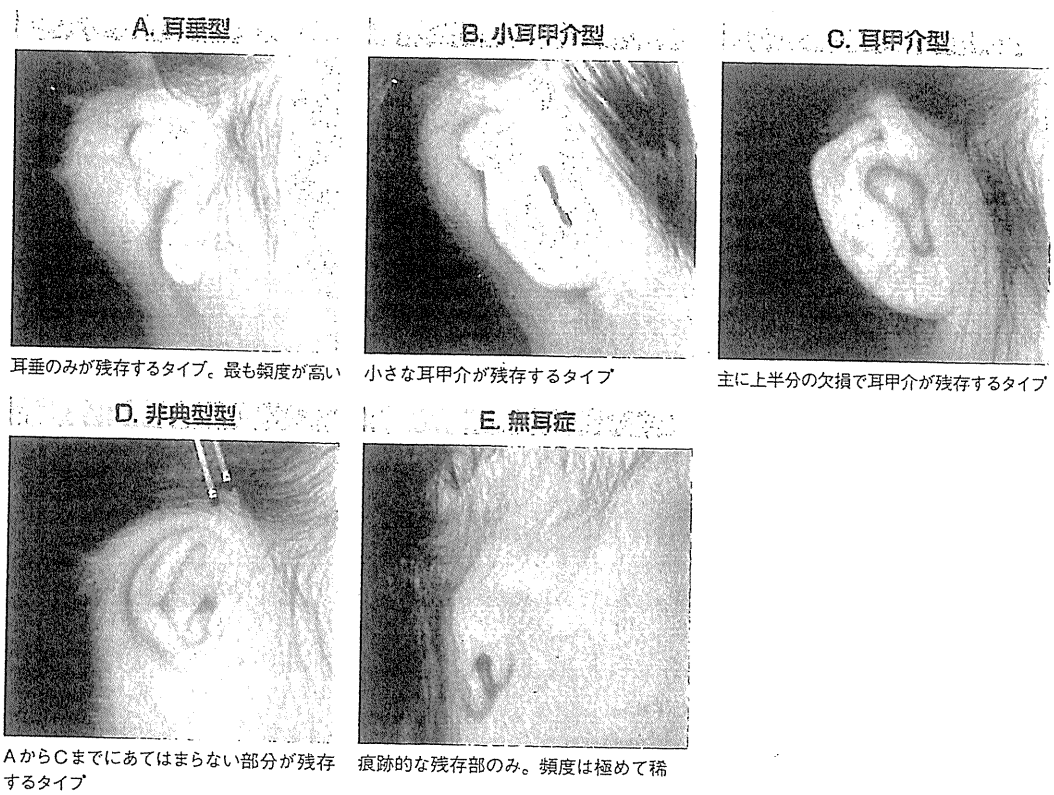
小耳症と無耳症

小耳症は耳介構成成分が欠損した状態で、頻度は約1万人に1人と言われている。男性が女性より多く、右側が左側より多く発生し、両側小耳症も約1割の発生頻度である。多くの場合、外耳道閉鎖を伴う。形態の異常によりマスクやめがねの装着に困難があり、外耳道閉鎖により伝音難聴を呈する。

残存する耳介の形状により、図8のA～Eの5タイプに分類される。

これらのうち最も頻度が高いのはAの耳垂型であり、Cの耳甲介型、Bの小耳甲介型がこれに次ぐ。A、B、Cのタイプに当てはまらないDの非典型型や痕跡的なEの無耳症は稀である。肋軟骨で耳介のフレームワークを作成する際に、Aの耳垂型の場合は下方の珠間切痕を回って前方の耳珠にいたるまでの全耳介を形成する必要があり、残存部分で利用できるのは耳垂の一部のみとなる。これに対して耳甲介が残存するタイプでは耳介の上半分を作成し、下半分や耳珠に関しては残存の部分をある程度利用することができるため、使用すべき肋軟骨の量は少なくて済む。ただし、いずれの場合でも肋軟骨は3本採取する必要がある。

他の合併症状を伴わない単独症例が多いが、中には第一第二鰓弓症候群やピエールロバン症候群に伴って下顎の低形成が見られたり、hairline（髪の毛の生え際のライン）が低下して耳介作成予定部位が有毛部となる症例があり、耳介形成の難度が高くなる。また、同側の先天性顔面神経麻痺を伴う症例も存在する。手術時期までの経過観察期間において、これらの合併症状が耳介形成術に及ぼす影響について十分に検討しておく必要がある。



耳垂のみが残存するタイプ。最も頻度が高い

小さな耳甲介が残存するタイプ

主に上半分の欠損で耳甲介が残存するタイプ

AからCまでにあてはまらない部分が残存するタイプ

痕跡的な残存部のみ。頻度は極めて稀

【図8】小耳症の分類

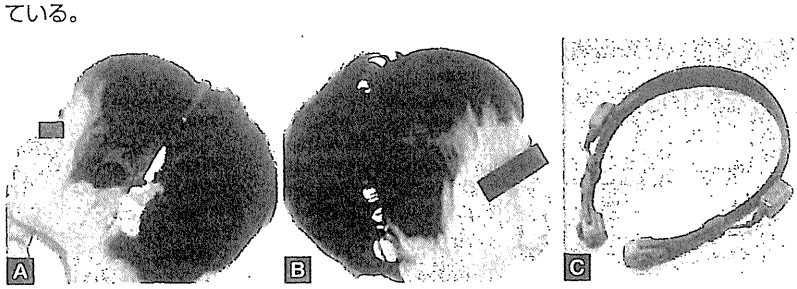
参考：Marx分類では、1度は耳介構成成分がかなり識別できるものとされ、constricted earやCの耳甲介型が含まれる。2度は構成成分が一部残存するもので、Bの小耳甲介型あるいはDの非典型型の一部が該当し、3度は単なる皮膚の隆起にとどまるもので、Aの耳垂型およびEの無耳症が該当する。

2 小耳症に対する手術までの耳鼻科医の対応（図9）

両側小耳症の場合、聴覚の発達の観点から補聴器の装着を必要とする。ほとんどの場合骨導聴力は正常に近いので、ヘアバンド式の骨導補聴器を使用する。これには箱型と耳掛型を改良したものがある。片側小耳症の場合は、健側の聴力が問題なければそのまま片側耳聴で生活する場合も多いと思われるが、健側の聴力も低下している場合など補聴器の装着を必要とする場合もある。BOA（behavioral observation audiometry），COR（conditioned orientation reflex audiometry）などの聴性行動反応聴力検査に加えて、気導や骨導刺激のABR（auditory brainstem response：聴性脳幹反応）検査を行って聴力を診断し、必要な場合は身体障害者手帳の交付申請を行う。1歳前後からの言語の発達を促すために、骨導補聴器は1歳頃には常時装用を目指すべきであろう。その後、小耳症に対する手術年齢（10歳前後）に至るまで、年1、2回ずつの経過観察を行い、患児の発達に合わせて適宜聴力検査を行う。

生後すぐ	出産した病院で「外耳道閉鎖症」と診断
3ヵ月	耳鼻咽喉科を受診 「両側小耳症・外耳道閉鎖症」と診断
6ヵ月	BOAやCORとABR検査にて左右ともに○○デシベルの聴力と診断 身体障害者手帳（聴覚障害）の交付申請
10ヵ月	骨導補聴器を装用、サポートプログラム開始 形成外科を受診（手術の時期などの説明）※
1歳～	年1～2回の面談と定期的な聴力検査 サポートプログラム継続
9歳 （術前6ヵ月）	術前の評価、手術に関する相談と説明
10歳	肋軟骨移植術、耳介挙上と外耳道形成の共同手術

※形成外科での対応：耳鼻科からの紹介を受けて、形成外科では手術時期や方法の説明を行い、同様に年1、2回ずつの経過観察を行う。耳介は肋軟骨を移植して形成するため、胸郭の発育状態も観察する。3本の肋軟骨で十分な大きさの耳介を形成するためには、おおよその目安として胸囲60cm以上が必要とされている。



△ 骨導補聴器と装用例
A・B：ヘアバンド式ワイヤレス片耳骨導補聴器
C：ヘアバンド式ワイヤレスデジタル両耳骨導補聴器（スターキージャパン提供）

- 両親に対する障害受容指導
- 言語および聴能指導
- 難聴や発育に関する情報提供
- 家族会の紹介
- 言語力、発音、聞こえの状態のチェック

△ サポートプログラムの主な内容

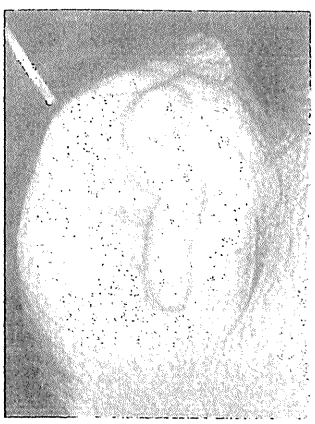
【図9】小耳症に対する手術までの耳鼻科医の対応（生後まもなく両側小耳症・外耳道閉鎖症と診断された一例）

②第1段階手術：肋軟骨移植による耳介形成術

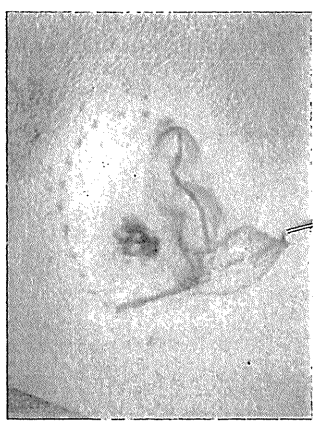
耳垂型小耳症を例にとり術式を紹介する（図15A）。まず残存耳垂の下部を後方へ移動し、耳垂上部は稜線に沿って切開する（図15B）。この切開線から肋軟骨フレームワークを挿入する皮下ポケットを作成するが、外耳道に相当する部位は皮下茎として温存し、皮弁の血行を保つようにする。そして遺残軟骨を直視下に摘出する（図15C）。

右胸部からⅥ、Ⅶ、Ⅷの肋軟骨を採取し、Ⅵ、Ⅶの肋軟骨でbase部分を作成し、Ⅷ肋軟骨で耳輪を作成する。対耳輪はⅦまたはⅧの一部で作成し、余剰の肋軟骨は耳介挙上の際の支柱とするため胸部皮下にbankingしておく（図15D）。フレームワークの作成には細いステンレスワイヤーを使用し、耳介フレームワークを作成する（図15E）。

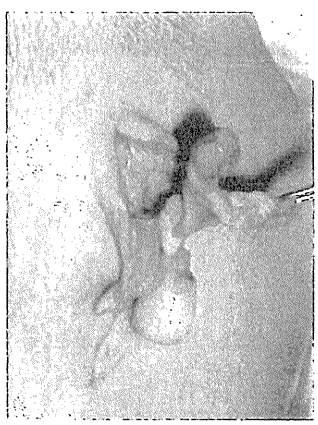
耳介部に持続吸引ドレーンを留置して、作成したフレームワークを皮下ポケットに挿入し固定する（図15F）。吸引を効かせながら余剰皮膚の切除を行い、縫合して手術を終了する（図15G）。ドレーンは2週間後に抜去して退院とし、抜糸はさらに1週間後に外来で行う。



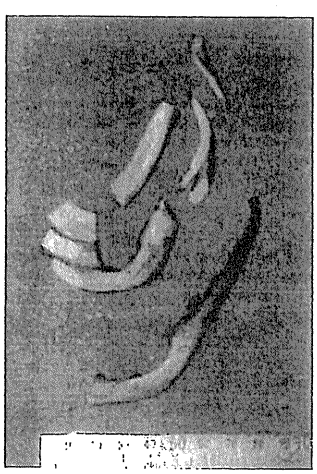
A：術前の状態（右耳垂型小耳症）



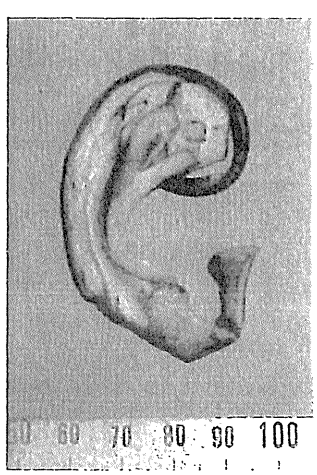
B：術前の状態（右耳垂型小耳症）耳垂下方は後方移動し、耳垂上方の稜線に切開を加える。丸く塗りつぶされた部分は皮下茎とし、点線の範囲を皮下剥離する



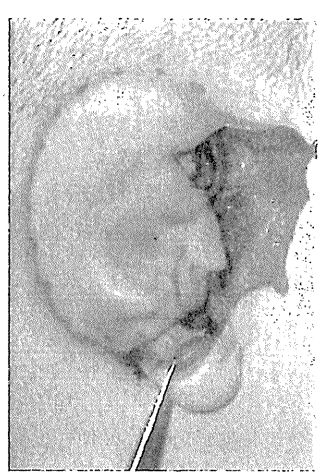
C：遺残軟骨を直視下に切除して皮下ポケットを作成



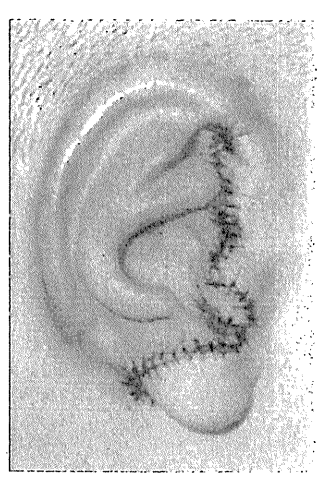
D：採取した肋軟骨のⅥ、Ⅶからbase部分と対耳輪、Ⅷから耳輪を作成している



E：作成した耳介フレームワーク



F：フレームワークを挿入し、皮膚をトリミングしながら縫合



G：手術終了時の状態

【図15】肋軟骨移植術

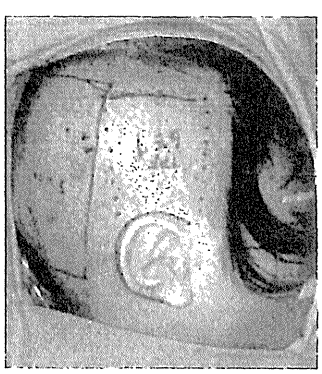
③第2段階手術：耳介挙上と外耳道形成共同手術

半年後に耳介挙上術と外耳道形成術の共同手術を行う（図16A）。耳介上方側頭部の横切開から浅側頭筋膜を挙上し、耳介周囲の切開と連続させて浅側頭筋膜下で再建耳介の裏面を剥離し、筋膜付きで耳介を挙上する。また同切開から深側頭筋膜を挙上しておく。電動式あるいは気動式デルマトームにより1000分の12インチの分層皮膚を採皮する（図16B）。耳甲介部分を切開して外耳道入口部を作成し、挙上耳介を反転して耳鼻科に交代する（図16C）。耳鼻科が外耳道形成を行う間、形成外科は胸部よりbankingしておいた軟骨を取り出し支柱を作成する。また採取した分層皮膚から外耳道用の皮膚管を作成しておく。外耳道形成に必要なコルメラはこの支柱軟骨の余剰部を利用し、鼓膜の代用には深側頭筋膜の一部を使用する。

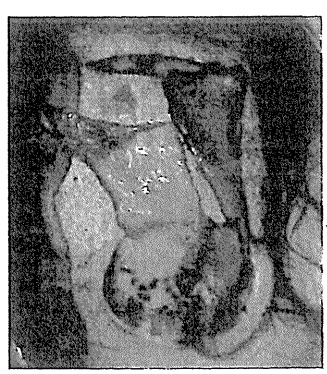
耳鼻科による外耳道形成術を行う。乳突部表層より骨板を採取し，新たに作成する外耳道の中心部とする。乳突洞口に向かって進みキヌタ骨の短脚を同定、閉鎖板を削除しツチ骨とアブミ骨を確認する。電気刺激により耳小骨の可動性をチェックした後、用意してあった骨板で外耳道後壁を作成する。肋軟骨で作成したコルメラをツチ骨−キヌタ骨のcomplexの上に立て、深側頭筋膜で鼓膜を形成する。挙上した深側頭筋膜で作成外耳道を被覆し（図16D）、皮膚管を挿入する（図16E）。

再度術者を交代し、形成外科が支柱の固定を行う（図16F）。浅側頭筋膜でこの支柱の前後面を被覆して、外耳道入口部と皮膚管を縫合し、耳介後面に先ほどの分層皮膚を用いて遊離植皮を行う（図16G）タイオーバー固定を行って手術を終了する（図16H）。2週間後にタイオーバーをはずして退院、さらに1週間後には外来で全抜糸を行う。

合併症として最も重要なものは術後の顔面神経麻痺である。手術は顔面神経刺激装置によってモニターしながら行うのであるが、骨削開のためのドリルによる熱が伝わり術後の浮腫によって麻痺が生じるものと考えられる。術後に麻痺症状が見られた場合はベル麻痺などの治療に準じてステロイド、ビタミンB_{12}を投与する。多くの場合、麻痺は一過性で数ヵ月以内に回復する。また、頸部の可動性が大きい小児の顔を横に向けて長時間の手術を行うため、術後に環軸椎亜脱臼を呈することがある。頸部を後屈しないよう術中の体位に注意しなければならない。



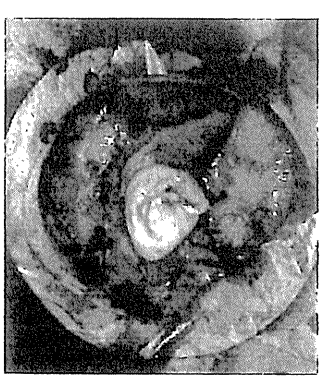
A：術前の形成外科のデザイン、耳介周囲と側頭部の横切開、分層採皮部を示す



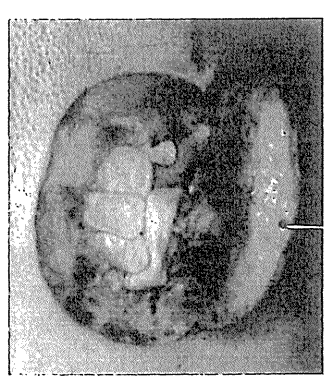
B：再建耳介を浅側頭筋膜とともに挙上反転し、深側頭筋膜も挙上する。この症例では鼓膜の代用として筋膜の一部を別に採取している

C：耳鼻咽喉科のデザイン、頭蓋底と乳様突起のライン、外耳道形成予定部位を示す

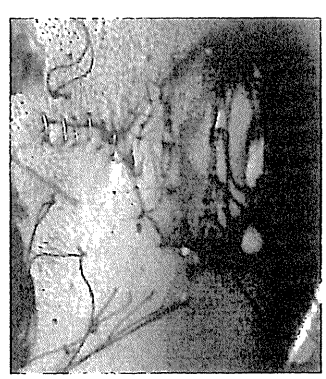
D：作成した外耳道周囲に沿って深側頭筋膜を挿入



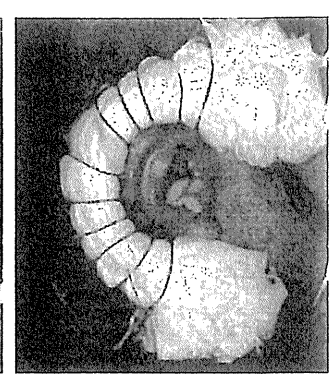
E：分層皮膚で作成した皮膚管を挿入



F：軟骨で形成した支柱を固定



G：浅側頭筋膜で被覆して耳介後面に植皮を行う



H：タイオーバー固定を行い、手術を終了したところ

【図16】耳介挙上と外耳道形成の共同手術

症例解説

I 両側の耳垂型小耳症および外耳道閉鎖の症例

症例1 10歳、男児

現病歴

◐術前は片耳にヘアバンド式箱型の骨導補聴器を使用して生活していた。

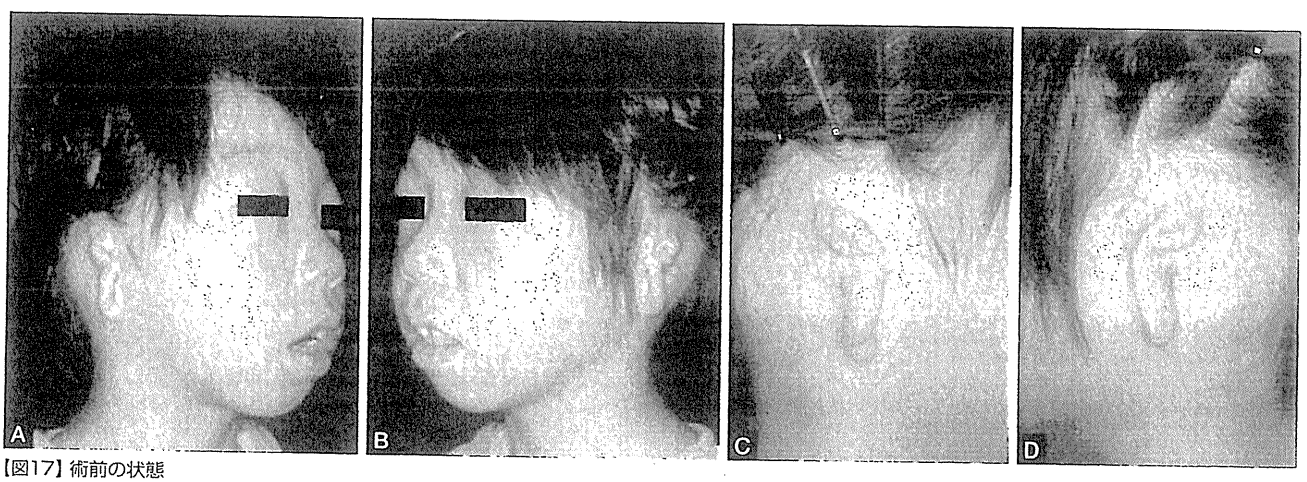
治療・経過

◐治療については片耳について2回ずつ、合計4回の手術を必要とするが、本人家族の希望はできれば小学生のうちに手術を終わらせたいとのことであった。残存耳介の後部は耳介再建の際に肋軟骨を埋め込んで輪郭を出すべき部位であるので、同部の皮膚を傷めないために、骨導補聴器の振動端子部分をこの部位からはずし毛髪内に当てるよう、また端子部を適宜左右入れ替えて生活し、手術によって一方が包帯でふさがれても支障を来さないように、手術の約1年前から指導を行っていた。同一部位での第1期手術と第2期手術との間は最低半年の期間を空ける必要があるため、手術は左肋軟骨移植術、右肋軟骨移植術、左耳介挙上・外耳道形成術、右耳介挙上・外耳道形成術の順に行った。

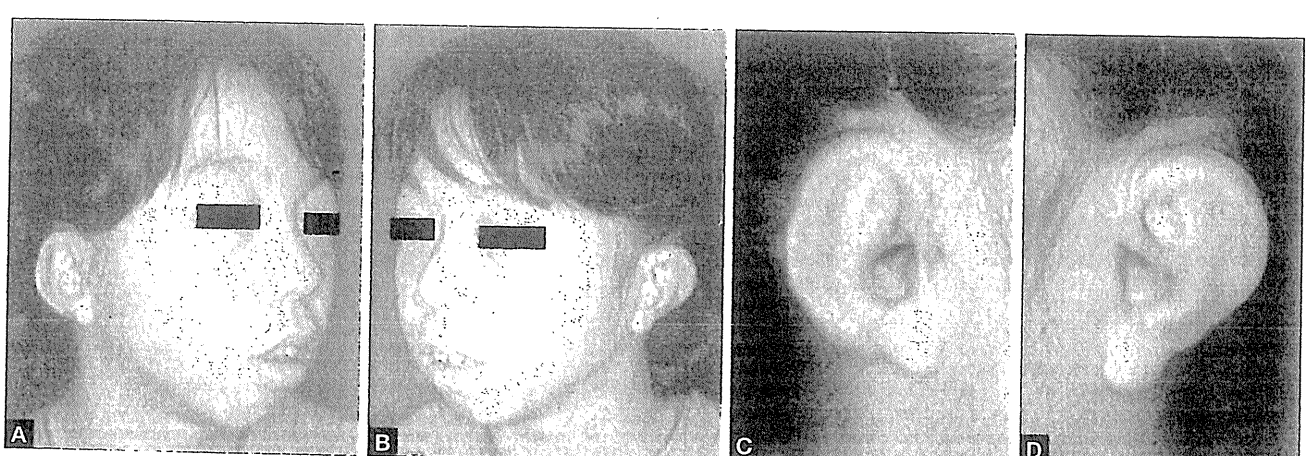
◐術後経過は順調で共同手術後左は4ヵ月、右は3ヵ月でそれぞれカナル型の気導式補聴器を装着することができるようになり両耳聴を享受できるようになった(図18A、B)。耳介の輪郭は良好に形成されており、メガネやマスクの着用にも問題なく生活している(図18C、D)。

診断・治療のポイント

両側ともに二段階の手術が必要で、治療期間短縮のため左肋軟骨移植、右肋軟骨移植、左耳介挙上・外耳道形成、右耳介挙上・外耳道形成の順に手術を行った。術後経過は順調で左右ともカナル型の気導式補聴器が装着可能となっている(図18A～D)。



【図17】術前の状態



【図18】術後の状態(右:共同手術1年3ヵ月後、左:共同手術1年9ヵ月後)

I 左の耳垂型小耳症外耳道閉鎖の症例

症例2 10歳、男児

治療・経過

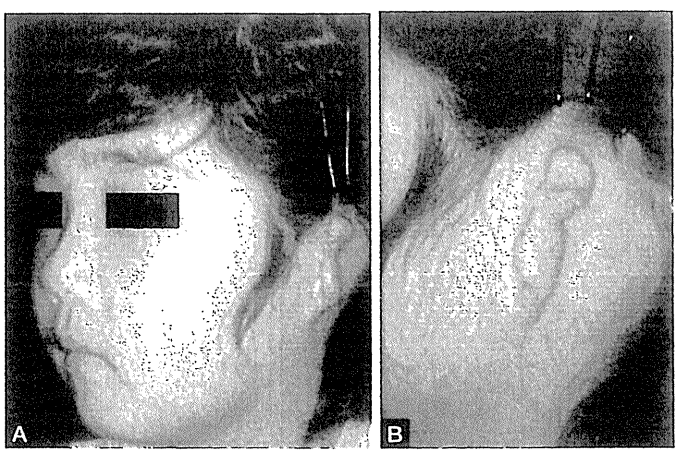
◎左の片側性耳垂型小耳症および外耳道閉鎖の症例で、下顎低形成など他の合併症は見られなかった(図19A、B)。術前の骨導聴力についてはほぼ正常領域であり、側頭骨CTの所見はJD score 9点と良好であった。

◎第1期手術として肋軟骨移植を行ったが、健側の耳介がやや大きめで、同じ大きさの耳介を形成するためには軟骨移植部位が若干hairlineにかかる状態であった。耳介挙上後にこの部分は耳介前面にくるため、後に脱毛を行うこととした。また本人家族と第2期手術の方針について話をしたところ、外耳道形成術の同時施行を強く希望したため、第2期手術は耳介挙上と外耳道形成の同時共同手術を行った。

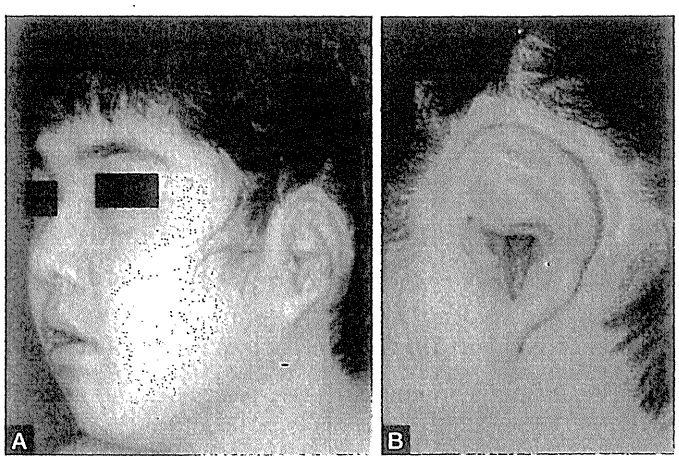
◎術後経過は順調で、外耳道部分が落ち着くにつれて患側の聴力も上がり、術後6ヵ月で平均聴力31.2デシベルの大幅な改善を見た(図20)。耳介の挙上が一部不十分であったのと、耳輪に沿って脱毛が必要であったため修正手術を行ったが、術後5年を経過した後も聴力は良好に推移しており、形態もよい状態を保っている(図21A、B)。

診断・治療のポイント

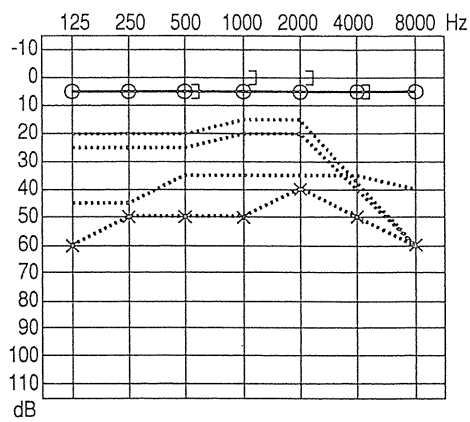
術前のJD scoreは9点で聴力改善を本人家族が希望して共同手術を施行した。術後の聴力改善は良好で(図20)、5年経過後も良好な形態と機能を維持している(図21A、B)。



【図19】術前の状態



【図21】術後5年の状態



【図20】術前後のオージオグラム所見
( )内は4分法による平均聴力

おわりに

耳介の先天性変形には種々のものがあり、形成外科的治療が必要となる。小耳症に対しては以前形成外科と耳鼻咽喉科が別々に治療を行ってきたが、両科が共同でチームアプローチを行うことによって、形態と機能の再建を両立させることが可能となっている。

この両科合同の共同手術にはいくつもの利点がある。耳鼻咽喉科にとっては、図16Cに示す如く非常に良好な術野が展開され、外耳道形成の手術操作が行いやすくなる点が、また形成外科にとっては、実際に外耳道が存在することによって作成耳介の凹凸がより強調されて輪郭がはっきりする点が挙げられる。しかしながら最も大きな利点は、患者にとって2つの内容の手術が同時に1回で済むことであり、これは患者の精神的・肉体的負担を大きく軽減すると考えられる。片側小耳症の場合には対側のみの聴覚で日常生活に不自由はなく、外耳道形成の適応はないという意見もあるが、音の動きの把握や聞きたい音に耳をすませるという両耳聴の意義は重要であり、中耳の発育のよい症例については外耳道形成の適応がある、というのがわれわれの考えである。小耳症外耳道閉鎖症の患者家族の希望は決して形態のみの再建にとどまるものではなく、聴力のみを求めるものでもない。本来その両方を獲得したいと願っているはずである。

また、現時点では外耳道形成を希望しない患者でも、将来本人が成人したときに希望するかもしれないし、現時点で中耳の発育が悪くても将来医学の進歩によってこれを克服できるかもしれない。これらを考えると第1期手術の際に正しい位置に耳介形成を行うことが形成外科にとっては最も重要である。この場合の正しい位置、というのは将来的に外耳道形成術を行うことの可能な位置であり、決してミリ単位での体表上の左右対称を意味するものではない。

形成外科・耳鼻咽喉科がお互いの立場を尊重しながら意見を出し合い、チームアプローチを行うことで大きく進歩を遂げた分野として頭頸部癌の外科治療が挙げられるが、小耳症に対する治療においても、形態と機能の再建の両立を目指した両科の緊密な連携が今後ますます重要になってくるものと思われる。

(2008年8月初出)

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The Auditory System

7

Hans J. ten Donkelaar and Kimitaka Kaga

Contents

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7.1 Introduction

The ear or vestibulocochlear organ is composed of external, middle and inner parts (Fig. 7.1). The external ear consists of the auricle and the external acoustic meatus with the outer layer of the tympanic membrane. The middle ear is formed by the tympanic cavity, the auditory ossicles and the inner layer of the tympanic membrane. The inner ear comprises the labyrinth, a series of fluid-filled spaces in the petrous part of the temporal bone. The auditory part of the inner ear consists of the cochlea with the organ of Corti, which contains hair cells as auditory receptors. Receptors sensitive to high frequencies are located near the cochlear base and those sensitive to low frequencies near the apex of the cochlea. The hair cells are innervated by the peripheral processes of bipolar ganglion cells in the spiral ganglion. Their central processes form the cochlear division of the vestibulocochlear nerve and terminate in the cochlear nuclei. The principal auditory pathway passes from the cochlea, via the cochlear nuclei, the inferior colliculus and the medial geniculate body (MGB) to the contralateral auditory cortex on the dorsal surface of the superior temporal gyrus. Each MGB is bilaterally innervated, so that each hemisphere receives cochlear input bilaterally. All of the components of the auditory pathway are tonotopically organized.

At birth, humans have about 20,000 inner and outer hair cells in the organ of Corti, which often do not last a lifetime as they do not regenerate when lost (Stone et al. 1998). By the age of 65–75 years, many individuals have a bilateral, high-frequency progressive hearing loss known as presbycusis associated with hair cell attrition. Hair cell loss is the most common cochlear defect causing hearing impairment in presbycusis and noise-induced hearing loss. Hearing disorders due to brain stem lesions are rare because of the bilateral projections of the central auditory pathways. Midline pontine lesions may result in impaired sound localization due to interruption of the input of the superior olivary complex (see Sect. 7.3.2 and ***Clinical case 7.2***). Disorders of

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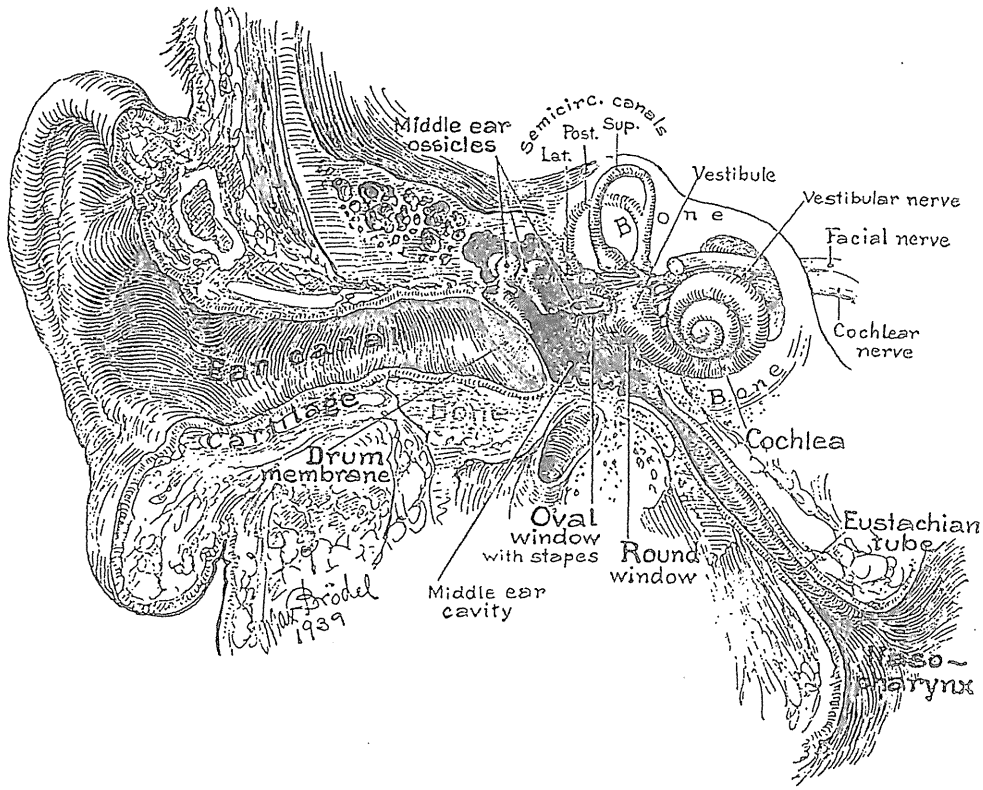

Fig. 7.1 Overview of the external, middle and internal ear (from Brödel 1946)

auditory perception may follow strokes in the territory of the internal carotid arteries or of the vertebrobasilar system. The central disorders of auditory perception may result from lesions of either the right and the left or both cerebral hemispheres, usually involving parietotemporal cortical areas as illustrated in *Clinical cases* (see Sect. 7.4.4).

7.2 The Cochlea and the Cochlear Nerve

7.2.1 The Middle Ear and the Cochlea: Mechanical Transmission of Sound

The **middle ear** comprises the tympanic cavity, the tympanic membrane, the three auditory ossicles, two middle ear muscles, air-filled cavities formed by the mastoid antrum and mastoid air cells, and the auditory tube. The **tympanic cavity** communicates with these air-filled cavities and through the auditory tube with the nasopharynx (Fig. 7.1). The three **auditory ossicles** are the hammer or **malleus**, the anvil or **incus** and the stirrup or **stapes**. The head of the malleus is anchored to the tympanic membrane, whereas the base of the stapes is connected to the fenestra vestibuli or oval window. Sound waves set the tympanic membrane into vibrating movements, which via the auditory ossicles are transmitted to the inner ear. The **inner ear** or cochlea is a fluid-filled tube that is coiled two and a half times. In cross-section, it has a broad base, a pointed apex and a central pillar called the modiolus. The osseous labyrinth communicates with the tympanic cavity through two openings in its medial wall, the oval window or **fenestra vestibuli** and the round window or **fenestra cochleae**. The oval window is closed by the base of the stapes, so that vibrations of the auditory ossicles are transmitted to the perilymph of the inner ear.

Motion of the auditory ossicles is modified by two small **middle ear muscles**, the tensor tympani and the stapedius. The **tensor tympani** is the largest of the two. It is attached to the handle of the malleus and is innervated by the trigeminal nerve. The smaller **stapedius** attaches anteriorly to the head of the stapes and is innervated by the facial nerve. The stapedius and tensor tympani motoneurons form separate cell groups, situated close to the facial and motor trigeminal nuclei, respectively (Lyon 1978; Mizuno et al. 1982; Shaw and Baker 1983). The stapedius functions to protect the auditory receptors of the inner ear against excessive stimulation caused by too strong sound pressure. The sound pressure depends on the amplitude of the waves: the greater the amplitude, the higher the sound pressure. The stapedius contracts in response to sounds above 70 dB (the intensity of loud conversation), damping the movements of the auditory ossicle chain. The tensor tympani contracts to louder sounds, especially impulse noises. The **acoustic middle ear reflex** includes projections from the ventral cochlear nucleus via the superior olivary nuclear complex to the motor nuclei of the trigeminal and facial nerves (Borg 1973). With electro-acoustic impedance measurements, **stapedius muscle contraction** can be readily detected in response to ipsilateral or contralateral sound, giving objective information

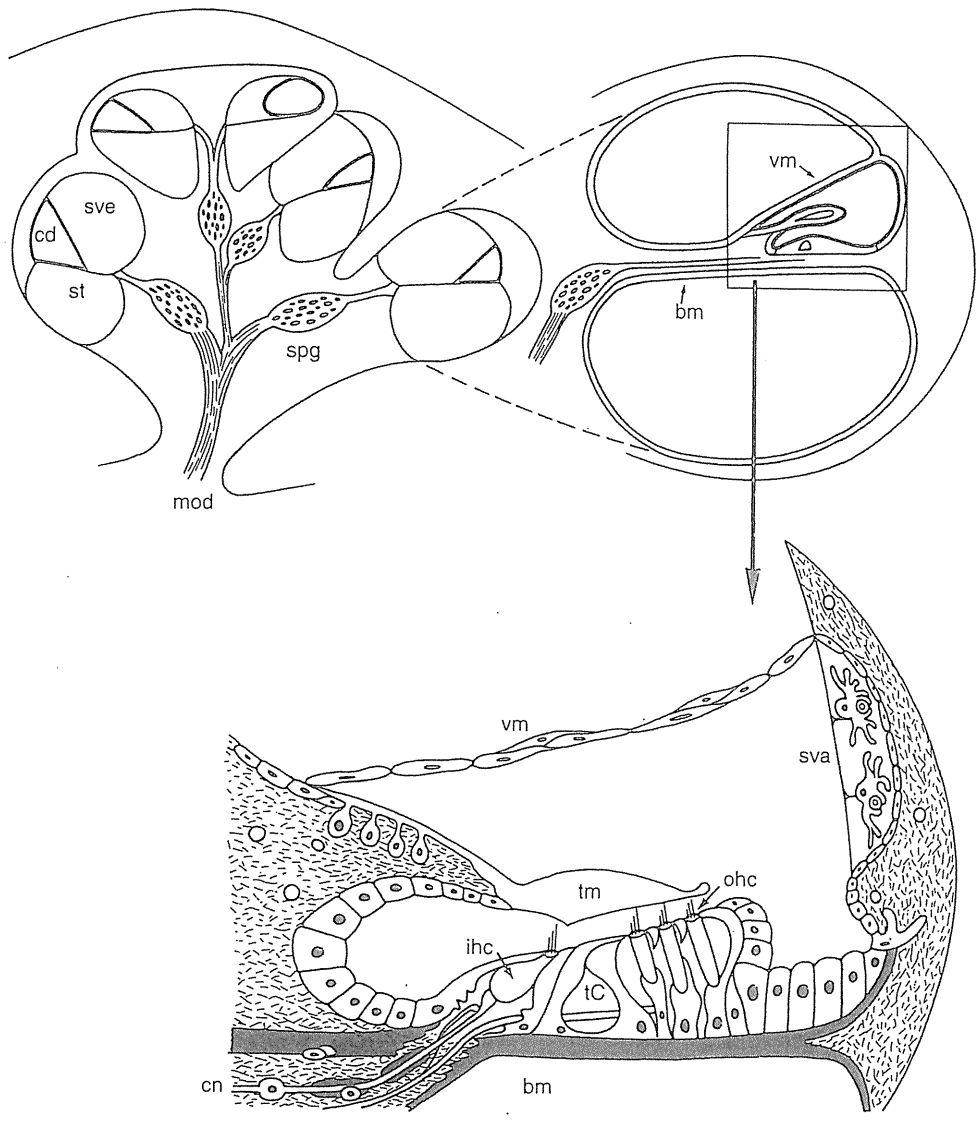

Fig. 7.2 The foetal cochlear duct. At 16 weeks of development, the cochlear nerve (*cn*) fibres pass through a central pillar, the modiolus (*mod*), whereas their cells of origin form the spiral ganglia (*spg*). Below, details of the spiral organ are shown for 25 weeks of development. *Abbreviations*: *bm* basilar membrane; *cd* cochlear duct; *ihc*, *ohc* inner and outer hair cells; *st* scala tympani; *sva* stria vascularis; *sve* scala vestibuli; *tC* tunnel of Corti; *tm* tectorial membrane; *vm* vestibular (Reissner) membrane (from ten Donkelaar et al. 2006)

about the functional state of the middle and the inner ear, the auditory and facial nerves and the central auditory pathways in the lower brain stem. Ipsilateral and contralateral measurements can distinguish between right, left and midline lesions of the lower brain stem (Hayes and Jerger 1981).

The **cochlea** is composed of three chambers or scalae: the scala vestibuli, the scala media and the scala tympani, separated from each other by the vestibular membrane of Reissner and the basilar membrane (Fig. 7.2). The inner scala media is filled with endolymph, which is rich in potassium and has the character of intracellular fluid. The perilymph of the outer scalae vestibuli and tympani has approximately the same composition as the cerebrospinal fluid. The two perilymph compartments form one space, since they are continuous with each other at the apex of the cochlea (the helicotrema). The perilymph drains to the subarachnoid space. The **scala media** or **cochlear duct** contains the organ of Corti, which rests on the basilar membrane (Fig. 7.3). The superior wall of the cochlear duct (the membrane of Reissner) angles downwards from lateral to medial, making the cochlear duct wedge shaped. The lateral wall is the stria vascularis. The thickened epithelium that constitutes the **organ** of **Corti** can be divided into hair cells and supporting cells. The hair cells are the sensory receptor cells of which there is a single row of inner hair cells and three rows of outer hair cells. The supporting cells include the inner and outer pillar cells, which are separated by the tunnel of Corti extending the length of the cochlea. Both the hair cells and the supporting cells are overlaid by the gelatinous tectorial membrane. In humans, there are 12,000 outer hair cells in three rows at the basal turn, increasing to four to five rows in the second and apical turns, and 3,500 inner hair cells in a single row (Retzius 1884; Bredberg 1968; Kimura 1975). On their apical side, the hair cells contain contractile proteins, including an actin cuticular plate, and about 100 stereocilia, graded in length, which extend to the overlying tectorial membrane. The stereocilia

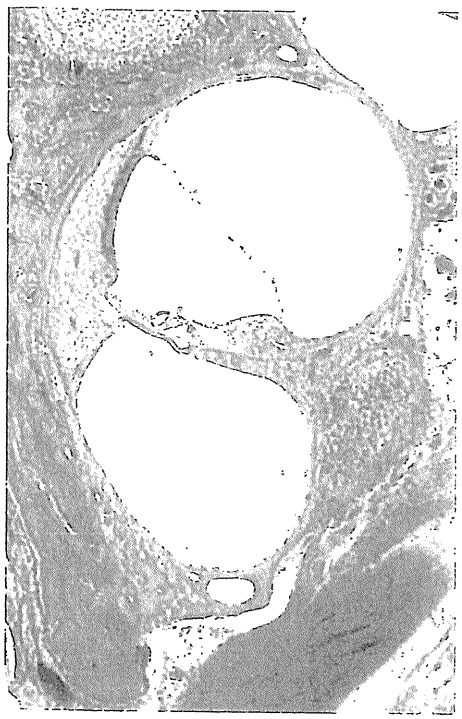

Fig. 7.3 Photomicrograph of the human cochlea (from ten Donkelaar et al. 2006; courtesy Jo Curfs, Nijmegen)

are composed of the active contractile proteins actin and myosin (Flock 1980; Corwin and Warchol 1991). The afferent fibres from the hair cells pass from the organ of Corti through small openings in the osseous lamina into the modiolus. Their cell bodies are located in the modiolus in Rosenthal's canal as the spiral cochlear ganglion (Sect. 7.2.3). The inner ear is vascularized by the **internal auditory artery** which in some 80% is a branch of the anterior inferior cerebellar artery (Kim et al. 1990; Schuknecht 1993).

7.2.2 Cochlear Hair Cells: Transduction and Amplification

The human ear can detect sound waves with frequencies between 20 and 20,000 cycles per second or Hertz (Hz), i.e. approximately ten octaves of sound. The human ear has the greater sensitivity for sounds around 1,000 Hz. The greater the frequency, the higher the pitch. The sound vibrations that enter the scala vestibuli and the perilymph at the oval window produce displacement of the basilar membrane before they finally dissipate back to the middle ear by movements of the membrane covering the round window. **Transduction** of **sound** occurs in the sensory cells of the organ of Corti. Oscillations of the basilar membrane produce a shearing force on the stereocilia of the receptor cells, which are in firm contact with the non-oscillating tectorial membrane. The tilting of the stiff cilia is the adequate stimulus for the auditory receptor cells. The inner and outer hair cells have different roles in the transduction of energy within the cochlea. Inner hair cells provide direct input to almost all of the axons in the cochlear nerve. Their activity is modified by local **amplification** of the motion of the basilar membrane produced by the outer hair cells and hair-cell related supporting cells (Flock et al. 1999; Moore and Linthicum 2004). Several molecules have been identified as having a vital role in hair-cell transduction. They are specifically expressed in and around the stereocilia and mutations in their genes lead to deafness (Steel and Kros 2001; ten Donkelaar et al. 2006).

7.2.3 Spiral Ganglion Cells and the Cochlear Nerve: Neural Transmission

The transition from hair cell activity to neural activity occurs within the cochlea. Activation of the stereocilia results in changes in the intracellular potential that lead to the release of a neurotransmitter from synaptic vesicle clusters at the base of the hair cells. Opposite such a cluster of synaptic vesicles, bulbous nerve terminals are found on the outer surface of the cell wall. Six to eight such terminals are present on the base of each inner hair cell, and a smaller number on each outer hair cell (Nadol 1990). These terminals continue as short unmyelinated processes, forming the "dendritic" segment of cochlear nerve fibres. They become myelinated when they enter the osseous spiral lamina. Here, they reach their cells of origin, the **spiral ganglion cells**. The **spiral ganglion** extends only halfway from the base of the cochlea to the apex. Therefore, the peripheral processes, containing hair cells in the apical and middle turns of the cochlea, extend down through the modiolus to reach the most apical ganglion cells. In humans, there are about 35,000 spiral ganglion cells (Hinojosa et al. 1985; Spoendlin 1985). Two types of ganglion cells are found (Spoendlin 1985). The majority (90–95%) are type I cells and contact inner hair cells. The unmyelinated peripheral processes of the remaining ganglion cells (5–10%), the type II cells, contact the outer hair cells. The central processes of both types of ganglion cells form the cochlear nerve (Spoendlin and Schrott 1989). The **cochlear nerve** enters the ventral cochlear nucleus on the ventrolateral side of the inferior cerebellar peduncle (see Fig. 7.6a). Upon entering the brain stem, primary auditory fibres bifurcate into equally sized ascending and descending branches (Moore and Osen 1979).

7.2.4 The Auditory Periphery: Generation of Evoked Activity

Neural activity is reflected in the **brain stem auditory-evoked potentials** or **responses** (**BAEPS** or **BAERs**), an externally recordable series of small amplitude and short latency wave-like potentials evoked by a transient stimulus

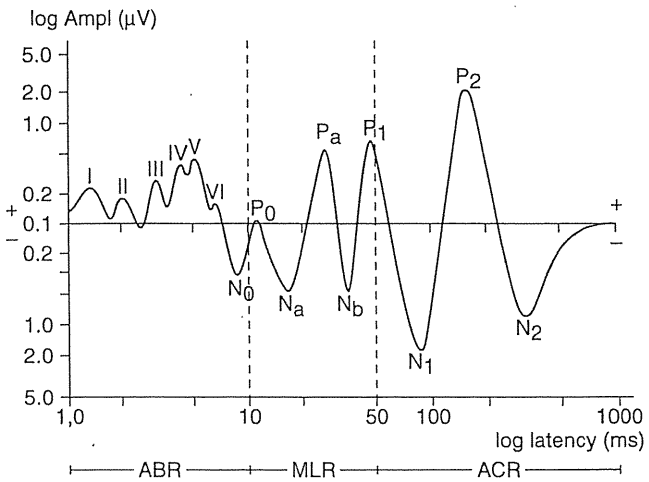

Fig. 7.4 Brain stem (ABR), middle latency (MLR) and cortical (ACR) auditory-evoked responses (from Pasman 1997; courtesy Jaco Pasman, Nijmegen)

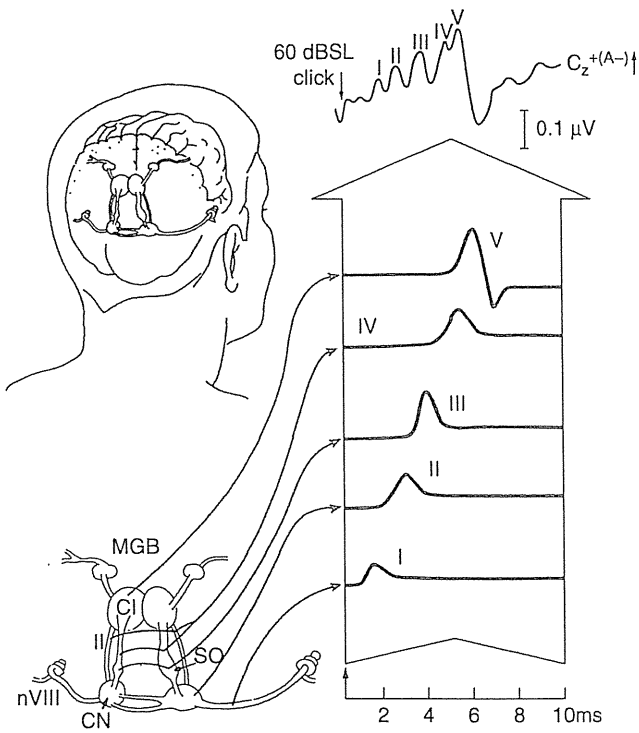

Fig. 7.5 Relationship between components of the brain stem-auditory evoked response and the auditory projection pathway. *Abbreviations*: *CI* colliculus inferior; *CN* cochlear nuclei; *ll* lateral lemniscus; *MGB* medial geniculate body; *nVIII* vestibulocochlear nerve; *SO* superior olive; *I–V* waves of BAEP (after Stockard et al. 1978)

such as a click (Jewett et al. 1970; Stockard et al. 1978, 1986). Auditory-evoked responses (AERs) can be subdivided according to their latency into brain stem (ABR), middle latency (MLR) and cortical (ACR) AERs (Fig. 7.4). In humans, the ABR or BAEP is characterized by six or sometimes seven deflections (I–VII) in the first 9 ms after the stimulus. Waves I, III and V are of greatest interest since they reflect volume-conducted activity from the levels of the acoustic nerve, pons and midbrain, respectively. The earliest BAEP waves (waves I and II) are generated by the cochlear nerve, prior to its entrance into the brain stem (Stockard et al. 1978, 1986; Moller and Jannetta 1982; Martin et al. 1995; Fig. 7.5). A potential corresponding to wave II of the scalp-recorded BAEP can be recorded intrasurgically from the surface of the human cochlear nerve as it passes through the internal auditory meatus and crosses the intradural space (Martin et al. 1995). This supports an earlier dipole localization study (Scherg and von Cramon 1985). Therefore, both waves I and II of the human BAEPs are generated by activity in axons of the cochlear nerve. BAEPs can distinguish between pathologies of the middle and inner ear, the auditory nerve and the brain stem. There are three major applications of BAEPs in adults: (1) the detection of tumours in the region of the posterior cranial fossa; (2) evolution of coma; and (3) assessment of patients with suspected demyelinating diseases such as MS. Acoustic neurinomas may cause complete loss of wave I on the side of the lesion or a significant increase in the I to III interpeak latency.

7.2.5 Hearing Loss

Two types of hearing loss can be distinguished: conductive and sensorineural. ***Conductive hearing loss*** is related to defects in conductive mechanisms in the middle ear, resulting from conditions such as otitis media and otosclerosis. ***Sensorineural hearing loss*** is caused by disease in the cochlea or its central connection, the cochlear nerve. Hearing loss of cochlear origin is common and can result from a variety of conditions, including tumours, infections, temporal bone fractures or from exposure to excessive noise or ototoxic drugs (Schuknecht 1993). In ***presbycusis***, the hearing loss of the aged, the loss begins with degeneration of outer hair cells at the basal end of the cochlea, but does not seriously affect hearing until the upper range of speech frequencies, around 3,000 Hz, is affected. Noise-induced hearing loss and severe blows to the head tend to affect the anterior basal turn of the cochlea, the region that processes 3,000–4,000 Hz (Moore and Linthicum 2004). ***Tinnitus***, characterized by noise in the ears such as ringing, humming or whistling, is a common symptom in disorders of the inner ear, but it can also occur in disorders affecting the VIIIth nerve such as an ***acoustic neurinoma*** (see ***Clinical case 7.1***) and with vertebrobasilar disease. Sudden onset of unilateral or bilateral deafness usually accompanied by dizziness or vertigo can be a sign of occlusion of the basilar artery (Huang et al. 1993; Levine and Häusler 2001).

Clinical Case 7.1 Cerebellopontine Angle Tumour

Electrocochleography and auditory brain stem response (ABR) are important electrophysiological tools for routine use in diagnosing vestibular schwannomas (Eggermont et al. 1980; Chandrasekhar et al. 1995). Kaga et al. (1997) reported a case of a vestibular schwannoma in which electrocochleography and ABR were correlated with temporal bone pathology (Fig. 7.6).

Case report: A 74-year-old female presented with a left hearing impairment. In 1975, she had undergone mastectomy of her left breast and in 1987, at the age of 73, she was treated with cobalt radiotherapy for a recurrence of the breast cancer. Pure tone audiometry revealed threshold elevation in the middle- and high-frequency range. ABR showed no response in the left ear but electrocochleography showed clear compound action potentials. CT scanning and MRI demonstrated the presence of a medium-sized cerebellopontine angle tumour in the left ear (Fig. 7.7a, b). Three years later, she died of metastatic lung cancer and sepsis. At autopsy, metastases of the breast cancer were found in the right upper lobe of the lung and in the right temporal lobe of the brain. The temporal bone pathology consisted primarily of a large schwannoma, originating from the left inferior vestibular nerve and occupying the left internal auditory meatus (Fig. 7.7c, d). The organ of Corti was well preserved in each turn. In the modiolus, the numbers of spiral ganglion cells and cochlear nerve fibres in each turn were decreased. These histological findings suggest that clear compound action potentials were recorded from the distal part of the cochlear nerve in spite of the presence of the vestibular schwannoma. ABR could not be detected because of the blockade of the proximal portion of the cochlear nerve by the vestibular schwannoma.

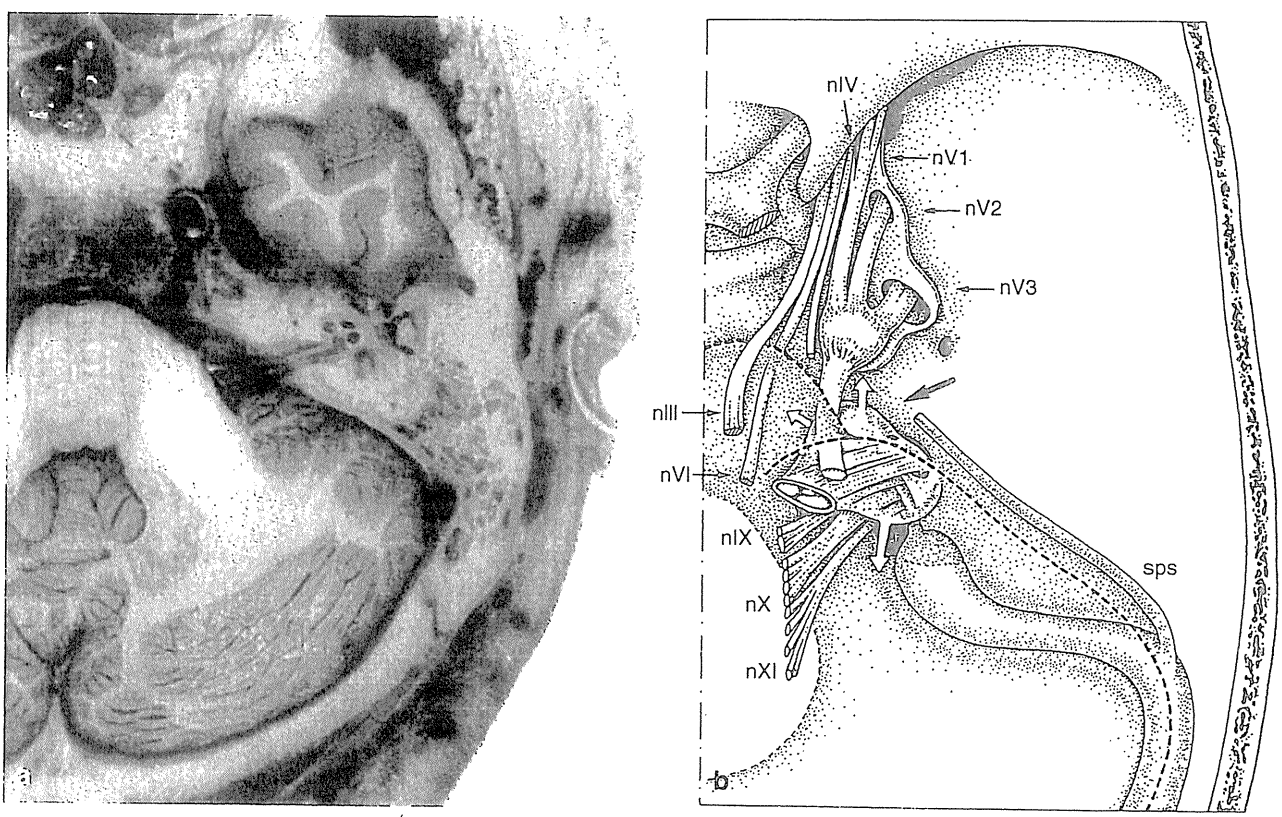

Fig. 7.6 (**a**) The course of the vestibulocochlear nerve from the inner ear to the brain stem on the right and (**b**) the cerebellopontine angle. The *black arrow* in (**b**) points at an acoustic neurinoma and the way it extends (*white arrows*). *Abbreviations*: *nIII* oculomotor nerve; *nIV* trochlear nerve; *nV1* ophthalmic nerve; *nV2* maxillary nerve; *nV3* mandibular nerve; *nVI* abducens nerve; *nIX* glossopharyngeal nerve; *nX* vagal nerve; *nXI* accessory nerve; *sps* superior petrosal sinus ((**b**) after ten Donkelaar et al. 2007)

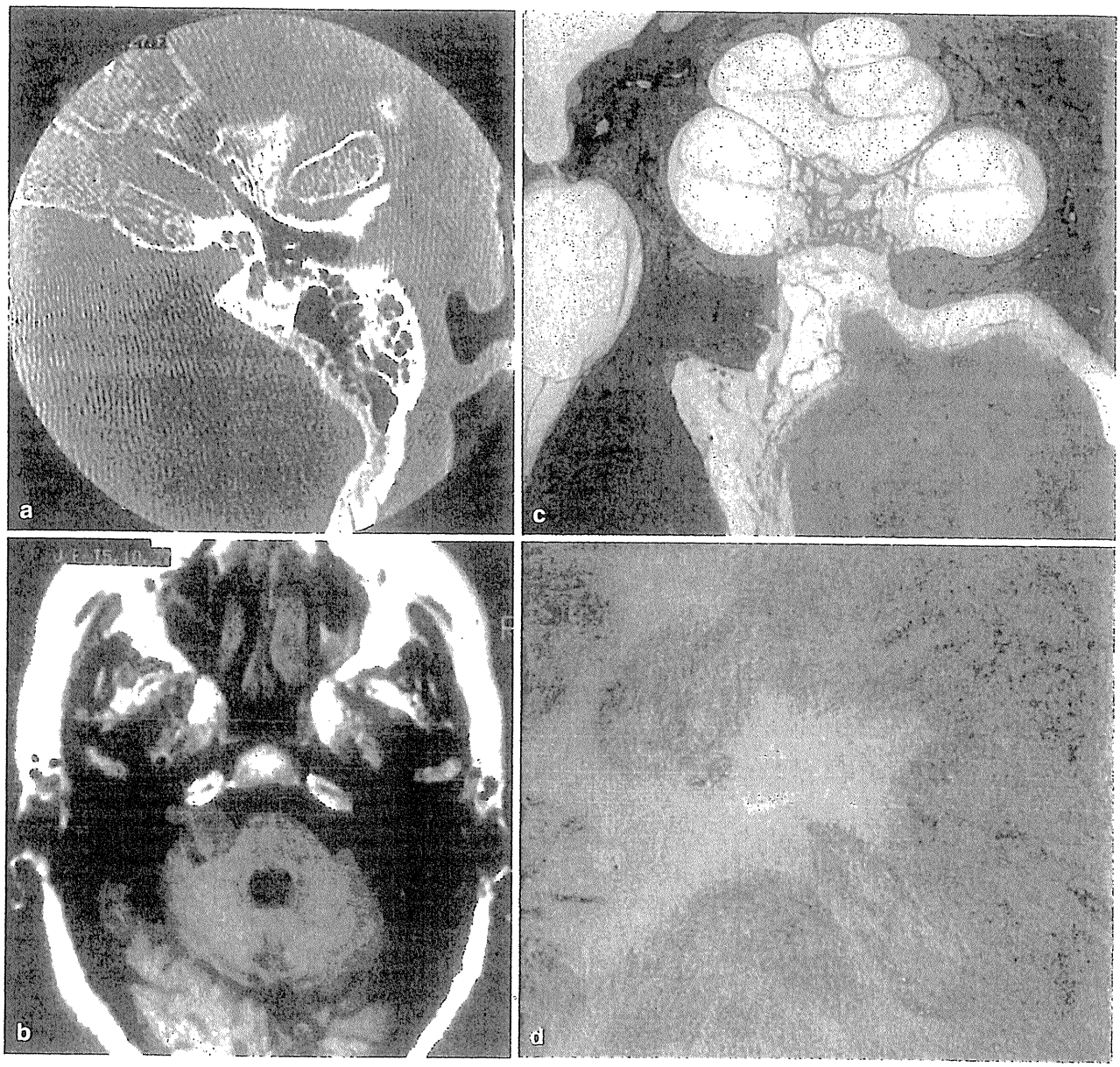

Fig. 7.7 (**a, b**) CT and MRI demonstrating the presence of a medium-sized tumour in the left internal auditory canal and the cerebellopontine angle. (**c**) Mid-modiolar section of the left ear showing enlargement of the internal auditory canal occupied by a vestibular schwannoma (HE stain). (**d**) Magnification of the vestibular schwannoma with mixed Antoni A and B cell types (HE stain; from Kaga et al. 1997)

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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

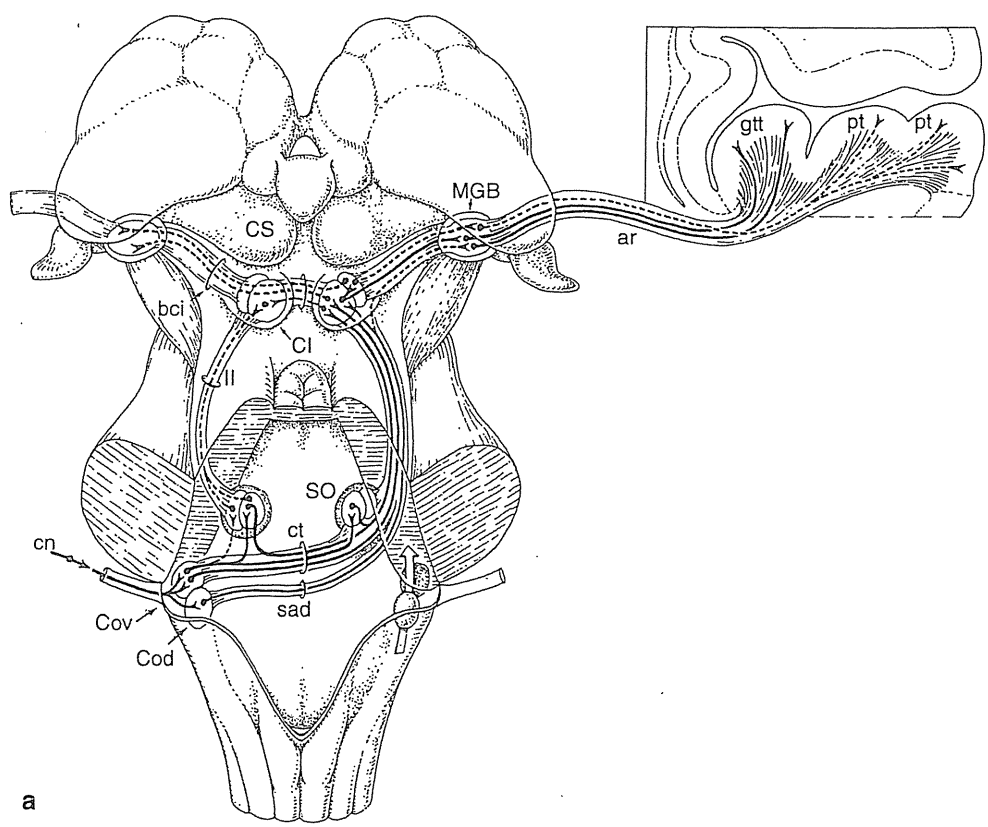

Fig. 7.8 (**a**) Overview of the auditory projections in the human brain (after ten Donkelaar et al. 2007); (**b–d**) the position of the cochlear nuclei (in *red*), the lateral lemniscus (in *light red*) and the colliculus inferior (in *red*) in horizontal sections of the brain stem (after Duvernoy 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

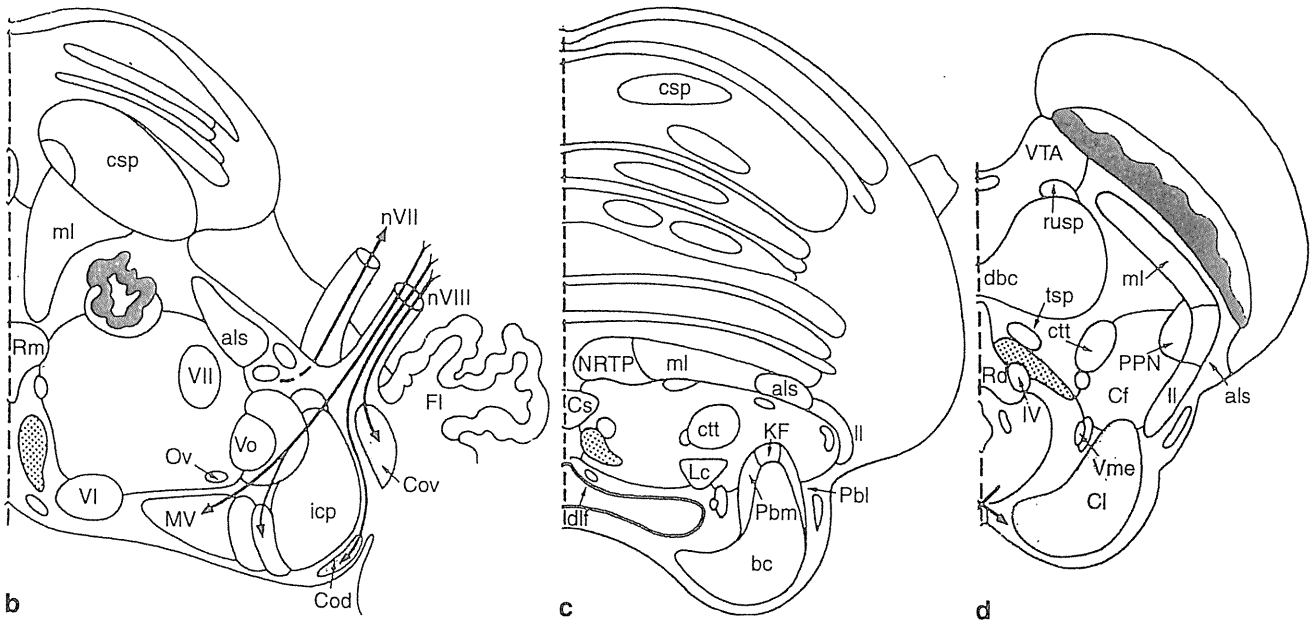

Fig. 7.8 (continued)

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.