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MAJOR AND MINOR CONGENITAL ANOMALIES OF THE EAR

classification and surgical results

E. Teunissen

MAJOR AND MINOR CONGENITAL ANOMALIES OF THE EAR

CLASSIFICATION AND SURGICAL RESULTS

MAJOR AND MINOR CONGENITAL ANOMALIES

OF THE EAR

CLASSIFICATION AND SURGICAL RESULTS

Een wetenschappelijke proeve op het gebied van
de Medische Wetenschappen, in het bijzonder
de Geneeskunde.

PROEFSCHRIFT

ter verkrijging van de graad van doctor
aan de Katholieke Universiteit Nijmegen,
volgens besluit van het College van Decanen
in het openbaar te verdedigen op
dinsdag 15 december 1992
des namiddags te 3.30 uur precies

door

Engelbert Teunissen

geboren op 7 juni 1958 te Velp

Promotor Prof. Dr. P. van den Broek

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Financial support for the publication was kindly provided by: Artu Biologicals BV, Astra Pharmaceutica BV, Brocades Pharma, Duphar Nederland BV, Elmed BV, Entermed BV, SmithKline Beecham Pharma.

aan mijn vader en in herinnering aan mijn moeder

The work presented in this thesis was performed in the Department of Otorhinolaryngology of the University Hospital Nijmegen.

CIP-DATA KONINKLIJKE BIBLIOTHEEK, DEN HAAG

Teunissen, E.

Major and minor congenital anomalies of the ear.
classification and surgical results / E. Teunissen. -
[S.l. : s.n.] (Meppel: Krips Repro). - Ill.
Thesis Nijmegen. - with ref. - with summary in Dutch.
ISBN 90 - 9005563 - 0
Subject headings: ears / congenital anomaly

Translations: Judith Abma - Hill
Press: Krips Repro, Meppel

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CHAPTER I

INTRODUCTION AND OUTLINE OF THE STUDY

INTRODUCTION

Major and minor congenital anomalies of the ear are rare. The incidence of major congenital anomalies is estimated to be about 1:10,000 births[1]. However, cases of slight aural atresia with a normal auricle are likely to be missed at birth. No reliable data are available on the incidence and prevalence of minor congenital ear anomalies. The generally accepted classification of anomalies into minor and major congenital ear anomalies has been employed in this thesis. Minor congenital ear anomalies are congenital ear anomalies confined to the middle ear. Major congenital ear anomalies involve the external ear canal, the middle ear and sometimes also the auricle. Anomalies and function loss of the inner ear are not listed separately in this classification. The division between minor and major congenital ear anomalies is very clear. Nevertheless exceptional cases do occur in which there is no sharp distinction.

HISTORICAL REVIEW OF SURGICAL TECHNIQUES

Micro-ear-surgery in the 1950s and 1960s

In the 1950s, micro-ear-surgery underwent enormous development through the introduction of myringoplasty in 1952 by Zollner[2] and Wullstein[3] and stapes replacement surgery in 1958 by Shea[4]. Previously, fixation of the stapes had been treated by fenestration of the horizontal semicircular canal or by mobilization of the stapes using a hammer and chisel. Although stapes replacement found general application for the treatment of otosclerosis in the early part of 1960, its ingression took much longer for the treatment of congenital stapes fixation. In many respects, the new technique of micro-ear-surgery met with great approval, but in the 1960s many surgeons still preferred to perform fenestration and stapes mobilization for congenital stapes ankylosis[5,6,7]. These developments are elaborated on in Chapter II-1 in a review of the literature on isolated congenital stapes ankylosis. This historical perspective is also applicable to stapes surgery for the treatment of non-isolated congenital stapes ankylosis, i.e. congenital stapes ankylosis with an associated ossicular chain anomaly, described in Chapter II-2. The technique of fenestration and mobilization which used to be the treatment of choice for minor congenital ear anomalies, was not applied to the series of patients described in this thesis, starting in 1964.

Minor ear anomalies

The historical development of surgery for middle ear anomalies with a mobile stapes is less impressive. The first reports and series of operated ears date from the latter part of the 1950s[5,7,8]. Worthy of mention, also with regard to the general development of middle ear surgery, is the use of homologous ossicular grafts for the reconstruction of the ossicular chain[9,10,11].

Major ear anomalies

Surgery for congenital aural atresia, synonymous with major congenital ear anomalies, is extremely complicated and forms a perpetual challenge for ear surgeons and plastic surgeons specialized in this field. Generally, surgery for congenital aural atresia is considered to be one of the most difficult forms of ear surgery. The aim of this form of surgery is to achieve a socially acceptable hearing level and a sufficiently wide, dry and stable ear canal. In addition, patients with auricular malformations will require cosmetic correction.

Important results were published by Livingstone[12], Lund and Phelps[13], Fenner et al[14], Colman[15] and Belluci[16]. The reported rates of postoperative restenosis and otorrhoea varied from 10% to 33%. The hearing gain was not always sufficient and a number of patients with bilateral major ear anomalies still needed to wear a hearing aid after the operation. It is recommended to fit these patients with a bone conduction hearing aid at a young age. The present surgical policy does not accept children with bilateral major ear anomalies until they are at least six years old. The reasons for this are that the mastoid must be adequately developed and the child must be sufficiently cooperative during the postoperative outpatient care of the ear. In contrast, the surgical policy employed in the Oxford studies accepted children from the age of 18 months. The extremely early operation and disregard of the varying grade of severity of the anomalies are presently considered to be the most important reasons why the results of the Oxford studies are so disappointing.

Cosmetic surgery for auricular malformations in combination with techniques to improve the patient's hearing is probably even more complex and disappointing than either of these forms of surgery on their own and the approach is only likely to be successful in few highly-experienced hands. This treatment method is performed in a staged series of operations by a plastic surgeon and an ear surgeon[17,18,19,20]. An alternative form of treatment for serious

congenital auricular anomalies has recently become available from a completely different angle: an auricular prosthesis can be fixed to titanium implants anchored to the mastoid. The percutaneous titanium implants can also be used to fit the patient with a bone anchored hearing aid[21,22].

The developments and improvement in the surgical results of major ear anomalies are due to improved microsurgical techniques, elucidation of middle ear and inner ear dynamics, a greater understanding of tympanoplastic reconstruction and the introduction of polytomographic X-ray techniques and computerized tomographic scanning. In addition, better preoperative patient selection, aided by classifying the severity of the anomaly, has also contributed to improving the results. A classification for major congenital ear anomalies, comprising a classification for auricular malformations and a classification for aural atresia, had been proposed in the earlier stages by Altmann[23] for ear canal atresia and supported by Nager[24] and Schuknecht[25]. This classification was supplemented by Nager for the auricular anomalies and by Cremers for aural atresia[26] and was used in this form in the presentation of the results of the Nijmegen series described in Chapter III.

Marquet[28] also applied a classification to categorize congenital aural atresia. Type I comprised a well-developed mastoid and middle ear and type II dysplasia of the mastoid and middle ear. This classification is somewhat similar to ours, but we further subdivided his type I into I and II A, and his type II was almost identical to our type II B and III. For better preoperative evaluation of the development of the mastoid and middle ear and subsequently better preoperative selection of patients with congenital aural atresia, Jahrsdoerfer[17,29] stressed the value of CT scanning and the accompanying scoring system.

Patients and methods

The results used in this study were all obtained at the Department of Otorhinolaryngology of the Nijmegen University Hospital. All the data from successive patients operated on between 1964 and 1990 with minor ear anomalies and between 1964 and 1988 with major ear anomalies were analysed. Reliable access to these data was possible through the ear control system which had been set up at the Nijmegen clinic. This system provides a record of each ear operation performed at the Nijmegen University Hospital and includes standardized postoperative follow-up and audiometric test results over a period of at least two years.

All the patients with minor ear anomalies had been suffering from typical conductive deafness since early childhood. In the patients with a clearly defined syndromal diagnosis, a minor congenital ear anomaly was very likely. In all the other cases, ears with tympanosclerosis, osteogenesis imperfecta[27] or otosclerosis were excluded. The following inclusion and exclusion criteria were gradually developed and employed:

1. The patient must be at least 10 years old at the time of the operation.
2. In children, intermittent periods of otitis media with effusion must be excluded by a sufficiently long follow-up.
3. The results of tone audiometry, speech audiometry and contralateral stapes reflexes must be available.
4. Preoperative CT scans must be available of the ossa petrosa, particularly in patients with mixed hearing loss.

Individual and mean preoperative and postoperative hearing levels are presented for the frequencies 0.5, 1 and 2 kHz. Hearing levels obtained one year postoperatively were used to calculate the hearing gain. The calculation of means was based on individual values. Preoperative bone conduction thresholds were not corrected if they changed postoperatively.

THE PURPOSE OF THE STUDY

The aim of this study was to gain more insight into the diversity of congenital anomalies of the ear operated on at the Department of Otorhinolaryngology of the Nijmegen University Hospital and to compare the results to those reported in other series in the literature.

Topics of special interest included the surgical findings, the postoperative results, the incidence and importance of a syndromal diagnosis and various audiological aspects in relation to surgery for congenital anomalies of the ear.

Analysing the anomalies encountered in ears with a major or minor congenital anomaly and the results obtained after ear surgery will contribute to increasing our insight into the type of ears with a sufficiently favourable surgical prognosis and promote the development of a valid classification system. In this way it will be possible to analyse the anomalies which lead to less favourable results and develop new surgical techniques. In addition, high-risk ears can be recognised preoperatively. This will enable us to identify the risks and potential success rates associated with these ear operations. The aim is to contribute to improving the quality

of the care for patients with a major or minor congenital ear anomaly.

OUTLINE OF THE THESIS

The surgical findings and results in patients with a minor congenital ear anomaly are described in **Chapter II**. The classification for minor ear anomalies presented in **Chapter II-4** was used in the preceding chapters when describing the findings and results. The classification for congenital minor ear anomalies is based on surgical data. Important criteria are a fixed stapes footplate as an indication for stapes replacement surgery, or a congenital anomaly with a mobile stapes footplate as an indication for some other type of ossicular chain reconstruction.

Aplasia or serious dysplasia of the oval or round windows are considered as separate entities in this classification. Using this system, the patients were classified into the groups shown in Table 1.

TABLE 1. Classification of minor anomalies of the ear.

Class	Main anomaly	Subclassification	No. of ears
I	Congenital stapes ankylosis		44
II	Stapes ankylosis associated with another congenital ossicular chain anomaly		55
III	Congenital anomaly of the ossicular chain but a mobile stapes footplate	A. Discontinuity in ossicular chain	22
		B. Epitympanic fixation	9
IV	Congenital aplasia or severe dysplasia of the oval or round window	Aplasia	10
		Dysplasia	
		Crossing facial nerve	3
		Persistent stapedial artery	1

The initial series comprised 104 ears operated on between 1964 and 1986. In **Chapter II-4** the series is extended to 144 ears operated on in the period between 1964 and 1990. **Chapter II-5** reports the long-term results of the initial series of 104 operated ears.

A discussion on the historical development of stapes surgery for minor congenital ear anomalies is part of **Chapter II-1**. It shows clearly that the development of the fenestration technique towards stapedectomy at that time was delayed in comparison with the developments in otosclerotic surgery.

Chapter III describes the findings and results in the patients with major congenital ear anomalies and **Chapter III-2** reports the long-term results. Once again, the classification system was used in the presentation. Contrary to the minor ear anomaly classification, the classification for major congenital ear anomalies is based mainly on the severity of the aural atresia encountered. The milder forms of congenital aural atresia, generally without auricular malformations, formed group I and IIA, while the more serious cases of atresia formed groups IIB and III.

The fascinating problems surrounding syndromes in relation to major and minor congenital ear anomalies are described in **Chapter IV**, and in **Chapter IV-1** a new syndrome is proposed on the basis of this series of patients.

The degree to which it is desirable and advantageous to operate on unilateral congenital ear anomalies is a long-standing subject of discussion[12,13,15,16,26]. In **Chapter V** several audiological aspects of major and minor ear anomalies are incorporated into this discussion with the aim of evaluating not only whether surgery is able to achieve a measurable improvement in the patient's hearing to the level of the unaffected ear, but also whether the patient actually experiences any measurable benefit. Deprivation of the audiovestibular system may have occurred in the years prior to surgery and should receive consideration in this question.

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CHAPTER II

MINOR EAR ANOMALIES

CHAPTER II-1

ISOLATED CONGENITAL STAPES ANKYLOSIS SURGICAL FINDINGS AND RESULTS IN 32 EARS AND A REVIEW OF THE LITERATURE

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Laryngoscope 1990; 100: 1331-1336

ABSTRACT

Isolated congenital stapes ankylosis is described in 32 operated ears from 28 patients. In 27 ears a regular stapedectomy was performed. In the remaining 5 ears, 2 had stapes gushers, 2 had bony stapedial tendons and 1 had an aberrant facial nerve crossing the oval window. The average individual hearing gain was 20 dB. The mean hearing gain for the 24 successful stapedectomies was 29 dB.

In 24 out of 32 ears (75%) an essential hearing gain of at least 15 dB Fletcher's index hearing threshold could be achieved. A Fletcher's index not exceeding 30 dB could be achieved in 19 out of 32 ears (60%), in spite of several cases with a sensorineural component in the hearing loss.

A review of the literature and overview of longer series with isolated congenital stapes ankylosis is presented.

INTRODUCTION

Congenital anomalies of the middle ear are rare. There is a large variability in the degree of malformation of the ossicular chain. In the Nijmegen Department of Otorhinolaryngology, 104 ears in 86 patients with congenital anomalies of the ossicular chain, excluding those with osteogenesis imperfecta, were explored from 1964 to 1985. The hearing loss in these patients was sometimes part of a syndrome.

A syndromal diagnosis can provide information about the spectrum of anomalies to be expected. We have, therefore, already separately published the surgical findings and results in a number of patients with syndromes such as Pfeiffer's[1], branchio-otorenal or earpits-deafness[2], cervico-oculoacoustic[3] syndromes, X chromosome-linked progressive mixed deafness with perilymphatic gusher during stapes surgery,[4,5,6] proximal symphalangia and stapes ankylosis[7], Klippel-Feil syndrome[8], frontometaphyseal dysplasia[9] and osteogenesis imperfecta.[10,11] From this series, the data from a girl with Crouzon's disease and several cases with Treacher Collins Syndrome are still unpublished. Recently, a report presenting a new autosomal dominant syndrome with stapes ankylosis, hypermetropia, syndactyly of the 2^{e} and 3^{e} toes with broad first toes and thumbs has been published.[12]

Since there is a wide spectrum of congenital anomalies of the middle ear involved, a

classification of these anomalies is required for a clear presentation and evaluation of the results. This presentation is, therefore, limited to ears with an isolated congenital stapes ankylosis without any other malformations of the malleus or incus and without total aplasia of the oval window.

PATIENTS AND METHODS

From 1964 to 1985, 104 ears in 86 patients with congenital anomalies of the ossicular chain were operated on in the Nijmegen Department of Otorhinolaryngology. All had a typical history of conductive deafness since childhood. Cases of suspected otosclerosis or tympanosclerosis were excluded from this series. Of the 104 ears, 32 ears in 28 patients appeared to have isolated congenital stapes ankylosis without any other malformation of the malleus or incus. Four patients underwent surgery on both ears for stapes ankylosis. The degree of the malformation of the stapes, the kind of stapes surgery and the type of teflon prosthesis used were found in the surgical reports.

The preoperative and postoperative hearing levels were calculated separately and as the mean of frequencies 0.5, 1, 2, kHz. Hearing gain calculations are based on individual values. Speech audiometry was performed preoperatively in all cases and, in many cases, also postoperatively. The following preoperative requirements were developed during these years for these ears. However, not all operated ears totally met these requirements, especially in the years before 1978.

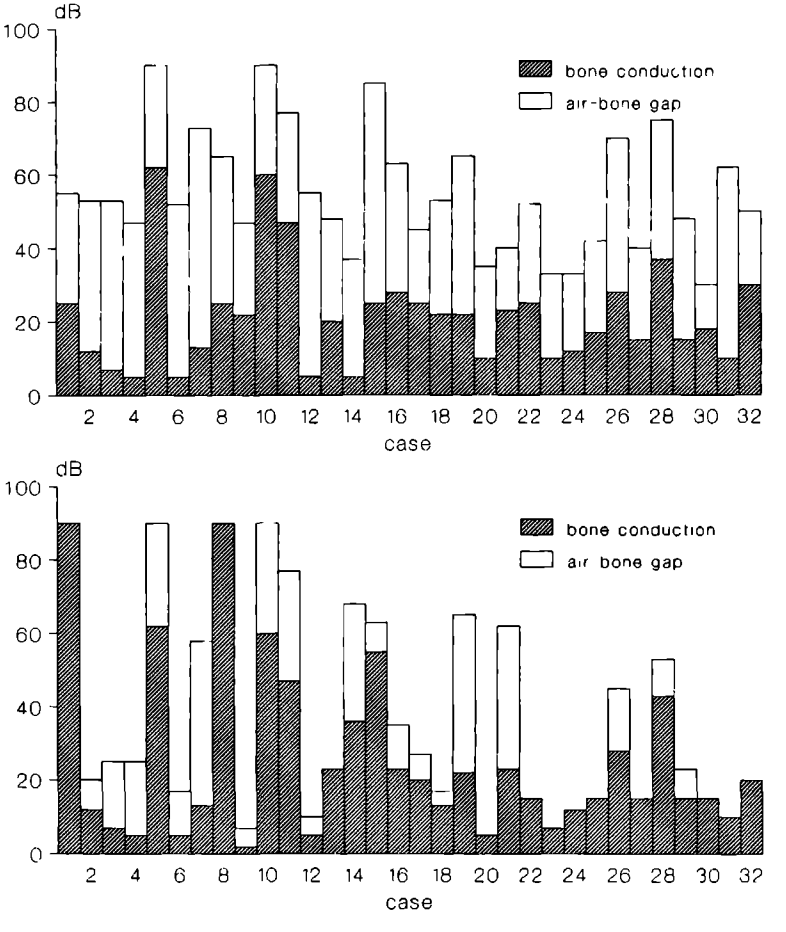
1. The patient has to be at least 10 years old at the time of operation.
2. In children, intermittent periods of serous otitis media must have been excluded by a sufficiently long follow-up.
3. Tone audiometry, speech audiometry, and examination of the contralateral stapedial reflexes must be performed.
4. Petrous bones must be computed-tomography scanned, especially in patients with a mixed hearing loss.

The age at which a child can be accepted for stapes surgery is arbitrary and mainly based on the fear of postoperative complications as a result of immediate postoperative or subsequent otitis media. The CT-scanning of the petrous bone was mainly performed to trace inner ear anomalies, especially those which appeared to be related to stapes gusher.[5,13]

Preoperative hearing levels are presented in Figure 1. A typical Carhart notch was seen in only 11 of 32 preoperative audiograms. Contralateral stapedial reflexes were routinely examined from 1978 and proved to be absent in 17 of 19 ears.

Postoperative speech audiometry levels were available in nearly all cases and were as could be expected from the pure-tone audiometry hearing levels. Tone audiometry hearing levels were used for the interpretation of the hearing results.

Figure 1: Preoperative and 1 year postoperative hearing levels for all 32 operated ears seperately (0.5, 1, and 2 kHz dB HL). Preoperative audiometry was troublesome in cases 5, 8, and 10. Surgery was accepted based on preoperative audiograms shown.



In one patient operated on both ears, an autosomal dominant inherited syndrome was present. Klippel-Feil syndrome was found in two other patients (three operated ears) without clear histories of familial hearing loss. Clear histories of familial hearing loss were present in only three patients, including the patient with the autosomal dominant inherited syndrome.

Contralateral hearing loss was found in 29 of 32 operated ears. Only three patients had unilateral hearing losses.
Of all operated ears, 8 ears had undergone surgery between 10 and 20 years of age, 15 after 20 years and 9 before 10 years of age. The 9 ears in these younger patients were all operated on before 1978.
The stapes suprastructure had a normal configuration in 25 of these 32 ears. In 7 cases, malformations of the suprastructure of the stapes was found.

Regular stapedectomies were performed in 27 ears: 8 stapedotomies, 12 partial platinectomies and 7 total stapedectomies were performed. All-Teflon prostheses were used in 23 of these stapedectomies. Teflon-wire prostheses were used in 4 ears. Bony stapedial tendons caused the stapes ankylosis in both ears of 1 patient (Figure 1: no. 20 and 23). Removal of the bony bridge between the pyramidal eminence and the stapes resulted in a closed air-bone gap in both ears.[14]
In two cases, a stapes gusher was encountered after opening the footplate by drilling a safety hole (Figure 1: no. 11 and 14). Preoperative and postoperative audiograms have been previously published.[4] In another case, an aberrant facial nerve crossed the oval window (Figure 1: no. 5).

This series contains 8 ears without a hearing gains or aggravation of the hearing losses. Three ears were preoperatively almost deaf (Figure 1: no. 5, 8, 10). In none of these 3 cases was a hearing gain established after stapedectomy. Preoperative audiograms were doubtful, and masking problems must have complicated the determinations of the conductive component in the hearing loss of these 3 young patients. One of these patients is a 5-year-old boy (Figure 1: no. 8). He was probably already deaf before his stapedectomy; furthermore, we have also noted a progressive sensorineural hearing loss in his contralateral ear. In 2 other cases with perilymphatic gushers, the hearing loss increased mainly as a result of an increased airbone

gap. We have no explanation for the lack of hearing gain in the remaining 3 cases.

Only in patient 31 did a failure to attain the expected hearing gain result in re-exploration of the middle ear. The Teflon prosthesis had slipped off, as was found at re-exploration for a renewed hearing loss after previous successful surgery. We have inadequate information about the reasons for the other failures.

In Table 1, the preoperative and postoperative hearing levels of the 24 successful ears are presented separately for the group with and without malformations of the stapes. The preoperative and postoperative air-bone gaps are presented in Table 2. The overall mean hearing gain is 29 dB. The hearing gain is at least 11 dB larger in the group without malformed stapes compared with the group with malformed stapes (Student's t, $p < 0,05$). There is no explanation for this finding. Small sensorineural components already present in most of the preoperative audiograms have limited the interpretation of the final results.

This explains the differences between Tables 1 and 2. Table 3 presents the overall results for all 32 operated ears. In 3 of 4 cases, essential and substantial hearing gains of more than 15 dB mean hearing thresholds could be achieved. In 2 of 3 cases, the Fletcher Index was better than 30 dB. Another small group also experienced substantial hearing gains, although they still need the support of hearing aids.

Table 1: Hearing levels in 24 successful operations (0.5, 1, 2 kHz dB HL).

	Pre HL	1 Year Post HL	Hearing Gain
Nonmalformed stapes (n = 19)	53 dB (SD = 14)	22 dB (SD = 16)	30 dB (SD = 9)
Malformed stapes (n = 5)	49 dB (SD = 15)	29 dB (SD = 17)	20 dB (SD = 5)
Total (n = 24)	52 dB (SD = 14)	23 dB (SD = 16)	29 dB (SD = 9)

Note: Pre HL = preoperative hearing level; Post HL = postoperative hearing level; SD = standard deviation.

A possible explanation for the slightly enlarged air-bone gap in this series of stapedectomies, compared with the results of stapes surgery in otosclerosis, may be the presence of malleo-

incudal joint ankylosis even without an epitympanic fixation of one of these ossicles. An unnoticed fixation of the short process of the incus in the incudal fossa may be another explanation. The failure to achieve good hearing improvement may also be due to a nonfunctional round window, since dysplasia or aplasia of the round window may have gone unnoticed.

Table 2: Air-Bone Gaps in 24 Successful operations (0.5, 1, 2 kHz dB HL).

	Pre A-B	1 Year Post A-B
Nonmalformed stapes (n = 19)	35 dB (SD = 12)	6 dB (SD = 6)
Malformed stapes (n = 5)	35 dB (SD = 18)	15 dB (SD = 19)
Total (n = 24)	35 dB (SD = 13)	7 dB (SD = 10)

Note: Pre A-B = preoperative air-bone gap; Post A-B = postoperative air-bone gap; SD = standard deviation.

Table 3: Hearing levels in all 32 operated ears (0.5, 1, 2 kHz dB HL).

	Pre HL	1 Year Post HL	Hearing Gain
Nonmalformed stapes (n = 25)	56 dB (SD =17)	36 dB (SD =29)	23 dB (SD =19)
Malformed stapes (n = 7)	52 dB (SD = 14)	43 dB (SD =28)	10 dB (SD = 22)
Total (n = 32)	55 dB (SD = 16)	37 dB (SD = 29)	20 dB (SD = 20)

Note: Pre HL = preoperative hearing level; Post HL = postoperative hearing level; SD = standard deviation.

The follow-up period varied from 1 to 19 years, with an average of 5 years. The long term results proved to be the same as the results after 1 year within 5 dB limits.

In none of the ears in the series was there damage to the function of the facial nerve, nor was there any persistent dizziness after stapes surgery.

REVIEW OF THE LITERATURE

In the last 3 decades, no classification of congenital middle ear anomalies has been generally accepted. Congenital stapes fixation with or without other congenital anomalies of the middle ear has been the most discussed and has been presented in numerous publications[16-38,43,45]. Some authors, like Ombredanne[22-29] and Hough[20], published detailed reports of many cases in quite large series of operated patients with congenital middle ear anomalies.

Arslan and Giacomelli[32] suggested a classification with five types of congenital stapes ankylosis, but without separate inclusion of the combination with epitympanic fixation of the malleus and the incus (congenitally fixed malleus syndrome) or fixation of incus alone in the fossa incudis.

Although the stapedectomy procedure for otosclerosis was used more and more in the 1960's, this was not the procedure of choice for isolated congenital stapes ankylosis.

In 1952, Shambaugh[16] published his success with the fenestration operation in 5 cases of isolated congenital stapes ankylosis. Since then several publications have mentioned similar results[17,22-28,30,32,45-49,51].

In 1958, Howard House *et al.*[18] described their attempts to mobilize the stapes footplate by needle and chisel techniques in 23 cases with congenital footplate fixations. When no hearing improvement was achieved, an additional fenestration operation was performed. In 1969, Steel[34] clearly described the results of the exploratory tympanotomies performed by House in the 1960's for congenital stapes ankylosis. This shows that House rarely used the stapedectomy procedure for this indication during this period. In 1958, Hough[20] described a large series of congenital middle ear anomalies as seen in exploratory tympanotomies for stapes mobilization.

Confidence gradually increased with the increasing success and safety of stapes interposition and stapedectomy procedures.[19,33-42,46,52-56] This also led to the virtual abandonment of fenestration of the lateral semicircular canal and stapes mobilization. In this period stapes mobilization was also used by others for congenital stapes ankylosis.[19,20,22-28,31,32,34,47,57,58]

Fear of a perilymphatic gusher during stapes surgery, and with it the incidental occurrence

of a postoperative dead ear, may also have favoured the prolonged choice of stapes mobilization instead of a stapedectomy.[26,30,35,54,59]

Ombrédanne's[22-29] French publications in the 1960's show the success of stapes mobilization and fenestration operations of the lateral semicircular canal in those cases where stapes mobilization procedures did not result in a mobile stapes.

In those days, stapes mobilization was the procedure of choice. Partial stapedectomy with repositioning of the stapes on a vein graft and stapedectomy with interposition were considered to be second choice procedures, but appeared to be as successful as a stapes mobilization.[22,24-26] A series of publications confirmed the good results of stapedectomies with interposition using a prosthesis. The prosthesis was fixed to the long process of the incus or, when this was absent, to the long process of the malleus.[19,33-42,46,52-55]

More recent studies on the results of surgery for congenital deafness both with and without anomalies of the ossicular chain are listed below. In 1967, Scheer[56] published his surgical results in 17 ears. Eight cases suffered from the Treacher Collins syndrome. A stapedectomy was performed in 13 ears. Five ears had complete closure of the air-bone gap. Twelve operations resulted in functional hearing. There was no further loss of hearing in any case in that series and there were no postoperative complications. In 1970, Gerhardt and Otto[36] reported three successful stapedectomies performed for isolated congenital stapes ankylosis. Funasaka[39] published a series of 6 ears with stapedectomies for isolated congenital stapes ankylosis.

In 1980, Morimitsu, *et al.*[42] published their results in 3 cases of isolated congenital stapes ankylosis and 7 cases where the prosthesis was fixed to the malleus. In 1980, House, *et al.*[40] published the results of 13 stapedectomies in 20 patients with congenital conductive hearing losses. In 12 cases, this resulted in air-bone gaps smaller than 10 dB. An overview of these larger series with isolated congenital stapes ankylosis is presented in Table 4.

DISCUSSION

Congenital abnormalities of the middle ear are sporadic, mostly nonfamilial. Sometimes they are part of a congenital syndrome. The stapes, with its dual embryological development from the otic capsule and second branchial arch, is liable to distinct and separate abnormalities of the footplate and the suprastructure. The annular ligament may fail to develop completely;

this may be found as an isolated anomaly or associated with a more generalized abnormality of the otic capsule.

This series of 27 stapedectomies for the treatment of an isolated congenital stapes ankylosis presented here represents the largest series. The present results have been specifically limited to isolated congenital stapes ankylosis in order to enable a better comparison of this type of surgery with cases of stapes ankylosis associated with other middle ear anomalies. Our results clearly show that many of these ears have well over 10 dB sensorineural hearing losses. This sensorineural hearing loss alone already limits the final result. A postoperative air-bone gap that is not perfectly closed is not unusual in these cases and presents an additional disadvantage in achieving a good final result, as can be seen from Tables 1 and 2.

Studying the audiograms of similar patients published by other authors shows that this small sensorineural hearing loss also appears to be present in their patients, with the same effect on the final results.

In some patients with isolated stapes ankylosis, we noted a remarkably larger sensorineural component in the hearing losses. There seems to be an unfavourable correlation between a larger sensorineural component and the possibility of achieving a decrease in the air-bone gap. A clear example is the X chromosome-linked progressive mixed deafness with a perilymphatic gusher during stapes surgery. Although stapes surgery has never been successful in this syndrome, recent studies have stressed the importance of preoperatively establishing other factors of this syndrome, such as the type of audiogram, eliciting stapedial reflexes and the radiological anomalies of the vestibule and internal acoustic canal.[4,5,6] There has been recent success with gene-linkage studies (to DFN_3) in this syndrome.[44,50]

CONCLUSION

We conclude that the results from the present study and the results obtained by others indicate that surgery for isolated congenital stapes ankylosis is favorable, especially for almost pure conductive hearing losses, that such surgery should be continued. Surgical treatment can also be considered in cases with unilateral pathology. We also consider it acceptable to operate upon the second ear of a patient after previously successful surgery on the first ear.

Table 4: Larger series with congenital stapes ankylosis in the literature.

Author	Year	No out of a Total Series	Surgical Procedures	Ears with Gain > 15 dB	Mean gain in ears with Gain > 15 dB	Other congenital middle ear anomalies in series
Shambaugh[16]	1952	5/5	5 Fenestrations	4	30 dB	None
House, et al [18]	1958	23/23	18 Mobilizations 3 Fenstrations 2 Terminated	12 > 10 dB	NS	None
Hough[20]	1958	1/25	1 Mobilization	1	25 dB	Various
Ombredanne[22 29]	1958 - 1968	24/77	14 Mobilizations 3 Fenestrations 7 (partial) stapedectomies	17	22 dB	27 nonisolated stapes ankylosis, various other anomalies
Arslan and Giacomelli[32]	1963	6/6	4 Mobilizations 1 Fenestration 1 Terminated	5	40 dB	
Gunderson[33]	1967	2/10	2 Stapedectomies	" closed air bone gap"		8 nonisolated stapes ankylosis
Steele[34]	1969	NS/32	11 Stapedectomies 19 Mobilizations 2 Fenestrations	25	29 dB	Isolated and nonisolated stapes ankylosis NS
Zuhlke[35]	1969	NS/21	9 Stapedectomies	6	27 dB	9 isolated and nonisolated stapes ankylosis not separated, 12 oval window aplasias and/or inner ear anomalies
Gerhardt and Otto[36]	1970	3/8	3 Stapedectomies	3	NS	3 aplasias of oval window, 1 persistant stapedial artery , 1 malleus-incus anomaly
Funusaka[39]	1979	6/18	6 Stapedectomies	4	21 dB	1 aplasia of oval window, 11 anomalies with mobile stapes
Morimitsu,et al[42]	1980	3/13	3 Stapedectomies	2	39 dB	6 nonisolated stapes ankylosis 3 aplasias of oval window 1 anomaly with mobile stapes
House, et al[40]	1980	4/20	4 Stapedectomies	10 closed air-bone gaps in 13 stapedectomies		Isolated and nonisolated stapes ankylosis not separated
Present study	1989	32/104	27 Stapedectomies 2 Gushers 1 Seventh cranial nerve crossing over 2 Bony stapedial tendons			

Note NS = not specified

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CHAPTER II-2

SURGERY FOR CONGENITAL STAPES ANKYLOSIS WITH AN ASSOCIATED CONGENITAL OSSICULAR CHAIN ANOMALY

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International Journal of Pediatric Otorhinolaryngology 1991; 21: 217-226

ABSTRACT

The surgical findings and results are presented on 32 ears with congenital stapes ankylosis with an associated congenital anomaly of the ossicular chain. One third of the patients had a syndromal diagnosis. In 26 ears, stapedectomy could be performed. In 2 other ears, stapes ankylosis to the bony facial canal was mobilized successfully. In the 4 remaining ears, surgical intervention had to be limited to an exploratory tympanotomy for various reasons. The average hearing gain was 23 dB for the 28 ears on which stapes surgery had been performed. A substantial hearing gain of at least 15 dB was achieved in 19 of these 28 ears (68%).

The end result was limited to a small extent by an average preoperative sensorineural component of 16 dB in the hearing loss.

A review of the findings and results from other larger series in the literature is presented.

INTRODUCTION

It is exceptional that conductive hearing loss in the young child is caused by a congenital ear anomaly. Very few series are available in the literature on the surgical findings in subjects with congenital middle ear anomalies.[1,11,13-27,29,30,31,34]

Although the hearing loss is usually a solitary congenital anomaly without any recognizable hereditary components, it is not unusual for a conductive hearing loss to form part of a syndrome or have a hereditary origin. A series of publications on the findings and results of middle ear surgery in patients with a congenital middle ear anomaly and a syndromal diagnosis have been published before.[2,3,6-10,28,32]

In a previous study we published our results on a group of 32 ears with isolated congenital stapes ankylosis, without an associated middle ear anomaly.[33]

This paper is limited to 32 ears in 31 patients with congenital stapes ankylosis and an associated congenital anomaly of the ossicular chain.

PATIENTS AND METHODS

From 1964 to 1986, exploratory tympanotomy was performed on 104 ears in 86 patients at

the Department of Otorhinolaryngology of the University Hospital Nijmegen, the Netherlands. A large variety of anomalies were found, whether or not in combination with a syndromal abnormality. Groups of similar ossicular chain abnormalities could be distinguished. For the purpose of this study, the patients with osteogenesis imperfecta were excluded,[4,5,13] leaving 104 ears for the study.

In 32 ears a congenital stapes ankylosis was noted associated with another congenital deformity, or fixation of the ossicular chain. The anomalies of the ossicular chain associated with congenital stapes ankylosis were divided into type A and type B. Type A for the ears with congenital stapes ankylosis and an incus or malleus configuration which was considered anomalous and type B for ears with congenital stapes ankylosis and combined fixation of the malleus or incus in the epitympanum.

A history of hearing loss since early childhood without evidence of long-term middle ear pathology was present in all cases. The majority of these patients were known to our department since there early childhood, so that long-term middle ear pathology could be excluded. In the other cases long-term middle ear pathology could be excluded by history, otological examination and information of the referral centre. A few patients had also experienced periods of serous otitis media. The insertion of grommets in these cases did not lead to any improvement. Therefore, the suspicion arose that the hearing loss may have a congenital origin.

During the study period, the following preoperative criteria have gradually been developed:

1. The patient must be at least 10 years of age.
2. In children, a sufficiently long follow-up period must exclude intermittent periods of serous otitis media.
3. Results of tone audiometry, speech audiometry and examination of the contralateral stapedial reflexes must be available, plus
4. CT scans of the petrous bones, especially in patients with mixed hearing loss.

CT scanning of the petrous bones provides useful information on the pneumatization of the mastoid and the size of the middle ear cavity and forms a sensitive means of tracing inner ear anomalies, especially those which are related to a stapes gusher.[8]

The preoperative and postoperative hearing levels were calculated for the frequencies 0.5, 1 and 2 kHz. The medical histories of all the cases included detailed surgical reports on all the ears, describing the anomalies of the ossicles, the middle ear and surgical reconstruction and,

where relevant, the diagnosis of congenital hearing loss, syndromal diagnosis or family history of hearing loss. The unilateral or bilateral appearance of the hearing loss was of special interest.

SURGICAL FINDINGS AND RESULTS

A stapedectomy was performed in 26 out of the 32 explored middle ears. The age at first surgery varied from 5 to 49 years, with a mean of 20 years. In 4 ears (Nos 12, 15, 27 and 28), the middle ear was explored, but reconstruction of the ossicular chain was not attempted for the following reasons.

- At the exploratory tympanotomy, ears No. 15 and 28 showed stapes ankylosis and aplasia of the long process of the incus, with hypermobility of the malleus. In these cases, the surgeon preferred not to perform stapedectomy with interpositioning of a malleus footplate prosthesis.
- Radiological examination of a patient with Crouzon's disease (No. 27) showed a very small mastoid. She had congenital stapes ankylosis and fixation of the malleus and incus in a very small epitympanum. Owing to the small size of the mastoid, an endaural epitympanotomy was performed to free the malleus and incus. During a second-stage procedure, in which it was intended to perform a stapedectomy, the malleus and incus proved to be blocked again in the epitympanum. The stapedectomy was not performed because recurrent fixation was expected.
- A canal wall down technique was performed on ear No. 12 to correct chronic otorrhoea and long-term hearing loss. No cholesteatoma was found. The middle ear showed a total congenital anomaly with fixation of the ossicular chain. No corrective surgery for the ossicular chain was carried out in this case.

In 2 ears of patients with the Treacher Collins syndrome (Fig. 1 and 2, Nos 5 and 19), the stapes and long process of the incus were ankylotic with the bony wall of the facial canal, but could be detached and mobilized successfully. In 10 patients (10 operated ears) a well-defined syndromal diagnosis was made. Branchio-oto-renal syndrome was seen in 5 patients, Treacher Collins syndrome was seen in 2 patients and Proximal symphalangia and stapes ankylosis, Pfeiffer syndrome and Crouzon syndrome each seen in 1 patient.

Figure 1 and 2 show the individual preoperative hearing levels of the 32 ears. The data are

presented in chronological order according to the date of first surgery.

Fig. 1. Individual preoperative hearing levels for bone and air conduction in 32 operated ears, according to date of first surgery.

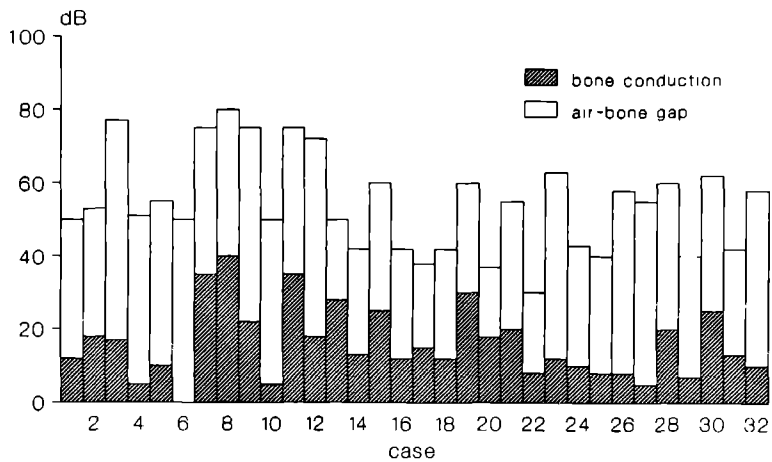
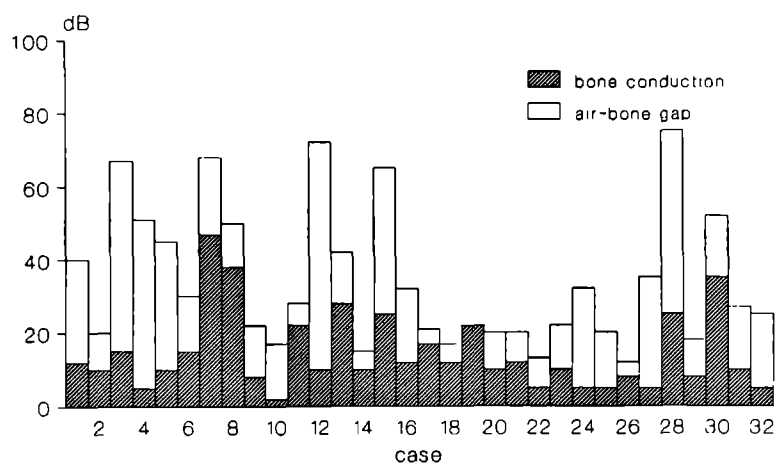


Fig. 2. Individual 1 year postoperative hearing levels for bone and air conduction in 32 operated ears, according to date of first surgery.



We divided anomalies of the ossicular chain associated with congenital stapes ankylosis into type A and type B. For type A ears with congenital stapes ankylosis and an incus or malleus configuration which was considered anomalous, was in most cases the long process smaller and malformed. In two cases (Nos 21, 23) we even found total aplasia of the long process. Type B, congenital stapes ankylosis and combined fixation of the malleus or incus in the epitympanum, was encountered in 9 out of the 32 ears. Malformation of the malleus or incus was found in 4 of these 9 ears.

Stapedectomy together with the necessary amount of mobilization of the malleus and/or incus was performed in one procedure in 6 ears (Nos 1, 11, 25, 26, 30 and 32), whereas the two procedures were separated into two stages in the other 3 ears (Nos 6, 17 and 22).

A total of 26 operations of the stapes were carried out, 13 stapedotomies, 4 partial stapedectomies and 9 total stapedectomies. A Teflon prosthesis was used in 19 ears and a Teflon-wire prosthesis was used in 7 ears.

Table 1 Preoperative and 1 year postoperative hearing loss and bone conduction levels in 28 ears

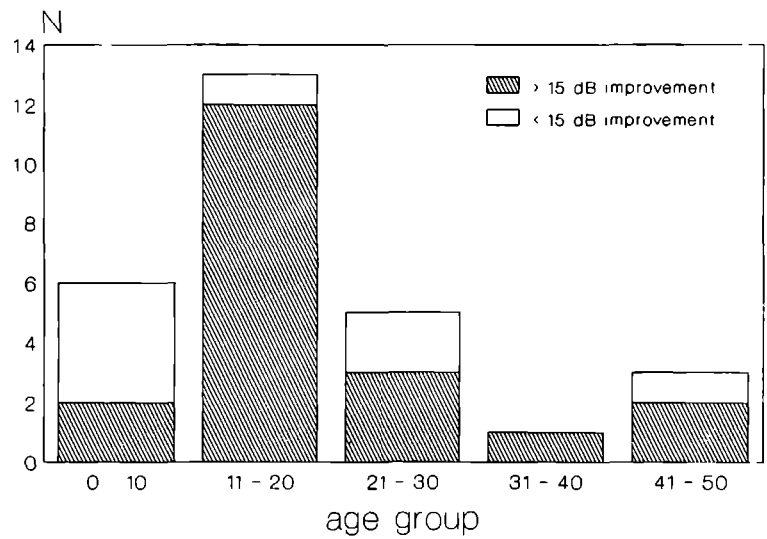
	n	Preoperative	Postoperative	Gain
Type A	17	54 dB	32 dB	22 dB
Type B	9	51 dB	27 dB	24 dB
Ankylotic stapes head	2	58 dB	34 dB	24 dB
Total	28	53 dB	30 dB	23 dB
Bone conduction	28	16 dB	15 dB	-

The postoperative hearing levels are presented in Fig. 2. The mean preoperative hearing levels and the mean postoperative hearing levels after one year follow-up were calculated for the frequencies 0.5, 1 and 2 kHz (see Table 1). The results are subdivided into type A, type B and the 2 ears with a stapes head fixed to the bony canal of the facial nerve. The average hearing gain for type A was 22 dB and for type B, 23 dB. Preoperative audiometry showed a clear Carhart notch in the hearing loss in only 5 cases. In 11 patients the anomaly was

unilateral, whereas in 21 operated ears, the contralateral ear was also involved.

Table 2 shows the postoperative success rate in the 28 patients who underwent stapes surgery. The data are presented in age groups, according to the age at the time of surgery. A hearing gain of at least 15 dB at the frequencies 0.5, 1 and 2 kHz was considered to be successful. The success rate was highest in the patients in their second decade. The average preoperative and postoperative sensorineural component in the hearing loss was 16 dB (range 5 - 40 dB) and 15 dB (range 5 - 40 dB), respectively.

Table 2: Improvement of hearing per age group.



Note: Shaded areas show the ears in which the hearing level improved by 15 dB or more.

In 6 cases (19%), revision surgery was necessary, two times even more than once. In 3 ears (Nos 1, 3 and 24) the prosthesis proved to have slipped off the malformed long process of the incus. Re-exploration was performed in two ears (Nos 22 and 25) because no hearing improvement was noted after the previous stapedectomy. Congenital fixation of the short process of the incus in the fossa incudis was found, which had gone unnoticed during the

earlier operation. Revision of the malleus footplate prosthesis was necessary in one other ear with stapes ankylosis and aplasia of the long process of the incus (Nos 21), because the prosthesis proved to be too short and there were initial problems with the fixation of the prosthesis to the malleus. This ultimately resulted in an essential hearing gain.

DISCUSSION

Our study group of 32 ears with congenital stapes ankylosis in combination with other associated congenital middle ear deformities, formed part of a larger series of 104 ears.
The surgical results of 32 ears with isolated congenital stapes ankylosis have been presented in a previous publication.[33] Patients with osteogenesis imperfecta and a conductive hearing loss were excluded from this study.[4,5,12]
Although these patients were suffering from hearing loss with a hereditary origin, this progressive hearing loss does not manifest itself before the first decade.
In the 9 ears of patients operated after the age of 21 years the anomaly of type A or Type B was in all cases of congenital origin rather than of inflammatory origin. The fact that some patients have either a hereditary hearing loss, a syndromal diagnosis, multiple external congenital features or a clear congenital anomaly like e.g. a monopodial stapes suprastructure and a negative medical history for long standing middle ear pathology makes a inflammatory cause of the anomaly extreme unlikely.

It is striking that there was such a low incidence of total failures in our series. None of the patients showed a serious decrease in hearing level or even a temporary postoperative deaf ear. Serious complications, such as persistent facial paralysis, were not encountered in this series. However, the hearing of one patient (No. 1) suddenly deteriorated 14 years postsurgery, after he had been parachuting. Explorative middle ear surgery revealed a perilymphatic fistula along the teflon prosthesis, which was closed using a venous graft; the shortened teflon prosthesis was reinserted.
In 4 ears it was not possible to complete surgical reconstruction of the ossicular chain. In 2 cases, it later appeared possible to perform interpositioning between the malleus and stapes footplate.
Table 2 shows that the results of reconstructive ear surgery in children younger than 10 years

of age in our series, were less favourable. This finding supports our present opinion that reconstructive ear surgery should be postponed until the age of 10 years in children with this type of abnormality. An additional advantage of waiting until the child has reached this age is that the results of hearing tests are more reliable, especially in cases with larger, mixed bilateral hearing loss. The years before successful surgery in patients with bilateral hearing loss were and should be covered by using hearing aids and intensive training. Acceptation of prolonged use of hearing aids after unsuccessful or partly successful surgery will be better in preoperative well supported patients.

Table 1 shows that the average preoperative and postoperative perceptive component of hearing loss is fairly large. A number of well-known syndromes are often accompanied by perceptive hearing loss. For example, a perceptive component is often encountered with the Branchio-Oto-Renal syndrome, in which 2 cochlear convolutions are found instead of 2½.[10] The phenomenon of improvement of the perceptive threshold after successful stapes surgery, was not observed in our series of ears with congenital stapes ankylosis.

Table 3: Larger series in the literature on surgery on ears with congenital stapes ankylosis and an associated anomaly of the ossicular chain.

Author	Year	No of ears	Surgical Procedures	Ears with gain > 15 dB	Mean gain in ears with gain > 15 dB	No and specification of other described middle ear anomalies in their series
Ombredanne	1959	27	10 fenestrations 10 mobilizations 7 stapedectomies	16	25 dB	24 isolated stapes ankylosis 26 various
Hough	1958	2	2 mobilizations	2	27 dB	23 various
Scheer	1967	3	3 stapedectomies	not specified		14 various
Gunderson	1967	8	8 stapedectomies	not specified		2 isolated stapes ankylosis
Morimitsu	1980	6	6 stapedectomies	5	28 dB	3 isolated stapes ankylosis 3 aplasia of oval window 1 discontinuity in chain
House	1980	9	9 stapedectomies	10 closed air-bone gaps in a series of 13 stapedectomies		11 various
Present study	1989	32	28 stapedectomies 4 terminated	19	30 dB	32 isolated stapes ankylosis 27 anomalies with a mobile stapes 13 aplasia of oval window

During surgery we observed that one third of the patients were suffering from a well-defined syndrome. It appears that separate syndromes are associated with distinguishable,

characteristic middle ear deformities. This observation is supported, for example, by the repeated finding of a fixed stapes head in cases with the Treacher Collins syndrome and also by data in the literature, theoretical embryological considerations and our own observations. Our classification into type A or type B was based on the nature of the associated ossicular chain deformity. There was no difference between the postoperative results for type A and type B.

Table 3 gives an overview of the larger series in the literature and shows the wide variation in congenital middle ear deformities. In a number of series, the thorough description of the deformities enabled us to distinguish between various corresponding deformities. Therefore, it was possible to isolate the surgical findings and results of the patients who were suffering from middle ear deformities with stapes ankylosis in combination with ossicular chain deformity, from those of the total series. These data are shown in Table 3.

In experienced hands, reconstructive middle ear surgery will generally lead to a considerable improvement in hearing in ears with congenital stapes ankylosis and an associated deformity of the ossicular chain. This conclusion is supported by the results reported in the literature (see Table 3) and by our own experience.

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CHAPTER II-3

SURGERY FOR CONGENITAL MIDDLE EAR ANOMALIES WITH A MOBILE STAPES

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European Archives of Otorhinolaryngology 1992 (submitted).

ABSTRACT

In a series of 104 patients with congenital middle ear anomalies operated on in the period from 1964 to 1986, 27 cases were found in which the stapes footplate was mobile and the conductive deafness was due to an anomaly in the remaining part of the ossicular chain.
In 8 cases the congenital middle ear anomaly was caused by discontinuity of the ossicular chain owing to congenital malformation. In the other 19 ears, epitympanic fixation of the ossicular chain was observed, whether or not in combination with malformation of the stapes, incus or malleus.
Ossicular chain reconstruction produced an improvement of at least 15 dB in 6 of the first 8 ears with discontinuity of the ossicular chain. The mean gain was 31 dB. Epitympanotomy with exposure of the fixed ossicle and if necessary ossicular chain reconstruction, led to an improvement of at least 15 dB in 12 of the 19 ears. The mean gain was 28 dB.

INTRODUCTION

Congenital middle ear anomalies are rare, as are reports in the literature on large series of congenital middle ear anomalies.[1,8,9,10-16,19]
The surgical findings showed a wide variety of anomalies in the middle ear, ranging from stapes ankylosis owing to a calcified annular ligament of the stapes, to complete aplasia of the ossicular chain and the oval and round windows. Histological investigation of congenital malformations of the temporal bone have shown similar severe anomalies.[9]
Owing to the large variety of anomalies encountered and for the sake of clarity, the surgical findings have been divided into groups.
In two earlier publications, we presented the results of surgery in 32 ears with isolated congenital stapes ankylosis and in 32 ears with stapes ankylosis in association with an anomaly of the incus or malleus.[19,20]

This paper is limited to the congenital middle ear anomalies in which a mobile stapes footplate was found during surgery and the remaining ossicular chain was either fixed or showed discontinuity. Various surgical techniques were employed for reconstructing the ossicular chain, whether or not in combination with exposure of the ossicular chain in the

epitympanum. It was not necessary to perform stapedectomy on any of these ears.

PATIENTS AND METHODS

In this series of 104 congenital middle ear anomalies, 27 ears in 24 patients showed a congenital anomaly of the middle ear in which the stapes footplate was mobile and the actual anomaly was found in the remaining part of the ossicular chain. Two types of anomaly could be distinguished:

A)Discontinuity of the ossicular chain, usually between the incus and stapes.

B)Congenital fixation of the ossicular chain in the epitympanum, sometimes in association with malformation of the ossicular chain.

For the analysis following data were collected: the otological history, the age at first surgery and the presence of other associated congenital characteristics which may form a symptom complex for a syndromal diagnosis. Particular attention was paid to family histories of hearing impairment and whether or not these were unilateral or bilateral. For the calculation of the results and for their presentation in tables and figures, mean hearing thresholds for air and bone conduction at 0.5, 1 and 2 kHz were employed. The postoperative airconduction thresholds measured 1 year after the last operation was used to calculate the hearing gain. The preoperative values for bone conduction were not corrected in retrospect if the postoperative values were more favourable.

A detailed surgical report was available for all 27 ear operations, which was used for the classification of the congenital middle ear anomaly and for judging the results of surgery on the ossicular chain.

SURGICAL FINDINGS AND RESULTS

The average age at first surgery was 26 years, ranging from 4 to 59 years. A syndromal diagnosis was made in 5 ears (19%): 2 cases of Treacher Collins (Fig. 1; table 2, nos 11 and 12); one patient with Klippel-Feil's syndrome who was operated on both ears (Fig. 1; table 1, nos 2 and 4) and one case with Fronto-Metaphyseal Dysplasia (Fig. 1;table 2, no. 18).

A Carhart notch was present in the preoperative audiogram in 6 ears. The middle ear

anomaly was unilateral in 13 operated patients and bilateral in 14. A definite family history was present in 2 patients (2 ears), one of whom had a syndromal diagnosis.

The type A anomaly with a mobile stapes footplate and discontinuity of the ossicular chain between the incus and stapes, was observed in 8 ears. The surgical findings during exploratory tympanotomy and the kind of ossicular reconstruction are shown in Table 1.

The mean gain in hearing level of the type A ears is presented in Table 3. In 6 out of the 8 ears the gain was at least 15 dB. The individual preoperative and postoperative airconduction thresholds are shown in Figures 1 and 2, respectively. A malformed stapes was found in 5 ears.

TABLE 1: Anatomical findings during exploratory tympanotomy in type a anomalies with a mobile stapes.

case	stapes	incus	malleus	reconstruction
1	malformed	dysplastic bony spine	rudimentary	Type III tympanoplasty with allogenous cartilage on malformed stapes
2	missing head	dysplastic long process	normal	allogenous incus transposition
3	aplastic crurae	normal	normal	prothesis on footplate
4	missing head	dysplastic long process	normal	allogenous incus transposition
5	normal	aplastic long process	normal	allogenous incus transposition
6	missing suprastructure	aplastic long process	normal	allogenous incus between footplate and tympanic membrane
7	normal	disconnection incus-stapes	normal	reconnection with bone ship
8	normal	aplastic long process	normal	allogenous malleus interposition

The mean hearing gain in these 5 cases was 19 dB (SD=14); in the 3 ears with a normal stapes the mean hearing gain was 35 dB (SD=4).

The type B congenital ossicular chain anomaly was seen in 19 ears. In 2 out of the 19 ears (Fig. 1; table 2, nos 10 and 17) the fixation of the malleus was due to a bony atretic plate

Fig. 1: Individual preoperative hearing levels for bone and air conduction in all 27 operated ears with a congenital anomaly of the ossicular chain and a mobile footplate (0.5, 1 ,2 kHz dB HL).

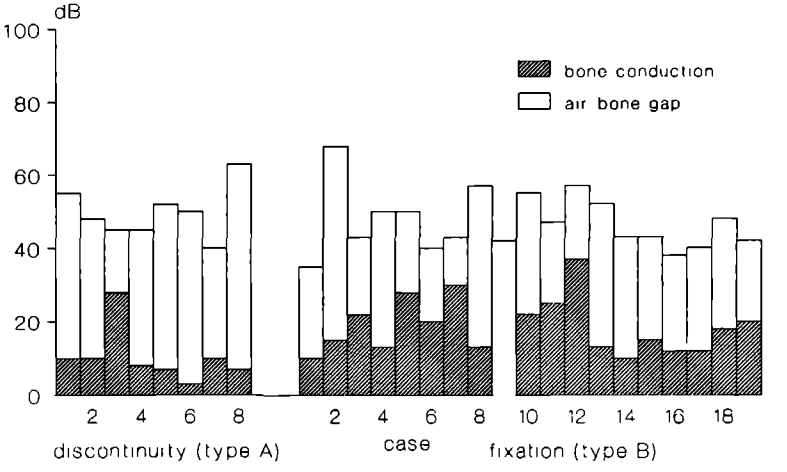

Fig. 2: Individual 1 year postoperative hearing levels for bone and air conduction in all 27 operated ears (0.5, 1, 2 kHz dB HL).

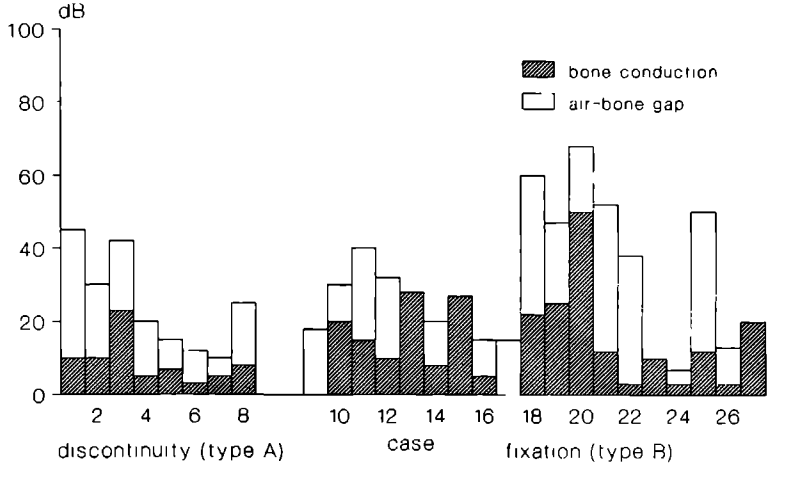

TABLE 2: Anatomical findings during exploratory tympanotomy in type B anomalies with mobile stapes.

case	Stapes	Long process of incus	Malleus	Fixation	Reconstructive surgery
9	malformed	dysplastic	malformed	epitympanic fixated incus	autogenous incus interposition
10	normal	malformed	malformed	atretic plate to malleus	mobilization of malleus by removal of atretic plate
11	normal	slightly malformed	normal	epitympanum	type III tympanoplasty
12	malformed	aplastic	normal	epitympanic fixated incus	allogenous incus interposition
13	normal	normal	normal	epitympanum	removal malleus head, interposition autogenous incus
14	normal	malformed	normal	epitympanum	removal malleus head, interposition autogenous incus
15	normal	normal	normal	fixed malleus	release of malleus handle, autogenous incus interposition
16	normal	dysplastic	normal	epitympanic	autogenous incus interposition
17	normal	almost normal	normal	atretic plate	mobilization by removal atretic plate
18	malformed	normal	normal	due to small epitympanum	allogenous malleus on stapes footplate
19	normal	normal	normal	epitympanic	removal long process of incus, type III tympanoplasty
20	normal	malformed	malformed	epitympanic	mobilization by removal of atretic plate
21	normal	normal	normal	epitympanic	mobilization of malleus handle and allogenous malleus interposition
22	normal	normal	normal	due to small epitympanum	mobilization of malleus and incus complex by epitympanotomy
23	normal	malformed	normal	epitympanic	mobilization of malleus handle by epitympanotomy and autogenous incus interposition
24	normal	normal	normal	epitympanic	mobilization malleus handle, autogenous incus interposition
25	normal	normal	normal	due to small epitympanum	mobilization by epitympanotomy
26	malformed	aplastic	malformed	epitympanic	allogenous malleus head
27	malformed	normal	normal	epitympanic, fused incus/malleus	mobilization by epitympanotomy

in the upper-anterior part of the tympanic membrane, which connected the handle of the malleus to the bony anterior annulus. This finding was classified as a congenital anomaly of the ossicular chain; it can also be considered as a mild form of bony atresia type IIA.[2] In the remaining 17 ears, the malleus and/or incus were fixed in the epitympanic region. In 10 out of the 19 ears, fixation was accompanied by an anomaly in the shape of the malleus and incus, giving in some cases also a disconnection of the ossicular chain. The surgical findings and the results of reconstructive surgery are shown in Table 2. The surgical procedure performed in most of these ears was anterior epitympanotomy, i.e. mobilization of the fixed ossicles.

The mean hearing gain in the 19 type B ears was 17 dB (Table 3). In 12 out of the 19 ears a substantial hearing gain of at least 15 dB was achieved, with a mean hearing gain of 28 dB in the 12 successful ears. The individual preoperative hearing levels and the postoperative hearing levels are shown in Figures 1 and 2, respectively.

Table 3: Hearing results for air-conduction thresholds 1 year postoperatively in 27 congenital middle ear anomalies with a mobile stapes footplate. Between brackets ears with hearing gain > 15 dB (0.5, 1, 2, kHz dB HL; SD = standard deviation)

	n (gain > 15 dB)	preoperative	postoperative	mean gain in all ears and in ears with gain > 15 dB ( )
discontinuity ossicular chain	8 (6)	50 dB SD = 7	25 dB SD = 13	25 dB (31 dB) SD = 13 (8)
epitympanic fixation	19 (12)	47 dB SD = 8	31 dB SD = 18	17 dB (28 dB) SD = 17 (9)
total	27 (18)	48 dB SD = 8	29 dB SD = 17	19 dB (29 dB) SD = 16 (9)

Revision surgery was carried out in 6 ears (Fig. 1, table 1 and 2, nos 5, 9, 11, 17, 21, 26) which resulted in a hearing gain in 4 of them. In 3 ears (nos 5, 9, and 26) a second reconstruction of the ossicular chain was necessary because the previous operation was non-

functional. In case 17 a previously unnoticed atretic bony plate in the upper-anterior section of the tympanic membrane was removed also. Hearing results after revision surgery were used in the hearing gain statistics. There were no cases of temporary or persistent facial nerve paralysis, severe deterioration of the hearing loss or other serious complications in this series.

DISCUSSION

There appears to be a wide variety of congenital anomalies of the ossicular chain. For the sake of clarity, this paper only deals with certain ears from the series, namely those in which a mobile stapes footplate was found, with either ossicular chain discontinuity (type A) or ossicular chain fixation (type B). In the type B anomaly, fixation can be accompanied by discontinuity of the ossicular chain.

Some of the findings on the patients with a syndromal diagnosis in this series have been published separately.[2-7,17,18] However, the majority of congenital anomalies are solitary and have a non-hereditary etiology. A small proportion (19%) of the anomalies were syndromal. This series of operated ears was selected on the basis of the type of anomaly, which also determined the surgical procedure required for the ossicular chain and formed the motive for distinguishing between types A and B. The fact that the stapes footplate was mobile in all the ears meant that stapedectomy was unnecessary.

In the 8 ears with discontinuity of the ossicular chain, reconstruction surgery was performed using autogenous or allogenous ossicles. The state and form of the stapes suprastructure is of importance to the surgeon and may influence the postoperative hearing gain. The hearing gain in the 3 ears with a normal stapes suprastructure was clearly better than in those with malformation of the stapes suprastructure.

In the 19 cases of epitympanic fixation, the problem of reconstruction surgery was much more complex. It was not always possible to expose the fixed incus and malleus sufficiently, owing to the epitympanum being too narrow. This problem was also encountered in the patient with craniometaphyseal dysplasia. In these cases an alternative approach can be employed, for instance, making a direct connection between the stapes and handle of the malleus, that has been detached before from its corpus.

Table 4: Longer series with congenital middle ear anomalies with a mobile stapes in literature.

Author	Year	No out of a total series	Surgical procedures	Ears with gain > 15 dB	Mean gain in ears with gain > 15 dB	Other congenotal middle ear anomalies in series
Ombredanne	1959/ 1968	10 (77)	3 reconstructions 7 mobilizations of ossicular chain	5	23 dB	24 isolated stapes ankylosis
Hough	1958	3 (25)	not specified	not specified		various
Manolidis	1972	6 (12)	5 reconstructions 1 type III tympano-plasty	6	32 dB	various
Funusaka	1979	11 (18)	6 reconstructions (silicon tubes) 2 reconstructions (gelfoam wire) 3 mobilizations of ossicular chain	10	33 dB	6 isolated stapes ankylosis 1 aplasia of oval window
Cousins	1988	36 (56)	21 reconstructions 5 mobilizations (3 with reconstruction) 10 no reconstruction	not specified		1 isolated stapes ankylosis 11 stapes ankylosis and associated ossicular chain anomaly
Present study	1990	27 (104)	19 mobilizations (12 with reconstruction) 8 reconstructions of ossicular chain	18	29 dB	32 isolated stapes ankylosis 32 stapes ankylosis and associated ossicular anomaly 13 aplasia of oval window

In this way the fixed corpus mallei and corpus incudis can remain in situ. This technique was chosen instead of mobilizing epitympanotomy in 5 ears (Fig. 1, nos 5, 19, 21, 23, 24) using an endaural approach; epitympanotomy was not performed in any of the cases. Owing to the small numbers it was not possible to determine which was the most successful technique on the basis of the gain in hearing levels.

In 2 ears with type B anomalies, a bony atretic plate was found in the upper quadrant of the tympanic membrane. This anomaly may be regarded as a mild variation of congenital meatal atresia and therefore fades the difference between major and minor anomalies of the ear.

If the air conduction loss remains too large after surgery, we feel that it is worthwhile to consider a second operation. In our series, 6 patients underwent reoperation one or more times. In 4 of them it was still possible to achieve or restore a gain in the hearing level.

We compared our results to those from large series in the literature (Table 4). Although the success rate varied to some degree, the mean hearing gain in successfully operated ears was

comparable (Table 4: mean gain in ears with at least 15 dB hearing gain). Large series of operated congenital middle ear anomalies are rare. Often the series reported in the literature are intermingled with results and data on congenital external auditory meatus atresia. However, Table 4 only shows data on congenital middle ear anomalies; particular attention is paid to anomalies in which the stapes footplate was mobile.

The present classification system applied to our series has a particularly strong surgical background and is mainly based on whether or not the stapes footplate is fixed. Moreover, further differentiation of the classification appeared to be possible. In anomalies with a mobile stapes, such as those described in this paper, the essential surgical consequence is that stapedectomy is superfluous in these cases. Nevertheless, in this series of ears with a wide range of anomalies, the necessary surgical reconstruction techniques were complex and each individual case differed greatly from the next with regard to the final results.

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CHAPTER II-4

CLASSIFICATION OF MINOR CONGENITAL EAR ANOMALIES. A REPORT ON 144 EARS

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Annals of Otorhinolaryngology 1992 (in press).

ABSTRACT

The surgical findings in 144 successive ears operated on for congenital conductive hearing loss were analysed and the results were evaluated in terms of hearing gain. All the patients underwent middle ear surgery at the University Hospital Nijmegen between 1964 and 1990. A classification system was developed with four main categories to analyse the findings and results in a more easily surveyable manner than formerly.

Class I comprises ears with congenital isolated stapes ankylosis; Class II comprises ears with congenital stapes ankylosis in combination with a congenital anomaly of the ossicular chain; Class III comprises congenital anomalies of the ossicular chain and at least a mobile stapes footplate; Class IV comprises ears with aplasia or severe dysplasia of the oval or round windows.

INTRODUCTION

There are many different types of congenital middle ear anomalies. In one quarter of the cases, the congenital hearing loss forms part of a syndrome. The great diversity of anomalies of the middle ear which are described in the literature makes it difficult to compare and analyse them without prior classification. Various classification systems have been proposed or suggested in the literature, but in our opinion they leave much to be desired. Therefore we developed a new classification and employed it in the presentation of the findings and results of classes I, II and III of the series of ears operated on up to 1986. The application of this classification to the series published by other authors allows reliable comparison of the results. The classification system presented in Table 1 is based on the different surgical reconstructive techniques available for the anomalies encountered in patients. For example, it makes a difference whether or not reconstructive stapes surgery was performed because the vestibulum was opened. In addition, stapes replacement surgery is more difficult if the incus-malleus complex is malformed. Consequently, a separate group of anomalies is distinguished in which the incus-malleus complex forms the only cause of hearing impairment. Another separate group comprises ears with aplasia of the oval or round windows, because creating a neo-oval window involves more risk to the inner ear and it also requires a different surgical procedure.

TABLE 1. Classification of minor congenital anomalies of the ear.

Class	Main anomaly	Subclassification	No. of ears
I	Congenital stapes ankylosis		44
II	Stapes ankylosis associated with another congenital ossicular chain anomaly		55
III	Congenital anomaly of the ossicular chain but a mobile stapes footplate	A. Discontinuity in ossicular chain	22
		B. Epitympanic fixation	9
IV	Congenital aplasia or severe dysplasia of the oval or round window	Aplasia	10
		Dysplasia	
		Crossing facial nerve	3
		Persistant stapedial artery	1

The objective of this classification is to provide more insight into the frequency with which these isolated or associated congenital middle ear anomalies occur with other anomalies and subsequently to analyse the various surgical techniques. The results of such analyses will provide guidelines for surgical indications, the required technique or the need to develop new techniques.

In an earlier publication we presented the surgical findings and results of the ears from this series which were operated on in the period up to 1986 because of congenital stapes ankylosis or a congenital anomaly of the ossicular chain and a mobile stapes footplate.[1,2,3] An analysis of the impact of a syndromal diagnosis has already been published.[4] Between 1986 and 1990, 40 operated ears in 31 patients were added to the series.

PATIENTS AND METHODS

From 1964 to 1990, 144 consecutive ears from 117 patients were operated on because of a congenital middle ear anomaly.

The study comprised a retrospective analysis of the surgical findings in the period up to 1986 and a prospective part in which the data were incorporated directly into the ongoing study. All the cases had a typical history of conductive hearing loss since early childhood. The

following inclusion and exclusion criteria were gradually employed:

1. The patient must be at least 10 years old at the time of the operation.
2. In children, intermittent periods of otitis media with effusion must be excluded by a sufficiently long follow-up.
3. The results of tone audiometry, speech audiometry and contralateral stapes reflexes must be available.
4. Preoperative CT scans must be available of the ossa petrosa, particularly in patients with mixed hearing loss.

Over the past 10 years, we have been paying extra attention to a syndromal diagnosis to evaluate whether such a special diagnosis has any diagnostic or therapeutic impact in relation to ear surgery.

The preoperative and postoperative hearing levels were calculated separately and the mean levels are presented for the frequencies 0.5, 1 and 2 kHz. Pure tone audiometry was performed routinely 0.5, 1 and 2 years after surgery in all the patients and in most cases also once a year for many years thereafter. The hearing levels measured one year after surgery were used for statistical analysis of the hearing gain. The air conduction and bone conduction levels were used for the presentation of the results of individual cases and the air-bone gap levels for the mean hearing gain results. Hearing gain calculations were based on individual values. Preoperative bone conduction levels were not changed in the calculations of cases whose postoperative bone conduction levels improved. Speech audiometry was performed preoperatively in all the patients and, in most cases, also postoperatively.

RESULTS

In this study, the data from 144 operated ears were classified into four groups. The number of ears per group is shown in Table 1.

Preoperative hearing levels and the levels measured at one-year follow-up are presented in Figures 1, 2a, 2b, 3 and 4 in chronological order. The individual results are shown separately for the four groups. The figures present the results of the ears admitted to the study since 1986. The individual findings and results of the ears in classes I, II and III operated on prior to 1986 have been reported in earlier publications.[1,2,3] The preoperative hearing levels of the

ears in class IV are presented for the whole period from 1964 to 1990 (Fig. 4), because the results of these ears with aplasia of the oval window, operated on prior to 1986, have not been published before. In four out of the 14 ears in class IV (Fig. 4: cases 1, 6, 12 and 14), an attempt was made to improve the patients' hearing; these operations were conducted in 1965, 1975, 1983 and 1989, respectively. The postoperative results at one-year follow-up are also presented. The number of ears per class and the percentages of air-bone gap closure are shown in Table 2.

Fig. 1. Preoperative and 1 year postoperative individual hearing levels for bone and air conduction in 12 operated class I ears between 1986 and 1990.

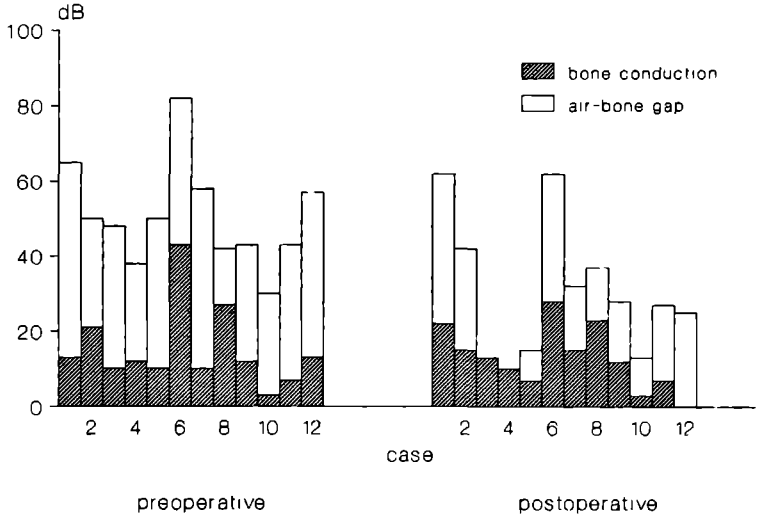


Table 2: Air - bone gap 12 months after reconstructive surgery in 127 minor congenital ear anomalies.

Classification	No. of ears		Air-Bone Gap < 10 dB (%)	Air-Bone Gap 10-20 dB (%)	Air-Bone Gap > 20 dB (%)
	per class	reconstructive surgery			
Class I	44	41	20 (49%)	9 (22%)	12 (29%)
Class II	55	51	20 (39%)	17 (33%)	14 (28%)
Class III	31	31	9 (29%)	14 (45%)	8 (26%)
Class IV	14	4	1 (25%)	2 (50%)	1 (25%)

Fig. 2a. Preoperative individual hearing levels for air and bone conduction in 23 operated class II ears between 1986 and 1990.

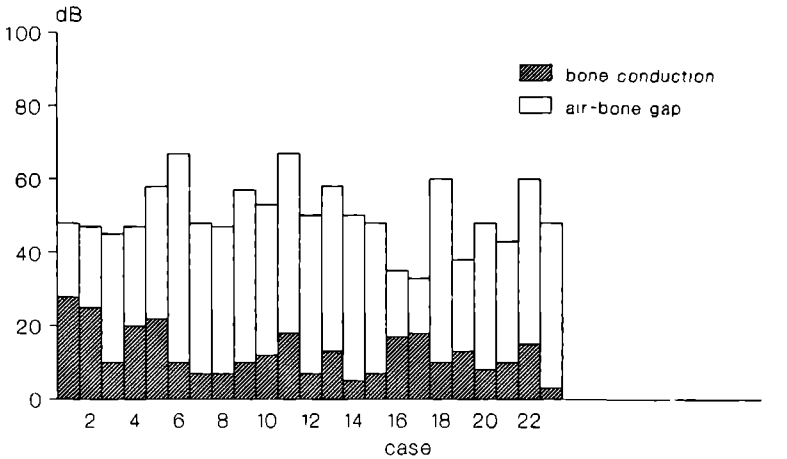


Fig. 2b. One year postoperative individual hearing levels for air and bone conduction in 23 operated class II ears between 1986 and 1990.

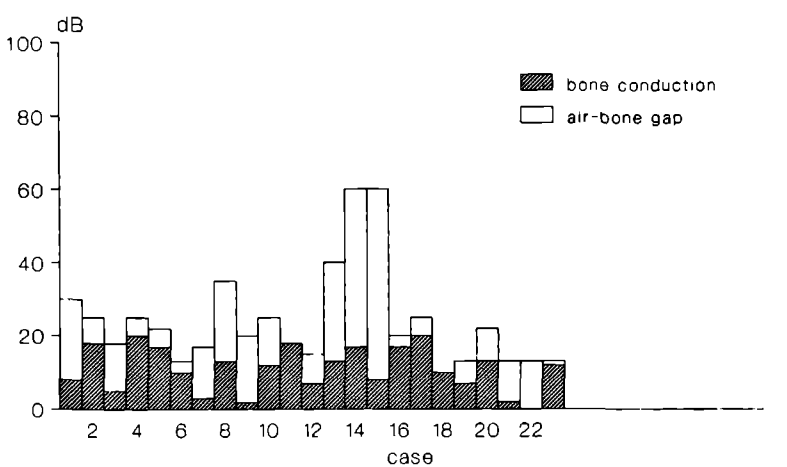


Fig. 3. Preoperative and 1 year postoperative individual hearing levels for bone and air conduction in 4 operated class III ears between 1986 and 1990.

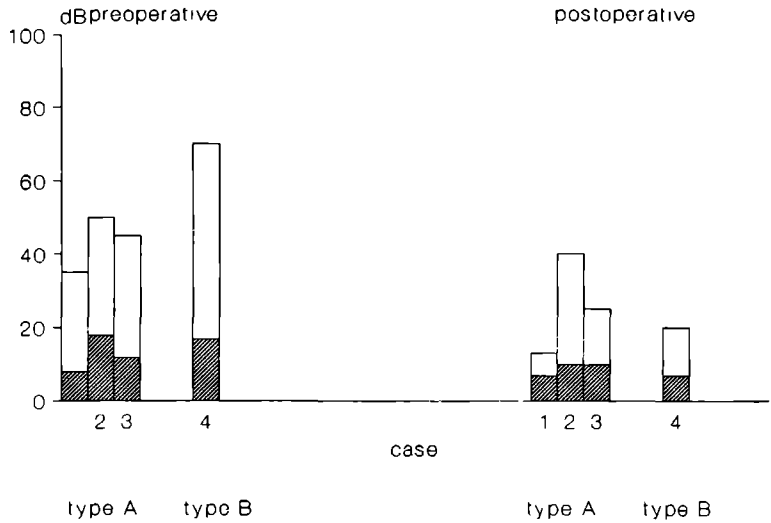
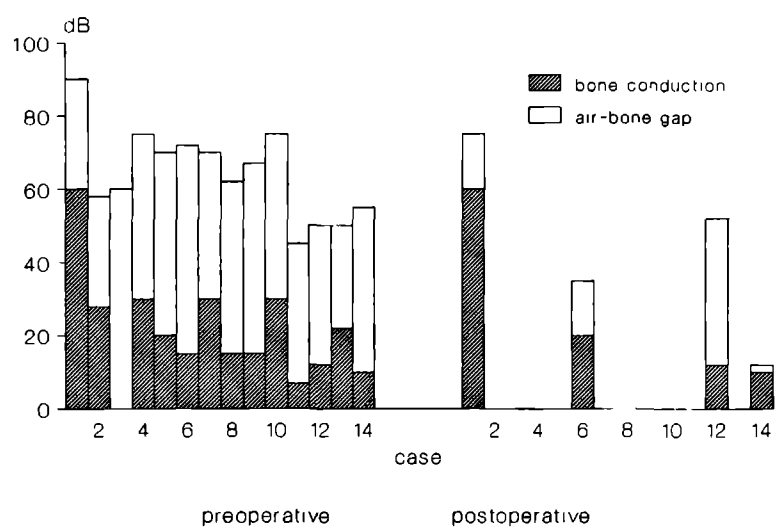


Fig. 4. Preoperative and 1 year postoperative individual hearing levels for bone and air conduction in 14 operated class IV ears including 4 ears with a neo-oval window between 1964 and 1990.



Stapes surgery was performed in 88 out of the 99 ears in class I and II. A different type of stapes surgery was necessary to mobilize the stapes in four other ears, two with a bony stapedial ligament[6] and two patients with the Treacher Collins syndrome in whom the stapedial head was fixed to the bony facial canal. In seven ears, no reconstructive surgery was performed because of a stapes gusher[7] or an aberrant facial nerve.[8] In 26 out of the 88 stapedectomies, stapes surgery was combined with additional surgical treatment for the associated congenital anomaly of the ossicular chain. All 26 ears were classified as class II. In some of the other operated ears in class II stapes surgery was complicated by form anomalies during insertion of the replacement stapes prosthesis or even by the absence of the incus.

If the long process of the incus is too short, it may be necessary to place a malleus footplate prosthesis. A recently developed technique which is only performed in exceptional cases is myringo-chorda-vestibulopexy. In this technique the prosthesis is fixed to the chorda tympani by interposing cartilage between the tympanic membrane and the chorda.[5]

In class IV, four neo-oval windows were created. In three cases teflon or teflon-wire was interposed between the incus and the neo-oval window which had been opened in the promontory[9] (Fig. 4: cases 1, 6 and 12) and in one case myringo-chorda-vestibulopexy was performed[5] (Fig. 4: case 14).

When the results of stapes surgery in the 12 ears in class I were analysed, it was striking that there was no improvement in one ear (no. 1) and that the postoperative threshold remained poorer than 35 dB in four ears and poorer than 30 dB in five ears. The postoperative bone conduction thresholds in three ears (Fig. 1: nos 1, 3 and 7) were less favourable after the operation, ranging from a few dB Fletcher index to 10 dB.

Comparing the results of stapes surgery in class II in Fig. 2a and Fig. 2b showed that there was no hearing gain in two ears (nos 14 and 15). A considerable hearing gain was achieved in all of the 21 other ears. In two ears (nos 22 and 23) there was only very slight improvement in the perceptive component of 10 dB or less postoperatively.

Only four ears were added to class III after 1986 and the hearing of all of them improved. In one ear (Fig. 3 no. 2) the hearing gain remained limited, with a Fletcher index of more than 40 dB. There was no increase in the perceptive threshold postoperatively.

In class IV we confirmed the current view in some of the reports in the literature that creating a neo-oval window is associated with a greater risk of inner ear hearing loss. Fairly recently, a shift has occurred in these limitations and the creation of an neo-oval window can once again be considered as a form of ossicular chain reconstruction.
In 1965 an attempt was made to create a neo-oval window in one case (no. 1), without any notable success and no other attempts were made until 1975 (no. 6). The hearing threshold improved from 70 dB to 35 dB and the perceptive threshold remained about the same. In case no. 14 (1989) of this series, the good result is evident.

DISCUSSION

This series of 144 operated middle ear anomalies showed great variation in the nature and severity of congenital anomalies. Therefore, diverse surgical procedures were applied to improve the patients' hearing. To achieve a more easily surveyable presentation of the findings and results, we sought a practical classification system. In the literature, various authors have proposed a classification based on their own operated cases (Table 3).
Within this great variation, we distinguished four main classes. Firstly, this classification preselects the anomalies on the basis of the condition and mobility of the stapes and the stapes footplate. Classes III and IV are subdivided further.
The clinical findings and results of ear surgery have been published separately for three out of the four main classes.[1,2,3] The most important reason for this is to achieve a surveyable presentation and description of the great diversity of congenital middle ear anomalies and at the same time to present the results in relation to ear surgery for each separate ear and for each class. This made it much easier to compare the results of minor congenital ear anomalies and also the various types of reconstructive surgery of the ossicular chain.

Class I comprises ears with isolated congenital stapes ankylosis and class II the operated ears with congenital stapes ankylosis and an associated congenital anomaly of the rest of the ossicular chain.
This group of anomalies has the same embryological basis. The earlier induced thinning of the bony division between the otic capsule and the stapes, develops into the stapes footplate. Subsequently, further de-ossification occurs of the annular ring around the stapes and it

Table 3: Earlier classifications proposed for minor congenital ear anomalies in literature.

Henner[15]: Class I: minor ear anomalies
Classes II and III: major ear anomalies

Class I:
1. ossified stapedial tendon
2. ossified ligament between the incus and epitympanum
3. synostose incus-malleus joint
4. discontinuity of the ossicular chain
5. agenesis of the long process of the incus
6. agenesis of the stapes crurae
7. agenesis of the annular ligament of the stapes footplate (= stapesankylosis)
8. agenesis of the stapes and footplate (= aplasia of the oval window)

House[12]:
1. congenital stapes ankylosis
2. congenital ossicular chain discontinuity
3. congenital fixed malleus syndrome
4 congenital fixed incus syndrome
5. congenital ossified stapedial tendon.

Arslan and Giacomelli[13]:
A. normal
B long process of the incus absent
C. aplasia of the oval window and footplate
D. aplasia of the oval window, footplate and stapes
E. aplasia of the oval window, footplate, stapes and fixation of the incus-malleus joint

Ombredanne[17]:
1. Congenital ossicular chain discontinuity
2. Congenital ossicular chain ankylosis
3. Congenital aplasia of the oval window and stapes

becomes mobile. Disruption of this process during the development of the ear leads to this anomaly: the footplate is often found to be thickened and fixed to the annular ring.[10]

Stapes ankylosis has also been accepted as a separate entity by other authors. Several reports have appeared in which the results of operative treatment for stapes ankylosis were published separately.[11-14] However, none of the previous authors have distinguished between patients with isolated stapes ankylosis and patients with stapes ankylosis with an associated congenital ossicular chain anomaly.

The surgery of choice for isolated stapes ankylosis is regular stapedectomy, in which the prosthesis is attached to the mobile incus. It was striking that the mean postoperative hearing gain in the patients with a deformed stapes suprastructure and isolated stapes ankylosis was

11 dB lower than that in the patients with a normal suprastructure.[1] We could not find any explanation for this.

Class I comprises two additional special forms of isolated stapes fixation other than at the level of the stapes footplate itself: fixation of the stapedial head to the bony facial canal and fixation of the stapes owing to a bony stapedial ligament. In both these situations, there were no other anomalies of the ossicular chain. The anomaly in which the stapedial head was fixed to the bony facial canal was only encountered in patients with the Treacher Collins syndrome. The application of laser surgery to such cases in the near future may achieve greater hearing gain.

Class II comprises operated ears with congenital ankylosis and an associated anomaly of the incus or malleus. It is sometimes difficult to recognize this anomaly during surgery. For instance, fixation of the short process of the incus in the fossa incudis can easily be overlooked. Such ears are then wrongly classified as class I ears, because the associated anomaly was missed by the surgeon, and will not number among the successfully operated ears in class I.
Surgical treatment of this associated anomaly is essential to achieve the required level of hearing gain. Fixing the prosthesis during malleo-vestibulopexy will lead to problems in a number of cases particularly when the incus and malleus handle are completely absent. Moreover, fixation of the incus and/or malleus will have to be dispelled first, which can take place in the same session. Various modifications have been made to the malleo-vestibulopexy technique over the past ten years, including the availability of a 0.4 mm teflon-platinum piston and an alternative method for fixing the malleus.

Class III is characterized by a mobile stapes footplate. The congenital anomaly in these cases lies in a deformed stapes, incus or malleus suprastructure, which causes discontinuity of the ossicular chain (type III A). Anomalies involving fixation of the incus or malleus but a mobile stapes are also classified as class III (type III B). These anomalies are considered to be variations of the developmental disorders of the first and second branchial arches.
Very few published series are available in the literature which are comparable to this class III.[15] From an otological point of view, the surgical treatment for this anomaly is

fundamentally different from that for classes I and II, because the vestibulum is not opened. Reconstruction of the ossicular chain can be achieved in various ways, whether or not in combination with mobilization of another part of the ossicular chain. There is so much variation between the anomalies and the required surgical techniques that we decided not to make any further subclassifications. An anomaly in which fixation of the ossicular chain is caused by a bony plate in the anterior upper quadrant of the tympanic membrane between the malleus handle and the bony anterior annulus, is included in class III. This anomaly can also be considered as a mild form of congenital aural atresia. It therefore occupies the transitional area between the major and minor congenital ear anomalies.

Class IV comprises aplasia of the oval and/or round windows. This anomaly forms such a clear clinical entity that it was desirable to create a separate group for its classification.
There is no clear embryological explanation for this anomaly. It has been suggested that during normal development, the bony division between the inner and middle ears gradually becomes thinner starting at the otic capsule at the level where the stapes develops in the middle ear.[10] At the same time, the stapes takes on its definitive form. It is probable that the developing stapes induces the thinning of the bony division which later becomes the stapes footplate. In the same way, the otic capsule will influence the development of the stapes. If this complex mechanism is disrupted during the course of development, various congenital anomalies of the oval window and stapes may arise. The most extreme form, total aplasia of the oval window, is often seen in association with aplasia or severe deformity of the stapes, in which the stapedial processes, if present, hardly ever reach the region of the footplate. This observation supports the above-described embryological development. However, other authors still regard aplasia as a separate entity.[16,17] So far, there is no definite answer to the question of which surgical technique is the best treatment for aplasia of the oval window. The greatest drawback is that opening the vestibulum can lead to considerable damage to the inner ear function. Ombrédanne usually performed fenestration of the semicircular canal.[17] Later on an interpositioning procedure was applied, in which the promontory was opened.
In our own series, there were 14 cases of aplasia of the oval window. A neo-oval window was created in four of them with teflon interpositioning, the first in 1965, two of which were successful.[5,9]
Class IV also comprises a number of anomalies which are denoted as serious dysplasia of the

oval window. In these anomalies, the oval window is sometimes completely obscured by the overlying facial nerve or a persistent stapedial artery. Successful neo-oval window procedures were described by Sterkers and Sterkers in six ears with aplasia of the oval window and an aberrant facial nerve.[18,19] The neo-oval window was not created on the promontorial side, but on the cranial side of the facial nerve.

In about one in every four operated ears, the congenital middle ear anomaly is not isolated, but forms part of a syndromal diagnosis.[4] It is also useful for the ear surgeon to be aware of the syndromal diagnosis. For instance, at present it is possible to diagnose the X recessive syndrome with stapes gusher preoperatively and therefore not perform surgery.[20] In the case of the branchiogenic syndrome, the surgeon will anticipate more complex and more severe anomalies of the middle ear and inner ear. The diagnosis craniosynostosis implies that the epitympanum may be too small and stapes surgery may have to take the form of malleo-vestibulopexy.[4]

Our series of 144 consecutive congenital middle ear anomalies is striking owing to its size and the proposed classification system. We have arranged our data in such a way that for each ear the preoperative and postoperative hearing thresholds for air and bone conduction are presented, partly in earlier publications on the surgical results of the subclasses,[1,2,3] which makes it possible to evaluate the results in relation to the actual middle ear anomaly and the surgical technique applied.

A survey of the results of the whole series showed that the most disappointing results occurred at the beginning of the series in the mid 1960s, in ears with major mixed hearing losses. It appears that in later years, the level of the sensorineural part in the hearing loss formed an unobtrusive selection criterion. Another interesting finding is that nearly all the ears had a sensorineural hearing loss of 10 to 20 dB. In the operative evaluation of such patients, the surgeon may feel inclined to consider this component as an artefact of the bone conduction threshold measurement, while this is not the case. The bone conduction thresholds of patients with a minor congenital abnormality are seldom normal, in contrast with those of patients with a major congenital ear anomaly. It is important to realize this when predicting the degree of success of an operation. Our results have shown that the hearing gain achieved, even in cases with a unilateral major or minor congenital anomaly, is also highly beneficial in terms of tone and speech recognition.[21]

The total results demonstrate unmistakably that ear surgery is a worthwhile treatment for minor congenital ear anomalies, particularly when intervention is limited to classes I, II and III and the selection criteria are strictly adhered to. In a few specific problematical cases requiring ossicular chain reconstruction or malleo-vestibulopexy, because of a minor congenital ear anomaly, such as aplasia of the oval window and/or round window, it is not yet clear what the best treatment is and if surgery is indicated, which surgical technique is the most appropriate. Independent of this, the cogency of our results would be enhanced in the future by similar studies from other centres.

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CHAPTER II-5

A CLASSIFICATION OF MINOR CONGENITAL EAR ANOMALIES AND SHORT AND LONG TERM RESULTS OF SURGERY IN 104 EARS

C.W.R.J. Cremers, E. Teunissen
In: Long-Term Results and Indications in Otology and Otoneurosurgery, pp. 11-12
Proceedings of the Politzer Society Conferences in Ibiza, 1989 and Courchevel, 1990
edited by R. Charachon and E. Garcia-Ibanez
1991 Kugler Publications, Amsterdam/Milano/New York.
and in: Polisch Journal of Otolaryngology 1991; suppl: 116-118.

INTRODUCTION

During the 1989 september meeting of the Politzer society we presented the short term results for surgery in 64 ears with a congenital stapes ankylosis. These 64 ears were part of a total of 104 ears with a minor congenital ear anomaly, that were operated from 1964 to 1986 in the Nijmegen Institute of Otorhinolaryngology. In this paper we will present the surgical findings and the short and long term results of all 104 operated ears with a congenital middle ear anomaly.

PATIENTS AND METHODS

Findings and results are based on a retrospective analysis of surgical reports and audiometrical investigations.

RESULTS

In the proposed classification there proved to be a large variability in congenital middle ear anomalies. Therefore the following classification for congenital middle ear anomalies is suggested, which is based on the findings at 104 consecutive ear operations of this study.
The first class is an isolated stapes ankylosis, and the second class is a stapes ankylosis in association with another congenital anomaly of the ossicular chain. The ears with a stapes ankylosis are separated into ears with an isolated stapes ankylosis and an associated anomaly of the ossicular chain. The ears with an isolated stapes ankylosis are then further differentiated depending on the malformations of the stapes suprastructure.
The third class of congenital middle ear anomalies are the anomalies with a mobile stapes. The mobile stapes may be normal shaped or congenitally malformed.
The hearing loss however in this type of anomaly is caused by a congenital anomaly somewhere in the ossicular chain, which can be either the result of a congenital fixation or a congenital discontinuity, or both.

The hearing loss in the 13 remaining ears, the fourth class, proved to be the result of an aplasia of the oval window, discovered at exploratory tympanotomy in these ears.

In ears with a stapes ankylosis, a stapedectomy, if possible was carried out. A stapedectomy was performed in 53 out of 64 ears with a stapes ankylosis. In 4 ears another kind of stapes surgery could be performed, while in the remaining 7 ears no attempt for hearing improvement was carried out for various reasons.

In ears with a congenital anomaly of the middle ear with a mobile stapes 2 sub-categories of anomalies are recognized.

A discontinuity in the ossicular chain as a result of a congenital anomaly of the shape of the ossicles was discovered in 8 ears of this subcategory. The shape of the stapes might also be somewhat malformed but there was a normal mobility.

The other sub-category counted 19 ears with a mobile stapes but with a congenital fixation of the incus and malleus, often together with a congenital malformation of there shape.

In these anomalies with a mobile stapes the surgical procedure was to reconstruct the ossicular continuity and to mobilize the fixated ossicular chain, in the epitympanum and in the fossa incudis.

The fourth category counts 13 ears with a congenital aplasia of the oval window, discovered at exploratory tympanotomy. In 3 ears an attempt for reconstructive surgery was carried out, with some success in one. In the 2 other ears no postoperative alteration of the hearing level was seen. In the remaining 10 ears no fenestration of the promontory was performed since severe deterioration of the hearing loss was feared.

SHORT AND LONG TERM FOLLOW-UP

The short term results, that is to say, the results after 1 year, and the long term results for a postoperative follow-up varying from 2 to 22 years are for the group with a congenital stapes ankylosis presented in Table 1 and for the group with a congenital ossicular chain anomaly in Table 2.

In ears with a congenital stapes ankylosis the average hearing gain after 1 year follow-up was in isolated stapes ankylosis 27 dB and in ears with a stapes ankylosis and an associated anomaly of malleus and incus 23 dB (Table 1). The hearing results after a long term follow-up do not significantly differ from the short term results.

Table 1: Congenital stapes ankylosis

	Short-term 1 year	Long-term average 6 years
Isolated stapes ankylosis		
25 ears in calculations	27 dB	28 dB
Stapes ankylosis with an associated ossicular chain anomaly		
28 ears in calculations	23 dB	21 dB

Table 2: Congenital ossicular chain anomaly and mobile stapes

	Short-term 1 year	Long-term 5 to 6 years
Hearing gain	21 dB	19 dB

In ears with a congenital anomaly of the malleus and or incus and a mobile stapes the short and long term results are a mean hearing gain of 21 and 19 dB respectively (Table 2).

CONCLUSION

In conclusion we investigated the findings and results of over 100 consecutive operations for congenital middle ear anomalies. For clarifying the diversity in the many different congenital middle ear anomalies a classification in distinct types of anomalies is suggested. Knowledge about the frequency of the different anomalies is considered as useful for the surgeon exploring these ears.

The results were satisfying for the 1 years postoperative results. Long term results were available for many of the operated ears and the average hearing gain in these ears does not significantly differ from the 1 year results.

CHAPTER III

MAJOR EAR ANOMALIES

CHAPTER III-1

CLASSIFICATION OF CONGENITAL AURAL ATRESIA AND RESULTS OF RECONSTRUCTIVE SURGERY

C.W.R.J. Cremers, E. Teunissen, E.H.M.A. Marres
Adv. Oto-Rhino-Laryngology 1988; 40: 9-14

Introduction

Congenital aural atresia varies from a mild abnormality with narrowing of the external auditory canal and hypoplasia of the tympanic membrane and middle ear space to the entire absence of the middle ear cleft in conjunction with anotia, bony atresia of the external canal and even hypoplasia of inner ear structures.

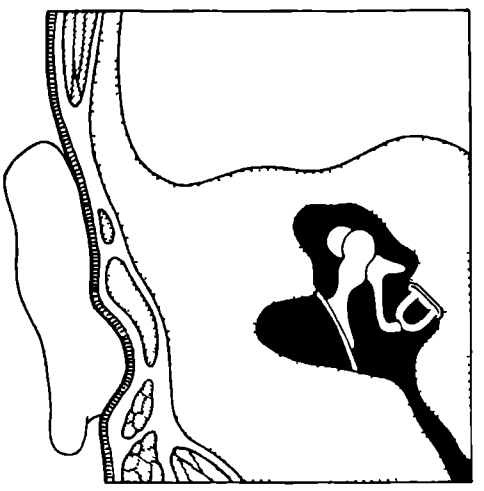
In the early days of surgery for congenital aural atresia, failure to improve hearing, stenosis of the newly formed canal, facial nerve paralysis, discharging cavities and cosmetic disadvantages limited the rare good results. Improved microsurgical techniques, elucidation of middle ear and inner ear dynamics, understanding of tympanoplastic reconstruction and the introduction of polytomographic roentgenographic techniques and nowadays computerized axial tomography for identification of middle ear and inner ear structures are essential in the improvement of surgical results.

Altmann[1] was the first to propose an anatomical classification according to the severity of the atresia and Nager[2] and Schuknecht[3] considered his classification helpful from the surgical point of view. Nevertheless, the second class of the three classes proposed by Altmann represented the vast majority of cases. Nager[2] proposed an additional classification of the developmental anomalies of the auricle, since severe anomalies of the auricle were thought to be related to the degree of middle ear anomalies.

Additional classification of type II congenital aural atresia

The type I aural atresia is presented in figure 1. The additional classification of atresia we proposed concerns type II which has to be divided in a class IIA (fig. 2, 3) and type IIB (fig.4). In type IIB there is a total bony stenosis over the full length of the meatus, while in type IIA (fig. 2, 3) there is a total bony stenosis over only a part of the lenght of the meatus or the canal is partially aplastic and ends blindly with a fistula tract sometimes leading to a rudimentary tympanic membrane.

In type III aural atresia the auricle is usually severely malformed or completely absent. The external auditory canal is absent and the tympanic cavity is either very small or missing. The ossicles are rudimentary or missing and the mastoid is not pneumatized. There are frequently associated anomalies of the inner ear and severe cranial malformations. These cases are to



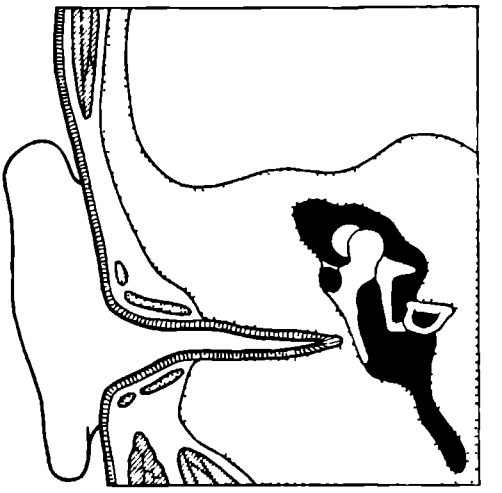
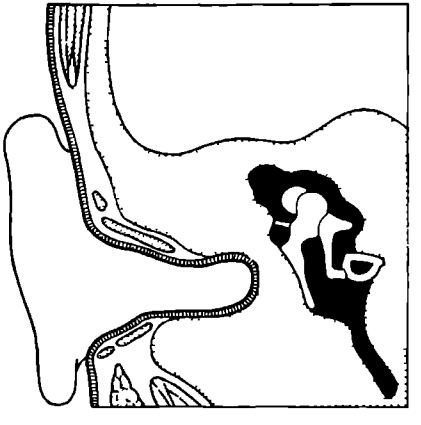


Fig. 1: Type I congenital aural atresia (according to Altmann[1]), with an intact tympanic membrane and bony atresia of the lateral part of the external auditory canal.

Fig. 2: Type IIA congenital aural atresia with a fistula tract ending blindly.



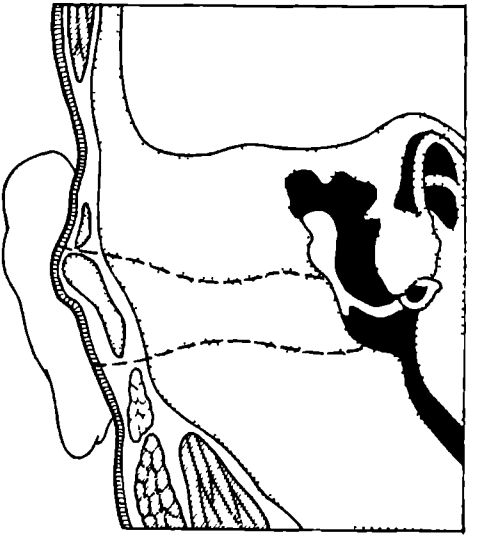


Fig. 3: Type IIA congenital aural atresia with a total bony atresia over only a part of the canal.

Fig. 4: Type IIB congenital aural atresia.

be excluded from surgery of the middle ear.
Sometimes a canal plasty procedure can be performed for rehabilitation with a hearing aid in the rare case the auricle is so well developed that a hearing aid can be adapted. This additional subclassification for congenital aural atresia type II has been published previously[4-7].

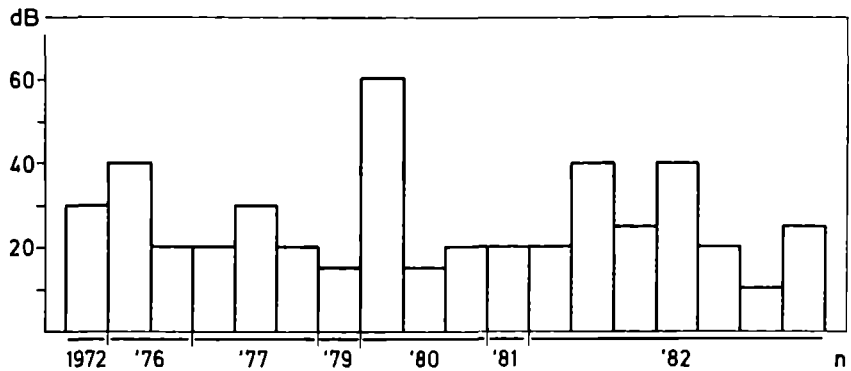
General management and preoperative evaluation

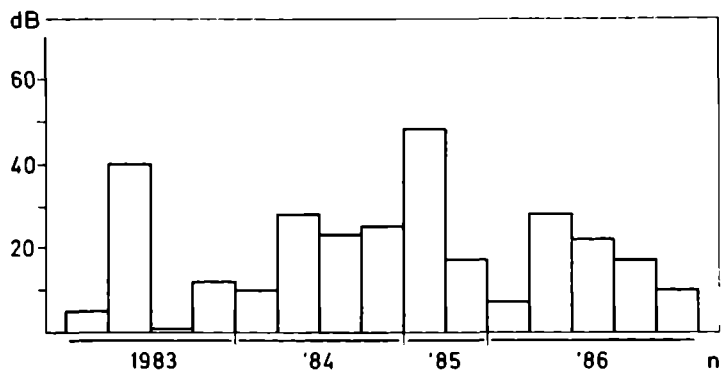
The main purpose of surgery is to give the patient serviceable hearing, corresponding to an average hearing threshold level below 30-35 dB ISO for 0.5 and 2 kHz. In cases of unilateral atresia it can result in binaural hearing, with the best results when the hearing level approaches that of the other ear.
In our opinion prerequisites for surgery are: (1) normal cochlear function in the ear to be operated upon; (2) polytomography or CT scan to show an ossicular chain and a clearly well-developed middle ear cavity; (3) some pneumatization of the mastoid, and (4) in unilateral cases a cosmetic appearance of the auricle that can be accepted as a good end result with or without cosmetic surgery.

Surgical technique, patients and results

Nowadays fenestration of the semicircular canal has been abandoned as method of choice for surgery of congenital aural atresia. Currently used methods are the type III tympanoplasty[3], the canal plasty[4] and the canal plasty with allograft tympanoplasty. The canal plasty procedure we use has been described previously[5]. The results obtained in the first 36 ears with a type II atresia over the period 1972-1983 have been reported.[4-6]
Of 55 ears of 48 patients with a type I or II anomaly, operated upon in the Nijmegen Department of Otorhinolaryngology over the period 1972-1986, 2 ears had a type I anomaly, 33 a type IIA and 20 a type IIB anomaly. The average preoperative threshold for the speech frequencies 0.5, 1 and 2 was kHz 44 dB (range 38-50 dB) in group I, 47 dB (range 25-77 dB) in group IIA, and 58 dB (range 30-70 dB) in group IIB atresia. The postoperative average threshold was 21 dB (range 20-22 dB) in group I, 26 dB (range 10-60 dB) in group IIA, and 38 dB (range 15-55 dB) in group IIB.

Detailed postoperative results are presented separately for class IIA (fig. 5, 6) and class IIB (fig. 7). The average hearing gain was 21 dB for group IIA and 20 dB for group IIB.





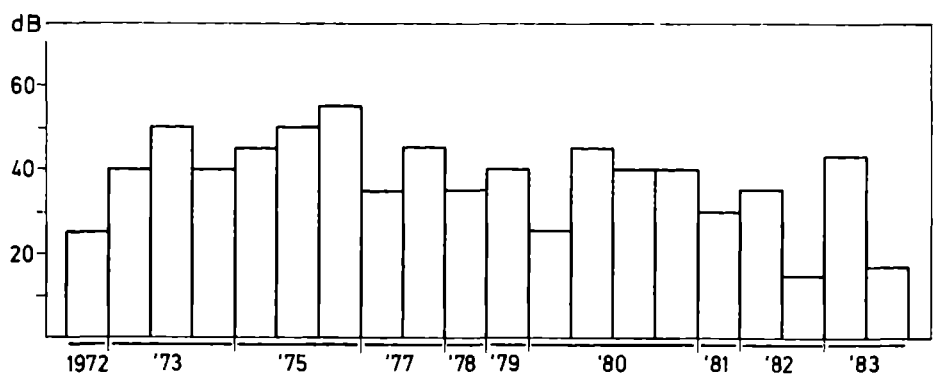


Fig. 5: Postoperative air-bone gap in 18 ears with a type IIA congenital aural atresia over the period 1972-1982.

Fig. 6: Postoperative air-bone gap in 15 ears with a type IIA congenital aural atresia over the period 1983-1986.

Fig. 7: Postoperative air-bone gap in 20 ears with a type IIB congenital aural atresia over the period 1972-1986.

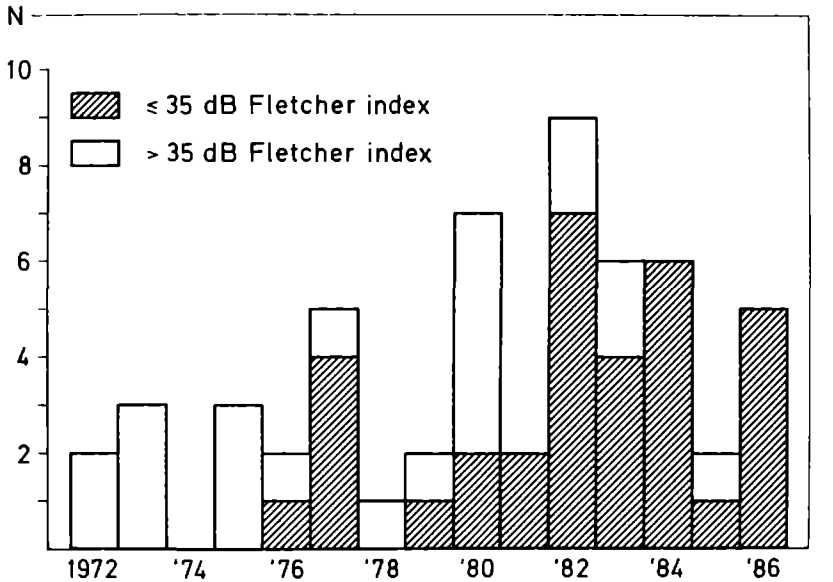


Fig. 8: Postoperative hearing loss in 55 ears with a type I, IIA or IIB congenital aural atresia, since 1972.

An overview of the ears with a type I, IIA and IIB classification operated on from 1972-1986 with a postoperative hearing loss of 35 dB or less or larger then 35 dB Fletcher Index is shown in figure 8.

Postoperative complications of this surgery can be sensorineural deafness, facial paralysis, perforation of the tympanic membrane, restenosis and otorrhea. None of these patients had a sensorineural hearing loss or facial paralysis following surgery.

In some patients with bilateral atresia a sensorineural component was only recognized in the audiogram after previous surgery elsewhere and our end results were therefore restricted. A perforation of the tympanic membrane occurred only twice. A total restenosis was seen in 4 patients and in 9 treatment was given for long-standing otorrhea, leading to dry ears in 8 of them. In the last 3 years we have not had any of these side effects.

Comment

Additional classification and strict criteria for surgery, such as age and the absence of a

sensorineural component in the hearing loss are required to select the successful cases preoperatively. This improved our rate of success to such an extent that a small group of patients with unilateral atresia has also had benefit from surgery in recent years. Since the group of unilateral atresia is 4-5 times as frequent as the bilateral group, this has opened possibilities for a large number of patients with unilateral aural atresia, type IIA. Possibly, this can also be expanded to type IIB unilateral atresia since the surgical experience may improve yet further.

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CHAPTER III-2

LONG-TERM RESULTS OF SURGERY FOR UNILATERAL AND BILATERAL CONGENITAL AURAL ATRESIA

C.W.R.J. Cremers, E. Teunissen
In: Long-Term results and Indications in Otology and Otoneurosurgery, pp. 13-15
Proceedings of the Politzer society conferences in Ibiza 1989 and Courchevel 1990
edited by R. Charachon and E. Garcia-Ibanez
1991 Kugler Publications, Amsterdam/Milano/New York

Introduction

In the past we have presented the Nijmegen results of surgery for the treatment of congenital aural atresia[1-5].

At that time, our study group comprised 36 ears which had been operated on in the period from 1972-1982. Presently the group comprises 64 ears operated on over the period from 1972-1988. Our earlier retrospective study showed that an additional classification system was useful if we want to estimate the chance of achieving a good postoperative hearing level. To recap, the classification was as follows:

Type I: Congenital atresia of the external auditory canal (EAC) with intact tympanic membrane and bony atresia of the lateral part of the EAC.

Type IIA: Incomplete bony atresia such that the beginning of a membranous EAC is viseble, possibly with a funnel-shaped, blind passage, or full bony atresia particularly in the medial part of the bony EAC.

Type IIB: Total bony atresia over the whole lenght of the EAC.

Cases with types IIA and IIB nearly always have a well pneumatized mastoid and middle ear. Therefore, the middle ear can be reached along the bony dural plate and any pneumatized cells which are present.

Type III: Atresia with: (a) generally absent or badly malformed auricle; (b) severely malformed middle ear, sometimes including the inner ear. Mostly there is a very limited pneumatization of the mastoid, therefore exploration of the middle ear has to be abondoned.

This classification is a useful aid when deciding whether reconstructive surgery is desirable in cases with congenital aural atresia. Marquet[6] and Jahrsdoerfer[7,8] put forward other classifications which also support the relationship between the severity of the malformation and the ultimate hearing results.

An important aspect in the decision regarding surgical intervention is whether the atresia is unilateral or bilateral. In cases with bilateral atresia, achieving social hearing in just one ear is often sufficient.

In case of a unilateral atresia, the ultimate gain in hearing level must be sufficient to provide

the patient with the advantage of bilateral hearing. Owing to the fact that physicians tend to have reservations about advising reconstructive surgery in cases with unilateral aural atresia, we would like to present our results seperately.

Patients and Methods

Between 1972-1988, 64 ears in 56 patients were operated on for types I, IIA, or IIB congenital aural atresia at the ENT Department of the University Hospital Nijmegen. There were 39 cases of unilateral atresia and 25 with bilateral atresia.
The preoperative and postoperative hearing threshold is presented as a hearing loss of 0.5, 1 and 2 kHz. The follow-up period was at least two years. None of the patients were lost to follow-up.

Results

Table 1 shows the preoperative and postoperative hearing thresholds. The ears have been classified in accordance with our classification system and we have distinguished between unilateral and bilateral cases.

Table 1: Preoperative and postoperative hearing results in unilateral congenital aural atresia, 1972-1988

	n	Preoperative	Postoperative	Gain
Type I	1	50 dB	15 dB	35 dB
Type IIA	23	50 dB	27 dB	23 dB
Type IIB	15	57 dB	35 dB	22 dB

Air-conduction levels for 0.5, 1 and 2 kHz

The mean gain in hearing level in ears with type IIA and type IIB was very similar, namely 23 and 22 dB, respectively. The preoperative hearing loss for type IIB ears was considerably higher than for type IIA ears. Thus the mean postoperative hearing thresholds for IIA ears was 25 dB and for type IIB ears 35 dB. Whether the aural atresia was unilateral or bilateral did not influence the results to any significant extent.

Table 2: Preoperative and postoperative hearing thresholds in bilateral congenital atresia, 1972-1988.

	n	Preoperative	Postoperative	Gain
Type I	3	49 dB	21 dB	28 dB
Type IIA	15	43 dB	23 dB	20 dB
Type IIB	7	56 dB	35 dB	21 dB

Air-conduction levels for 0.5, 1 and 2 kHz

We did not observe any postoperative side-effects, such as facial paresis, complete hearing loss or serious detoriation of the perceptive threshold.

In this series the most favorable results were achieved particularly from 1980 onwards. This can largely be attributed to the selection criteria we used, which gave preference to cases with type IIA atresia.

Discussion

It is still an open question why the results in the group of patients with type IIB atresia remain disappointing. In our opinion, two factors are mainly responsible:

1. As a result of the serious degree of malformation, the middle ear and particularly the epitympanum will be narrowed. Preservation of the malleus and incus may lead to an increased risk of new postoperative fixation of the chain in the epitympanum.
2. Particularly the manubrium of malleus is often too short and too thick. If the chain is left intact, this can have a negative influence on the result.

At present, we are more likely than we used to be in these cases, to give preverence to removing the malleus and incus and to heightening the mobile stapes with the transformed autologous head of the malleus.

Our recent results are more encouraging. Therefore we feel that patients with type IIB atresia including unilateral cases should no longer be excluded from surgical treatment.

Our results for unilateral type IIA congenital aural atresia also show that unilateral atresia forms a good indication for reconstructive surgery.

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CHAPTER IV

SYNDROMES AND MAJOR AND MINOR EAR ANOMALIES

CHAPTER IV-1

AN AUTOSOMAL DOMINANT INHERITED SYNDROME WITH CONGENITAL STAPES ANKYLOSIS

E. Teunissen, C.W.R.J. Cremers
Laryngoscope 1990; 100: 380-384

ABSTRACT

A newly recognized autosomal dominant inherited syndrome associated with congenital conductive deafness, hyperopia, broad thumbs, broad first toes, short distal phalanges, and syndactyly is reported. The conductive loss was the result of congenital stapes ankylosis and, in two cases, was associated with fixation of the short process of the incus in the fossa incudis. Stapedectomy improved hearing in these patients. Fused cervical vertebrae are also an associated feature.

INTRODUCTION

Congenital stapes ankylosis is a rare anomaly. The medical history is usually adequate to exclude otosclerosis or other diseases of the middle ear. Congenital stapes ankylosis is mostly an isolated anomaly. It can be part of a congenital syndrome, with or without other congenital anomalies of the ossicular chain.

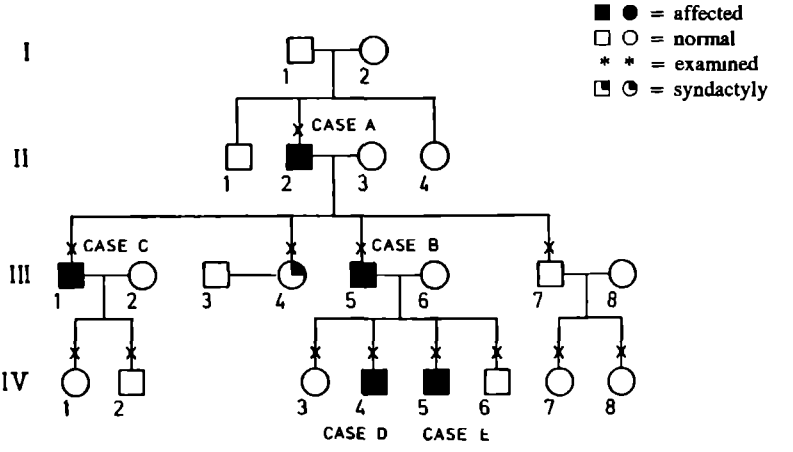
A family is reported with an autosomal dominant inherited conductive deafness, mostly as a result of isolated congenital stapes ankylosis. this congenital conductive deafness is part of a new syndrome that also includes hyperopia and syndactyly.

Associated features are broad thumbs, short distal phalanges (brachiotelephalangy), broad first toes, and fused cervical vertebrae. Exploratory tympanotomy revealed congenital stapes ankylosis that, in two cases, was associated with a fixed short process of the incus in the fossa incudis. Stapedectomy was successful in restoring normal hearing in three out of four ears, and on two cases, included widening of the fossa incudis. The result in the fourth ear was limited by a preexisting sensorineural component as a result of a previous stapes mobilization procedure. The syndrome reported in this family has not been previously described to our knowledge.

FAMILY CASE HISTORIES

The family members' main genetic features are presented in the pedigree (Fig. 1). The clinical findings of the affected family members are presented in Table I.

Fig. 1. Pedigree of the family



Case A (II-2)

A 57-year-old man was referred in 1959 because of his early childhood deafness. The medical history and especially the otological medical history were otherwise normal. There proved to be a bilateral flat conductive hearing loss of 50 to 60 dB. Radiological examination of the temporal bones showed no abnormalities. However, some associated congenital anomalies were noted, such as syndactyly of the second and third toes, hyperopia, and fused cervical vertebrae at C6-7. A possitive correction was used for the hyperopia. The correction in diopters is unknown. An exploratory tympanotomy was not performed.

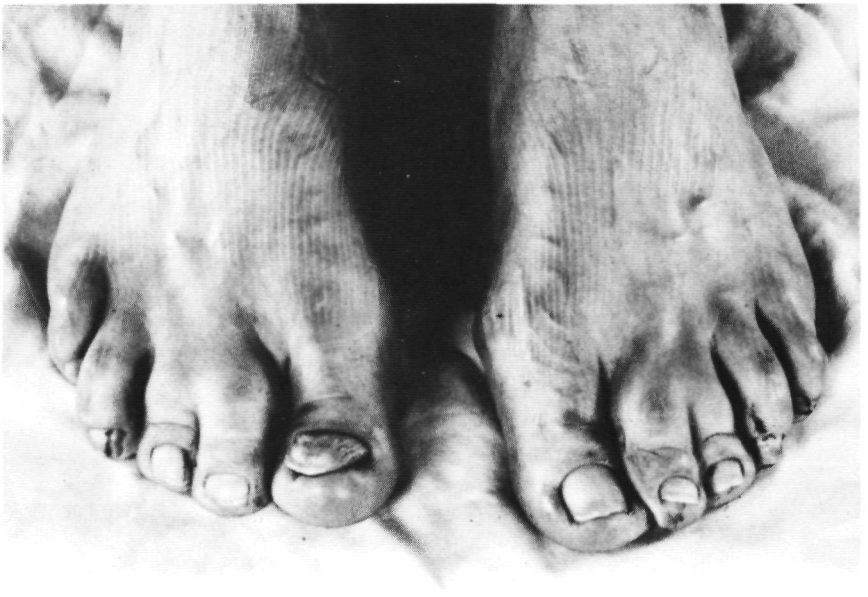
Case B (III-5)

A 44-year-old man who had been refered in 1959 at the age of 15, and his father (case A) were examined at the Nijmegen University Department of Otorhinolaryngology because of childhood deafness. He had a bilateral flat conductive hearing loss of 50 to 60 dB. His

membranes and well-aerated middle ear clefts. An exploratory tympanotomy was performed on the right ear, and a congenital stapes ankylosis was found. The footplate was white and thicker than usual, and the crurae were heavy. The malleus and incus looked normal and the mobility was intact. A stapes mobilization in 1959 had not been successful, and a small sensorineural hearing loss was subsequently found. The hearing loss was reinvestigated in 1978. The bilateral conductive hearing loss was unchanged. Speech discrimination was 100% at 80 dB. Contralateral stapedial reflexes could not be elicited. Radiological investigations of the temporal bones revealed no abnormalities. The right ear was re-explored in 1978. A stapedectomy and an all-Teflon piston interposition resulted in an essential hearing gain, although some sensorineural hearing loss remained at 2, 4 and 8 kHz as a result of the stapes mobilization in 1959. An air-bone gap of 10 to 20 dB also remained.

In 1981, an exploratory tympanotomy of the left ear was performed. The surgical findings were identical to those in the right ear. A stapedectomy with an interposition of an all-Teflon piston almost completely closed the air-bone gap.

Fig. 2. Syndactyly of the second and third toe with broad first toes in case B.



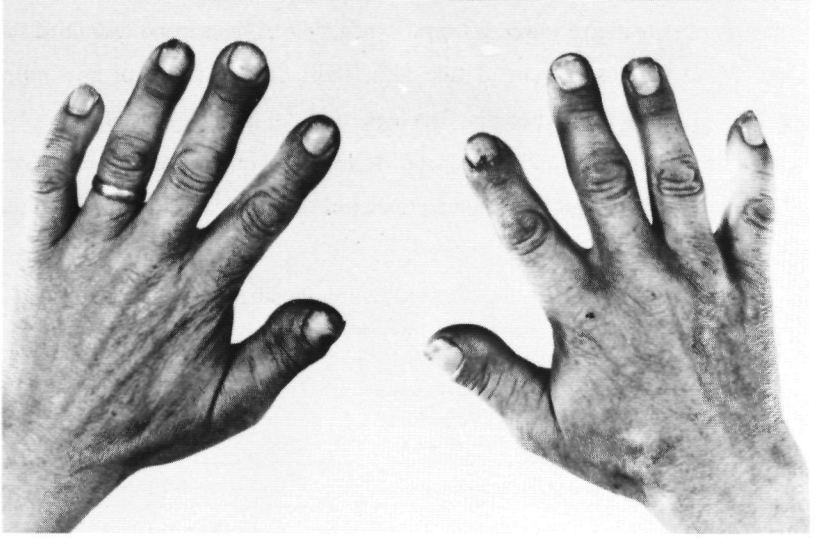


Fig. 3. Broad thumbs in case B.

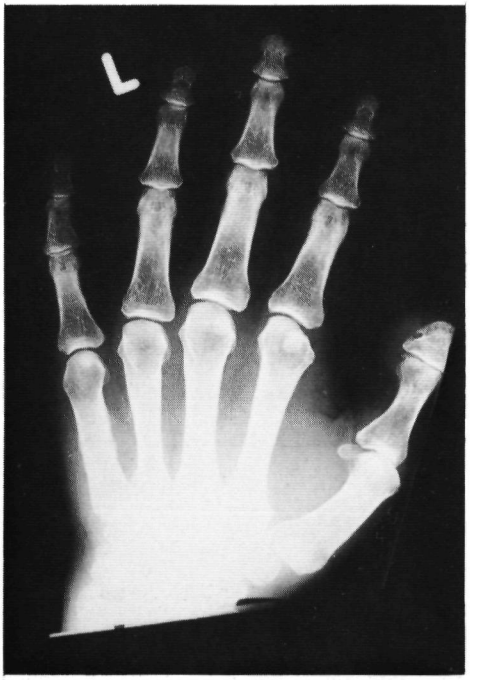


Fig.4. Radiological impression of the left hand in case B demonstrating short distal phalanges. Symphalangism or carpal fusions are absent.

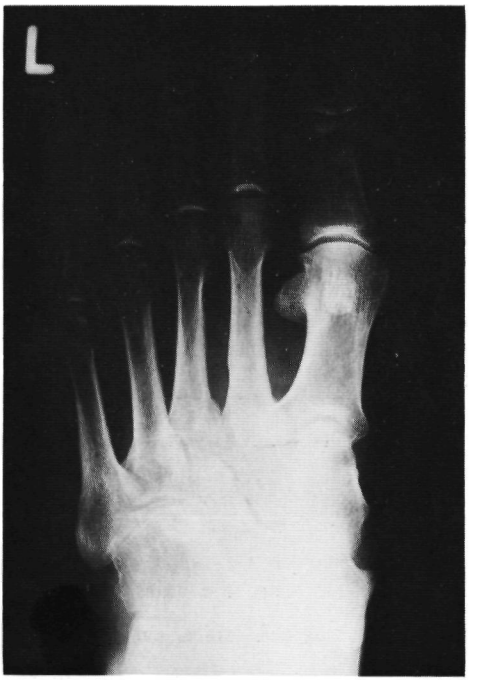


Fig. 5. Radiological impression of the left foot in case B demonstrating short distal phalanges. Symphalangism or tarsal fusions are absent.

Recent clinical investigation showed bilateral syndactyly of the second and third toes (Fig. 2), bilateral broad thumbs and broad first toes, short distal phalanges, as confirmed by radiology (Figs. 3, 4, 5), and hyperopia. The hyperopia required a correction of +5 diopters and has remained unchanged since childhood. Routine ophthalmologic investigations were normal. Radiological investigations showed fusion of the cervical vertebrae (C-6 and C-7).

Table 1: Clinical findings in the affected family members

Case	Hearing loss	Findings at operation		Syndactyly of 2nd and 3rd toe	Broad thumbs and first toes with Brachio-telephalangia
		Stapes Ankylosis	Fixed Short Process		
A	60 dB	not operated upon		+	+
B	60 dB	+	-	+	+
C	60 dB	not operated upon		+	+
D	40-50 dB	+	+	+	+
E	60 dB	+	+	+	+

Case	Correction for Hyperopia	Diopters	Fused cervical Vertebrae on Roentgenograms	Symphalangism
A	+	?	C6-C7	-
B	+	+ 5	C6-C7	-
C	+	+ 11	-	-
D	+	+ 5	-	Fifth fingers
E	+	+ 4	-	-

Case C (III-1)

A 53-year-old man, the eldest son of the patient in case A, presented with bilateral conductive deafness since early childhood. The medical and otological history were uneventful. Otological examination showed normal tympanic membranes and well-aerated middle ear clefts. The audiogram showed a bilateral flat conductive hearing loss of 50 to 60 dB. Further clinical investigations revealed all the signs of this syndrome (Table I).

Case D (IV-4)

A 7-year-old boy was refered for otological and audiological examination in 1977. The medical and otological history were normal. There was a left-sided-50 dB flat conductive deafness and a lesser conductive hearing loss of 40 dB in the right ear. Contralateral stapedial reflexes could not be elicited in either ear. Table I presents typical clinical, otological, audiometrical, and ophthalmological findings. There was also proximal symphalangism of both fifth fingers. Computed tomographic (CT) scans of the temporal bones showed no abnormalities.

A retroauricular mastoidectomy and posterior tympanotomy had recently been performed and showed a congenital stapes ankylosis with a slightly abnormal suprastructure. The short process of the incus was congenitally fixed in the fossa incudis. Restoring the mobility of the incus and a stapedectomy with Teflon-wire interposition resulted in an almost complete closure of the air-bone gap.

Case E (IV-5)

A 12 year-old boy had been refered for otological and audiological examination at the age of 4, because of suspected serious hearing loss. The medical, and especially, otological history were unremarkable. Normal tympanic membranes and well-aerated middle ear clefts were seen at otoscopy. A flat bilateral conductive hearing loss of 60 dB (Fig. 6) has been confirmed by recent investigations. Contralateral stapedial reflexes could not been elicited in either ear. CT scans of the temporal bones showed no abnormalities. The other clinical findings are summarized in Table I.

Recently at the age of 12, exploratory tympanotomy was performed on the right ear. A congenital stapes ankylosis was found without typical anomalies of the stapes suprastructure. The malleus and incus looked normal, although the mobility of the incus seemed less than usual. A retroauricular mastoidectomy and posterior tympanotomy were performed to free the short process of the incus, which was congenitally fixed in the fossa incudis. After restoring the mobility of the incus, a stapedectomy and a Teflon-wire interposition were performed, which resulted in a closed air-bone gap (Fig. 7).

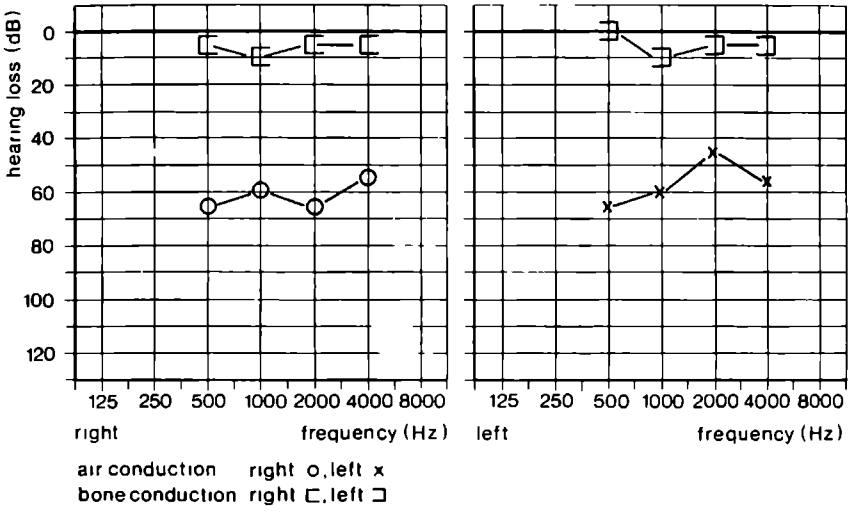


Fig. 6. Preoperative conductive hearing loss in case E (IV-5).

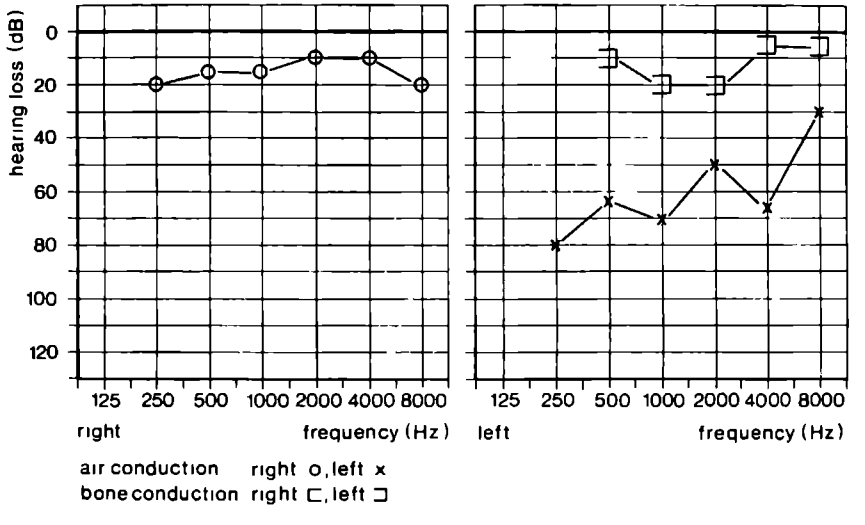


Fig. 7. Postoperative audiogram 6 months after restoring the mobility of the incus and a stapedectomy of the right ear in case E.

In one patient (III-4), a syndactyly of the second and third toes, and normal hearing and vision was found.

All other family members have undergone clinical and audiometrical investigation to trace symptoms of this syndrome. They are free of symptoms and therefore considered unaffected.

DISCUSSION

The affected members of this family show a well-delineated clinical entity that has not been reported previously.

Three generations have had affected family members. The repeated transmission of this disease from father to son is proof of an autosomal dominant pattern of inheritance. By this male to male transmission the X-dominant pattern of inheritance is ruled out, because it would require only a transmission from a father to daughter, and so possibly again to her son. The very rare Y-dominant pattern of inheritance is also excluded since this pedigree shows affected fathers having unaffected sons.

Congenital conductive deafness as a result of anomalies of the ossicular chain, especially stapes ankylosis, hyperopia, broad thumbs and broad first toes, short distal phalanges (brachiotelephalangy), and syndactyly of the second and third toes are constant features of this new syndrome. Associated features are fused vertebrae and proximal symphalangism of the fifth finger.

Stapedectomy resulted in normal hearing with a closed air-bone gap in three out of four ears. On two occasions, congenital fixation of the short process of the incus in the fossa incudis could be resolved during the same operation.

Recent reviews of syndromes associated with genetic deafness do not describe the syndrome reported in this study.[1-4] Congenital stapes ankylosis is part of an autosomal dominant inherited syndrome with proximal symphalangism.[5] Several somewhat similar syndromes with stapes ankylosis and anomalies of the hand and feet have also been described as the facio-audio-symphalangism syndrome[6] and the Nievergelt-Pearlman syndrome.[7] Carpal and tarsal fusion, symphalangism, and syndactyly are also features of the syndrome. Although

syndromes with congenital conductive deafness and proximal symphalangism are similar to this syndrome, detailed reviews of the literature indicates that the syndrome in this report is a seperate entity.

Stapes ankylosis is also a feature of syndromes with craniosynostosis like Crouzon's disease,[8] Apert's syndrome,[9] and Pfeiffer's syndrome.[10] The absence of a craniosynostosis and of the typical hand anomalies of some of these syndromes is sufficient to exclude them.

Congenital conductive deafness as a result of stapes ankylosis, is also symptomatic of the Wildervanck syndrome[11] and the Klippel-Feil syndrome.[12] Hyperopia has been reported as part of several syndromes, but not with congenital conductive deafness resulting from stapes ankylosis.[4]

The other features of the syndrome reported here and the autosomal dominant pattern of inheritance exclude other possible diagnoses. We therefore conclude that the clinical findings described represent a new syndrome.

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CHAPTER IV-2

THE IMPACT OF A SYNDROMAL DIAGNOSIS ON SURGERY FOR CONGENITAL MINOR EAR ANOMALIES

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International Journal of Pediatric Otorhinolaryngology, 1991; 22: 59-74.

ABSTRACT

Between 1964 and 1986, 104 ears of 86 patients with a minor congenital ear anomaly underwent an exploratory tympanotomy at the Institute of Otorhinolaryngology of the University Hospital Nijmegen. A classification of these anomalies is proposed based on the surgical findings and results. The 4 groups in this classification are: isolated stapes ankylosis, stapes ankylosis associated with an anomaly of the malleus and incus, an isolated anomaly of the malleus and incus with a mobile stapes footplate and finally, aplasia of the oval and/or round window.

In a total of 29 ears (22 patients) out of these 104 ears, the anomaly formed part of a syndrome. The various syndromes and the anomalies encountered are discussed. The impact of a syndromal diagnosis on the outcome of reconstructive ear surgery is discussed per syndrome.

INTRODUCTION

A congenital anomaly of the middle ear is a rather rare condition. Owing to the large variation in anomalies, we classified them into the following four groups:[22,23,24]

- class I: ears with isolated stapes ankylosis,
- class II: ears with stapes ankylosis associated with an anomaly of the malleus and/or incus,
- class III: ears with an isolated anomaly of the malleus and/or incus and a mobile stapes footplate
- class IV: ears with aplasia of the oval and/or round window.

The results of surgery are presented for each of the first three categories. The syndromes in this series of 104 ears are listed together with the anomalies encountered and the results of surgery.

PATIENTS AND METHODS

Between 1964 and 1986, 104 ears of 86 patients with a minor congenital ear anomaly underwent exploratory tympanotomy at the Institute of Otorhinolaryngology of the University

Hospital Nijmegen. A retrospective study was conducted based on the medical files, including extensive surgical reports.

All the participants had a typical history of mainly conductive deafness, which had been present since childhood. Many of them had been seen recently to extend the follow-up. Since the mid seventies, special attention has been paid to the possible existence of a syndromal diagnosis in patients with congenital hearing loss at our department. This approach also revealed that previously missed syndromal diagnoses could still be made at a later date. Many such findings have already been published separately in relation to their syndromal diagnosis, including clinical genetic studies on these syndromes.[3,4,5,6,10,11,12,13,18,19,21] An overview is given below, together with the unpublished syndromal data on this series.

Patients with osteogenesis imperfecta were excluded from this series, mainly because of the late onset of the hearing loss.[7,15,16] Cases of suspected otosclerosis or tympanosclerosis were also excluded.

The preoperative and postoperative hearing levels were calculated as the mean values at frequencies 0.5, 1 and 2 kHz. Hearing gain calculations were based on individual values. Tone audiometry and speech audiometry was performed preoperatively in all patients. Preoperatively elicited stapedial reflexes were not available in all cases. All the patients had a follow-up of at least 1 year; the majority had a much longer follow-up.

The impact of being aware of a syndromal diagnosis to evaluate the profit of reconstructive surgery will be discussed.

RESULTS

A syndromal diagnosis was made in 29 ears of 22 patients from a series of 104 ears of 86 patients. These ears were explored between 1964 and 1986.

The syndromal diagnoses are listed in Table 1. We have subdivided this list of syndromal diagnoses into the following 4 subgroups:

(1) Stapes gusher syndrome

(2) Branchiogenic syndromes

(3) Craniosynostosis syndromes

(4) Skeletal syndromes

Table 1: Syndromal diagnosis in congenital minor ear anomalies. Series of 104 ears from 1964-1986.

Syndrome	Number of ears	Number of patients
Stapes gusher syndrome		
Stapes gusher	2	2
Branchiogenic syndromes		
Branchio-oto-renal syndrome (earpits-deafness syndrome)	7	6
Treacher Collins syndrome	5	3
Preauricular sinus, external ear anomaly, commissural lip pits	2	1
Hemifacial microsomia	1	1
Craniosynostose syndromes		
Crouzon syndrome	1	1
Pfeiffer syndrome	1	1
Skeletal syndromes		
Klippel-Feil	5	3
Wildervanck	1	1
Frontometaphyseal dysplasia	1	1
Proximal symphalangia	1	1
Stapes ankylosis, hypermetropia, Brachytelephalangia	2	1
	29	22

In our experience, the syndromes in these 4 subgroups show typical anomalies which also influence the outcome of surgery. We found the following two new syndromes in our series:

- the autosomal dominant inherited syndrome of preauricular sinus, external ear anomaly and commissural lip pits and
- the autosomal dominant inherited syndrome of congenital stapes ankylosis, brachytelephalangia and hypermetropia.

The results and syndromes are presented per subdivision of the four groups of syndromes.

STAPES GUSHER SYNDROMES

Stapes gusher is a rare, but avoidable complication of surgery for presumed stapes ankylosis. After the stapes footplate has been opened, a gush of excess perilymph and cerebrospinal fluid (CSF) occurs, which fills the ear speculum and may wet the postoperative dressings for several days as it discharges via the external auditory meatus.[17,20]

Stapes gusher occurs in cases with an X-recessive hereditary syndrome who have been suffering from mixed, progressive hearing loss since childhood. The perceptive part of the hearing loss is progressive. The conduction loss is strikingly high for the frequencies 250, 500 and 1000 Hz and strikingly low for 2000 and 4000 Hz.[4] It is also noteworthy that the stapes reflex can be induced contralaterally.[4,9] Polytomography and CT scanning can be employed to show that the lateral part of the internal auditory meatus is widened. During exploratory middle ear inspection, we noticed that a small amount of perilymph is secreted when the mobility of the stapes is tested. It is usual for the female carriers to have a slight hearing loss.[11] In the meantime, the gene for this syndrome has been linked to the X chromosome, which before too long can be expected to provide support for a clinical diagnosis.[2]

If stapedectomy is unsuccessful, the postoperative hearing loss is generally greater owing to the discontinuity of the ossicular chain. In addition, the perceptive component can increase as a result of persistent leakage of perilymphatic fluid. In our series, two cases of stapes gusher were observed.[19] This induced a search for characteristics which can be recognized preoperatively, such as those described above.

Stapes gusher can occur in persons in whom the communication between the perilymphatic space and the internal auditory meatus is openly accessible. In some cases, widening of the internal auditory meatus is visible on the X-ray.[14]

BRANCHIOGENIC SYNDROMES

In view of the fact that the middle ear is derived from branchiogenic structures, it is understandable that particularly in these syndromes, in which external characteristics of branchiogenic malformation are also visible (fig. 1, 2), this background can lead to

Table 2: Specification of congenital minor ear anomalies and surgery in ears with syndromal diagnosis. Preoperative (Preop) and at least 1 year postoperative (postop) bone conduction (bone) and air conduction (air) thresholds for 500, 1000 and 2000 Hz.

Branchio-oto-renal syndrome

Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
1	rudimentary anterior crus, long posterior crus, stapesankylosis	broad lenticular process	normal	stapedectomy	15/77 dB	30/60 dB
2	absent stapedial tendon, stapes ankylosis	broad crus longum	normal	stapedectomy	28/50 dB	28/42 dB
3	absent stapedial tendon and broad crurae, stapes ankylosis	broad, small and curved crus longum	normal	stapedectomy	40/80 dB	40/50 dB
4	aplasia of oval window	unknown	unknown	none	30/60 dB	30/60 dB
5	absent, aplasia of oval window	absent lenticular process	normal	none	30/75 dB	30/75 dB
5	absent, aplasia of oval window	strongly curved crus longum	normal	none	15/72 dB	15/72 dB
6	fragile, aplasia of oval window	broad crus longum	fixed to incus	none	40/70 dB	50/70 dB

Prae-auricular sinus, external ear anomaly, commissural lippits

Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
7	malformed suprastructure and stapes ankylosis	too small crus longum	normal	stapedectomy	18/53 dB	10/20 dB
7	malformed suprastructure and stapes ankylosis	absent crus longum	normal	stapedectomy maleus-footplate prosthesis	20/55 dB	12/20 dB

Treacher Collins Syndrome

Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
8	head fixed to bony facial canal	bony spine	normal	mobilisation of stapes head	10/55 dB	10/42 dB
8	rudimentary	bony spine	rudimentary	mobilisation of bony spine	10/55 dB	10/45 dB
9	head fixed to bony facial canal	ankylotic I-S joint	fixed	mobilisation; interposition allologous malleus	30/60 dB	22/22 dB
9	malformed and fragile window	rudimentary	fixed	mobilisation; interposition of allologous incus on stapes	13/50 dB	10/32 dB
10	absent crurae very small oval window	bony spine	absent	none; facial nerve crossing oval window	5/50 dB	5/50 dB

Hemifacial microsomia

Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
11	normal	strongly curved long process	fixed to bony annulus through atretic plate	mobilisation of malleus and removal of atretic plate	15/68 dB	20/30 dB

Klippel-Feil syndrome

Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
12	ankylotic; not malformed	normal	normal	stapedectomy	25/52 dB	15/15 dB
12	ankylotic; not malformed	normal	normal	stapedectomy	18/40 dB	17/17 dB
13	missing stapes head	dysplastic long process	normal	allologous incus interposition	10/48 dB	10/30 dB
13	missing stapes head	small long process	normal	allologous incus interposition	8/45 dB	5/20 dB
14	aplasia of oval window	short long process	normal	stapedectomy in fenestrated oval window region	10/50 dB	12/50 dB

Permission for publication granted by Ann Otol Rhinol Laryngol.[18]

Wildervanck syndrome

Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
15	ankylotic not malformed	normal	normal	stapedectomy	25/45 dB	18/25 dB

Crouzon syndrome

Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
16	ankylotic	epitympanic fixation	epitympanic fixation	epitympanotomy of congenital too small epitympanum	7/55 dB	5/40 dB

Pfeiffer syndrome

Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
17	ankylotic and malformed	fixed; malformed long process	normal	epitympanotomy for congenital small epitympanum; stapedectomy	15/38 dB	17/21 dB

Fronto Metaphyseal Dysplasia

Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
18	reduced mobility, malformed	reduced mobility in small epitympanum	normal	epitympanotomy and allologous malleus interposition	22/55 dB	22/55 dB

Proximal symphalangism

Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
19	ankylotic and malformed	fixed in small epitympanum	normal	epitympanotomy and stapedectomy in separate procedures	8/30 dB	5/13 dB

Stapesankylosis, hypermetropia, brachiotelephalangia, syndactyly

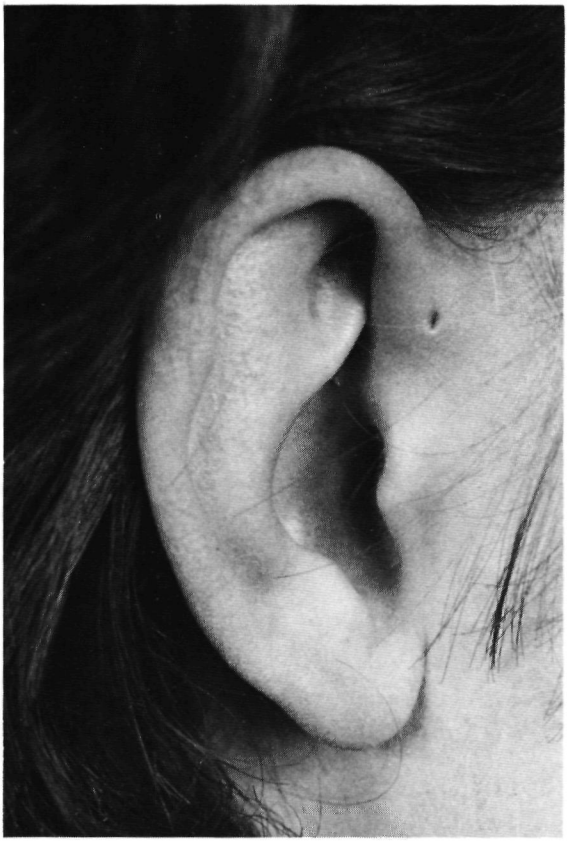
Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
20	ankylotic	normal	normal	stapedectomy	28/63 dB	23/35 dB
20	ankylotic and slightly malformed	normal	normal	stapedectomy	22/53 dB	13/17 dB

Stapes gusher

Patient	Middle ear anomaly			Reconstruction	Hearing loss	
	Stapes	Incus	Malleus		Preop. bone/air	Postop. bone/air
21	ankylotic	normal	normal	none	47/75 dB	47/75 dB
22	ankylotic	normal	normal	none	5/45 dB	40/85 dB

malformation of all 3 ossicles as well as other middle ear structures, such as aplasia of the oval and round windows and deviation of the facial nerve. Consequently, the chance of surgery being successful depends to some extent on the severity of the malformation.

Fig. 1. Preauricular pits in branchio-oto-renal syndrome. Permission for publication granted by Karger, Basel.[12]

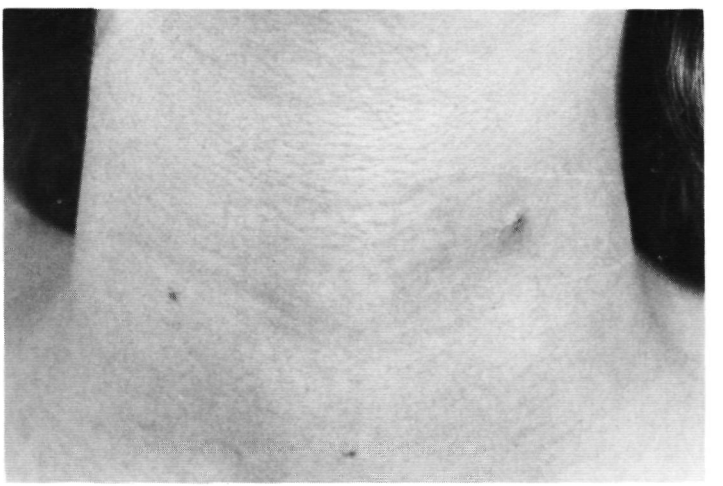


In 7 ears of cases suffering from the branchio-oto-renal syndrome, formerly referred to as the earpits deafness syndrome, variations were observed in the type and severity of the middle ear anomalies described and in the generally somewhat disappointing surgical results (Table 2).[6,12] It nevertheless seems justified to perform middle ear inspection on the basis of these results, although the chance of success is small.

The Treacher Collins syndrome is associated with an even larger variety of middle ear

anomalies. The whole middle ear can be absent in combination with meatal atresia and anotia. In our experience, in two out of 5 operated ears a considerable hearing gain could be achieved. The long process of the incus was absent in these cases. This discontinuity in the ossicular chain was restored by fixing a transformed malleus head to the mobile stapes. It was striking that the mobile stapes was so malformed in

Fig. 2. Branchiogenic fistula in branchio-oto-renal syndrome. Permission for publication granted by Karger, Basel.[12]



some cases that it pressed against the facial nerve or had formed an ankylotic bridge, while the stapes head stuck out even further above the facial nerve. This can lead to difficulties during reconstruction surgery of the ossicular chain.

The autosomal dominant hereditary syndrome with preauricular sinus, external ear anomaly and commissural lip pits was recently recognized as a new syndrome during a specific clinical genetic study.[18] The middle ear anomalies found varied from a shortened long process of the incus to isolated stapes ankylosis and aplasia of the oval window with serious malformation of the remaining ossicles.

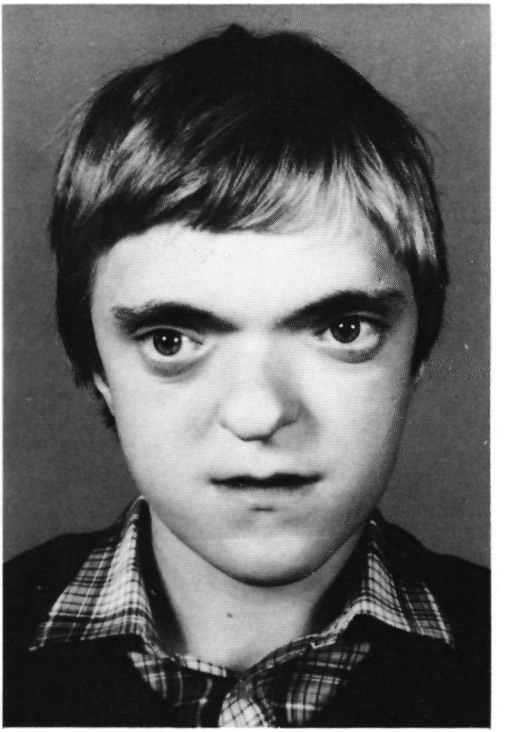
Hemifacial microsomia is generally so serious that there is a considerable degree of meatal atresia, which cannot often be corrected surgically. In one case we observed a conductive

hearing loss in the contralateral ear resulting from fixation of the malleus to the tympanic membrane by means of a small atretic bony plate. The conductive hearing loss disappeared after the bony plate had been removed from the tympanic membrane. Such an anomaly forms the transition between major and minor congenital anomalies of the ear.

CRANIOSYNOSTOSIS SYNDROMES

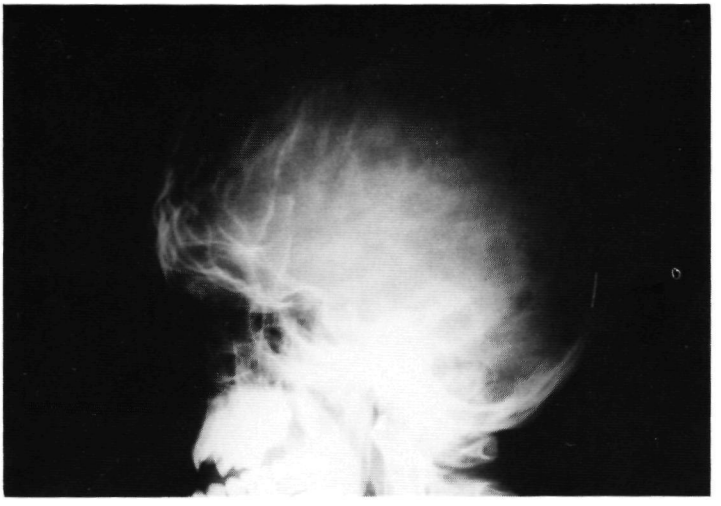
Very few results on ear surgery in patients with craniosynostosis have been reported in the literature.[1] Surgery may be indicated on account of isolated ossicular chain fixation or bony meatal atresia. Our experience is limited to one patient with the Pfeiffer syndrome (Fig. 3,4)[3] and one patient with the Crouzon syndrome.

Fig. 3. Facial appearance of Craniosynostosis syndrome (Pfeiffer syndrome).



In cases of craniosynostosis, the middle ear, epitympanum and mastoid are smaller than usual and the mastoid may even be absent. The ossicular chain was intact in both cases, but in addition to stapes ankylosis, the malleus and incus were fixed in the narrow epitympanum (Table 2). In the patient with the Pfeiffer syndrome, the epitympanum was wide enough to allow the successful separation of the ossicular chain using the epitympanic approach, followed by successful stapedectomy.[3] In the patient with the Crouzon syndrome, the mastoid was underdeveloped and the sigmoid sinus was situated too far forward. Although it was possible to free the ossicular chain in this area from its bony fixation during the first of a two tempo operation, during the second operation for stapes ankylosis, it was decided not to perform stapedectomy because new bony growth had led to recurrence of the epitympanic fixation.

Fig. 4. Rontgenographic appearance of skull in craniosynostosis syndrome (Pfeiffer syndrome). Note the absence of bone sutures and impressions of cerebrum in the skull bone.



SKELETAL SYNDROMES

KLIPPEL-FEIL / WILDERVANCK SYNDROME

The fusion of multiple hemivertebrae into one osseous mass is the only characteristic (anomalad) of the Klippel-Feil syndrome. It can be accompanied by perceptive or conductive deafness. If abducens paralysis is also present, the condition is referred to as the Wildervanck syndrome. The findings in our patients were described recently by Van Rijn and Cremers[18] together with a review of the literature. The anomaly can vary from aplasia of the oval window to isolated stapes ankylosis or a shortened long process of the incus with a mobile stapes. One case of stapes gusher was described following stapedectomy.[14]
In the remaining syndromes, other skeletal malformations appear to form part of the characteristics. Frontometaphyseal dysplasia involves extra bony apposition which gradually reduces the size of the middle ear cavity and epitympanum. At first the ossicles are mobile, but as the cavity becomes smaller owing to bony outgrowth, they become fixed. Once the stapes has become fixed at a later stage, surgical intervention is no longer successful.[5]

A striking new finding has been made in patients with the proximal symphalangia syndrome (fig 5)[13] and the stapes ankylosis syndrome with hypermetropia and brachytelephalangia.[21]
It appears that stapes ankylosis is accompanied by isolated fixation of the short process to the surrounding bone in the fossa incudis, whereas there is no fixation of the malleus or incus in the epitympanum.
Such fixation of the short process in the fossa incudis is easily missed if one is not expecting it and can be the cause of insufficient improvement in the patient's hearing after successful stapedectomy.[21]

DISCUSSION

In this series of congenital middle ear anomalies, over 25% of the ear anomalies formed part of a congenital syndrome. If the cases of osteogenesis imperfecta had been included, the percentage would be even higher. Our studies have shown that it is possible to recognize the X-recessive hereditary syndrome with a mixed progressive hearing loss preoperatively and the results of gene linkage studies will help to improve this in the near future. Thus it should

be possible to avoid stapes gusher as a surgical complication in these cases.

In the branchiogenic syndromes, the degree to which the ossicles are deformed is greater than we would normally expect in congenital middle ear anomalies. This has a negative influence on the results of reconstructive surgery.

The craniosynostosis syndromes with small-sized middle ear and epitympanum, are also typical in their anomaly. The degree to which the mastoid is absent and the epitympanum is too small, appears to influence the final surgical results.

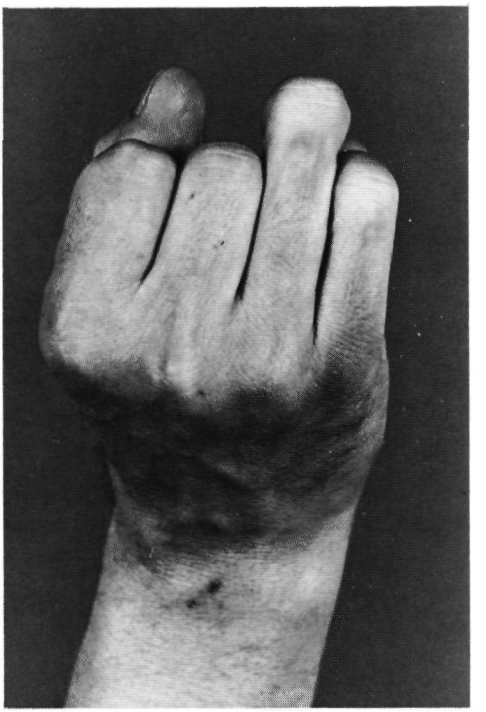
The remaining syndromes with an associated skeletal anomaly generally display a perfectly formed ossicular chain which has become fixed at one or more places. Consequently, good results can be expected with surgical reconstruction.

The Klippel-Feil syndrome is associated with a wide variety of middle ear anomalies; this can limit the surgical possibilities.

CONCLUSION

If we review the present series, we can conclude that a syndromal diagnosis provides some preoperative indications as to the type of middle ear anomaly present. It also appears to be possible to recognize patients with the X-recessive stapes gusher syndrome prior to surgery. The clinical genetic aspects of disabilities with congenital middle ear anomalies are not the only reason to consider a syndromal diagnosis. A syndromal diagnosis also provides information on the type of middle ear anomaly present and on the chance that surgery will lead to hearing gain.

Fig. 5. Proximal symphalangism in patient with normal facial appearance.



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CHAPTER V

SOME AUDIOLOGICAL ASPECTS IN MAJOR AND MINOR EAR ANOMALIES

CHAPTER V-1

FREQUENCY RESOLUTION AFTER SUCCESSFUL SURGERY IN CONGENITAL EAR ANOMALIES

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Scandinavian Audiology 1991; 20: 265-267

ABSTRACT

Frequency resolution was studied in patients with a unilateral congenital ear defect (atresia), who were successfully operated on. Psychophysical tuning curves (PTCs) were obtained. A comparison of the shape of the PTCs revealed that the average curve for all the patients was much alike for the operated and nonoperated (normal) ear: broad tuning curves, as reported from animal studies, were not found. After appropriate corrections were made for the small remaining air-bone gaps, as seen in all patients, the PTCs of all the operated ears lay reasonably well within the 2 S.D. range of the average PTC obtained in subjects with normal hearing.

INTRODUCTION

Good hearing results have been reported after reconstructive surgery for unilateral and bilateral aural atresia. These good results are obtained more frequently in ears with a lighter degree of bony atresia of the external aural canal. Consequently, Cremers et al.[1] proposed an additional classification. Hence reconstructive surgery was also advocated in unilateral cases with a less severe anomaly. Clinical experience has shown that patients with a good postoperative pure tone hearing threshold profit from this hearing improvement, even in noisy circumstances. Nevertheless it has been suggested that suspected auditory deprivation would impede these patients from profiting from this improved hearing, especially in cases with unilateral atresia.

In the late 1970s, Webster & Webster[2] published a study on the histological findings in the auditory neural system of animal ears which had been occluded directly after birth. The induced conductive hearing loss proved to lead to deprivation of the auditory neural system. Clopton[3] studied several functional properties of such ears: e.g. he found that the physiological tuning curves obtained were broad which indicated that frequency resolution was poor.

In the present study, frequency resolution was determined in a number of patients with unilateral, succesfully operated, congenital atresias, by measuring (psychophysical) tuning curves (PTCs). PTCs were measured so that the results could be compared with Clopton's psychophysical tuning curves[3] and because a simple procedure was available to obtain

PTCs.[4,5]

PATIENTS AND METHODS

Eight patients and ten subjects with normal hearing participated in the experiments. All the patients had a unilateral conductive hearing loss due to congenital anomalies. The average hearing losses at 500, 1000, 2000 and 4000 Hz before and after surgery are presented in Table 1. The average loss in the contralateral, normal ear is presented too. In all the patients, some residual conductive loss was still present after surgery. To be included in this study, the difference in postoperative air-conduction thresholds between the two ears had to be 20 dB or less at 500, 1000 and 2000 Hz and 25 dB or less at 4000 Hz.

Table 1. Preoperative and postoperative audiological results in the group of 8 patients

Ear	Average loss* (dB HL)	SRT (dB)	SRS (%)
Preoperative	51 ± 12	52 ± 10	
Postoperative	20 ± 4	19 ± 5	95 ± 6
Normal ear	7 ± 3	4 ± 3	99 ± 1

* Average loss at 500, 1000, 2000 and 4000 Hz.

The average age of the patients was 27 years, ranging from 11 to 45 years. Patients were operated on at the age of 8 or later. Six of the patients were male, 2 female. The time between the measurements and the previous surgery varied from 5 months to 8 years. None of the patients had used a hearing aid.

All the subjects with normal hearing (n=10) had thresholds of 15 dB HL or better. Their age ranged from 18 to 38 years with an average of 30 years.

Pure tone and speech audiometry were performed, using standard audiological procedures. Phonetically balanced monosyllable word lists were used to obtain the speech audiogram. From the speech audiogram, the speech recognition threshold (SRT) was derived as being the displacement of the speech intensity-recognition curve measured at 50% phoneme recognition. Furthermore, the speech recognition score (SRS) was derived as being the phoneme score at an intensity level of 35 dB above the SRT value.

The PTCs were determined using the method described by Zwicker & Schorn[4]. All tuning curves were determined at a fixed test-tone frequency of 2000 Hz. The test tone was presented intermittently (switched on and off smoothly, cycle time 1 s, duty cycle 50%) at a low sensation level (10 dB above threshold). Simultaneously, a second (continuous) tone, the masker, was presented to the same ear. At six masker frequencies (three above and three below the test tone frequency) the level of the masker which just masked the test tone, the so-called masker level, was determined. The curve of the masker levels versus the frequency, represents a simplified PTC.

To mask the combination tones, low pass noise was used in all the experiments.[5]

The test-tone, the noise as well as the maskers was generated by an Interacoustics AC-5 audiometer calibrated according to ISO 389. The calibration values for the frequencies not specified in the ISO 389 were derived by interpolation on a sheet of logarithmic paper (as suggested in the AC-5 manual Interacoustics), using the specified calibration values. As a result, all levels will be expressed in dB HL. All the experiments were performed in a double-walled, sound-proof room, using the AC-5 audiometer with a Telephonics TDH-39P headphone with a MX-41/AR cushion.

The test tone with noise had been recorded previously on (professional) tape (Snik & Horst 1991). During a measurement, this recorded signal was processed by channel 1 of the audiometer. The maskers comprising pure tones at frequencies of 812, 1562, 1812, 2187, 2437 and 2687 Hz were generated by channel 2 of the audiometer.

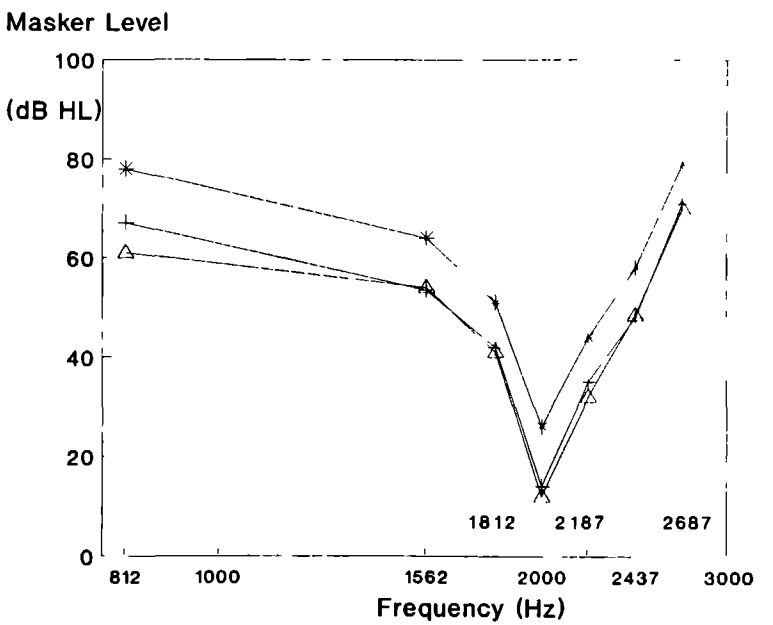
To obtain a PTC, first the threshold for the test tone was determined. Next, the test tone was set at 10 dB SL (sensation level) and the masker levels were determined at all masker frequencies in the following way: The masker was presented for 3 seconds. The subject indicated whether the test tone could be heard in the presence of the masker, in which case the level of the masker was increased (in 5 dB steps). This procedure was repeated until the test tone could no longer be heard (the masker level). To increase the reproducibility, all masker levels were obtained twice. Average values are presented.

To be able to make a comparison with the values in the literature, the d_{1oct} value was used. According to Bonding[6], the d_{1oct} value is the distance in dB between the tip of the PTC (estimated by the test tone level) and the level at which the PTC is one octave wide.

RESULTS AND DISCUSSION

In Table 1, average preoperative and postoperative pure tone thresholds and speech recognition thresholds are presented. On average, the air-bone gap of the operated ear was reduced by 31 dB, ranging from 22 to 45 dB. The SRS of the operated ear was poorer than that of the normal ear (Table 1): the SRS value of the operated ear varied from 83 to 100%. Reduced frequency resolution may have played a part: it is known that frequency resolution is one of the fundamental properties of the ear that contributes to speech discrimination.

Fig. 1. Average psychophysical tuning curves of subjects with normal hearing ( ), normal ears (+) and operated ears (*) of the patients.

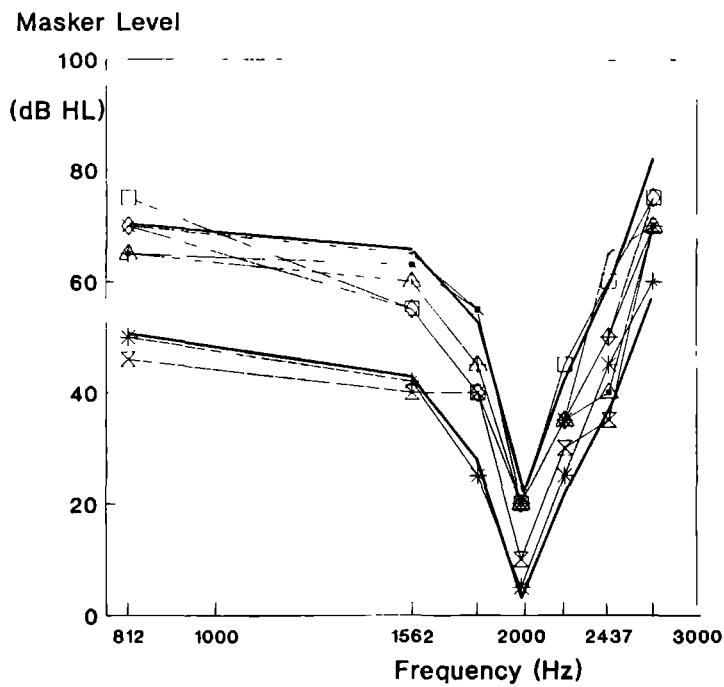


In Figure 1, the average PTCs of the subjects of normal hearing, the normal ear of the patients and the operated ear are presented. The average PTCs of the subjects with normal hearing were in good agreement with those of the normal ear of the patients.

The average d_{1oct} value of the subjects with normal hearing and the normal ear of the patients was 44.3 $\pm$ 2.0 dB and 45.5 $\pm$ 4.6 dB, respectively. Both values are in good agreement with the values in the literature.[7]

The PTC's of the operated ears had the same shape as those of the other two PTC's, although all the masker levels as well as the test tone levels were about 10 dB higher (Figure 1). This was caused by the set-up chosen: the test tone was presented at 10 dB SL, leading to higher values of the test tone in dB HL for the operated ear than in the normal ear as a result of the remaining small air-bone gap.

Fig. 2. Individual psychophysical tuning curves (n=8) of the operated ears after corrections were made for remaining air-bone gaps. The two thick lines connect the average masker levels plus and minus 2 S.D. of the subjects with normal hearing. Different symbols refer to different patients.



In order to compare the individual PTCs of the patients to normal PTCs, the PTCs were corrected for the above-mentioned shift caused by the remaining air-bone gap. The test tone level and masker levels were corrected by subtracting the measured air-bone gap. As only

the air-bone gap was known at the octave frequencies, the masker level at 812 Hz was corrected by subtracting the air-bone gap obtained at 1000 Hz; the other masker levels and the test tone level were corrected by subtracting the air-bone gap obtained at 2000 Hz. In this way, corrections were made for the remaining middle ear losses.

The corrected PTCs of all eight patients are presented in Figure 2. To make a comparison, two more PTCs are presented (thick lines), based on the average masker levels plus and minus 2 s.d. of the subjects with normal hearing. We can conclude that almost all the masker levels of the patients lay within the 2 s.d. range, which leads to the further conclusion that reduced frequency resolution, as reported by Clopton[3], was not present in our patient group.

Acknowledgments:

The authors wish to thank Ir. G. Hombergen for reading the manuscript.

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CHAPTER V-2

SPEECH RECOGNITION IN PATIENTS AFTER SUCCESSFUL SURGERY FOR UNILATERAL CONGENITAL EAR ANOMALIES

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Laryngoscope 1992 (submitted).

ABSTRACT

It is generally assumed that auditory stimulation since birth is important for the proper development of the central auditory nervous system. Studies on animal ears occluded directly after birth show that the induced conductive hearing loss leads to deprivation (immaturity) of the auditory neural system. Whether auditory deprivation also occurs in man and therefore whether it may be considered as a contraindication to surgery in unilateral congenital middle ear anomalies and atresias, is the subject of the present study. Speech recognition during both monaural and binaural presentation was studied in patients with a unilateral congenital ear anomaly, following successful surgery.

In binaural speech recognition tests, the average results of the patient group (n=13) and a group of subjects with normal hearing proved to be comparable, although the standard deviation in the patient group was larger in one test (binaural fusion test). Using monaural presentation, the speech recognition of both ears for each patient was compared using bandpass filtered words and sentences with a competing noise. The average speech recognition score using bandpass filtered speech was 84 $\pm$ 8% and 77 $\pm$ 10% for the unoperated (normal) and operated ears, respectively. This difference was statistically significant. Speech in noise ratios of -5.6 $\pm$ 0.7 dB and -3.9 $\pm$ 1.6 dB were found in the normal and operated ears, respectively. This was also a statistically significant difference. Correlation analysis showed that the greater the pre-operative hearing loss, the poorer the operated ear performed.
It was concluded that in general, the speech recognition scores of the operated ears were satisfactory, but poorer than those of the normal ears, most probably owing to auditory deprivation. Binaural interactions were found. Therefore, the present results suggest that auditory deprivation is not a contraindication to surgery for unilateral congenital ear anomalies.

INTRODUCTION

The value of surgery for unilateral, major or minor congenital ear anomalies often has been questioned in the literature[1]. In the sixties and seventies, surgery was not recommended

because of poor long-term results and it was argued that one good ear was enough for normal speech and language acquisition. During recent years, studies on new surgical techniques and better predictors for successful surgery, based on the classification of the anomaly, have been published.[2,3,4,5] If the anomaly is mild, surgery is recommended in children aged 6 years and older.[2] The need for surgery in children with unilateral ear anomalies has become more urgent as a result of recent studies indicating that problems caused by unilateral hearing in children may have been underestimated.[6,7]

A problem that should be considered is whether or not a conductive hearing loss with a congenital onset leads to auditory deprivation. It is generally assumed that auditory stimulation is important for the proper development of the central auditory nervous system, particularly during certain critical periods in the development. From animal studies, it is known that a conductive hearing loss provoked just after birth, leads to an immature auditory system.[8,9] No human studies have been performed on the effect of auditory deprivation on speech recognition following successful surgery for unilateral congenital ear anomalies. On the other hand, several studies have been published on the negative effect of recurrent otitis media with effusion (OME) during early childhood on auditory perception[10-13] and on late onset auditory deprivation.[14,15]

The present paper addresses the effects of auditory deprivation on speech recognition in successfully operated unilateral, congenital anomalous ears. Poorer speech recognition in the operated ears when compared to the unoperated (normal) ears was taken as an indication of auditory deprivation resulting from the unilateral anomaly. For this purpose, speech recognition performance of the operated and normal ears of the patient group and within each patient were compared. Filtered speech and speech-in-noise tests were performed because speech recognition in quiet may be an insensitive measure due to the redundancy of the speech signal. Furthermore, such tests often are applied for assessment of central auditory function.[10,11,13,16] In addition to these monaural tests, binaural interaction tests using speech were performed.[13,17] Special attention was given to the effect of the duration of deprivation and the preoperative hearing loss on the performance of the operated ear.

METHOD

Subjects

Our study group was comprised of 13 patients with successfully operated unilateral congenital

ear anomalies; 6 patients had minor ear anomalies (congenital fixations of middle ear ossicles) and 7 patients had major ear anomalies (bony atresia of the meatus but with middle ear cavity and ossicles). The average age of the patients was 24 years, ranging from 10 to 45 years; four of them were female. The participants had to fulfil several criteria. First, the difference between the post-operative air-conduction pure tone thresholds for the two ears was not permitted to be more than 20 dB at 500, 1000 and 2000 Hz, and 25 dB at 4000 Hz. These criteria were based on binaural hearing experiments; binaural interactions have been found in ears even when they differed by as much as 25 dB.[17] Second, the difference between the masked bone-conduction thresholds for the two ears was not permitted to be more than 10 dB at the above-mentioned frequencies. This criterion served to exclude patients with asymmetrical sensorineural hearing losses. Furthermore, patients who had used a hearing aid on the anomalous ear were also excluded. A (symmetrical) sensorineural hearing loss was not an exclusion criterion; one patient had a mild high frequency hearing loss (thresholds of 40 dB at 4 kHz).

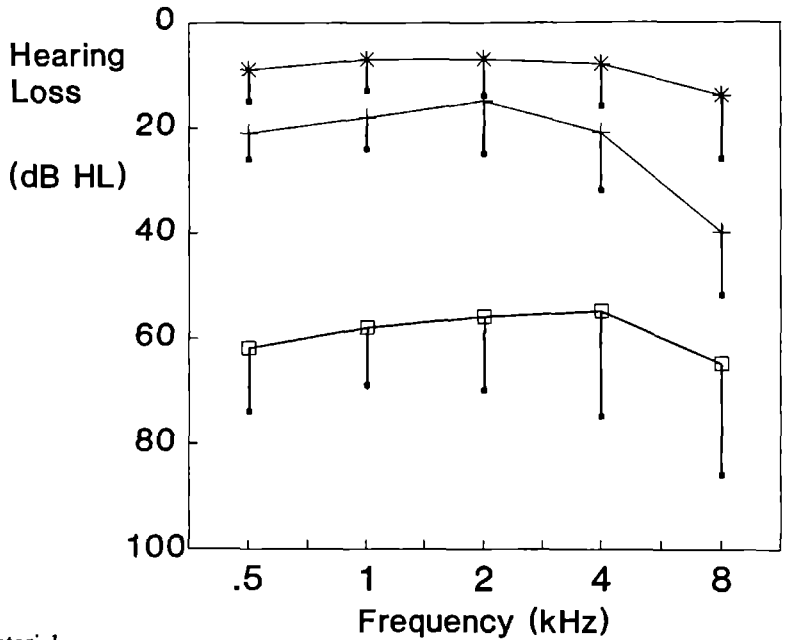
All the patients with congenital unilateral ear anomalies operated between 1980 and 1990 and who fulfilled these criteria, were tested. The surgery was performed at an average age of 20 years, ranging from 7 to 44 years. Long term test results are presented, obtained at least one year after the surgery. The mean time interval between the surgery and testing was 4.2 years, ranging from 1 to 9 years. To compare the short and long-term results, three of the patients were also tested within 1 year after the surgery, at 1, 3 and 7 months, respectively. In the statistical evaluations, only the long-term results were used.

The pre-operative and post-operative audiogram for the operated and the opposite, normal ear, averaged for all the patients, is shown in Figure 1. For most of the patients, a small air-bone gap remained after surgery.

The results of a questionnaire, used to gather the patients' opinion about the effect of surgery on their binaural hearing are presented in APPENDIX 1.

Ten subjects with normal hearing were also tested. All of them had thresholds of 15 dB HL or less bilaterally. The average age was 29 years, ranging from 24 to 38 years; four were female.

Figure 1. The average hearing loss as a function of frequency for the normal ears (*), anomalous ears before surgery (□) and after surgery (+). The standard deviations (+1 SD) are indicated.



Material

All the experiments were performed in a double walled sound-isolated room. Audiometry was performed using standard equipment (Interacoustics AC 5 audiometer with TDH 39P headphones with supra-aural cushions, MX-41/AR). Calibration was performed according to the ISO (International Standards Organization) 389 standard.[18]

Speech audiometry was performed using standard Dutch PB word lists consisting of 10 monosyllables (CVC words) recorded on professional quality tape. The special speech tests, i.e., the filtered speech test, the speech-in-noise test and the binaural fusion test, were also recorded on tape. The speech-in-noise test used was the sentence test in noise constructed by Plomp and Mimpen.[19] This test makes use of lists of 13 short conversational sentences of 8 or 9 syllables each and a steady state noise signal with a spectrum equal to the long-term spectrum of the sentences. The level of the noise is fixed, the level of the sentences is varied using an adaptive procedure (2 dB down after a correct response, 2 dB up after an incorrect response). The 50% recognition threshold obtained in this way has a standard deviation of approximately 1 dB.

For the filtered speech test and the binaural fusion test, PB word lists were bandpass filtered

in two bands; one band had a central frequency of 400 Hz and a bandwidth of 85 Hz (1/3 octave filter, Bruel & Kjaer 2121) and the other band had a central frequency of 2000 Hz and a bandwidth of 200 Hz (10% bandwidth filter, Bruel & Kjear 2121). Similar filtering was applied to a Dutch binaural fusion test by Pot and Verschuure.[20] To obtain equivalent filtered word lists, selection was made as described in the APPENDIX 2.

The calibration of the special speech tests was accomplished by adjusting the input level so that the frequent peaks of the speech signal reached 0 dB on the VU meter of the audiometer. The same procedure was followed for the noise in the speech-in-noise test.

Procedure

First, air- and bone-conduction thresholds were obtained followed by speech audiometry. Phoneme scores were obtained as a function of the presentation level of the word list.[21] Initially, the presentation level for each new list was lowered in steps of 10 dB from a starting point of 70 dB HL. When the phoneme score was below 100%, the step size was reduced to 5 dB. Thus, a speech audiogram was obtained. From the speech audiogram, the speech recognition threshold (SRT) was obtained, representing the displacement of the recognition-presentation level curve compared to the normal curve at 50% phoneme recognition.[20] The SRT values were expressed in multiples of 5 dB.

The difference in speech recognition between the operated and normal ears was determined using speech with and without noise and bandpass filtered speech. Each ear was tested separately. The speech stimuli were presented at a sensation level of 40 dB above the SRT of each ear. This was done to compensate for any residual hearing loss, if present. In a pilot experiment with subjects with normal hearing, this sensation level was generally stated to be a comfortable listening level. The speech recognition in quiet score (SQ) was the phoneme score 40 dB above the SRT; SQ was obtained twice. The speech-in-noise score was obtained using the test described by Plomp and Mimpen.[19] The noise was presented at 40 dB above the SRT, and the level of the sentences was varied using an adaptive procedure. The difference between the noise level and the 50% recognition threshold for the sentences was the speech-in-noise ratio (SN). The SN ratio was obtained 3 times for each ear.

The filtered speech score (FS) and the binaural fusion score (BF) were obtained from the experiment with the bandpass filtered words. Measurement was conducted as follows: first, the phoneme score of each ear was obtained while presenting both bands to the same ear

(diotic presentation). In this way, the FS score of each ear was obtained. Second, the low band was presented to one ear (at 40 dB above its SRT) and the high band to the other (at 40 dB above its SRT, dichotic presentation) and this measurement was repeated once with the bands reversed. The average score of both dichotic presentations was calculated. Binaural fusion was considered to have occurred when the phoneme score in the dichotic presentation was comparable to that in the diotic presentation. The BF score was defined as the dichotic score minus the highest diotic score for each subject. The FS and BF tests were administered twice during a given test session.

The binaural summation score was obtained as follows: the phoneme score was obtained using monosyllables presented binaurally at the SRT level (not at 40 dB above the SRT to avoid ceiling effects) of each ear. The binaural score was compared to the highest of the monaural scores at the SRT. The difference, binaural minus the highest monaural score, was the binaural summation (BS) score. The monaural scores and the binaural score at the SRT were administered twice. The BS score presented is the average value from both determinations.

The testing took approximately 50 minutes; before each test, the subjects were given instructions and allowed to practice the test until they were accustomed to performing the task. In all the tests, the normal ear was tested first. Therefore, if learning effects occurred, they were to the advantage of the results of the operated ear.

Data Analysis

Average speech recognition scores of the operated and the opposite, normal ear were tested for significance using the Student-t test. If the variances of the normal and operated ears were unequal (tested with the F test), the approximate method, as described by Cochran and Cox,[23] was used.

To study learning effects in the tests which were administered to each ear (speech-in-quiet and in noise, and filtered speech), a signed test-retest evaluation was performed. The average test-retest differences and the intra-subject standard deviations were calculated. Ceiling effects were encountered with the SQ; most of the patients had scores from 95 to 100%. The results of the test-retest evaluation are presented in Table 1. Using the Student-t test, it was found that none of the average differences for test-retest of SQ, SN or FS deviated significantly from zero; therefore, learning effects were not observed.

As it was our aim to compare the differences in test scores between the ears in each patient, the critical difference was used. Values for the 95% confidence level calculated from the standard deviations, representing the critical differences, are also presented in Table 1. If the difference between the ears for a particular test exceeded the critical difference, it was considered to be significant.

To investigate the strength of the relationship between the test scores and audiological data, correlational analysis was performed using Pearson's correlation coefficients.

Table 1. Test-retest differences

Measure	No. of Subjects	Mean of differences		Critical difference
		M	SD	
SQ (%)	15	-0.1	0.9	
FS (%)	15	-0.3	4.1	8.8
SN (dB)	15	-0.2	0.6	1.3

Note: SQ = speech-in-quiet score; SN = speech-in-noise score; FS = filtered speech score; M = mean; SD = standard deviation.

RESULTS

Audiometry

For all the patients, the pure tone average (PTA) of their air-conduction hearing thresholds at 500, 1000, 2000 and 4000 Hz was calculated. The average PTA values of the unoperated (normal) and operated ears, before (PTApre) and after (PTApost) surgery for all the patients are presented in Table 2. The improvement in PTA values as a result of the surgery varied from 13 to 50 dB. In addition, SRT values are presented in the same Table. The SRT and PTA values are in good agreement.

Binaural interaction

The average binaural summation (BS) scores for the operated patients and the subject sample with normal hearing proved to be comparable, 13.0 $\pm$ 7.0% and 12.0 $\pm$ 7.0%, respectively. The average binaural fusion (BF) score for the patients and subjects with normal hearing was 1.7 $\pm$ 7.0% and 2.9 $\pm$ 3.1%, respectively. The average values were comparable, but the

SD for the patients was twice as high.

Table 2. Group means and standard deviations for the average hearing loss (PTA) and speech reception threshold (SRT).

	PTA (dB HL)		SRT (dB)	
	M	SD	M	SD
preoperative	49	14	48	11
postoperative	19	5	21	8
normal ear	8	5	6	7

Monaural speech recognition

Table 3 presents the average filtered speech scores (FS), speech recognition scores in quiet (SQ) and in noise (SN) for the group of operated patients and subjects with normal hearing. For the patients, the average scores were based on the whole group, except for the SN scores. No SN scores for the two youngest patients are available because the test used is not suitable for subjects under 11 years of age. As a result, the SN score is the average score of 11 patients.

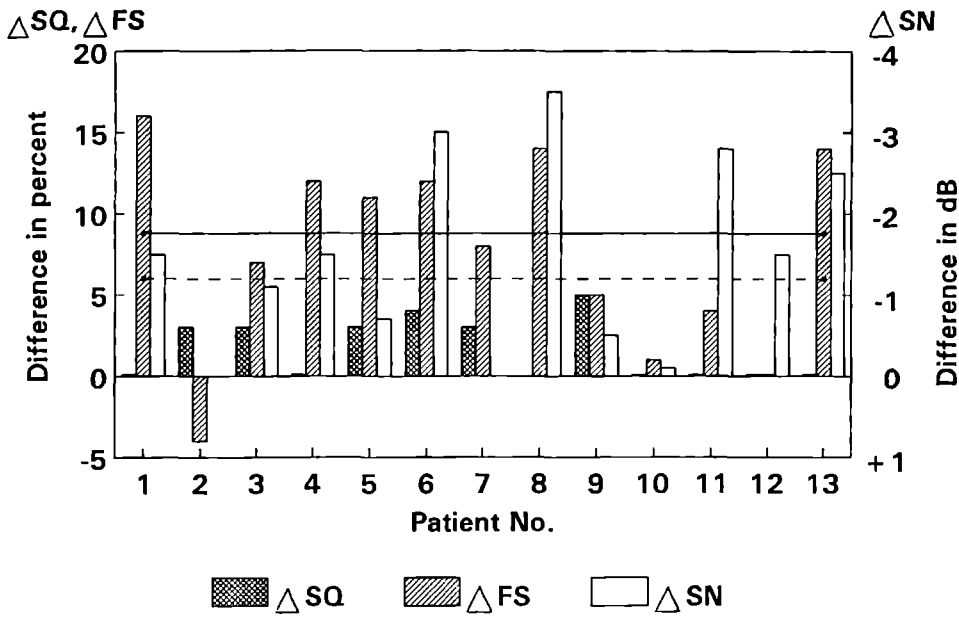
Table 3. Group means (M) and standard deviation (SD) for the speech-in-quiet score (SQ), the filtered speech score (FS) and the speech-in-noise score (SN).

	SD (%)		FS (%)		SN (dB)	
	M	SD	M	SD	M	SD
normal	99.8	0.8	83.9	8.1	-5.6	0.7
operated	98.3	2.4	76.7*	9.6	-3.9**	1.6
NH	100		85.5	8.5	-5.9	0.4

* $p < 0.05$, ** $p < 0.01$
note: NH = subjects with normal hearing.

The average SN score of the operated ears was significantly poorer than that of the normal ears: $p<0.01$ (Cochran-Cox test). For the FS score, a significant difference ($p<0.05$, t test) was also found. The scores for the patients' normal ears agreed well with those of the subjects with normal hearing.

Figure 2. The individual difference scores (normal minus operated ear) for the speech-in-quiet test (Δ SQ), the filtered speech test (Δ FS) and the speech-in-noise test (Δ SN). For patients nos. 2 and 7, no Δ SN values were available. The horizontal solid line indicates the critical difference for Δ FS, the dashed line the critical difference for Δ SN.



The scores of the operated ear were compared to those of the unoperated normal ear for each patient; the differences in the SQ, FS and SN scores (normal ear minus operated ear), called Δ SQ, Δ FS, Δ SN, were calculated. In Figure 2, the Δ SQ, Δ FS and Δ SN scores for each patient are presented. Note that Δ SN scores are negative.

Using the critical difference as given in Table 1, the FS score was poorer in 6 out of the 13 patients and the SN score was poorer in 7 out of the 11 patients older than 11 years. For three patients, none of the measures was significantly poorer in the operated ear.

Three patients were tested relatively shortly after surgery and a second time, 8 to 19 months later. Results are shown in Table 4. The Δ SQ of all 3 patients had improved; surprisingly, the Δ FS scores were poorer in the second measurement. It should be noted that the observed changes over time in the Δ FS and Δ SN for the three patients were small compared to the critical difference, which leads to the conclusion that no systematic improvements were found.

Table 4. Speech recognition scores from the initial measurement (within 7 months post surgery) minus those from the repeated measurement (more than 1 year post surgery) in 3 patients. By definition, a positive outcome for Δ SQ and Δ FS and a negative outcome for Δ SN means an improvement over time.

	Patient No.		
	1	2	3
$\Delta SQ_i - \Delta SQ_r$ (%)	+5	+3	+10
$\Delta FS_i - \Delta FS_r$ (%)	-5	-4	-5
$\Delta SN_i - \Delta SN_r$ (dB)	-0.2	-0.2	+0.4

Note: Δ = difference score, normal minus operated ear; SQ = speech-in-quiet score; FS = filtered speech score; SN = sound-in-noise score; indices i and r refer to initial and repeated measurement scores respectively.

Correlation analysis

The correlation coefficients between Δ SQ, Δ FS and Δ SN for all the patients were calculated. The coefficients were not statistically significant (see Table 5, rows 1-3). Correlations between the Δ SQ, Δ FS and Δ SN scores and the hearing thresholds were studied. For this purpose, the PTApre and PTApost of the normal ear minus that of the operated ear, called Δ PTApre and Δ PTApost, were calculated. Correlation coefficients for Δ SQ, Δ FS, Δ SN and for Δ PTApre, Δ PTApost are shown in Table 5, rows 4 and 5. To interpret the sign of the correlation coefficients, it should be noted that owing to their definition, Δ PTApre, Δ PTApost and Δ SN values are generally negative (or zero), whereas Δ SQ and Δ FS values are generally positive (or zero). Significant correlations were seen between the speech scores Δ SN and Δ FS, and the Δ PTApre values. As expected, the correlation between Δ SN and Δ PTApre was positive and that between Δ FS and Δ PTApre

negative. The correlations between the Δ SN, Δ FS and Δ PTApost values were also significant. The correlations between Δ SQ, and Δ PTApre and Δ PTApost scores were not significant. Similarly, no significant correlations were found between the Δ SQ, Δ FS, Δ SN scores, and age at surgery, or the time interval between the surgery and testing (test interval); see Table 5, rows 6, 7.

Table 5. Correlation matrix of speech recognition scores, audiological data and other data.

	Δ SQ	Δ FS	Δ SN
Δ SQ	1.0		
Δ FS	-0.16	1.0	
Δ SN	0.26	-0.45	1.0
Δ PTApre	0.34	-0.58*	0.82**
Δ PTApost	0.15	-0.64*	0.72**
age at surgery	0.18	0.45	-0.28
test interval	0.35	-0 49	0.34

* $p < 0.05$, ** $p < 0.01$

note: Δ = difference in scores, normal minus operated ear; SQ = speech-in-quiet score; FS = filtered speech score; SN = speech in noise score; PTApre = preoperative average hearing loss; PTApost = postoperative average hearing loss; test interval = time interval between surgery and testing.

DISCUSSION

Although the patient group was of limited size, some preliminary conclusions on speech recognition can be drawn. The BS scores of the patients and subjects with normal hearing were comparable. The average BF scores in both groups were also comparable, although the SD in the patient group was larger. At present, there is no explanation for the difference in SD values. It is concluded that the binaural scores of the patient group compare well with those of the subjects with normal hearing. The satisfactory binaural scores in the patients were in agreement with the patients' comments regarding better intelligibility after surgery, especially in noisy situations (see APPENDIX 1).

Table 3 shows that the average FS and SN scores for the operated ears were poorer than those for the unoperated (normal) ears. This cannot be due to poorer hearing; if a hearing loss was still present after surgery, it was compensated for by adjusting the presentation level of the speech. Since patients with asymmetrical sensorineural hearing loss were excluded from the study, auditory deprivation might have caused the poorer FS and SN test scores of the operated ears.

For statistical analysis, difference scores Δ FS, Δ SQ and Δ SN were introduced. By taking difference scores, the results of the operated ears were presented relative to those of the normal ears; therefore, effects of such variables as verbal intelligence and auditory skills were ruled out.

The correlations between Δ SQ, Δ FS and Δ SN were low. Low correlations between speech-in-noise and speech-in-quiet tests have often been found.[22] Concerning the Δ SQ, it should be noted that ceiling effects may have negatively influenced the correlations. For 8 patients, the SQ value of both ears was 100%.

Correlations between the Δ SN and Δ FS scores and Δ PTApre were statistically significant. This result suggests that the larger the air-bone gap before surgery, the poorer the operated ear performed. The significant correlation between Δ SN and Δ FS, and the Δ PTApost may be ascribed to the high, significant correlation between the Δ PTApre and Δ PTApost values ($\rho = .80$, $p < 0.01$). The dependency between Δ PTApre and Δ PTApost is plausible; a high Δ PTApre value (large pre-operative air-bone gap) indicates a more severe type of anomaly and therefore a greater chance of an air-bone gap remaining after surgery (relative high Δ PTApost value). No significant correlation was seen with either between the Δ SQ value and the Δ PTApre or Δ PTApost (see Table 5). Ceiling effects may have played a role.

The correlations between the Δ SN, Δ FS and Δ SQ values, and the time interval between the surgery and testing (test interval) were not significant. This was in agreement with the results of repeated measurements in 3 patients over time as presented in Table 4; no systematic improvements were found. The absence of a significant improvement over time may be explained by the fact that sensory deprivation from birth (immaturity) may be an irreversible phenomenon. The correlations between the age at surgery and Δ SQ, Δ FS and Δ SN were not significant either.

In conclusion, the present results indicate that as in animal studies, auditory deprivation from birth may lead to an irreversible auditory processing deficit. Although the results of the

operated ears on the FS and SN tests were generally poorer than those of the normal ears, the operated ear should be considered far from useless because it contributed to speech recognition, both objectively and subjectively (see APPENDIX 1). Therefore, auditory deprivation may play a role, but it is not a contra-indication for surgery in patients with unilateral congenital conductive hearing loss.

APPENDIX 1

A questionnaire was used to gather the patients' opinions about the effect of the surgery on their binaural hearing. The patients were asked whether they had experienced any change in hearing, directional hearing and hearing in noisy situations as a result of the surgery and whether they considered the result to have been worth the surgery.
All 13 patients reported an improvement in their hearing after surgery; 9 of them reported better directional hearing and 11 experienced better hearing in noisy situations. Twelve out of the 13 patients stated that the benefits of the improved hearing had made it worth undergoing the surgery. The remaining patient had been suffering from recurrent external otitis in the operated ear since the surgery.

APPENDIX 2

To select equivalent word lists for the determination of the filtered speech score and the binaural fusion score, 15 different (standard) PB word lists, filtered as described in the Methods/Materials section, were presented to five subjects with normal hearing in a separate experiment. The filtered lists were presented to the right ear at 40 dB above the SRT. For each subject, the overall average score was calculated and the lists for which the scores deviated by no more than plus or minus 2 phonemes from the average value, were selected. Combining the selected lists of all 5 subjects produced 6 common lists, which were used for the measurements. The 6 lists were used in a random order, 2 lists for each measurement condition, i.e., the filtered speech score of the right and left ear and the binaural fusion score.

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CHAPTER VI

SUMMARY AND CONCLUSIONS

This thesis describes the surgical findings and results in patients with a major or minor congenital ear anomaly. The two central themes were to establish a classification system for the many forms of major and minor anomalies and to evaluate the value of surgical treatment. During various stages, many questions were raised and where possible answered in the light of a mainly retrospective study on 144 operated ears with a minor ear anomaly and 55 operated ears with a major anomaly.

Chapter II introduces a classification for minor congenital ear anomalies. The classification enabled us to gain an insight into the frequency and prevalence of the various types of anomaly in our own series of operated ears. It was also possible to apply the classification to series of operated ears reported in the literature and to compare the results to ours.
The classification is not based directly on the severity of the anomaly, but instead it distinguishes between whether or not the stapes footplate is fixed and between whether or not an associated anomaly of the ossicular chain is present. As a consequence, the surgeon can decide whether stapes replacement surgery is justified, because opening the vestibulum involves a higher risk of complications than other reconstructive surgical procedures.
The higher risk of complications was reflected in the results obtained in the early years of middle ear surgery in our series. It is also important to realize that the frequency of stapes fixation is about 70% and the frequency of aplasia of the oval window about 10%.
Despite the application of a simple and clear classification, there are very few large series in the literature which provide comparable data. Nevertheless, it was possible to compare congenital stapes ankylosis, and the results from the various series were found to be very similar. Comparison with other series reported in the literature showed that so far, we have compiled the largest consecutive series of operated ears. It has been our aim to present the preoperative and postoperative hearing thresholds in an easily surveyable manner for each individual ear, as mean values for the frequencies 500, 1000 and 2000 Hz. In many cases where strikingly positive or negative results were obtained, reports on the pathological findings and the surgical reconstruction technique were available. Presenting the individual results in this way together with the mean hearing gain for the total group of operated ears, provides a facilitating overview of the technical possibilities. For example, it became clear that a considerable hearing gain can be achieved, although not as often as is usually encountered after stapes surgery in patients with otosclerosis. However, in view of the

complex nature of the anomaly, this is not an unexpected result.
When the total series was analysed in chronological order, we consistently found a small perceptive component of 10 dB to 15 dB on average in the preoperative audiogram and also in the postoperative audiogram. This inner ear component influenced the ultimate level of hearing gain that could be achieved. The analysis also gave the impression that the introduction of preoperative patient selection soon after the initial stages, led to the gradual exclusion of ears with a large mixed hearing loss from reconstructive surgery. A major inner ear component can be an expression of an associated inner ear anomaly, which may have an unfavourable influence on the operative results. In the early years of ear surgery in our series, some of the patients had disappointing results. In the following period, there were no great disappointments regarding the ultimate gain in hearing level. In our opinion, the inclusion and exclusion criteria listed in the introduction, were partly responsible for this.

Attention is drawn to congenital aural atresia, synonymous with major ear anomaly, by presenting a small adjunctive series whose operative results have been published previously. For this group of ears, a new supplementary classification was proposed in an earlier paper, which proved not only easier to use but also made meaningful predictions regarding the chance of achieving improvement in the patients' hearing. This classification, like most other classifications, is based on distinguishing the severity of the anomaly.
The long-term results are presented for the major and minor ear anomalies. The value of the surgical techniques discussed in this thesis is not only evaluated using the short-term results, but also the long-term results.
We were not able to answer the question as to why the results in class IIB were so often below par. Marquet described similar results for his type II, while Jahrsdoerfer reported that the results declined sharply in patients with a score below 7 or 8 on the basis of his radiological classification with scores from 1 to 10. Our latest results in class IIB are favourable, which is probably the result of the insight that the middle ear and epitympanum in these ears is narrower and can easily lead to refixation through bony growth, particularly in the epitympanum. At present, there is no hesitation to remove the incus and malleus head and increase the height of the stapes using a homologous malleus head if this is indicated. It can be expected that this modification to the surgical technique will produce a socially acceptable hearing threshold in a sufficient number of class IIB ears with congenital atresia.

Our as yet unpublished experience justifies these expectations.

The analysis of such a series of complicated congenital ear anomalies will benefit from clear classification. Therefore new classifications are proposed in Chapters II and III and subsequently applied to our series. This classification produced a framework which offers a better understanding of the pathology and prospects, risks and impossibilities of surgery.

Another way of arranging the data is based on the presence of a syndromal diagnosis, as discussed in Chapter IV. In 25% of the minor congenital ear anomalies, the patient is suffering from a syndrome. A syndromal diagnosis has predictive value regarding the congenital anomaly present and consequently, the chance of being able to improve the patient's hearing using surgery. Such an analytical approach goes far beyond the importance of presenting a simple case report. A syndromal diagnosis will often also form a guide to a genetically-determined background of the disorder. We have found that establishing a syndromal diagnosis can contribute to minimizing the risk of complications during surgery and that some special ear anomalies can be recognized preoperatively and thus prepare the surgeon for what he is likely to encounter.
A number of publications have appeared on this subject outside the monograph based on this series of ears. For instance, it is now generally accepted that the disorder X recessive progressive mixed deafness with stapes gusher during surgery can be recognized in advance on the basis of audiological and radiological criteria. In addition, family studies have made it possible to perform gene coupling, which has brought carrier detection within reach.

Future developments

It has become clear that syndromes with a branchiogenic component are often associated with a serious anomaly of the middle ear, or of the inner ear such as in the branchio-oto-renal syndrome. Particularly in the more complicated group, there is presently a great need for new and improved surgical techniques. Recent, incidental examples of new techniques may provide the necessary progression. For example, the results with malleo-vestibulopexy, in which a piston is placed between the vestibulum and the tympanic membrane, can be expected to improve now that it has become clear how it can be surgically prevented that the

piston becomes disengaged from the malleus handle.
More insight has also been gained into how interpositioning can be performed between the tympanic membrane and the vestibulum in ears in which the malleus handle is missing or inadequate. The first and foremost example is the procedure in which the piston is fixed to the chorda tympani, while homologous cartilage is placed between the tympanic membrane and the chorda. Another technique has been described in which a donor incus fixed in a sheet of fascia, is implanted under the tympanic membrane, while the fascia is fixed between the membranous and bony aural canal. In this way, second-stage interpositioning is possible by fixing a piston to the implanted incus, between the vestibulum and the tympanic membrane. In cases with craniosynostosis syndromes, the epitympanum is smaller than usual and such ears therefore seem to benefit from the improved techniques of vestibulo-malleopexy.
Laser surgery which has recently been introduced for middle ear surgery, may be useful for the treatment of the typical anomaly encountered in the Treacher Collins syndrome. In this syndrome, the stapedial head of an in itself mobile footplate, may tend to become fixed or to lean too hard against the (bony) facial canal. Laser surgery may also prove to be useful for other forms of unusual fixation of the ossicular chain.

We were not able to answer the question as to whether sufficient hearing gain can be achieved in a sufficient number of patients with aplasia of the oval or round windows and it is not clear how great the risk is of damaging the inner ear or vestibulum. However, there are indications that good results have been achieved by other authors in small consecutive series. This shows that further refinements are being made to ear surgery techniques, particularly for congenital anomalies and that greater success can be expected in the future, also for patients who on the grounds of selection criteria obtained from the classification, are presently excluded from surgery. The recent acquisition of the bone anchored hearing aid as an alternative treatment, particularly for cases with bilateral anotia, should not go unmentioned.

The final chapter presents the results of hearing measurements from patients operated on because of a unilateral major or minor ear anomaly. An ear with a congenital anomaly of the aural canal and/or middle ear is potentially a good ear. In many cases, it can be expected that only very slight or no deprivation is associated with this type of congenital hearing

impairment because conduction via bone conduction of the affected, but particularly via the unimpaired contralateral ear, provides adequate stimulation of the inner ear, auditory nerve, central nuclei and cortex. The test results showed that a successfully operated ear can make a valuable contribution, especially in noisy circumstances, to binaural hearing and to the sensory integrated hearing of the operated ear and to the ear with normal hearing since birth.

Seeking answers to a scientific question generally gives rise to new questions and this study was no exception. We have aimed to present our data on the surgical approach to the problem of major and minor congenital anomalies in a systematic and integral manner.
On the basis of the size of this series and the incorporation of all the successive operated ears, clinical evidence has been provided that in experienced hands, otological surgery for major or minor congenital anomalies is worthwhile in relation to the complications reported. Moreover, further improvement of the results for various types of anomaly are within our reach through further refinement of new and existing surgical techniques.

SAMENVATTING EN CONCLUSIES

In de voorgaande hoofdstukken van dit proefschrift zijn de bevindingen en resultaten tijdens chirurgische behandeling van congenitale minor en major afwijkingen van het oor besproken. Het centrale thema is daarbij enerzijds een inventarisatie en classificatie van de vele vormen van minor en major oor anomalieen en anderzijds het onderzoeken van de waarde en de mogelijkheden van de chirurgische behandeling. Stapsgewijs komen hierbij vele vragen aan de orde, die zomogelijk beantwoord worden aan de hand van een voornamelijk retrospectieve studie van 144 geopereerde oren met een minor oor anomalie en 55 geopereerde oren met een major oor anomalie.

In hoofdstuk II wordt een classificatie voor congenitale minor anomalieen van het oor voorgesteld. Voor het eerst wordt nu inzicht verschaft in de frequentie van voorkomen van de verschillende typen anomalieen in de eigen serie geopereerde oren. Door toepassen van de classificatie op series geopereerde oren uit de literatuur werd vergelijking van resultaten mogelijk. Bij deze classificatie is niet direct naar de ernst van de anomalie ingedeeld, maar is onderscheid gemaakt tussen het al of niet gefixeerd zijn van de stapesvoetplaat en het al of niet aanwezig zijn van andere geassocieerde anomalieen van de gehoorbeenketen. Gevolg hiervan is het al of niet plaatsvinden van een stapesvervangende operatie, die vanwege het openen van het vestibulum een hogere kans van complicaties kent vergeleken met andere reconstructieve operaties van het middenoor. Deze hogere kans op complicaties voor stapesvervangende chirurgie werd ook gevonden in de eerste jaren van deze serie geopereerde middenoren. Het is derhalve van belang te beseffen dat de frequentie van stapesfixatie in de orde van 70% bedraagt en de frequentie van aplasie van het ovale venster in ongeveer 10% van de oren gevonden wordt. Ondanks het toepassen van een eenvoudige en heldere classificatie valt op dat maar weinig grotere series uit de literatuur beschikbaar zijn voor vergelijking van de resultaten. Voor de congenitale stapes ankylose was deze vergelijking evenwel goed uit te voeren, waarbij is gebleken dat de resultaten van de verschillende series elkaar weinig ontlopen. Bij deze vergelijking met de eerdere resultaten uit de literatuur valt de grote omvang van deze achtereenvolgend geopereerde oren direct op. Daarenboven is deze serie uniek vanwege de voor ieder oor individueel gepresenteerde preoperatieve en postoperatieve gehoordrempels, weliswaar als gemiddelde over de frequenties 500, 1000 en 2000 Hz. Veelal kon voor opvallende resultaten, zowel in positieve als in negatieve zin, een omschrijving van aangetroffen pathologie en uitgevoerde reconstructieve chirurgie gegeven

worden. Het op deze wijze presenteren van individuele resultaten naast gemiddelde gehoorwinsten voor een hele groep geopereerde oren geeft een veel reeler beeld van de mogelijkheden. Aldus valt op dat vaak een aanzienlijke gehoorwinst te bereiken is, alhoewel niet zo vaak als gebruikelijk is bij stapeschirurgie in geval van otosclerose. Gezien de gecompliceerdheid van de ooranomalie ligt dat ook niet in de lijn der verwachting.
Wanneer de serie over de loop der jaren geanalyseerd wordt valt voorts op dat er steeds een kleine perceptieve component van gemiddeld 10 dB tot 15 dB bestaat in het preoperatieve, maar ook in het postoperatieve audiogram. Een dergelijke binnenoorcomponent beinvloedt de mogelijkheden van de uiteindelijk te bereiken gehoordrempel. Verder ontstaat de indruk dat tengevolge van preoperatieve selectie na de eerste beginperiode het aanvaarden van reconstructieve chirurgie voor oren met grotere gemengde gehoorverliezen steeds meer is ontweken. Een belangrijke binnenoor component kan inderdaad uiting zijn van een tegelijkertijd bestaande binnenooranomalie die de resultaten van chirurgie ongunstig zouden kunnen beinvloeden. In de eerste begin jaren van ooroperaties bij patienten uit deze studie zijn enkele teleurstellingen genoteerd. Grote teleurstellingen voor wat betreft het uiteindelijk resultaat op de gehoordrempel zijn evenwel in de hierop volgende periode uitgebleven. Het hanteren van de in de inleiding genoemde in- en uitsluitingscriteria heeft daartoe naar onze mening bijgedragen.

Aan de congenitale gehoorgangatresie, synoniem voor major ear anomaly, wordt aandacht besteed door presentatie van een vervolgserie op een in delen eerder gepubliceerde kleinere serie oren aangaande resultaten van gehoorverbeterende chirurgie. Voor deze groep oren werd eerder een nieuwe aanvullende classificatie voorgesteld die niet alleen eenvoudig te hanteren bleek, maar ook betekenisvol bij het voorspellen van de kans op een gunstig resultaat van de uiteindelijke gehoordrempel. Deze classificatie geeft evenals de meeste andere classificaties als vermeld in de inleiding een indeling naar ernst van de anomalie.
Voor zowel de minor als major congenital ear anomalies worden de lange termijn resultaten gemeld. De waarde van de in dit proefschrift besproken chirurgie wordt immers niet alleen bepaald door de korte termijn resultaten, maar vooral ook de resultaten op langere termijn. Onbeantwoord blijft de vraag waarom de resultaten zo vaak voor de klasse II B achterbleven. Eenzelfde ervaring vond Marquet in zijn type II, terwijl ook Jahrsdoerfer op basis van zijn radiologische classificatie met een score van 1 tot 10 aangeeft dat onder een score van 7 a

8 de resultaten sterk teruglopen. Wij menen dat onze jongste gunstige resultaten met klasse II B het gevolg zijn van het inzicht dat het middenoor en epitympanum in deze oren nauwer zijn en zo eerder een benige refixatie kan ontstaan door botaangroei met name in het epitympanum. Op dit moment wordt veel eerder besloten de incus en de hamerkop uit te nemen en de mobiele stapes op te hogen met een gemodificeerde allologe hamerkop. Het is de verwachting dat door deze aanpassing van de toe te passen chirurgische techniek de II B congenitale atresie oren alsnog voldoende vaak tot een sociaal aanvaardbare gehoordrempel gebracht kunnen worden. De hierover thans nog niet gepubliceerde ervaringen rechtvaardigen het in deze koesteren van hoopvolle verwachtingen.
Een goede analyse van een dergelijke gecompliceerde serie congenitale ooranomalieen is gebaat met een heldere rangschikking, reden waarom nieuwe classificaties in hoofdstuk II en III zijn voorgesteld en voor deze serie toegepast. Deze classificering maakt het mogelijk een raamwerk te verschaffen, waarbinnen een beter begrip van de pathologie en mogelijkheden, risico's en onmogelijkheden van chirurgie verkregen kan worden.

Een andere wijze van rangschikken is te baseren op een aanwezige syndroom diagnose, zoals dat in hoofdstuk IV ter sprake komt. In 25% van de congenitale minor ear anomalieen blijkt er bij de patient sprake te zijn van een syndroom. Deze syndromale diagnose heeft een voorspellende waarde ten aanzien van de te verwachten oor anomalie en de daaruit volgende kansen op gehoorsverbetering na chirurgie. Een dergelijke analytische benadering overstijgt verre het belang van een eenvoudige casus beschrijving. Een syndromale diagnose wijst bovendien vaak de weg naar een genetisch bepaalde achtergrond van het ziektebeeld. Het blijkt inderdaad zo te zijn dat het vaststellen van een syndromale diagnose kan bijdragen tot verkleinen van de kans op complicaties bij chirurgie en dat sommige bijzondere ooranomalieen preoperatief herkend kunnen worden en daarmee de problemen voor de oorchirurg kunnen verkleinen. Eerder verschenen buiten de monografie gebaseerd op deze serie oren een aantal publicaties over dit onderwerp. Zo is het thans algemeen aanvaard dat het ziektebeeld van de X-recessieve progressive mixed deafness met een stapesgusher tijdens stapes chirurgie tevoren herkend kan worden op audiologische en radiologische criteria. Bovendien is het door familiestudies mogelijk geworden het gen te koppelen en daarmee komt carrier detectie op termijn binnen handbereik.

Het is duidelijk geworden dat syndromen met een branchiogene component vaker gepaard gaan met een ernstige anomalie van het middenoor en zoals bij het Branchio-Oto-Renaal syndroom met anomalieen van het binnenoor. Juist in die meer gecompliceerde groep is thans behoefte aan verbeterde en nieuwe chirurgische technieken. Recente incidentele voorbeelden van nieuwe technieken kunnen mogelijk hier de gewenste verbetering brengen. Zo is bijvoorbeeld te verwachten dat de resultaten van malleo-vestibulo-pexie, waarbij een piston tussen vestibulum en trommelvlies geplaatst wordt zullen verbeteren nu duidelijk wordt hoe een uitstoten van de aan de hamersteel gefixeerde piston chirurgisch voorkomen lijkt te kunnen worden.

Ook ontstaat inzicht hoe in oren zonder een aanwezige of bruikbare hamersteel een interpositie tussen trommelvlies en vestibulum verricht kan worden. Allereerst is er het voorbeeld waarbij de piston aan de chorda tympani gefixeerd kan worden terwijl autoloog kraakbeen geplaatst wordt tussen het trommelvlies en de chorda. Daarnaast is een techniek gemeld waarbij een donor incus gefixeerd in een fascielap onder het trommelvlies geimplanteerd wordt terwijl deze fascie gefixeerd wordt tussen de vliezige en benige gehoorgang. Aldus is ook in tweede tempo een interpositie tussen vestibulum en trommelvlies mogelijk door fixatie van de piston aan de geimplanteerde incus.

Bij craniosynostose syndromen is het epitympanum kleiner dan gebruikelijk en dergelijke oren lijken daarom in het bijzonder te kunnen profiteren van deze verbeterde technieken van vestibulomalleopexie.

Laserchirurgie, sedert kort ook toegepast in het middenoor, lijkt van nut te zullen zijn om de bij het Treacher Collins syndroom typische voorkomende afwijkingen te behandelen. Immers het stapeskopje van een op zich mobiele voetplaat neigt bij dit syndroom nog wel eens gefixeerd te zijn of al te zeer aan te leunen tegen het al of niet benig beklede facialis kanaal. Laser chirurgie is mogelijk ook nuttig inzetbaar bij andere ongewone fixaties van de gehoorbeenketen.

Onbeantwoord is nog de vraag of bij aplasie van het ovale of ronde venster voldoende vaak een voldoende grote gehoorwinst geboekt kan worden en onduidelijk is nog hoe groot het eventuele risico is op schade aan het binnenoor en evenwichtszintuig. Echter ook hier zijn

er voortekenen dat goede resultaten in kleine opeenvolgende series door anderen bereikt kunnen worden. Daarmee is aangegeven dat verdere verfijningen van oorchirurgie, in het bijzonder voor congenitale anomalieen nog gaande zijn en daarmee ook nog hoop gegeven kan worden op nieuwe successen in de toekomst, ook voor die patienten die thans op grond van selectiecriteria volgend uit de classificatie niet voor chirurgie in aanmerking komen. Ook de recente verworvenheid van de bone anchored hearing aid als alternatief, vooral in de gevallen met een bilaterale anotie mag hier niet onvermeld blijven.

Als afsluitend hoofdstuk worden de resultaten van gehoormetingen gepresenteerd voor personen geopereerd aan een unilaterale congenitale minor of major oor anomalie. Een oor met een congenitale anomalie van de gehoorgang en/of het middenoor is potentieel een goed oor. In het algemeen kan een veel geringere of zelfs geen deprivatie verwacht worden bij deze typen aangeboren slechthorendheid doordat voortgeleiding via de beengeleiding van het aangedane, maar vooral ook van het goed horende contralaterale oor, voldoende stimulatie geeft van binnenoor, gehoorzenuw, centrale kernen en cortex. Het is gebleken uit deze gehoormetingen dat het met succes geopereerde oor een nuttige bijdrage kan leveren aan horen, vooral in rumoerige omstandigheden, ook wanneer het gaat om binauraal horen en het zintuiglijk geintegreerde horen van het geopereerde oor en het reeds van geboorte af goed horende oor.

Het oplossen van een wetenschappelijk vraagstuk roept doorgaans nieuwe vragen op en deze studie is hierop geen uitzondering. Deze hier gepresenteerde studie is uniek door zijn omvang en systematische, integrale aanpak van het chirurgische probleem van de congenitale minor en major anomalieen.
Samenvattend kan geconcludeerd worden dat door de grootte van deze serie en het betrekken van alle opeenvolgend geopereerde oren de overtuiging wordt gegeven dat gehoorverbeterende chirurgie in met deze oorproblematiek vertrouwde en ervaren handen zinvol is in relatie tot de gerapporteerde complicaties. Daarenboven lijkt een verdere verbetering van de resultaten voor sommige typen anomalieen bereikbaar door een verdere verfijning van en nieuwe mogelijkheden in de oorchirurgie.

Dankwoord

Dit proefschrift kwam tot stand dankzij de medewerking en inzet van velen aan wie ik veel dank verschuldigd ben. In het bijzonder wil ik dank zeggen aan:

Prof. Dr. P. van den Broek, mijn promotor, die mij tijdens mijn opleiding aanmoedigde en de mogelijkheden gaf om deze studie te verrichten.

Dr. C.W.R.J. Cremers als stuwende kracht achter deze studie. Vooral dankzij zijn enthousiasme en onvermoeibare inzet en hulp kon deze studie door mij worden voltooid.

Dr. Ir. A.F.M. Snik dank ik voor de bijzonder prettige en vruchtbare samenwerking bij het laatste deel van dit proefschrift.

Mevr. M.W. Helsper-Peters voor haar hulp bij de tekstverwerking gedurende de studie en afwerking van het proefschrift.

G.M. van de Meerakker, mijn associé, die mij het afgelopen jaar voldoende ruimte kon laten in onze drukke dagelijkse praktijk om dit proefschrift af te ronden.

Tenslotte mijn vrouw Anneke die geduldig moest toezien hoe ik mij inspande om deze dissertatie te schrijven.

Curriculum vitae

De auteur van dit proefschrift werd geboren op 7 juni 1958 te Velp. Na het behalen van het diploma atheneum β aan het Rhedens Lyceum te Velp, studeerde hij 1 jaar Civiele techniek aan de Technische Universiteit Delft. In 1978 ving hij aan met de studie Geneeskunde aan de Katholieke Universiteit Nijmegen. Op 28 juni 1985 behaalde hij zijn artsexamen en aansluitend van juli 1985 tot october 1986 vervulde hij zijn militaire dienstplicht als reserve officier arts op de afdeling Keel-, Neus- en Oorheelkunde van het Militair Hospitaal Dr. A. Mathijsen in Utrecht. Van October 1986 tot februari 1987 werkte hij als AGNIO op de afdeling Keel-, Neus- en Oorheelkunde van het Academisch Ziekenhuis Nijmegen, waar hij van 1 februari 1987 tot 1 februari 1991 werd opgeleid tot Keel-, Neus- en Oorarts (opleider: prof. dr. P. van den Broek). Sedertdien is hij werkzaam als Keel-, Neus- en Oorarts in het Carolus-Liduina Ziekenhuis te 's Hertogenbosch in associatie met G.M. van de Meerakker. Hij is getrouwd met Anneke Lammers en zij hebben een nu 1 jaar oud dochtertje, Emma.

Stellingen

1. Congenitale major en minor anomalieën van het oor dienen afzonderlijk geclassificeerd te worden. Voor beide groepen is een verdere classificering mogelijk.

(dit proefschrift)

2. Classificering van congenitale minor anomalieën op grond van chirurgische bevindingen lijkt beter bruikbaar dan classificering op grond van embryologische ontwikkelingsstoornissen.

(dit proefschrift)

3. Een kenmerkende congenitale middenoor anomalie bij het Treacher Collins syndroom is een naar het facialiskanaal neigende of zelfs daarmee gefixeerde stapes suprastructuur.

(dit proefschrift)

4. Deprivatie van het auditieve systeem speelt een rol bij unilaterale congenitale anomalieën van het oor, doch is geen contraindicatie voor chirurgische behandeling gezien de postoperatieve spraakverstaanscores.

(dit proefschrift)

5. Bij systemische antibiotische behandeling van maligne otitis externa is monotherapie onvoldoende.

6. Behandeling van allergische rhinitis voor boom- en graspollen door middel van hyposensibilisatie met gepolymeriseerde allergenen heeft voordelen ten opzichte van behandeling met geadsorbeerde allergenen.

7. Volgens de huidige inzichten moet het vitamine D_3 derivaat calcipotriol gezien worden als een eerste keuze therapie bij lichte tot matig ernstige psoriasis.

8. Het gebruik van thiaziden ter voorkoming van nierstenen bij hypercalciurie is vooral geindiceerd bij de renale vorm van hypercalciurie en minder geschikt bij behandeling van absorptieve vormen van hypercalciurie.

9. Intraoperatieve echografie van de galwegen is minstens zo betrouwbaar als cholangiografie voor beoordeling van choledochuspathologie.

10. Decentrale opwekking van electriciteit met gelijktijdig gebruik van resterende warmte, de zogenaamde warmte - krachtkoppeling, is een belangrijke energiebesparende techniek.

11. Gezien de veranderde internationale verhoudingen en om maatschappelijke en economische redenen heeft in Nederland een beroepsleger de voorkeur en dient de militaire dienstplicht te worden afgeschaft.

Stellingen behorend bij het proefschrift " Major and minor congenital anomalies of the ear. Classification and surgical results."

Vught, december 1992
E. Teunissen