American Journal of Otolaryngology and Head and Neck Surgery

Research Article
Published: 06 Apr, 2021

Minor Malformations of the Middle Ear

*Rim Zainine**

Department of ENT, La Rabta Hospital, Tunisia

Abstract

Introduction: Minor malformations of the middle ear are abnormalities that almost exclusively involve the middle ear and ossicles, while the external ear is normal.

Objectives: To report the clinical and radiological features of this disease, describe the therapeutic management and evaluate treatment outcomes.

Material and Methods: Retrospective study on 17 patients (20 ears) with a minor malformation of the middle ear, treated between 1992 and 2010.

Results: The mean age of the patients of this series was 32 years. According to the classification proposed by Teunissen and Cremers, these malformations were classified as Type I in 5 cases, IIa in 5 cases, IIb in 1 case, IIIa in 4 cases, IIIb in 2 cases, IVa in 2 cases and IVb in 1 case. Sixteen patients underwent exploration of the tympanic cavity with a successful surgical result in 9 cases.

Conclusion: The treatment of minor malformations of the middle ear is surgical and management depends on whether the stapes footplate is mobile or fixed.

Keywords: Malformations; Middle ear; Surgery

Introduction

Minor malformations of the middle ear, also called minor aplasia or ossicular dysplasia, are very rare abnormalities almost exclusively affecting the middle ear and ossicles, while the external ear is normal. They can be either unilateral or bilateral. The diagnosis is suspected on the basis of clinical findings (personal history and clinical interview) and audiometry and confirmed intraoperatively during exploration of the tympanic cavity. This article reports the clinical and radiological features of minor malformations of the middle ear, describes the therapeutic management and evaluates the results of treatment.

Material and Methods

This retrospective study was based on 17 patients (20 ears) with a minor malformation of the middle ear followed in the Otorhinolaryngology and maxillofacial surgery department of La Rabta hospital in Tunis, during the 19-year period from January 1st, 1992 to December 31st, 2010. Computed tomography of the petrous temporal bones was performed in only 6 patients, as CT has become a part of routine practice only during recent years. Sixteen patients underwent endoscopic exploration of the tympanic cavity. Lesions were classified according to the classification proposed by Teunissen and Cremers [1] (Table 1).

Results

Patients in this series had a mean age of 32 years (range: 11 to 60 years) with a female predominance (sex ratio of 0.4, i.e. 12 females for 5 males). According to the classification proposed by Teunissen and Cremers, abnormalities were classified as type I in 5 cases, type IIa in 5 cases, type IIb in 1 case, type IIIa in 4 cases, type IIIb in 2 cases, type IVa in 1 case and type IVb in 1 case.

A family history of hearing loss was reported in 2 cases and a malformation of the external ear in a brother was reported in one case. A personal history of congenital heart disease (ventricular septal defect) was reported in one case. The mean age at the time of presentation was 2 years 6 months (range: 5 months to 12 years). Hearing loss was the presenting complaint in every case and was unilateral in 9 cases and bilateral in 8 cases with more severe hearing loss on one side in 4 cases. Tinnitus was present in 11 cases.

Physical examination revealed low implantation of the ear in one case (type IVa malformation) and a pretragal fistula ipsilateral to the hearing loss in 2 cases. On otoscopy, the tympanum was

OPEN ACCESS

Correspondence:
Rim Zainine, Department of ENT, La Rabta Hospital, Jabbari 1007, Tunis, Tunisia,
E-mail: rimzainine@yahoo.fr

Received Date: *05 Mar 2021*
Accepted Date: *02 Apr 2021*
Published Date: *06 Apr 2021*

Citation:
Zainine R. Minor Malformations of the Middle Ear. Am J Otolaryngol Head Neck Surg. 2021; 4(3): 1127.

Table 1: Classification according to Teunissen and Cremers [1].

Type	Malformations
Type I	Isolated congenital fixation of the stapes footplate
Type II	Fixed footplate with another congenital abnormality of the ossicular chain
IIa	Disruption of the ossicular chain
IIb	Epitympanic fixation
IIc	Tympanic fixation
Type III	Congenital anomalies of the ossicular chain with mobile stapes footplate
IIIa	Disruption of the ossicular chain
IIIb	Epitympanic fixation
IIIc	Tympanic fixation
Type IV	Congenital aplasia or dysplasia of the oval window or round window
IVa	Aplasia
IVb	Dysplasia
B1	Prolapse of the facial nerve
B2	Persistence of stapedial artery

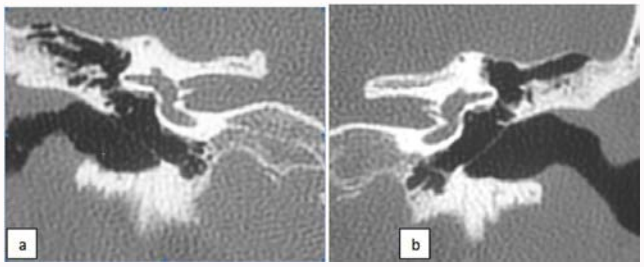

Figure 1: CT scan of the right (a) and left (b) temporal bones, coronal section: bilateral agenesis of the stapes and vestibular window.

normal in every case. Pure-tone audiometry, performed in every case, showed a mean air conduction of 56 dB. Hearing loss was unilateral in 9 cases and bilateral in 8 cases. Bone conduction was impaired in 3 cases. The mean air-bone gap was 45 dB. Speech audiometry was performed in 12 cases. Intelligibility was impaired in 11 cases with a mean threshold of 66 dB.

Hearing of the contralateral ear was affected in 7 cases with unilateral malformations. Tympanometry was classified as type A in 13 cases, type Ad in 2 patients and type B in 5 cases. The acoustic reflex was absent in 18 cases and present in 2 cases.

Computed tomography of petrous temporal bones was performed in only 6 patients (8 ears) and showed obvious lesions in 4 cases of type IIa and in the case of type IIIa (the patient presented osteolysis of the long crus of the incus, masked on CT scan by a fibrous bridge ensuring the incudostapedial contact). CT scan was normal in one case of type I malformation and in one case of bilateral type IVa malformation, consisting of bilateral agenesis of the stapes and windows (Figure 1, 2). This poor diagnostic performance was certainly due to the poor resolution of the CT scanner used in these old cases.

Bilateral hearing aids were indicated in the patient with a type IVa malformation. Endoscopic transcanal exploration of the tympanic cavity was performed in 16 patients, with deferred bilateral endoscopy in 2 cases (after an interval of 1 year).

The stapes footplate was fixed in 11 cases and mobile in 7 cases. Calibrated stapedotomy with insertion of a stapedial piston was

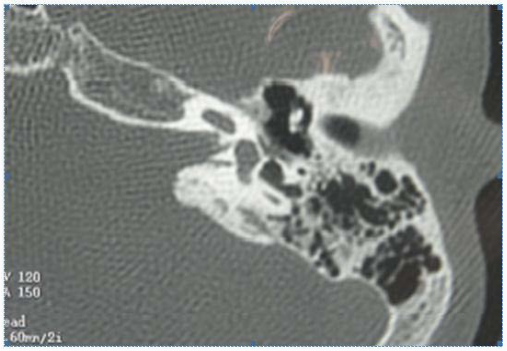

Figure 2: CT scan of the left temporal bone, axial section: agenesis of the vestibular window.

Table 2: Mean hearing gain and residual air-bone gap according to the type of malformation.

Type of malformation	Mean hearing gain (dB)	Mean residual air-bone gap (dB)
Type I (5 cases)	30	15
Type IIa (3 cases)	13.3	21.6
Type IIb (1 cases)	0	90
Type IIIa (4 cases)	22.5	35
Type IIIb (2 cases)	15	20
Type IVb (1 case)	10	50

performed in 6 cases (4 cases of type I malformation, with a deferred bilateral procedure in 1 case, 1 case of type IIa and one case of type IVb). Accidental stapedectomy occurred in 4 cases: partial posterior in 2 cases and total in 2 cases. Ossiculoplasty was performed in 4 cases of type III malformation (2 type IIIa and 2 type IIIb), corresponding to a type II ossiculoplasty in 3 cases and a type III ossiculoplasty in one case. Myringostapediopexy was performed in one case of type IIIa malformation. Three cases were treated conservatively (2 cases of type IIa malformation with a long crus in a short incus in one case and dislocation of the incus in one case and one case of type IIIa malformation, in which the stapes was malformed with absence of the long crus of the incus).

Immediate postoperative complications were marked by sensorineural hearing loss in one case and loss of balance in another case.

The postoperative results were evaluated for only 14 years after a mean follow-up of 12 months, with a successful result in 9 cases, with a mean auditory gain of 26 dB and a mean residual air-bone gap of 11.7 dB. A failure was observed in 5 cases: One case of type IIa malformation, one case of type IIb malformation with immediate postoperative sensorineural hearing loss and 3 cases of type IIIa malformation (Table 2).

Analysis of the results according to the type of procedure performed revealed a successful result in 5 cases of stapedotomy with stapedial piston, one case of hemistapedectomy with venous interposition piston, 2 cases of total stapedectomy with venous interposition piston, and the case of myringostapediopexy. Failure of surgery was observed in 5 cases: One case of stapedotomy, one case of total stapedectomy with postoperative sensorineural hearing loss, 2 cases of type II ossiculoplasty and one case of type III ossiculoplasty (Table 3).

Table 3: Audiometric results according to the procedure performed.

Procedure performed		Hearing gain (dB)	Residual air-bone gap (dB)
Stapedotomy (6 cases)		15.5 (mean)	31 (mean)
Stapedectomy	Partial (1 cases)	45	15
	Total (3 cases)	20 (mean)	12.5 (mean)
Myringostapediopexy (1 cases)		40	10
Type II ossiculoplasty (3 cases)		5 (mean)	30 (mean)
Type III ossiculoplasty (1 case)		10	20

Discussion

Minor malformations of the middle ear, also called minor aplasia or ossicular dysplasia, are abnormalities almost exclusively affecting the middle ear and ossicles with a normal external ear. Embryologically, an anomaly of the first branchial arch is responsible for anomalies of the head of the malleus and the body of the incus, while an anomaly of the second branchial arch is responsible for anomalies of the manubrium of the malleus, the long crus of the incus and the stapes superstructure [2]. Several classifications have been proposed in order to guide surgical or prosthetic management: Altmann, Colman, Ombrédanne, Funusaka, Causse, Charachon-Barthez. We have used the classification proposed by Teunissen and Cremers [1], which is the most widely accepted classification. Minor aplasia is rare with an estimated incidence of one case per 10,000 to 20,000 births according to the rare ENT malformation reference centre in France. These abnormalities are dominated by incudostapedial dislocations and stapedovestibular ankylosis [3], representing 40% and 44% of cases, respectively, in the series by Hashimoto et al., followed by fixation of the malleus or incus, observed in 8% of cases [4].

In the study by Kuhn JJ et al. [5], 8 cases of malformations of the stapes and incudostapedial complex were observed over a period of 13 years in a series of 185 cases of exploration of the tympanic cavity, i.e. 4.2%, including two cases associated with otospongiosis. The malformation rate in our series was 0.82%, which can be attributed to a problem of coding and filing of medical records.

Minor aplasia is bilateral in 30% to 40% of cases according to the rare ENT malformation reference centre in France. Two cases of bilateral minor aplasia were observed in our series. These malformations are often missed and are only diagnosed at the age of 5 or 6 years, in the case of unilateral malformations, sometimes in the context of a school screening examination, or at a younger age in the case of bilateral malformations in a context of abnormal language acquisition [3].

The mean age of the patients in this series was 32 years. This late diagnosis can be explained by delayed referral to the specialist, usually for socio-economic reasons, and the absence of systematic screening of schoolchildren. Several malformation syndromes have been described: the most classical syndrome is Wildervanck or cervico-oculo-acoustic syndrome, which comprises Duane syndrome (abducens nerve palsy), Klippel-Feil syndrome and possibly pure or mixed sensorineural hearing loss with fixed stapes footplate [3]. In the series of 104 ears published by Teunissen and Cremers, minor aplasia of the ear as part of a malformation syndrome represented 25% of cases [1].

In the study by Arnold et al. [6], published in 1986, comprising 50 cases of severe cardiac malformations, 16% were associated with hearing loss. A ventricular septal defect, either isolated or associated with other malformations (pulmonary artery stenosis, atrial septal defect, coarctation of the aorta) was present in every case. Ventricular septal defect was observed in one case in our series. Hearing loss is the most common presenting complaint, sometimes in the form of abnormal language acquisition especially in children. Pretragal fistula and low implantation of the ear are associated with minor aplasia of the ear in 13% to 30% of cases [1].

Minor aplasia is usually associated with conductive hearing loss with air conduction between 30 and 60 dB. Bone conduction is generally normal or, when abnormal, predominantly affects the high frequency range [3]. Tympanograms differ according to the type of malformation and the acoustic reflex [3]. High-resolution spiral computed tomography with 2D reconstruction is the method of choice for investigation of the middle ear, especially the ossicular chain, and its performance for the detection of the various abnormalities has been clearly demonstrated [7]. CT can also guide the surgical indications and allow planning of the surgical procedure by predicting possible difficulties [8]. Virtual endoscopy is a more recent noninvasive method of investigation, preferred by some authors because it provides three dimensional visualization of the tympanic cavity, thereby simulating the surgeon's intraoperative vision [9-11]. Except in the case of malformation syndromes, minor aplasia of the ear is isolated with no identified etiology at the present time. A systematic assessment to exclude the presence of any associated anomalies must be performed, comprising at least echocardiography and renal ultrasound, ophthalmological examination and X-rays of the cervical spine. The risk of isolated aplasia when a sibling is already affected is low, about 3/1,000 [12]. Surgery can be proposed from the age of 7 years when local conditions are favourable, depending on the degree of the child's hearing loss, generally when the loss on air conduction is greater than or equal to 30 dB. The surgical procedure depends on whether or not the stapes footplate is fixed to the vestibular window, in order to avoid the risk of "gusher" ear during the stapes footplate procedure. "Gusher" ear is a syndrome corresponding to the abnormal presence of cerebrospinal fluid in the internal ear cavities, which are often anomalous. A cerebrospinal fluid fistula from the oval window may be observed with CSF leak around the footplate or through an orifice in the footplate itself.

In the presence of a fixed stapes footplate, calibrated stapedotomy or stapedectomy can be performed, but these procedures are associated with a considerable cochlear risk. When the footplate is non-existent or inaccessible (prolapsed facial nerve), lateral semicircular canal fenestration techniques are no longer proposed due to the considerable risk of labyrinthitis (25% to 30%). Some authors propose transposition of the facial nerve [13], while others propose the creation of new oval window on the cochlear promontory [14, 15]. Isolated fixation of the stapes footplate is a rare condition. Teunissen and Cremers reported two cases of fixation the incudostapedial complex to the facial canal [2]. Henner and Kinsella reported cases of fixation of the stapes to the pyramidal process via a bone bridge (agenesis of the stapedius muscle) [16, 17]. Nandapalan and Tos [18] described a case of fixation of crus of the stapes to the cochlear promontory, which was mobilized by a micro-hook allowing complete mobility of the footplate.

In the presence of a mobile footplate, various types of ossiculoplasties can be performed depending on the type of malformation [4]. The postoperative results are generally good with a success rate of 88% according to the series published by Hashimoto

et al. [4], in which various types of ossiculoplasties were performed according to the type of malformation [4].

Conclusion

Since the advent of CT scan of the petrous temporal bone, the diagnosis of minor malformations of the ear has become considerably easier, allowing prediction of the most appropriate management. Treatment is surgical and the procedure depends on the mobile or fixed nature of the stapes footplate to the vestibular window.

References

1. Cremers CWRJ, Teunissen E. The impact of a syndromal diagnosis on surgery for congenital minor ear anomalies. Int J Pediatr Otorhinolaryngol. 1991;22(1):59-74.
2. Mayer TE, Brueckmann H, Siegert R, Witt A, Weerda H. High resolution CT of the temporal bone in dysplasia of the auricle and external auditory canal. AJNR Am J Neuroradiol. 1997;18(1):53-65.
3. Teissier N, Benchaa T, Elmaleh M, Van Den Abbeele T. Malformations congénitales de l'oreille externe et l'oreille moyenne. EMC oto-rhino-laryngologie. 2008;20-055-A-10.
4. Hashimoto S, Yamamoto Y, Satoh H, Takahashi S. Surgical treatment of 52 cases of auditory ossicular malformations. Auris Nasus Larynx. 2002;29(1):15-8.
5. Kuhn JJ, Lassen FL. Congenital incudostapedial anomalies in adult stapes surgery: A case-series review. Am J Otolaryngol. 2011;32(6):477-84.
6. Arnold SA, Brown OE, Finitzo T. Hearing loss in children with congenital heart disease: A preliminary report. Int J Pediatr Otorhinolaryngol. 1989;11(3):287-93.
7. Truy E, Bossard D, Coudert JL, Aslam S, Froehlich P, Trunde F, et al. Evaluation of 3D imaging software to study computed tomography images of middle ear defects. Ann Otolaryngol Chir Cervicofac. 2009;126(5-6):243-9.
8. Reisser C, Schubert O, Weidauer H. 3-dimensional imaging of temporal bone structures using spiral CT. initial results in normal temporal bone anatomy. HNO. 1995;43(10):596-600.
9. Tomandal BF, Hastreiter P, Eberhardt KE, Nimsky C, Grees H, Huk WJ, et al. Rapid interactive 3-D imaging of temporal bone with "direct volume rendering". HNO. 2001;49(8):618-25.
10. Neri E, Caramella D, Battola L, Cosottini M, Scasso CA, Bruschini P, et al. Virtual endoscopy of the middle and inner ear with spiral computed tomography. Am J Otol 2000;21:799-803.
11. Neri E, Caramella D, Panconi M, Berrettini S, Sellari Franceschini S, Forli F, et al. Virtual endoscopy of the middle ear. Eur Radiol. 2001;11(1):41-9.
12. Manac'h Y. L'aplasie de l'oreille. Des principes au cas particulier. Arch Pediatr. 1995;2:413-4.
13. Jahrsdoerfer RA. Transposition of facial nerve in congenital aural atresia. Am J Otol. 1995;16(3):290-4.
14. Han D, Zhao S, Wang D, Guo J, Dai H. Vestibulotomy above a severely displaced facial nerve. Acta Otolaryngol. 2005;125(9):962-5.
15. Lambert PR. Congenital absence of the oval window. Laryngoscope. 1990;100(1):37-40.
16. Henner R. Congenital middle ear malformations. AMA Arch Otolaryngol 1960;71:454-8.
17. Kinsella JH. Familial stapes superstructure fixation. J Laryngol Otol. 1993;107(1):36-8.
18. Nandapalan V, Tos M. Isolated congenital stapes superstructure fixation. J Laryngol Otol. 1999;113(9):798-802.