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OSSICULOPLASTY IN HEARING LOSS TREATMENT OF PATIENTS WITH OSTEOGENESIS IMPERFECTA

Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

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Abstract

Osteogenesis imperfecta is a connective tissue disease manifested by abnormalities within organs and structures rich in collagen. Typically, symptoms arise from the osteoarticular system. Excessive brittleness of the bones causes multiple fractures. Among patients with hearing loss, osteogenesis imperfecta manifests as changes to the stapes: the legs are broken and the footplate is immobilized and thickened. Changes to the malleus and incus are rarely reported. This location is associated with difficult conditions during surgery, difficult access to the ossicles. This research presents the characteristics and hearing results of patients who have undergone ossiculoplasty as a treatment of hearing loss in osteogenesis imperfecta.

Two case reports present massively altered conditions within the middle ear in patients with type III osteogenesis imperfecta – the most severe among live births, who have undergone multiple surgeries due to hearing loss, resulting in a moderate improvement in hearing.

Key words: osteogenesis imperfecta, hearing loss, ossiculoplasty

OSICULOPLASTIAS EN EL TRATAMIENTO DE LA HIPOACUSIA EN PACIENTES CON FRAGILIDAD ÓSEA CONGÉNITA

Resumen

La osteogénesis imperfecta como enfermedad del tejido conectivo se manifiesta con irregularidades en los órganos y estructuras ricas en colágeno. Los síntomas más comunes están relacionados con el sistema óseo y articular. La excesiva fragilidad ósea resulta en múltiples rupturas de los huesos y su posterior fusión, lo cual produce deformaciones. En caso del oído, normalmente se observan alteraciones del estribo: sus ramas se ven rotas, la platina del estribo queda inmovilizada y engrosada. Actualmente en estos casos el tratamiento de elección es la estapedotomía. Rara vez se refieren alteraciones en el área del martillo o yunque. Tal localización implica acceso quirúrgico dificultado. En estos pacientes, el tratamiento de elección son las osiculoplastias. El presente trabajo describe las características y los resultados auditivos de los pacientes que se han sometido a osiculoplastias durante el tratamiento de la hipoacusia en la osteogénesis imperfecta.

Los 2 casos clínicos descritos a continuación presentan muestran condiciones masivamente alteradas dentro del oído medio en pacientes con la osteogénesis imperfecta de tipo III: la más grave de entre los que nacieron vivos y que se han sometido a múltiples cirugías por hipoacusia, consiguiendo una mejora moderada de la audición.

Palabras clave: osteogénesis imperfecta, hipoacusia, osiculoplastia

ОССИКУЛОПЛАСТИКА ПРИ ЛЕЧЕНИИ ТУГОУХОСТИ У ПАЦИЕНТОВ С ВРОЖДЁННОЙ ЛОМКОСТЬЮ КОСТЕЙ

Резюме

Несовершенный остеогенез (Osteogenesis imperfecta), будучи болезнью соединительной ткани, проявляется в виде аномалий в органах и структурах, содержащих большое количество коллагена. Наиболее типичные симптомы касаются костно-суставной системы. В связи с повышенной ломкостью костей происходят их многократные переломы с последующим сращением, что приводит к деформациям. В случае уха типично изменено стремя — ножки сломаны, пластинка обездвижена и утолщена. В таких случаях в настоящее время выбирается лечение в форме стапедотомии. Очень редко появляется информация об изменениях в области молоточка или наковальни. Их расположение связано со сложностью операционного доступа. Выбирающимся видом лечения среди таких пациентов является оссикулопластика. Настоящая работа представляет слуховую характеристику и результаты пациентов, которые прошли оссикулопластику в лечении тугоухости при неовершенном остеогенезе.

Два нижеследующих описания клинических случаев представляют массивно изменённые условия в пределах среднего уха у пациентов с III типом несовершенного остеогенеза — самым сложным среди живорожденных — которые прошли многократные операции по причине тугоухости с достижением улучшения слуха посредственной степени.

Ключевые слова: врождённая ломкость костей, тугоухость, оссикулопластика.

OSSIKULOPLASTYKI W LECZENIU NIEDOSŁUCHU PACJENTÓW Z WRODZONĄ ŁAMLIWOŚCIĄ KOŚCI

Streszczenie

Osteogenesis imperfecta jako choroba tkanki łącznej objawia się nieprawidłowościami w obrębie narządów i struktur bogatych w kolagen. Najbardziej typowe objawy dotyczą układu kostno-stawowego. W związku z nadmierną kruchością kości dochodzi do ich wielokrotnych złamań i następczego zrastania, co skutkuje deformacjami. W przypadku ucha typowo zmienione jest strzemiączko - odnogi są złamane, płytka strzemiączka jest unieruchomiona i pogrubiała. W takich przypadkach leczeniem z wyboru aktualnie jest stapedotomia. Bardzo rzadko raportuje się zmiany w obrębie młoteczka bądź kowadełka. Ta lokalizacja wiąże się z utrudnionym dostępem operacyjnym. Leczeniem z wyboru są ossikuloplastyki pośród tych pacjentów. Niniejsza praca przedstawia charakterystykę i wyniki słuchowe pacjentów, którzy przeszli ossikuloplastyki w leczeniu niedosłuchu w Osteogenesis imperfecta.

Poniższe 2 opisy przypadków klinicznych przedstawiają masywnie zmienione warunki w obrębie ucha środkowego u pacjentów z III typem Osteogenesis imperfecta - najcięższym spośród żywourodzonych, którzy przeszli wielokrotne operacje z powodu niedosłuchu, uzyskując poprawę słyszenia miernego stopnia.

Słowa kluczowe: wrodzona łamliwośc kości, niedosłuch, ossikuloplastyka

Introduction

Osteogenesis imperfecta is a connective tissue disease that affects organs and structures rich in collagen, including the auditory organ. Due to the etiology of the disease-disturbed connective tissue structure, it manifests as excessive bone brittleness, fusion of bone structures, osteophyte formation, and callus [1,2,3]. In the ear, stapes immobilization is typically found: thickening, footplate fixation, obliteration,and broken crura [4-12]. Individual studies report pathologies of the other auditory ossicles, the malleus and incus, such as epitympanal fixation [5], although the most common is a bone adhesion between the lateral wall of the epitympanum and the incus.

Typically, ossiculoplasty is used for pathologies within the malleus and incus, usually to deal with ossicular destruction occurring in chronic otitis, congenital malformation of the ossicles, or defects acquired in traumatic ossicular chain discontinuity. Treatments include the use of PORP and TORP prostheses, interpositions, and malleus head resections. The procedures are performed using a transcanal or, rarely, a retroauricular approach. A posterior atticotomy is most frequently performed using a retroauricular approach with the ossiculoplasty mobilizing the I and II ossicles in cases of recurrent fixation in the epitympanum.

The present study aims to describe patients who present immobilization of the I and II ossicles in the context of patients with stapes pathology. The analysis describes intraoperative abnormalities, the effects of treatment, and the characteristics of patients with osteogenesis imperfecta; in all of whom ossiculoplasty was performed.

Material and method

Out of 20 patients with osteogenesis imperfecta treated for hearing loss in the Institute of Physiology and Pathology of Hearing, 18 underwent surgery (Table 1). Of these, 3 had ossiculoplasty. The study excluded ears operated in other centers. The analysis here describes 2 patients. They were male, the beginning of hearing loss was typically noticed when they were in their mid-20s or 30s. Each had undergone several ear operations from ages 2 to 7. The patients were classified as type III osteogenesis imperfecta. The following study presents the characteristics of those two patients who underwent ossiculoplasty and their auditory results.

Patient OI III

The first case report concerns OI III patient with hearing loss first observed at age 22 (Table 2). The history of

Table 1. Patients with osteogenesis imperfecta and hearing loss under the care of the Institute of Physiology and Pathology of Hearing. Patients with ossiculoplasty and described here are marked in grey

Initials	Sex	Duration of hearing loss	Age at 1st operation
OI I	M	From 15–16 y/o, ailment intensification after a car accident	17*
OI II	M	From 9–10 y/o	14
OI III	M	From 22 y/o	32*
OI IV	M	From 15–16 y/o	25
OI V	F	From 25 y/o, after childbirth	61
OI VI	M	From approx. 40 y/o	63
OI VII	F	From approx. 25 y/o	32
OI VIII	F	From approx. 6 y/o	30
OI IX	F	From approx. 13 y/o	17
OI X	M	From approx. 30 y/o	49
OI XI	M	From 18 y/o	37
OI XII	M	From 14 y/o	16*
OI XIII	F	From 13 y/o	23
OI XIV	M	From 17–18 y/o	27
OI XV	M	From 14 y/o	20
OI XVI	M	From 15 y/o	22
OI XVII	F	From 50 y/o	56
OI XVIII	F	From 25 y/o	45
OI XIX	F	From 18 y/o	-
OI XX	F	From 20 y/o, after otitis	-

* One ear underwent surgery in another ORL clinic

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present illness was extensive: 30 bone fractures, deformities of the upper and lower limbs, blue sclera, keeled chest, and dentinogenesis. In the family, the mother showed similar symptoms. His first two surgeries due to hearing loss were performed in another center. These were exploratory operations, which due to the significantly altered anatomical conditions within the middle ear did not extend to ossiculoplasty or stapedotomy. In the first surgical procedure, a pathologically altered tympanic segment of the facial nerve was found; a biopsy specimen was taken and histopathological examination undertaken. Degenerative changes of the facial nerve were indicated in the examination description. After the procedure, the patient had left side facial nerve paresis. After the second left ear surgery, the patient was referred to the Institute of Physiology and Pathology of Hearing with mixed bilateral hearing loss for further surgical treatment.

In the physical examination, the tympanic membranes were preserved bilaterally, but were dull. In the auditory examination, there was mixed hearing loss of a significant degree on both sides, a type A tympanogram, and no stapedius muscle reflex on either side. The patient was referred for surgery of the right ear (not previously operated on). During the first surgical procedure, immobilization of the incus and stapes was found. It was assumed this may be a result of earlier fracture and fusion of the ossicles. In addition, the facial nerve was exposed and overlying the oval window; the tympanic cavity ependyma was prone to bleeding. Stapedotomy with ossiculoplasty was performed, mobilizing the incus by eliminating the bone union with the attic. An opening for a prosthesis was made at the border of the footplate and the promontory. During the treatment, there was intense bleeding from the thickened ependyma.

Because the operation had no auditory effect, the patient underwent reoperation using a retro-auricular approach. Attico-antro-mastoidectomy was performed with removal of lesions and reossiculoplasty, again finding incus immobilization due to bone deposits near the attic and adhesions in the area of the oval window niche, which were removed in stages. After a year, the patient was re-scheduled for reoperation: a restapedotomy was performed with atticotomy and posterior tympanotomy. A prosthesis was correctly placed in the vestibule. Other findings were adhesions, immobilized incus, a short process and body of the incus merged by callus with the lateral semicircular canal. Adhesions and callus lesions were removed, but there was intense bleeding from the ependyma cavity.

Lack of an effect of the procedure resulted in rescheduling after about a year. The patient underwent stapedotomy after revision. Abundant adhesions around the prosthesis and malleus immobilization in the attic were found, and the adhesions were removed. Finally, it was decided to attempt reoperation for the last time: a revision after stapedotomy was performed, again numerous adhesions around the prosthesis were found and removed, and there was incus immobilization (fusion with the attic lateral wall was found), so bone chips from the body of the incus and lateral wall of the attic were removed.

In a control CT of the temporal bones, there was massive bilateral otospongiotic changes in the medial wall of the middle ear – including the promontory, oval and round window frame, lamella of the facial canal, alteration near the cochlear base bend, and in the cochlear section of the internal auditory canal.

Patient OI XI

The second case report concerns patient OI XI with hearing loss since the age of 18 (Table 2). He was a very short patient (102 cm), who moved in a wheelchair due to numerous deformations of the spine and bones, had flaccid skin, and excessively flexible joints. There were no symptoms of osteogenesis imperfecta in the family. Because of bilateral significant mixed hearing loss, type A tympanogram, and lack of reflexes from the stapedius muscle on either side, the patient was scheduled for exploratory tympanotomy of the right ear with possible stapedotomy or ossiculoplasty. During surgery, bone growths were found on the posterior-upper wall of the external auditory canal (possibly a mark after fracture), immobilization

Table 2. Summary of the types of surgery and intraoperative changes observed during surgery

Initials	Age at 1st operation	Types of ear operations and descriptions of lesions found during operations
OI III	32	– exploratory tympanotomy of the left ear in 2001* – myringotomy of the left ear in 2002* – stapedotomy with ossiculoplasty of the right ear in 2008 (immobile incus and stapes, most probably due to previous fracture and fusion, exposed facial nerve overlays the oval window, intense bleeding of ependyma) – attico-antro-mastoidectomy with removal of lesions and reossiculoplasty of the right ear in 2008, immobile incus) – restapedotomy with atticotomy and anterior tympanotomy of the right ear in 2009 (prosthesis with adhesions, correctly placed in the vestibule, immobile incus, rigid connection of the short process of incus and body with lateral semicircular canal (callus), exposed facial nerve, intense bleeding) – revision after stapedotomy of the right ear in 2010 (numerous adhesions around prosthesis, incus immobilized in the attic) – revision after stapedotomy of the right ear in 2012 (numerous adhesions around prosthesis, incus immobilization due to bony union of incus body with lateral wall of attic)
OI XI	37	– ossiculoplasty of the right ear in 2013 (features of fractures or osteophytes on anterior- superior wall, bony union of lateral wall of attic and long process of incus and neck of malleus, I and II ossicles were mobilized) – reossiculoplasty of the right ear in 2014 (bone deposits between the long process of incus and lateral wall of attic, immobilization of malleus and incus, mobile stapes)

* surgery in another ORL clinic

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of the I and II ossicles due to merger of the long process of the incus and neck of the malleus with the lateral wall of the attic. The malleus and incus were mobilized and adhesions from the area of the mobile stapes were removed. Due to a small auditory effect, the patient was scheduled for reoperation. During the reossiculoplasty, immobilization of the I and II ossicles was confirmed due to bone deposits reaching the lateral wall of attic, but the stapes was correctly mobile. Bone deposits and adhesions were removed in stages.

A CT of the temporal bones showed marked areas of impaired calcification. There was extensive decalcification of the malleus and incus, sclerosis of the oval window with local deossification of the anterior frame of the window, and ossification disorders in the capsula otica.

Results

Patient OI XI

The patient underwent two right ear operations. The air-bone gap after two operations on the right ear increased on average by 10 dB (Table 3) and hearing thresholds gain was on average 2.5 dB (Table 4). The patient refused to be scheduled for reoperation of the right ear or for a first operation of the left ear. The patient was also not interested in implants and decided to use a hearing aid.

Table 3. Analysis of the air-bone gap (ABG, in dB) after right ear operations in OI XI patient

	500 Hz	1000 Hz	2000 Hz	4000 Hz	Average
ABG preop1	60	45	15	15	33.75
ABGpostop1	55	50	30	45	45
ABG 1 difference	5	–5	–15	–30	–11.25
ABG preop2	55	50	30	45	45
ABGpostop2	55	55	35	30	43.75
ABG 2 difference	0	–5	–5	15	1.25
ABGX difference	5	–10	–20	–15	–10

ABGX- change in air-bone gap (dB) after the last operation compared with preoperative results

Table **4.** Analysis of hearing gain (HG, in dB) after right ear operations in OI XI patient

	500 Hz	1000 Hz	2000 Hz	4000 Hz	Average
HG1	5	5	–10	–10	–2.5
HG2	0	0	5	5	2.5

Patient OI III

The patient underwent five right ear operations. Ultimately, the air-bone gap decreased by 5–15 dB (6.25 dB on average) and improvement of hearing thresholds by up to 15 dB (10 dB on average), as shown in Tables 5 and 6. The patient was not interested in implants, and it was decided to fit conventional hearing aids.

Discussion

Available studies only mention single cases of epitympanal fixation in osteogenesis imperfecta [5]. Typically, the pathology is reported within the stapes and oval window niche as a thickening and immobilization, and even an obliteration of the stapes footplate. The results of crurotomy, stapedectomy, and, recently, stapedotomy have been reported for surgical treatment of conductive or mixed hearing loss associated with osteogenesis imperfecta [5,6,10,11,13]. There is only one study available regarding the auditory results of ossiculoplasty in cases of malleus and/or incus immobilization in patients with III type of osteogenesis imperfecta and it shows unsatisfactory effects of the treatment [5].

Our study discovered advanced bone lesions in the ears of patients with type III osteogenesis imperfecta. Extensive areas of bone alterations, deossification, and ossicular malformations indicate that extremely difficult surgical conditions can occur in patients with congenital bone fragility. In the presented patients, immobilization of the I and II ossicles in the attic was found, which, despite removal, recurred. This indicates continuous bone alteration in patients with type III osteogenesis imperfecta. The described patients underwent multiple ear surgeries, with patient OI III undergoing 5. All surgical procedures were performed by an otosurgeon who had the largest clinical experience. An improvement in hearing was achieved in both cases, although it was minor. To our knowledge, this is the one of the first reports regarding the results of ossiculoplasty in patients with osteogenesis imperfecta.

The literature has extensively described ossiculoplasty in middle ear malformations [14–17]. Typically, minor malformations are distinguished as those where the tympanic membrane is preserved, the tympanic cavity is of normal size, and pathology relates only to the ossicles; major malformations are those where, apart from an altered middle ear, the malformation also extends to the external ear, resulting in atresia of the external auditory canal and microtia [14,15,17].

The most typical middle ear anomaly is stapes fixation (stapes ankylosis) [15,22]. There is a wide range of anomalies found in the region of ossicles: bone unions of malleus and incus within the epitympanum, fusion of the malleus and incus, and aplasia of the long process of the incus [17]. Often the decision on ossiculoplasty for a minor congenital anomaly depends greatly on the course of the facial nerve, which frequently has a slight anterior position and overlays the stapes footplate [17].

Epitympanal fixation is believed to result from a bone bridge connecting the malleus head with the anterior-superior wall of the epitympanum [18–20]. Local inflammation in the attic, cholesterol granuloma, or tympanosclerosis may also result in attical fixation [18]. In the Nager study [21], temporal bone histopathology showed that active otosclerosis may also very occasionally lead to attical fixation. Fixation of the I and II ossicles is commonly revealed during exploratory tympanotomy, when otosclerosis is suspected

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Table 5. Analysis of the air bone gap (ABG, in dB) after subsequent right ear operations. ABGX: change in air-bone gap (dB) after the last operation compared with preoperative results

	500 Hz	1000 Hz	2000 Hz	4000 Hz	Average
ABGpreop1	50	45	30	35	40
ABGpostop1	45	35	25	35	35
ABGpostop2	60	55	35	35	46.25
ABGpostop3	60	40	30	45	43.75
ABGpostop4	50	40	30	25	36.25
ABGpostop5	35	40	30	30	33.75
ABGX	15	5	0	5	6.25

Table 6. Analysis of hearing thresholds after subsequent right ear operations. HG: hearing gain (dB); HGX: change in hearing thresholds (dB) after the last operation compared with preoperative results

	500 Hz	1000 Hz	2000 Hz	4000 Hz	Average
HG1	5	10	5	0	5
HG2	–5	–5	0	5	–1.25
HG3	–10	–10	–10	–20	–12.5
HG4	10	15	10	35	17.5
HG5	15	0	0	–10	1.25
HGX	15	10	5	10	10

[18]. Various surgical techniques have been described for mobilization of the ossicles: in the attic using a tray, widening the space around the malleus and incus with a diamond drill [18], resection of the malleus head, removal of the incus and malleus with the use of a malleovestibulopexy prosthesis, or interposition of cartilage or a PORP prosthesis between the incus and the stapes head [14,17,18]. Recurrences of epitympanal fixation are reported [18].

In minor middle ear malformations, improvement of air conduction thresholds to 30 dB by ossiculoplasty is deemed to be a criterion of success [14]. The limit has been set at 30 dB because below this level of conduction thresholds it is usually necessary to use a hearing aid. Other studies define a decrease of the air-bone gap to 20 dB or less as a successful ossiculoplasty [22]. In the case reports presented above, the auditory results indicate an unsatisfactory ossiculoplasty in type III osteogenesis imperfecta.

In the absence of effective treatment of hearing loss by means of classic reconstruction, it is possible to use bone conduction devices. There is a report on the application of the Baha Connect [23] in a patient with type III osteogenesis imperfecta and advanced ear malformation, after unsuccessful reconstructive surgeries. In addition, the literature also reports the use of the Vibrant Soundbridge middle ear implant, along with stapedotomy, in congenital bone fragility in deep mixed hearing loss [24].

Summary

Conductive or mixed hearing loss in osteogenesis imperfecta is an indication for reconstructive surgery – stapedotomy or ossiculoplasty. The most common abnormalities involve the stapes. Sometimes the pathology may include the I and II ossicles, and deossification lesions and bone alteration can be extended within the middle ear, especially in patients with the most advanced forms of osteogenesis imperfecta. Such location of immobilization of the ossicles indicates a much lower chance of obtaining a satisfactory auditory effect which will be durable over time.

References

1. McKusick VA. Heritable disorders of connective tissue. V: Osteogenesis imperfecta, Journal of Chronic Diseases, 1956, 3(2): 180–202.
2. Harrington J, Sochett E, Howard A. Update on the evaluation and treatment of osteogenesis imperfecta. Pediatric Clinics of North America, 2014 December; 61(6): 1243–57.
3. Van Dijk F, Sillence D. Osteogenesis imperfecta: clinical diagnosis, nomenclature and severity assessment. Am J Med Genet, 2014; 164A: 1470–81.
4. Shea JJ, Postma DS. Findings and long-term surgical results in the hearing loss of osteogenesis imperfecta. Arch Otolaryngol, 1982 Aug; 108(8): 467–70.
5. Van der Rijt AJ, Cremers CW. Stapes surgery in osteogenesis imperfecta: results of a new series. Otol Neurotol, 2003 Sep; 24(5): 717–22.
6. Kuurila K, Pynnönen S, Grénman R. Stapes surgery in osteogenesis imperfecta in Finland. Ann Otol Rhinol Laryngol, 2004 Mar; 113(3 Pt 1): 187–93.
7. Swinnen FK, De Leenheer EM, Coucke PJ, Cremers CW, Dhooge IJ. Audiometric, surgical, and genetic findings in 15 ears of patients with osteogenesis imperfecta. Laryngoscope, 2009 Jun; 119(6): 1171–9. doi: 10.1002/lary.20155.
8. Pedersen U, Elbrønd O. Surgical findings and results of stapedectomy in patients with osteogenesis imperfecta. J Laryngol Otol, 1979 Dec; 93(12): 1229–33.
9. Cremers C, Garretsen T. Stapes surgery in osteogenesis imperfecta. Am J Otol, 1989; 10: 474–6.
10. Doi K, Nishimura H, Ohta Y, Kubo T. Stapes surgery in Japanese patients with osteogenesis imperfecta. Adv Otorhinolaryngol, 2007; 65: 226–30.
11. Swinnen FKR, De Leenheer EMR, Coucke PJ, Cremers CWRJ, Dhooge IJM. Stapes surgery in osteogenesis imperfecta: retrospective analysis of 34 operated ears. Audiol Neuro-otol, 2012 April; 17(3): 198–206.
12. Vincent R, Wegner I, Stegeman I, Grolman W. Stapedotomy in Osteogenesis Imperfecta: a prospective study of 32 consecutive cases. Otol Neurotol, 2014 December; 35(10): 1785–9.
13. Garretsen AJTM, Cremers CWRJ. Osteogenesis imperfecta type I: otological and clinical genetic aspects. Arch Otolaryngol Head Neck Surg, 1990; 116: 317–23.
14. Philippon D, Laflamme N, Leboulanger N, Loundon N, Rouillon I, Garabedian EN, Denoyelle F. Hearing outcomes in functional surgery for middle ear malformations. Otol Neurotol, 2013 October; 34(8): 1417–20.

DOI: 10.17430/1002971

15. Vincent R, Wegner I, Kamalski DM, Bittermann AJ, Grolman W. Congenital Stapes Ankylosis in Children: Surgical Findings and Results in 35 Cases. Otol Neurotol. 2016 Apr;37(4):367-73.
16. Quesnel S, Benchaa T, Bernard S, Martine F, Viala P, Van Den Abbeele T, Teissier N. Congenital middle ear anomalies: anatomical and functional results of surgery. Audiol Neurootol, 2015; 20(4) :237–42.
17. Schwager K. Reconstruction of middle ear malformations. Curr Top Otorhinolaryngol Head Neck Surg, 2007; 6.
18. Martin C, Oletski A, Prades JM. Surgery of idiopathic malleus fixation. Otol Neurotol, 2009 Feb; 30(2): 165–9.
19. Armstrong BW. Epitympanic malleus fixation: correction without disrupting the ossicular chain. Laryngoscope, 1976 Aug; 86(8): 1203–8.
20. Seidman MD, Babu S. A new approach for malleus/incus fixation: no prosthesis necessary. Otol Neurotol, 2004 Sep; 25(5): 669–73.
21. Nager GT, ed. Pathology of the Ear and Temporal bone. Baltimore, MD: Williams and Wilkins, 1993, 952–7.
22. Park K, Choung YH. Isolated congenital ossicular anomalies. Acta Otolaryngol, 2009 Apr; 129(4): 419–22.
23. Coutinho MB, Marques C, Mendes GJ, Gonçalves C, Successful bone-anchored hearing aid implantation in a patient with osteogenesis imperfecta. J Laryngol Otol, 2015 Nov; 129(11): 1133–6.
24. Kontorinis G, Lenarz T, Mojallal H, Hinze AL, Schwab B. Power stapes: an alternative method for treating hearing loss in osteogenesis imperfecta? Otol Neurotol, 2011 Jun; 32(4): 589–95.

DOI: 10.17430/1002971