

## Case presentation:

A 16 year old girl presented to our clinic for reconstruction of a malformed right ear. She did complain of some hearing loss with difficulty in localizing sounds, but was doing fairly well with excellent school performance. The rest of her history was negative.

On physical exam she had a grade III microtia with a 0.5 x 3cm vertical sausage-shaped skin remnant, with poorly organized cartilage in the superior aspect, and a well formed lobule inferiorly. No pit or external auditory canal (EAC) opening was detected. The rest of her head and neck exam was otherwise unremarkable.

On tuning fork exam the Weber lateralized to the right; and the Rinne was positive on the left and negative on the right.

The pure tone audiometry revealed a right moderate conductive hearing loss with an air bone gap ranging from 30dB to 60dB with a speech discrimination score of 96% and a normal hearing on the left side (*fig.1*). A computed tomography (CT) scan of the temporal bones was consistent with bony atresia of the right external auditory canal with absent ossicles and a normal labyrinth (*fig.2*).

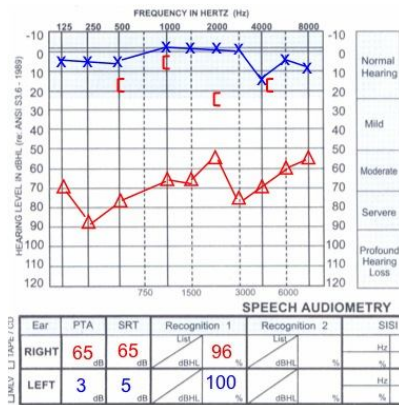
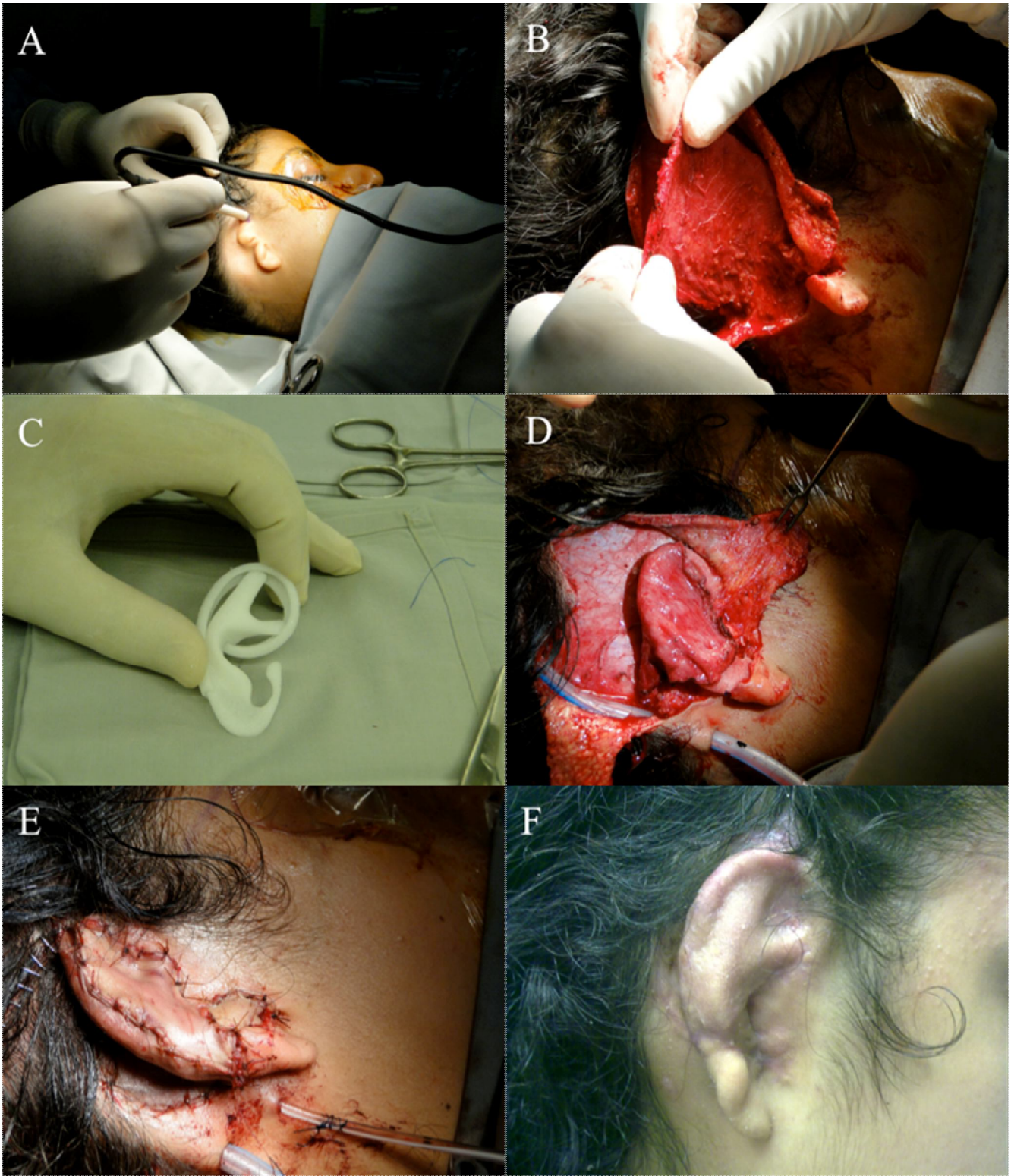


Fig.1 Pure tone Audiogram



Fig2. CT scan temporal bones

The patient underwent right auricular reconstruction using the MedPor (Porex<sup>®</sup>) prosthesis. A temporoparietal fascial flap was elevated and draped over the prosthesis, and then covered with a split thickness skin graft from the right lateral thigh and a full thickness skin graft harvested from the left post-auricular area.



*Fig.3 operative procedure of right microtic ear reconstruction with Medpor prosthesis. Superficial temporal artery detection with Doppler (A). Temporoparietal fascia elevation (B). Medpor prosthesis (C). Wrapping of the elevated fascia around the prosthesis (D). Closure with Full thickness skin graft from the left post-auricular area and split thickness skin graft from the right lateral thigh (E). End result at 4month follow up (F).*

## **Discussion:**

Microtia, or 'small ear', denotes a congenital malformation of the pinna, ranging from a smaller ear to a severely malformed pinna. The microtic auricle consists of a disorganized remnant of cartilage attached to a variable amount of soft tissue lobule. It has an incidence of 1 in 7000 to 8000 births in the general population [1][2] with a higher incidence in Hispanics and Asians [3]. Males are affected more commonly, and the reported ratio of right-to-left-to-bilateral is approximately 5:3:1 with bilateral deformity occurring in only 10% of patients [4][5][6][7].

Most cases of microtia are sporadic, with a suspected multifactorial etiology. Less than 15% of cases are considered familial [2], with both autosomal dominant and recessive forms with variable expression and incomplete penetrance being reported in the literature [8]. The suggested theory of in utero pro-bands has led to the use of 'malformation' and 'deformity' interchangeably in describing microtia. Other risk factors include maternal Rubella, thalidomide and some other teratogens exposure [8].

A brief review of the external ear embryology allows a better understanding of the pathophysiology of microtia. The auricle begins to develop at 5 weeks from 6 hillocks. The first branchial arch gives rise to the first 3 hillocks that will form the tragus, the helical crus, and the helix. The second arch gives rise to the other three hillocks that form the antihelix, the scapha and the lobule. In between the first and second branchial arches, the first branchial cleft will form the external auditory meatus [9]. Progressively the forming auricle migrates from a ventromedial to a more dorsolateral position as the midface and mandibular processes grow and push it outward and upward. Interruption in the proliferation or fusion of the hillocks at varying stages results in the different grades of microtia. Due to the intimate embryologic development of the auricle and the EAC, microtia is often associated with aural atresia [10]. It can also be associated with other malformations in around 15 to 60% of patients, especially in bilateral cases [11][12]. Whether isolated microtia is the mildest form of the 'Hemifacial microsomia spectrum' with Goldenhar syndrome being the most advanced stage, is still controversial [3]. Syndromes with microtia are frequently associated with anomalies such as facial cleft, facial asymmetry, renal abnormalities, cardiac defects, microphthalmia, polydactyly and vertebral anomalies [5][12].

There is no universally adopted classification system for microtia. A very popular grading system, the Marx classification [13], assigns a grade from I to III based on the severity of the deformity. Grade I denotes a slightly smaller than normal ear with overall normal features. Grade II is a rudimentary and malformed auricle but with recognizable components. Grade III, the most common type, includes the classic “peanut” ear, which is severely attenuated with usually associated aural atresia. Another commonly used classification system, proposed by Nagata [3] categorizes microtia into either ‘Lobule-type microtic ears’ or ‘Concha-type microtic ears’, according to vestigial structures present.

Addressing the hearing loss in these patients is an integral part of the overall management. The above described patient was considered a poor candidate for atresiaplasty and was thus offered a BAHA but declined for financial considerations. Timing of the surgery should take into consideration both the psychosocial aspect of microtia in a child that becomes evident by the age of 10 [14], and other factors such as normal auricle size. The normal auricle is 66% of its adult size at birth and reaches 85% and 95% at ages 3 and 6. Therefore repair before the age of 6 is not advised [3][14]. Whether rib cartilage or prosthetic technique is to be used for reconstruction also affects the timing of surgery with the prosthetic reconstruction possibly started earlier and the autogenous rib cartilage later, until enough cartilage can be harvested [15].

The standard microtia reconstruction with autogenous rib cartilage was first described by Tanzer in the early 70’s [16]. It usually requires a minimum of two and up to four or five staged surgical procedures. It has been since then subject to several modifications. The main drawbacks of this technique are the multiple procedures, donor site morbidity, post-operative pain, and variable results [15]. Interestingly a recent report by DellaCroce et al. has documented a quantitative growth in both the height (average 5 mm) and the width (average 2.75 mm) of the cartilaginous framework, but showed no statistically significant difference in the size of the patient’s reconstructed ears compared with the normal ears at follow-up [17].

With the advent of osseointegrated implants, prosthetic reconstruction became an alternative. However, despite the benefit of a single operative procedure, prostheses eventually need to be replaced. Also, a thin temporal bone and a growing bone are considered major obstacles to the insertion of such implants in children [15]. Failed autogenous reconstruction, severe soft

tissue-skeletal hypoplasia and low unfavorable hairline have been proposed as relative indications for prosthetic microtia reconstruction [18].

Recently, the use of an alloplastic auricular framework has been advocated as an alternative. Silicone was initially presented with great enthusiasm but revealed a high failure rate from implant exposure, and has been largely abandoned [3]. Porous polyethylene framework (Medpor, Porex Surgical, Inc., Newnan, GA) has recently been gaining a considerable popularity in microtia reconstruction. It is favored for its minimal tissue reaction and its porosity, which allows soft-tissue ingrowth and better stability. Despite some early reports on porous polyethylene that showed a similar failure rate to that of silicone [19][20], the complete coverage of the implant with a temporoparietal fascia flap kept it as a very promising surgical reconstructive option.

Another advantage of MedPor reconstruction is that atresioplasty, when indicated, can be performed at any time and does not have to wait for the aesthetic component to be completed. Results of atresia repair before MedPor microtia reconstruction compare favorably with results achieved with atresia repair after microtia reconstruction with autogenous rib cartilage and with atresia repair without microtia repair [21].

Microtia is a challenging entity to the otolaryngologist that requires careful pre operative planning. Both the aesthetic and otologic aspects should be addressed. Several reconstructive options are available and proper patient selection and meticulous surgical technique result in excellent outcomes.

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