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Editorial

A Short Review on Microtia and Bangladesh Perspective

Microtia is a congenital malformation of variable severity of the external and middle ear. The microtic auricle consists of a disorganized remnant of cartilage attached to a variable amount of soft tissue, which often is displaced from a position symmetrical with the opposite normal ear. The direction of displacement based on the degree of associated facial hypoplasia. Depending on the severity of the anomaly, there may be evidence of external meatus formation. Microtia commonly involves the external canal and middle ear; hence, hearing can be affected. Microtia may present within a spectrum of branchial arch defects (hemifacial microsomia, craniofacial microsomia) or may manifest as an independent malformation.

Practical Anatomy of the Ear

External ear is composed mostly by the auricle. Auricle is elastic cartilage covered with skin; exception is there is no cartilage in the lobe rather fibro-fatty tissue. Major cartilagenous framework of the auricle are the[2]: a) Helix lobule complex, b) Antihelix antitragus complex, c) Conchal complex and d) Tragus. Some important aesthetic measurements of the ear to be considered before planning the microtia correction[1,3] –

- Ear location: one ear length posterior to the lateral orbital rim
- Height (adult): 5.5-6.5 cm
- Width: 55% of its height
- Lateral protrusion: 1-2 cm from scalp
- Inclination: 21-25 degrees in vertical axis
- Long axis tilts: 20 degrees posteriorly

Epidemiology

Melnick and Myranthopoulos reviewed[4] auricular deformities and associated anomalies in a series of 56,000 pregnancies in an ethnically diverse population (Caucasian 46%, African American 46%, Latino 8%), commenting on the incidence of anomalies and the embryogenesis and etiopathology of the varying deformities. Ear deformities occurred in approximately 1.1% (11 in 1000) of births. Severe anomalies, such as microtia, occurred in approximately 3 in 10,000 live births. Occurrence has been reported to be 1 in 4000 in the Japanese population and as high as 1 in 900 to 1 in 1200 in the Navajo population.

Almost one half of the microtia patients in the Melnick and Myranthopoulos study[4] presented with craniofacial microsomia. In the same study, the right side was affected almost twice as often as the left, and bilateral deformity occurred in 10% of patients, with the reported ratio of right-to-left-to-bilateral of approximately 5:3:1.

Etiology

Both hereditary factors and vascular accidents in utero have been suggested as factors in the etiology of microtia. Several groups have studied their microtia patients as probands, finding evidence for familial craniofacial microsomia and patterns suggestive of multifactorial inheritance[5,6].

Specific causative factors also can include maternal rubella during the first trimester of pregnancy; Brent has reported thalidomide exposure during pregnancy as a cause[6]. Poswillo[5] points to the varied timing of teratogenic insults in patients with ear deformities associated with mandibulofacial dysostosis (Treacher Collins-Franceschetti syndrome) and more common forms of branchial arch anomalies in hemifacial microsomia.

Pathophysiology

A review of embryology allows a better understanding. Microtia often is associated with atresia or absence of the external auditory meatus, suggesting an arrest of development. The external and middle ear develop from the first (mandibular) and second (hyoid) branchial arches. In most patients with isolated microtia, the ear remnant is positioned with relative symmetry or somewhat superiorly to the contralateral ear[7,8].

The mandible, maxilla, facial musculature, and facial nerve, which also are derived from the same branchial arches, are affected in patients with craniofacial microsomia and microtia. In auricular dystopia, the microtic ear is placed inferiorly and anteriorly compared to the unaffected side. Because the inner (neural) portion of the ear develops on a different time scale and from different ectodermal tissue, most patients have some hearing in the affected ear. In the case of bilateral microsomia, acceptable hearing can be achieved with bone conduction hearing aids and eventual canal, ossicular chain, and tympanic reconstruction. Typically, auditory surgery is performed after auricle reconstruction[9,10].

Presentation

Most patients present as infants or children. The head and neck examination should be complete; search for other evidence of craniofacial microsomia including facial asymmetry, epibulbar dermoids, malocclusion, facial nerve weakness, and macrostomia. Consultation with a geneticist is often helpful to identify special subsets of microtia patients, such as those with Goldenhar syndrome. Genetic consultation is also helpful to the family to identify risk to future progeny of the parents and of the proband[1,2,9].

Indication of surgery

Psychologically, the absence of an ear is significant for both males and females. Even after adjusting hairstyles, the absence of an ear is noticeable to peers and others. Like other patients with blatant anomalies, many children with microtia have lower self-esteem and develop either behavioral problems or become excessively introverted[9].

Opinions vary between surgeons as to the optimal sequence of procedures for reconstruction of the microtic ear. While the approach championed by Brent[11] was well illustrated in the literature, with excellent long term results, newer approaches by Nagata[12] and Firmin[13] had been followed by others, with possibly better results. The basic steps required are similar, but the staging and manner in which those steps are accomplished vary.

The ear reaches approximately 85% of adult size at age 3 years. Growth continues into adulthood but little change in the width or distance from the scalp occurs in individuals older than 10 years. For practical purposes, the normal ear is developed fully by age 6-7 years. To perform an autologous reconstruction, sufficient cartilage must be available. Generally, the costal cartilages are adequate by the time the patient is aged 10 years. The surgeon must balance concerns regarding the psychological impact of the deformity with having sufficient cartilage to carry out the optimal reconstruction in the fewest number of surgeries[11,12].

Treatment

The reconstruction of microtia, regardless of the type and associated deformities, requires 2 main elements. The first is sculpture of a framework from autogenous rib cartilage to reproduce the contours of the ear, and the second is coverage of the framework with the cutaneous remnant and adjacent skin. The greater part, if not the complete reconstruction, can be accomplished in 2 stages, with only minor revisions generally required beyond these two surgeries[1,2,14].

Reconstruction of the external ear can be performed in 1 of 3 fashions: prosthetic replacement, reconstruction with a prosthetic framework, or local tissue and/or flap coverage or reconstruction with an autologous framework[14,15]. Infants should have baseline audiology evaluation of the affected and unaffected ears. Address hearing deficits promptly to prevent speech problems.

Prosthetic ear replacements

Prosthetic ear replacements, when made well, can appear quite natural. The newer osseointegrated anchoring systems are more reliable than older adhesive-based systems. The procedure requires placement of a titanium anchor within the drilled temporal bone. After integration of several titanium posts, the surface magnet system and prosthesis can be attached[16,17].

Prosthetic frameworks

Cronin and Brauer reported prosthetic frameworks for auricular reconstruction in the mid-1960s; they then abandoned them because of the likelihood of erosion and exposure necessitating explantation in virtually all patients if observed long enough. Newer alloplasts also have been marketed with the promise of better tissue integration and spontaneous healing because of vascular ingrowth. The junior author's experience with Porex, reported to heal spontaneously at small sites of exposure, has been disappointing[16,18].

Autologous reconstruction

The criterion standard for external ear reconstruction is autologous reconstruction with cartilage[19]. First reported by Pierce in the 1930s[20] and expanded upon by Converse[21,22] and Tanzer[23] in the 1940s, autologous cartilage reconstruction came to the forefront with Brent in the 1980s[11]. Refinements in framework carving and techniques by the senior author, Nagata[12], and Firmin[13] had increased the artistry and reliability of autologous ear reconstruction.

All autologous reconstructions have 3 main elements in common: (1) construction and placement of a cartilage framework; (2) lobule rotation, conchal excavation, and tragus formation; and (3) elevation of the pinna.

Long-term results of ear reconstruction are gratifying for both patients and surgeons. Autologus reconstruction of the ear can look quite lifelike but does not have the flexibility to touch. If indicated, auditory surgery can be performed after the second stage. Interestingly, growth has been reported in reconstructed ears in children[24,25].

Bangladesh Perspective

The history of microtia operation in Bangladesh is quite long. Different plastic surgeons used to operate on microtia cases sporadically and no proper documentation and publications

are found in this regard. There is no study present to describe the demography and incidence of the case or even the existing number. But if we consider the hospital records of tertiary level Dhaka Medical College Hospital, in last four years (2009-2013) around 43 patients were presented for treatment. Among them 37 were at the age appropriate for surgery. 19 cases were operated by different visiting overseas educator surgical teams, 8 operations by host surgeons and 10 patients were lost to follow-up.

Conclusion

Successful staged reconstruction of the ear requires meticulous attention for available auricular tissue, donor tissue and the surrounding area of planned reconstruction. Whether one can choose a Nagata, Brent or Firmin technique, careful attention to the vascular supply of the skin flaps minimizes the risk of flap ischemia and cartilage exposure. With considerable variation in auricular remnants, meticulous planning is the most important factor in reducing complications. In our country, there is no definitive data regarding microtia and it's reconstruction are present. So, the plastic surgeons of Bangladesh should come forward to place some footsteps in this challenging task. Building on lessons from the past, surgeons have decreased what was a 4-stage operation in the 1950s to a 2-stage reconstruction today. In the future, tissue engineering and newer alloplasts may replace autologous reconstruction; however, they must measure up to the criterion standard set by autologous reconstruction.

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References

1. Brent BD, 2013. Reconstruction of the ear. In: Peter C Neligan eds. Plastic Surgery. Third edition, Vol 3. Elsevier Inc, 187-225.
2. Brent BD, 2005. Reconstruction of the ear. In: Stephen J Mathes eds. Plastic Surgery. Second edition, Vol 4, Philadelphia, USA. Saunders Elsevier.
3. Tolleth H. Artistic anatomy, dimensions, and proportions of the external ear. *Clin Plast Surg*. Jul 1978;5(3):337-45.
4. Melnick M, Myranthopoulus NC, eds. External ear malformations: epidemiology, genetics and natural history. In: *Birth Defects*. Vol 15. 1979.
5. Poswillo D. Otomandibular deformity: Pathogenesis as a guide to reconstruction. *J Max Fac Surg*. 1974;2:64-72.
6. Llano-Rivas I, Gonzalez-del Angel A, del Castillo V, et al. Microtia: a clinical and genetic study at the National Institute of Pediatrics in Mexico City. *Arch Med Res*. Mar-Apr 1999;30(2):120-4.
7. Lipan MJ, Eshraghi AA. Otologic and audiology aspects of microtia repair. *Semin Plast Surg*. Nov 2011;25(4):273-8.
8. Poswillo DE. Discussion of Bennun RD, Mulliken JB, Kaban LB, Murray JE. Microtia: A microform of hemifacial microsomia. *Plast Reconstr Surg*. 1985; 76: 864-865.
9. Snively SL. Plastic surgery of the ear. *Select Read Plast Surg*. 1994;7:1-26.
10. Takahashi H, Maeda K. Survey of the familial occurrence of 171 microtia cases. *Jpn J Plast Surg*. 1982;15:310.
11. Brent B. The correction of mi-rotia with autogenous cartilage grafts: I. The classic deformity. *Plast Reconstr Surg*. Jul 1980;66(1):1-12.
12. Nagata S. A new method of total reconstruction of the auricle for microtia. *Plast Reconstr Surg*. Aug 1993;92(2):187-201.
13. Firmin F. Controversies in autogenous auricular reconstruction. *Presented at: Ear Reconstruction '98: Choices for the Future;* March 4-6, 1998;Lake Louise National Park, Alberta, Canada.
14. Im DD, Paskhover B, Staffenberg DA, Jarrahy R. Current management of microtia: a national survey. *Aesthetic Plast Surg*. Apr 2013;37(2):402-8.
15. Yang SL, Zheng JH, Ding Z, Liu QY, Mao GY, Jin YP. Combined Fascial Flap and Expanded Skin Flap for Enveloping Medpor Framework in Microtia Reconstruction. *Aesthetic Plast Surg*. Oct 7 2008;
16. Romo T 3rd, Morris LG, Reitzen SD, Ghossaini SN, Wazen JJ, Kohan D. Reconstruction of congenital microtia-atresia: outcomes with the Medpor/bone-anchored hearing aid-approach. *Ann Plast Surg*. Apr 2009;62(4):384-9.
17. Oyama A, Fujimori H, Funayama E, Yamamoto Y. Intraoperative simulation device using negative pressure for construction of framework in microtia reconstruction. *Plast Reconstr Surg*. Mar 2008;121(3):129e-130e.
18. Helling ER, Okoro S, Kim G 2nd, Wang PT. Endoscope-assisted temporoparietal fascia harvest for auricular reconstruction. *Plast Reconstr Surg*. May 2008;121(5):1598-605.
19. Lee LN, Boahene KD. A Novel Technique for Sculpting Costal Cartilage in Microtia Repair and Rhinoplasty. *JAMA Facial Plast Surg*. Jun 13 2013;1-3.
20. Pierce GW. Reconstruction of the external ear. *Surg Gynecol Obstet*. 1930;50:601.
21. Converse JM. Reconstruction of the auricle - Part I. *Plast Reconstr Surg*. 1958;22:150-163.
22. Converse JM. Reconstruction of the auricle - Part II. *Plast Reconstr Surg*. 1958;22:230-249.
23. Tanzer RC. Total reconstruction of the external ear. *Plast. Reconstr. Surg*. 1959;23:1-15.
24. Thomson HG, Winslow J. Microtia reconstruction: does the cartilage framework grow?. *Plast Reconstr Surg*. Dec 1989;84(6):908-15.
25. Tolleth H. A hierarchy of values in the design and construction of the ear. *Clin Plast Surg*. Apr 1990;17(2):193-207.