



RESEARCH ARTICLE

“HEAR FOR ALL”: A SPECIAL CLUB FOR PATIENTS WITH EXTERNAL EAR DEFORMITY

*Ratna Priya

Seth G S Medical College and KEM Hospital, India

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ABSTRACT

Hear for all club started at the KEM hospital, Mumbai is an autonomous and selfless association that is dedicated to patients with congenital external ear deformity. Here, we focus on evaluating all patients with microtia and providing them with the best treatment plan. It has been one of the first efforts of its kind in the country. it works as a support group.

Methods: Our interdisciplinary team consists of an ENT surgeon, a plastic surgeon, an audiologist, a dentist, a speech therapist, a paediatrician, radiologist, paediatric cardiologist and paediatric nephrologists and we all work together to give the child the best coordinated care. Activities include complete ENT and head and neck evaluation of the patient to rule out syndromic features, Baha trial, Baha surgery and holding meetings which give an opportunity to the patients and their families to interact, share their experiences and motivate each other

Results: Beginning in 2011, at present we have a group of 60 patients of congenital external ear deformity which include 40 children and the rest are adults. Out of the 60 patients, 12 patients have undergone Baha surgery.

Conclusion: Our club helps us to reach out to patients with microtia and their families and bring all of them together at one place and provide information, reassurance and guidance to deal with the microtia. Also, the presence of an interdisciplinary team helps in planning the best treatment options for the patient.

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INTRODUCTION

Congenital external ear deformity in children and adults is worrisome and perplexing for the patients and their family members. Though it may appear that the condition may not have a significant impact on speech development as it may be unilateral with other ear being normal. The prevalence of microtia is seen in 2.35 in 10,000 live births and thus is quite rare to encounter (Leoncini, 2011). The etiology is multifactorial and it may be idiopathic or occur either as a result of chromosomal abnormality or due to antenatal exposure to rubella and other viruses or teratogenic agents such as thalidomide and alcohol (Weerden, 2007). The physical deformity of microtia causes significant and emotional impact on patients and their families and is a constant cause of anxiety and concern among them. Parents of children are rightly concerned about the psychological health of their child, knowing that having only one ear carries a higher chance of being bullied and singled out during the school years (Granstrom *et al.*, 1993; van der Pouw *et al.*, 1998; Tjellstrom *et al.*, 1995).

The absence of ear or an abnormally looking ear provokes hatred in different communities and in some places they are even considered as evil and cursed. Such people are humiliated and have low self esteem, increased tension and may become a loner as they try to escape social meetings as much as possible (Schwentner, 2006). In addition to this, children with congenital external ear malformations get bullied a lot and this leads to psychological fear and poor performance at school. MacGregor studied and described this as “exquisite cruelty of young children towards children who happen to look different” (Macgregor, 1978) precisely known among psychologists as peer victimization (Kumpulainen, 1999). We also know that the hearing loss associated with microtia and external auditory canal atresia leads to speech and learning difficulties. Hear for all club started at the KEM hospital, Mumbai is an autonomous and selfless association that is dedicated to patients with congenital external ear deformity. Here, we focus on evaluating all patients with congenital external ear malformations and providing them with the best treatment plan. It provides a platform where patients with microtia and their family members can meet and talk about their problems, so that they don't feel let down that they are the only one who are affected by this condition.

*Corresponding author: Ratna Priya,
Seth G S Medical College and KEM Hospital, India

Objectives

It has been one of the first efforts of its kind in the country. Here, since all patients with microtia and their families face similar problems, they are brought together and thus it works as a support group. It helps to provide information, reassurance and guidance as most parents have never even heard of this condition before their child is born with it. Hence it helps in spreading awareness and better patient counselling regarding the deformity and its management. The thought came when at one given time, two to three very young children attended our outpatient department with bilateral microtia. We started counselling them for Baha band and one of the patient's parents got convinced and agreed for Baha band and started wearing it. With this example it was very easy to convince the other patients and this gave the idea of forming a group or association of them. This was also inspired from the microtia clinic at "The Royal Children's Hospital in Melbourne, Australia which constitute of plastic, maxillofacial and ENT surgeons who specially treat children with ear deformities since birth as well as after trauma and provide leading care to such patients. Another organisation working with the similar goal is 'The Ear Community' constituting of patients with microtia and various healthcare professional working towards their betterment.

MATERIALS AND METHODS

KEM being a tertiary care centre, our interdisciplinary team consists of an ENT surgeon, a plastic surgeon, an audiologist, a dentist, a speech therapist, a paediatrician, radiologist, paediatric cardiologist and paediatric nephrologists and we all work together to give the child the best coordinated care. Since, we have been getting referrals of patients with microtia from all over the country we have come with our protocol which involves few visits in which we plan treatment for the patients. In the first visit, details and relevant history are noted in a case record form. Complete ENT and head and neck examination is done to look for syndromic features if associated in the patient. Photographs are taken in frontal view as well as right and left lateral views as a part of documentation (Fig.1, Fig.2, Fig.3). Hearing is assessed using pure tone audiometry in adults and BC Bera in young children. According to the literature, microtia is associated either with a single congenital defect or may occur as part of syndrome and among them the most common anomalies are facial cleft, facial asymmetry, renal abnormalities, cardiac defects, microphthalmia, polydactyly, and vertebral anomalies (Simon *et al.*). Paediatric evaluation is done as part of routine evaluation and also to look for these congenital anomalies. Cardiology reference and two dimensional echocardiography is done to rule out congenital heart diseases if any. Ultrasonography of the abdomen is an important investigation in our protocol to detect renal anomalies as congenital external ear malformations have a higher risk of renal anomalies as depicted in various studies such as that of Wang *et al.* (2001). In the second visit, patients are given Baha trial so that we can give Baha soft band (Fig. 4) as early as possible to young children less than five years of age and also we can assess the improvement patients will be getting following Baha surgery. High resolution computerised tomography of the temporal

bone is done to measure bone thickness behind temporo-parietal suture line. This helps us to plan surgery as single stage surgery is done if bone thickness is more than 2.5 mm where fixture placement and abutment insertion are done in the same stage. In case the bone thickness is less than 2.5 mm, a two stage surgery is planned. After detailed investigations, work up and pre anaesthesia check up, patients are dated for Baha surgery. Till now we have successfully implanted 9 children with microtia, (Fig.5, Fig.6).

RESULTS

Beginning in 2011, at present we have a group of 60 patients of congenital external ear deformity which include 40 children and the rest are adults. Out of the 60 patients, 12 patients have undergone Baha surgery.



Fig.1. Child with Marx Grade 3 Microtia



Fig. 2. Child with Goldenhar syndrome with bilateral microtia, hemivertebra, cleft lip, cleft palate and hemimandibular agenesis



Fig. 3. Child with bilateral microtia with microphthalmia



Fig. 4. Child using Baha soft band



Fig. 5. Baha implant in place

We have been holding monthly meetings for these patients in which we counsel and assess the patient, get the necessary

specialist consultation and perform Baha trials. This gives an opportunity to the patients and their families where they interact, share their experiences, motivate each other and thus it functions as a support group. Since implant cost is an important factor, we also help and guide the families with donations from various government institutions and social groups for the surgery as well as Baha soft band.



Fig. 6. Baha implant with processor in place

DISCUSSION

Microtia patients and their families pay more attention to the obvious external deformity and they want it to be corrected as soon as possible. But the surgery for correcting external deformity takes place only after 7 years of age and needs technical expertise (Chandrasekhar, 1995; Im, 2013). Pinna reconstruction is an elective procedure and is multistaged. The deformity can usually be masked by a longer hair style. Ear prosthesis can also be an option for some individuals with microtia-antia. There can be difficulties in matching the appearance of the other ear, and it is just for cosmesis (Simon). Also external auditory canal reconstruction is a challenge in itself. The congenital hearing loss has to be dealt as early as possible as this will help in proper speech and language development. The Baha system can be a real solution for people with this type of impairment. The Baha implant is directly integrated to the skull bone. Because of this direct interface, the Baha offers significantly better sound quality than that of a traditional bone conductor. The Baha implant works without pressure on the skin avoiding headache and soreness associated with the conventional bone conductor hearing aid. Baha offers excellent wearing comfort and a better aesthetic result. In children with bilateral atresia, amplification with bone-conduction aids should be provided as soon as possible, preferably within the first few months of life. For children younger than 5 years of age, Baha soft band can be given so that early amplification and training is started. Bone thickness behind temporo-parietal suture line in patients with congenital external ear deformity is poor as compared to patients with normal looking ear and this becomes challenging. In case the thickness is less than 2.5mm we have to consider for two staged implant. Though Baha has many indications but we feel that this indication for bilateral congenital conductive

hearing loss is the best one. However, there are challenges as bone is also of poor quality and it mostly never matches the bone thickness of normal person of same age group.

Promotion

Our "Hear for all" club at the KEM Hospital was launched on 14 November 2011, which is celebrated as children's day as the birthday anniversary of our beloved prime minister Pandit Jawaharlal Nehru. He was extremely fond of children and became famous as Chacha Nehru. He strongly believed that children are the asset of the country and he worked very hard for their education, welfare and progress. The inauguration was done by Salman Khan, Bollywood actor and founder of Being Human Foundation, on a big scale at the KEM hospital and this was done keeping in mind that the publicity and wider mass media address would help us to spread awareness and at the same time attract donors and collect funds necessary for the implant of all our microtia children as the cost of Baha implant stands out as an important factor as most of the patients have poor socio economic status. Congenital external deformity is not covered in health insurance and we also lack a national health scheme related to the same, thus the availability of Baha implant to the patients is solely dependent on the mercy of the donors. The social worker at KEM hospital takes an active part in providing financial assistance to the patients for the implant.

Compliance with Ethical Standards: Disclosure of potential conflicts of interest: None of the authors have any conflict of interest

Ethical approval: All procedures performed were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

Informed consent: Informed consent was obtained from the patient included in this study.

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Conclusion

Thus, our club helps us to reach out to patients with congenital external ear deformity and their families and bring all of them together at one place and provide information, reassurance and guidance to deal with the microtia. Also, the presence of an interdisciplinary team helps in planning the best treatment options for the patient. This has also helped in spreading awareness regarding the deformity and the social stigma attached to it.

Not to forget its role in attracting donors for the implant. We would also like to mention that since pediatricians are the first ones to deal with such patients, they can be of great help in counselling and guiding the patients.

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