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Maxillofacial Prosthetic Management of a Patient with Hemifacial Microsomia

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The treatment of choice for patients with hemifacial microsomia has traditionally been surgical reconstruction. The use of craniofacial implants has afforded the option of an implant-retained prosthesis.¹ This article presents a case where this craniofacial anomaly was treated using an implant-retained auricular prosthesis for replacement of the pinna. This method of management offers a viable approach to the patient while involving a multidisciplinary team of clinicians. This provides comprehensive care, with improvement in the quality of life for such individuals.

Introduction

Hemifacial microsomia (HFM) is a disorder that results from the underdevelopment of structures within the first and second branchial arches. It is also the second most common craniofacial malformation, following cleft lip and palate.² Due to a wide range of phenotypic expressions, a variety of names have been used to describe HFM. They include Goldenhar syndrome, otomandibular dysotosis, first and second branchial arch syndrome, oculoauriculovertebral sequence, lateral facial dysplasia, and craniofacial microsomia.³

The etiology of HFM is complex. Teratogens, such as retinoic acid primidone and thalidomide, as well as genetic factors have been implicated. Both autosomal dominant and recessive inheritance patterns have been postulated to explain familial cases of HFM.

The clinical manifestations of HFM include a wide range of expressions. The disorder is especially recognized by its facial asymmetry, due to the agenesis, hypoplasia and/or displacement of the pinna. Maxillary, temporal, and malar bones on the involved side are often reduced in size and flattened. Furthermore, some patients may exhibit an underdeveloped mastoid region. The eye on the side of the defect may be slightly lower than its counterpart, and the external ear of the involved side may range from distortion to complete aplasia. In some cases, bilateral anomalous pinnas and conduction deafness due to middle ear abnormalities are observed. Intraorally, patients with HFM may have hyperplastic or aplastic enamel, significant delay of tooth development on the affected side, or absence of the mandibular third molar and other teeth on the affected side. Chalky opacities in enamel are often found on the maxillary central and lateral incisors on the affected side and serve as a distinguishing feature in individuals with HFM. Other craniofacial

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features associated with HFM include cleft lip and/or palate, hypoplasia of facial muscles, and unilateral colobomas of the superior eyelid. Approximately half of the patients with hemifacial microsomia exhibit occipitalization of the atlas, cuneiform vertebra, partial or complete synostosis of two or more cervical vertebrae, supernumerary vertebrae, spina bifida, and anomalous ribs.²

Case Report

A 26 year-old female with HFM presented to New York Presbyterian Hospital for treatment of atresia of the right ear. Her left ear had previously been surgically reconstructed using tissue from her thigh (fig. 1). Her past surgical history included orthognathic correction of a Class III asymmetric maxillary and mandibular relation. In this procedure, her left jaw was reset.

A multidisciplinary team, which included maxillofacial prosthodontists and otolaryngologists, evaluated the patient's options and assessed her extra-oral needs. Based on the literature and current methodologies, it was decided that the patient would benefit best from a bone anchored hearing aid (BAHA) and 3 craniofacial implants for an implant-retained prosthesis (fig. 2).

To achieve success, the following pre-surgical protocol is observed:

- 1. Identify the exact location of the atretic ear in relation to the other anatomic structures.
- 2. Make an impression of the defect.
- 3. Fabricate a diagnostic wax up of the ear.⁴
- 4. Make clinical modifications to the diagnostic ear in consultation with the patient.
- 5. Design a surgical template and determine the craniofacial implant location (20 mm from the external ear canal at 9 and 11 o'clock positions on the right side, and 1 and 4 o'clock on the left side.)²



Figure 1 Patient's left ear surgically reconstructed. Notice lack of detail of the tragus and lobe, and color/shading irregularities.

In the post-surgical stage, the procedure continues as follows:

- 1. Make the implant connection for the BAHA.
- 2. Make an impression incorporating the implant.
- 3. Fabricate the working cast of the ear.
- Design and fabricate the framework for the orienta tion of the prosthesis; this may include either a mag netic retention or a retentive bar and clip retained structure.

The prosthetic ear must then be sculpted and modified. A casting in silicone is made and extrinsic coloring is done. Afterwards, the patient is given instruction in the maintenance of the prosthesis and tissue around the implant abutments and told to return for regular follow up care (fig. 3).

In this case, the craniofacial implant was placed at 8 and 11 o'clock positions on the right side. Custom staining of the prosthesis was done, and the implantretained auricular prosthesis was made using 2 clips and a type III gold bar. The gold bar was cantilevered to retain the clips since the craniofacial implants were placed slightly anterior to avoid the mastoid air cells.

The patient's intra-oral needs are being treated in a multidisciplinary fashion between prosthodontics, orthodontics, oral and maxillofacial surgery, and periodontics.

Discussion

There are several concerns about using an implantretained auricular prosthesis. They present a challenge with respect to the adaptation of the visible margins, as this may adversely affect the esthetic results.⁵ With



Figure 2 Bar and clip assembly in place to receive prosthetic ear. Notice bone anchored hearing aid posterior and superior to the bar.

changes in head position and condylar movement, loss of tissue contact at the anterior margin of the silicone prosthesis with the underlying skin can occur. It is important for the exposed anterior margin of the prosthesis to maintain skin contact at all times for its esthetic value to be preserved. Besides the cosmetic value, an implant-retained prosthesis has the following distinct advantages: easily cleaned, more hygienic with less skin reactions than when sticky adhesives are used, and possible thinner margins. With retention no longer a problem, patients feel much more secure psychologically.⁶

Long term follow-up results of craniofacial titanium implants in adult subjects without irradiation are favorable. A long-term follow-up study of BAHA demonstrated that although 90 percent have stable implants after 8 years, adverse skin reactions (a complication of BAHA) do occur in 7% of the patients.⁷

After the delivery of the auricular prosthesis, it was noted on follow-up that the patient had changed her hairstyle to expose the prosthetic ear (fig. 4). This demonstrated her satisfaction of the new image that the auricular prosthesis helped create.

Psychosocial problems in children with craniofacial deformities, such as HFM, include lack of emotional attachment between parents and child, inadequate development of peer relationships, and shame related to poor body image.² Consequently, treatment should provide for patient confidence post-operatively.

Conclusion

With the introduction of craniofacial implants, an implant-retained auricular prosthesis is a viable option



Figure 3 Prosthetic ear in place. Notice anatomic details, color and overall esthetics.

for patients with HFM because of its predictable results. The advantages of an implant-retained auricular prosthesis are summarized as follows:⁸

- 1. It is a relatively short and less demanding surgical procedure.
- Ît can be done as an outpatient procedure under local anesthesia.
- 3. It creates a greater similarity to the contralateral ear.
- 4. It affords a greater ease of replacement or correction
- if the prothesis is unsatisfactory.

The disadvantages are as follows:

- It eventually needs replacement, since silicone products degrade with time.
- 2. It requires home care by the patient.
- 3. It requires continuous follow-up visits to assess the skin surrounding the implant abutments.
- It may be difficult to incorporate the prosthesis into body image, although this is also a disadvantage with conventional adhesive-retained prostheses.

This article discussed the prosthetic management of a patient with HFM emphasizing the use of an implantretained auricular prothesis for the replacement of the right ear. The prognosis for this patient treated with a BAHA and implant-retained auricular prothesis is very favorable.

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Figure 4 Patient satisfied with result. Her shorter haircut reflects confidence in her appearance due to the good esthetics of her new prosthetic ear

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