Case Report

Maxillofacial Prosthetic Management of an Auricular Defect for a Young Patient With Hemifacial Microsomia: A Clinical Report

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Abstract
Facial anomalies in hemifacial microsomia patients may have significant psychosocial impact even from a very young age. The management and fabrication of an auricular prosthesis replacement supported by endosseous craniofacial implants for a young patient with Goldenhar-Gorlin Syndrome has been reported. It is beneficial for the defects of a hemifacial microsomia patient to be managed as early as possible, consistent with the patient’s ability to manage the prosthesis. [Singapore Dent J 2011;32(1):33–38]

Key Words: craniofacial anomalies, hemifacial microsomia, Goldenhar-Gorlin syndrome

Introduction
Goldenhar-Gorlin Syndrome is a variant of the developmental disorder which falls under the umbrella of syndromes associated with the Oculo-Auriculo-Vertebral Spectrum (OAVS). It is associated with unilateral deformities embryologically related to the first and second brachial arch derivatives.1 An incidence of 1 in 5600 live births was proposed by Gorlin to be the most accurate prediction of its frequency.1 It is estimated to be the fourth most common craniofacial anomaly after cleft lip with or without cleft palate, cleft palate and craniosynostosis.2

OAVS is characterized by associated hemifacial microsomia, epibulbar dermoids, auricular appendages, blind-ended auricular fistulars, vertebral anomalies3 and hypodontia on the affected side of the head.4 These characteristics are often present in different combinations, and also varying degrees of severity along the spectrum. Auricular defects often with hearing loss, followed by unilateral facial and ocular deformities with right sided predilection, was found to be the most consistent findings among patients with OAVS.1

Various genetic5 and environmental factors such as assisted fertilization,6 paternal service in the Gulf Wars,7 maternal smoking and drug use,8 and diabetic status9 have been suggested as potential pathogenic mechanisms contributing towards fetal development of OAVS. It is highly likely that the aetiology of this developmental disorder is multi-factorial in nature.

Clinical Report
A 7-year-old healthy Chinese male diagnosed with Goldenhar-Gorlin syndrome was referred to the Graduate Prosthodontic clinic from the Otorhinolaryngology (ENT) clinic for prosthetic assessment and management at the National University Hospital, Singapore (Figure 1). The patient presented with complete aplasia of the right external...
auditory meatus and conduction deafness associated with middle ear abnormalities. It was the parents’ main concern to replace the missing right ear for the psychological well-being of the patient.

Clinical and radiographic evaluations were carried out. Preliminary clinical examination showed that the patient presented with gross facial asymmetry with reduced vertical facial proportion on the right side of the face in comparison to the contralateral side. Surgical and prosthetic replacement options were discussed with the parents during a multidisciplinary clinic. The treatment aim was to attempt concurrent replacement of the missing right ear and restoration of hearing function. An implant-retained prosthesis was planned.

**Treatment Sequence**

Two craniofacial implants (3.75 × 3.0 mm; Entific Medical System, Göteborg, Sweden) were placed in the right temporal bone (posterior and superior to the auricular canal) by the ENT surgeon under a General Anaesthesia. A surgical template was utilized to aid in the placement of the implants (Figure 2).

One craniofacial implant was placed in the right mastoid process to receive a Bone Anchored Hearing Aid (BAHA; Entific Medical System).

At second stage surgery, the implants placed in the temporal bone were exposed. The tissue overlying the implants was thinned surgically down to approximately 2 and 4 mm healing abutments (Standard abutment; Entific Medical System) were inserted. The subcutaneous tissue around the BAHA implant was also surgically thinned and skin-grafted.

After 6 weeks of soft-tissue healing, the patient was reviewed in the Graduate Prosthodontic clinic and an impression was made for the two anterior implants on the right temporal area using vinyl polysiloxane material (Aquasil Ultra LV; Dentsply Caulk, Milford, Del) to fabricate the implant retained auricular prosthesis (Figure 3A).

The impression was poured in type IV stone (Silky Rock; Whip Mix Corp, Louisville, Ky) (Figure 3B) and wax sculpting (Modeling wax; Dentsply, Konstanz, Germany) of the right ear was developed according to the contralateral ear. The wax sculpting was tried on clinically for esthetic assessment (Figure 4A–4C).

The sculpting was invested and the wax was boiled out before separating the cope and drag of the flask.

The tissue bar was designed on the master cast by visually checking with the cope to ensure sufficient space for acrylic resin housing and the silicone prosthetic material.

The tissue bar framework was established using gold cylinders (4 mm; Entific Medical System) and round plastic bar (Plastic bar; BIOMET 3i, West Palm Beach, FL, USA) and casted in noble alloy (Bond on-4; Degussa, Hanau, Germany). The framework was tried in clinically, sectioned and soldered to achieve passive fit over the implants (Figure 5).

The tissue bar was returned to the definitive cast and four metal clip attachments (Clip attachment 2 mm; Entific Medical System) were placed
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Figure 3. (A) Six weeks post-surgical placement of craniofacial implants. (B) Impression was poured in Type IV stone (Silky Rock).

Figure 4. (A) Wax sculpturing of the auricular prosthesis. (B) Patient’s contralateral ear. (C) Wax sculpting was tried on clinically for esthetic assessment.

An autopolymerized acrylic resin housing (Quick Resin, Shofu, GC, Japan) with 4 clip attachments was fabricated and the flask cope was used to visually check that 2 mm of space allowance was present for the silicone prosthetic material\textsuperscript{10} (Figure 6A). Retentive undercuts and perforations were made on the acrylic resin housing (Figure 6A and 6B).

The sculpting and acrylic resin housing were flanked and the wax was boiled out. The acrylic resin housing and tissue bar were finished and processed with silicone elastomer (Dow-Corning 2186; Factor II, Arizona, USA) to complete the auricular prosthesis.

The processed silicone auricular prosthesis was tried on clinically and was extrinsically colored (Earth Color; Factor II, Arizona, USA) to match the patient’s complexion. This process was observed and verified with the patient’s parents. The auricular prosthesis was delivered to the patient upon curing of the extrinsic coloration. Hygiene and
Figure 6. (A) Acrylic resin housing with four 2 mm clip attachments. (B) Assessment of space allowance for silicone material. (C) Completion of sculpture wax up.

maintenance instructions were given. At 6 months recall, the patient and parents were still satisfied with the cosmetic result achieved (Figure 7).

Discussion

This case report illustrates the role of the maxillofacial prosthodontist, and the importance of a multidisciplinary approach to management of a young patient with Goldenhar-Gorlin Syndrome. It also validates the method of using a thermoformed shell guide, to ensure proper spatial relationship among the implant tissue bar, retentive elements and external contour of the auricular prosthesis, while not encroaching onto the space of the Bone Anchored Hearing Aid implant.

Replacement of a missing ear in a pediatric patient may be achieved with either surgical reconstruction or prosthetic replacement. While autogenous reconstruction remains the treatment of choice in pediatric patients with microtia, prosthetic reconstruction of the auricle is considered under the following circumstances: (1) awaiting rib cartilage reconstruction, (2) failed autogenous reconstruction, (3) severe soft-tissue/skeletal hypoplasia, (4) a low or unfavorable hairline, (5) acquired total or subtotal auricular defect and (6) to avoid multiple and longer surgical procedures.

Surgical ear reconstruction in the pediatric patient has the advantage of providing a stable, low-maintenance ear reconstructed from autogenous cartilage framework. It also has the potential to have continued growth of the grafted cartilage over time. However, the patient will need to undergo multiple and longer surgical procedures. Acceptable facial symmetry and aesthetics is more difficult to achieve as compared to a sculptured auricular prosthesis.

On the other hand, prosthetic augmentation confers superior aesthetics at a considerably lower cost and risk to the young patient.

Prosthetic retention is generally achieved via use of anatomical undercuts, use of adhesives, or through the use of implants. In the replacement of an auricular prosthesis, anatomical undercuts usually do not provide any effective retentive elements. Adhesives have been shown to cause degradation and color changes to the silicone prosthesis. Repeated application and removal of the prosthesis may also result in damage to the prosthetic ear as well as tissue irritation to the patient.

Studies have shown greater patient satisfaction with implant retained over adhesive-retained
prosthesis. Osseointegrated craniofacial implants provide enhanced retention, stability, and ease of maintenance of a maxillofacial prosthesis. The use of a craniofacial implant-retained prosthesis has been shown to be a viable alternative to a surgical reconstruction approach. Surgical placement of craniofacial implants is relatively less demanding in comparison with the reconstruction procedure. Success of craniofacial implants has been shown to strongly correlate with anatomic sites and exposure to radiotherapy, with implants placed in the auricular sites displaying the highest success rates among other craniofacial anatomic sites.

On the other hand, failure of osseointegrated implants placed in the temporal bone is still possible. Some patients may also experience redness and irritation around the soft tissues surrounding the implant. Occasionally, granulation tissue may form around the abutment resulting in infection of the peri-implant soft tissues and subsequent implant loss. The importance of the patient’s compliance with hygiene measures and timely adjustments by the clinician at follow up visits cannot be over-emphasized. Ultimately, replacement of the prosthesis will still be required over time due to degradation of the silicone prosthetic material itself.

It was found that ear width reached its mature size by age 7 and attained its full length by about age 13 in males. The optimal age proposed for a child to begin wearing an auricular prosthesis is between the age of 6 and 9. It is postulated that the child should have attained a certain level of maturity sufficient to want the prosthesis and is also able to help care for it. Compared with surgical auricular augmentation, prosthetic reconstruction would allow periodic adjustment to account for the change in ear size in growing individuals. During the discussion of treatment options with the parents, special consideration was given to advise them on the necessity for several replacements of the prosthesis throughout the child’s growth phase.

Adolescents with craniofacial anomalies have demonstrated elevated risk for problems with academics and peer relationships. It has been reported that 75% of adolescents with craniofacial anomalies cited teasing or bullying about their appearance as causing considerable distress. Studies have shown that children with facial anomalies received lower preferences as playmates than other physical differences. This behavior of social avoidances among children, which appears to be similar in other parts of the world, may account for an increased risk of impaired psychosocial functioning and stigma experiences in children and young adults with craniofacial anomalies. It may be imperative that some form of surgical/prosthetic reconstruction be provided for even a young child to facilitate normal social interaction with peers and to improve their overall psychosocial well-being.

The external ear is a challenging prosthesis to fabricate. Aesthetic appearance has since gained greater emphasis in society, and a missing right ear will have a significant psycho-social impact on a growing child. It is therefore beneficial for the defects of a hemifacial microsomia patient to be managed as early as possible consistent with the patient’s ability to manage the prosthesis.

Summary

This article outlined the maxillofacial prosthetic management of a young individual with hemifacial microsomia.

References


