Hemifacial microsomia and treatment options for auricular replacement: A review of the literature

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Statement of problem. Although surgical reconstruction is the treatment of choice for auricular deformities that result from hemifacial microsomia, the implant-retained auricular prosthesis must be considered when surgical reconstruction is not possible. The competent and successful practitioner should be knowledgeable of the nature of this congenital disease.
Purpose. This article reviewed the first and second branchial syndrome, treatment approaches, and potential advances in surgical and prosthetic rehabilitation for patients with hemifacial microsomia.
Conclusion. Advantages and disadvantages of autogenous and alloplastic ear reconstructions are discussed. New research initiatives, such as tissue engineering and fabrication of auricular prosthesis by CAD/CAM, offer the potential for improved treatment for the future treatment of hemifacial microsomia. (J Prosthet Dent 1999;82:197-204.)

CLINICAL IMPLICATIONS

This article provides a review of the characteristics of hemifacial microsomia. Practical application of this information should enable the clinician to develop a better understanding of the disease and to work with other specialists in treating hemifacial microsomia patients.

The term *hemifacial microsomia* (HFM) was first used by Gorlin to refer to patients with unilateral microtia, macrostomia, and failure of formation of the mandibular ramus and condyle.[1] It is the second most common craniofacial malformation after cleft lip and palate. In the past, HFM has been the purview of various medical specialists, each preoccupied with 1 or 2 anatomic areas and each with a particular technical expertise. Reconstructive surgeons have struggled with the external ear anomalies, microphthalmia, and soft tissue and muscle defects. Otolaryngologists have been concerned with hearing disorders, middle ear anomalies, and airway obstruction. Oral surgeons and orthodontists have focused on occlusal and jaw abnormalities. These specialists, all concerned with HFM, are beginning to work together in craniofacial teams. Prosthodontists also become members of craniofacial teams to offer expertise in prosthetic reconstruction of the external ear using craniofacial implants as a support or as a retention system for prosthetic rehabilitation.

The purpose of this article is to review the literature regarding characteristics of HFM and to discuss treatment options available to patients who have auricular deformities or absence of the auricle to HFM.

HEMIFACIAL MICROSOMIA

Incidence

Poswillo[2] suggested a frequency of HFM was 1 per 3500 births, although there was no data to support this claim. Grabb[3] estimated an incidence of at least 1 per 5600 births. The male-to-female ratio is 3:2, and there is also a 3:2 ratio of right-side versus left-side involvement.[4-6] Because of an extraordinarily wide range of phenotypic expression, various nomenclature are applied to HFM such as Goldenhar-Gorlin syndrome,[7] first arch syndrome,[8] lateral facial dysplasia,[9] unilateral craniofacial microsomia,[10] otomandibular dysostosis,[11] oculoauriculovertebral dysplasia,[12] auriculo-branchiogenic dysplasia,[13] and oculoauriculovertebral spectrum.[14]

Etiology

The cause is unresolved and complicated. Teratogenic and genetic components have been examined by many investigators.[15-25] Reports indicate that several teratogenic agents, such as retinoic acid, primidone, and thalidomide, have produced HFM in infants born to pregnant women exposed to those agents. Poswillo[26,27] produced facial anomalies in laboratory animals by maternal intake of 10 mg/kg thalidomide or 60 mg/kg triazene. He believed that either total or

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Table I. The skeletal, auricle, and soft tissue (SAT) classification system of hemifacial microsomia[58]

Skeletal categories	Auricle categories	Soft tissue categories
S_1 = Small mandible with normal shape	A_0 = Normal	T_1 = Minimal contour defect with no cranial nerve involvement
S_2 = Condyle, ramus, and sigmoid notch identifiable but grossly distorted; mandible strikingly different in size and shape from normal	A_1 = Small, malformed auricle retaining characteristic features	T_2 = Moderate defect
S_3 = Mandible severely malformed, ranging from poorly identifiable ramal components to complete agenesis of ramus	A_3 = Rudimentary auricle with hook at cranial and corresponding to the helix	T_3 = Major defect with obvious facial scoliosis, possible severe hypoplasia of cranial nerves, parotid gland, muscles of mastication; eye involvement; clefts of face or lips
S_4 = An S_3 mandible plus orbital involvement with gross posterior recession of lateral and inferior orbital rims	A_3 = Malformed lobule with rest of pinna absent	
S_5 = The S_4 defects plus orbital dystopia and frequently hypoplasia and asymmetrical neurocranium with a flat temporal fossa		

incomplete development of stapedial artery caused by an expanding hematoma could trigger a localized necrosis in the derivatives of the first and second branchial arches which would cause HFM.[26,27] Granström[28-30] also induced microtia and other craniofacial defects in rats by injecting excessive dose of retinoic acid or etretinate to pregnant rats.

The association of chromosomal anomalies with hemifacial microsomia has been documented.[31,32] Clinical observations of chromosomal defects are likely to be those karyotypes. Discordance in monozygotic twins with HFM has been noted frequently.[33-36] Most cases of HFM are sporadic, but familial instance has been reported,[37] but successive generations of affected persons[38-41] and affected siblings with normal parents have also been reported. Autosomal dominant and autosomal recessive inheritance have both been hypothesized to explain various familial occurrences.[42-44]

Clinical manifestations

Though there is extreme variability of expression for HFM, it is especially recognized by facial asymmetry.[45] This is due in part to absence, hypoplasia, and/or displacement of the pinna, but the degree of involvement is markedly variable. Maxillary, temporal, and malar bones on the involved side are somewhat reduced in size and flattened. Malformation of the external ear may vary from a complete aplasia to a crumpled, distorted pinna that is displaced anteriorly and inferiorly.[46] Occasionally, bilateral anomalous pinnas are noted. Approximately 40% of patients with microtia have varying degrees of the syndrome. Conduction deafness due to middle ear abnormalities and/or absence or deficiency of the external auditory meatus has been noted in 30% to 50% of cases.[47] Supernumerary ear tags may occur anywhere from the tragus to the angle of the mouth.

Intraoral deformity include hyperplastic or aplastic teeth and enamel. Common oral manifestations of HFM include a significant delay of tooth development on the affected side, frequent absence of mandibular third molar on the affected side, and increased frequency of missing teeth on the affected side. The incidence of delayed tooth development with HFM is proportional to the extent of mandibular deformity.[48,49] Hypodontia is found particularly in the mandibular second premolars involving the affected side. Chalky opacifications of enamel are occasionally found on the maxillary central and lateral incisors of the underdeveloped side as a marker of development for HFM.[50]

Patients may have minimal underdevelopment of the condyle to unilateral aplasia of the mandibular ramus and/or condyle with absence of the glenoid fossa; 50% to 70% of HFM patients have agenesis of the ramus on the affected side.[51] The maxilla is narrowed on the involved side with decreased palatal width. Associated cleft lip and/or palate is found in 7% of HFM patients.[3]

Hypoplasia of facial muscles, such as the masseter, temporalis, pterygoideus, and those of facial expression on the involved side has also been observed. Narrowing of the palpebral fissure occurs on the affected side in about 10% of patients.[52] Clinical microphthalmia or anophthalmia has been reported and the ipsilateral eye may be at a lower level than that on the opposite side. Unilateral colobomas of the superior lid is a common finding. Skeletal alterations are other common anomalies of HFM; 40% to 60% of HFM patients exhibit occipitalization of the atlas, cuneiform vertebra, cervi-

Table II. The orbit, mandible, ear, facial nerve, and soft tissue (OMENS) classification system of hemifacial microsomia[59]

Orbit	Mandible	Ear	Facial nerve	Soft tissue
O_0 = Normal orbital size position	M_0 = Normal mandible	E_0 = Normal ear	$N^7{}_0$ = No facial nerve involvement	S_0 = No obvious soft tissue or muscle deficiency
O_1 = Abnormal orbital size	M_1 = Mandible and glenoid fossa are small with a short ramus	E_1 = Mild hypoplasia and cupping with all structures present	$N^7{}_1$ = Upper facial nerve involvement (temporal and zygomatic branches)	S_1 = Minimal subcutaneous/muscle deficiency
O_2 = Abnormal orbital position. An addition arrow denoted relative position of affected side (ie, $O_2{}^{\uparrow}$ for superior, $O_2{}^{\downarrow}$ for inferior)	M_2 = Mandibular ramus is short and abnormally shaped Subdivision A and B are based on relative positions of condyle and TMJ	E_2 = Absence of external auditory canal with variable hypoplasia of concha	$N^7{}_2$ = Lower facial nerve involvement (buccal, mandibular, and cervical branches)	S_2 = Moderate—between the 2 extremes, S_1 and S_3
O_3 = Abnormal orbital size and position	2_a = Glenoid fossa is in anatomically acceptable position with reference to opposite TMJ	E_3 = Malpositioned lobule with absent auricle; lobular remnant usually inferiorly and anteriorly displaced	$N^7{}_3$ = All branches of facial nerve affected. Other involved nerves were also analyzed, eg, trigeminal N^5 (sensory), hypoglossal N^{12}; remaining cranial nerves are signified by the appropriate number in superscript	S_3 = Severe soft tissue deficiency due to subcutaneous and muscular hypoplasmia
	2_b = TMJ is inferiorly, medially, and anteriorly displaced, with severely hypoplastic condyle			
	M_3 = Complete absence of ramus, glenoid fossa, and TMJ. Submental vertex views were used to distinguish mandibular type 2_A from type 2_B.[67] Type O, an apparently normal mandible has not been included in previous classification systems			

TMJ = Temporomandibular joint.

cal complete or partial synostosis of 2 or more vertebrae, supernumerary vertebrae, spinal bifida, and anomalous ribs.[53]

HEMIFACIAL MICROSOMIA CLASSIFICATIONS

The wide spectrum of anomalies associated with HFM has made systematic and inclusive classification difficult. Classification of the disease aids in diagnosis, treatment planning, prognostic predications, and data evaluation. Attempts have been made to classify HFM by concentrating on 1 aspect of the disease such as mandibular, auricular, and soft tissue deformities.[54-56]

Tables I and II present the 2 popular classification systems used for HFM, namely, the skeletal, auricular, and soft tissue (SAT) system, and the orbit, mandible, ear, nerve, and soft tissue (OMENS) system.[57,58] The objective of both systems is to delineate the condition of HFM to provide a rational basis for treatment choice. The OMENS system, a newer and revised HFM classification system, assesses 5 major dysmorphic manifestations and allows each to be graded separately, unlike the SAT system. Orbit is assessed independently from the mandible, and nerve involvement has been added to the system. Categories are completely independent, and every clinical situation, including normal, is represented in each group. Categorization of patients would enable clinicians to predict timing and specific treatment required.

The SAT and OMENS classification systems for HFM have been compared[59-62]; the results were concordant with current literature and demonstrated the phenotypic heterogeneity of HFM. Essentially, both classifications embody the major craniofacial defects, but the OMENS system is further refined by its differ-

entiation between soft tissue and nerve defects, and between orbital and mandibular defects.

TREATMENT OF HEMIFACIAL MICROSOMIA

Although there is a broad spectrum of clinical manifestation, patients with HFM deserve comprehensive care that addresses coexistent medical reconstruction of soft and hard tissues for maxillary and mandibular jaws, ears, and orbits. Some need orthodontic treatment, hearing aid, and language development to restore function and appearance. Psychologic well-being is an issue to be assessed for children with HFM during their growing stages.

Surgical treatment

There are numerous reports on surgical approaches to facial asymmetry for HFM.[63-74] It was once thought that definitive treatment would not be successful until facial growth was finished, but now more clinicians believe that early diagnosis and treatment of HFM is beneficial. Studies have shown HFM is a progressive skeletal and soft tissue deformity with the earliest skeletal manifestation in the mandible.[63,67,69] The hypoplastic mandible interferes with normal downward growth of the maxilla and is consistent with asymmetrical skeletal growth. Mandibular distortion becomes worse and produces secondary deformation of the maxilla, nose, and orbit as the contralateral side grows.[75] Thus, correction of the mandibular abnormality in childhood establishes a more normal "functional matrix" for symmetric midfacial growth and unlocks the growth potential of the adjacent structures, minimizing secondary deformity, and improves function and appearance.

Retrospective analyses of growth of reconstructed condyle/ramus in children with HFM have revealed that costochondral grafts sometimes enlarge in size and grow in length; this growth usually occurs after 2 years of costochondral grafts at a slow and irregular rate. Patients 5 to 6 years old are considered candidates for early surgical reconstruction of the maxilla and mandible, with probable necessity of the operation being partially repeated when growth is complete.[76]

Patients with HFM usually have approximately 50% conductive hearing loss associated with middle ear abnormalities.[77] Major ear anomalies such as total bony atresia over the meatus, partial aplastic canal, missing tympanic cavity, and absence of external meatus sometimes are not good indications for reconstructive surgery.[78] Positive results of hearing restoration can be achieved by using osseointegrated craniofacial implants as a bone-anchored hearing aid (BAHA).[79] The BAHA is considered to be a better device than the conventional hearing aid because it provides support and retention for the hearing aid, has less skin irritation, and is more cosmetically pleasing.[80-82]

Reconstructive ear surgery is one of the most demanding challenges for the plastic surgeon because of the ear's complex structure. With the pioneering work of Tanzer and Kirkham,[83,84] great surgical advances have been made in ear reconstruction over the years. Many authors have also demonstrated excellent and consistent results,[85-88] and Brent has set the standards of modern autogenous ear reconstruction.[89,90] The indications of autogenous ear reconstruction for HFM are classic microtia with no prior surgery and patients with normal lower third of an ear. A recent ear reconstruction conference (Lake Louise, Alberta, Canada, March, 1998) concluded that HFM is better surgically reconstructed rather than having a lifetime of prosthetic management, namely, continual remakes of the prosthesis over a life time.

Surgical techniques are various from Brent's 4-stage to Nagata's 2-stage surgery.[86,89-91] Even 1-stage surgery for total ear reconstruction has been used for patients with HFM starting at age 6 and up.[92,93] The use of autologous costal cartilage is considered the most reliable method for reconstruction of the pinna. For situations where there is insufficient spare skin, a tissue expander and/or temporoparietal fascia flap may be used.[90,94]

Principles of timing for surgical reconstruction of facial deformity are based on age, severity, and psychologic considerations. For severe deformity such as orbital dystopia, osteotomies can be conducted from the age of 2 and onward.[65] There are a number of concerns in regard to sequencing of microtia treatment relative to management of jaw deformity as part of the patient's overall management. For example, should the ear be constructed first or should a distraction osteotomy of the mandible and leveling of the occlusal plane be accomplished so the position of the replacement ear be symmetrically oriented to the unaffected side? Unfortunately, there is no set of rules to list the treatment sequence for HFM. Management of HFM patients is a multidisciplinary effort that is complex and needs to be tailored to an individual patient's unique requirements.

Prosthetic treatment

With the benefits of craniofacial osseointegration as a tool for enhanced retention of facial prostheses, appropriate patient selection is an essential component to the prosthetic management of HFM. Pediatric patients with auricular deformities or absence of the auricle that resulted from HFM should be considered surgical candidates first. If they are not surgical candidates for reconstruction because of, for example, high operative risk, severely compromised tissue, or failed previous autogenous ear reconstruction, then an implant-retained ear prosthesis should be considered as an alternative treatment. Criteria of osseointegrated alloplastic versus autogenous ear reconstruction for auricular defects have

Table III. Advantages versus disadvantages of autogenous ear reconstruction for HFM

Advantages	Disadvantages
Using patient's own tissue	Longer procedures with more surgical stages over a longer period, especially soft tissue expander or temporoparietal fasicia flaps needed
Likelihood of stable, long-term success of grafted tissues	Greater surgical mobility
Continued growth of grafted cartilage framework with age	Less similar to the normal opposite side ear than a sculptured prosthetic ear
Little ongoing maintenance	
Less chance of late complications	
No need of clinical and technical prosthetic support	
No significant continued costs incurred	

Table IV. Advantages versus disadvantages of implant-retained prosthetic ear reconstruction for HFM

Advantages	Disadvantages
Relatively short and less demanding surgical procedures	Replacement of the prosthesis required because of degradation of silicone materials
Outpatient procedures under local anesthesia	Regular aftercare needed by the patient
Greater similarity to the normal ear	Ongoing follow-up for assessment of the skin around implant abutments
Easy replacement or correction of an unsatisfactory prosthesis by some patients	Difficulty incorporating the prosthesis into the patient's body image

been thoroughly discussed by Wilkes and Wolfaardt.[87] Advantages and disadvantages of these 2 treatment options are listed in Tables III and IV.

Initial application of a bone-anchored auricular prosthesis was limited to patients over 15 years of age because of thicker temporal and mastoid bones for titanium implant anchorage.[95] Irradiated patients and younger populations who received bone-anchored auricular prostheses were also reported.[96,97] An implant-retained ear prosthesis offers significant improvements in the quality of life when compared with the tissue adhesive systems previously available for this type of prostheses.[98,99]

The prosthetic involvement in providing a patient with an implant-retained auricular prosthesis can be considered in 2 stages: presurgical and postsurgical phases; clinical and laboratory procedures are summarized in Table V.[100-102] Three to 4 months between implant placement and final prosthesis fabrication are required for osseointegration.

Table V. Prosthetic involvement for implant-retained ear prosthesis

(A) Presurgical phase
Examination and consultation
Diagnostic cast fabrication
Diagnostic waxup, clinical modification, and confirmation
Surgical template fabrication and determination of craniofacial implant location, 20 mm away from the external ear canal at 8 and 11 o'clock positions on the right side and 1 and 4 o'clock positions on the left side of the ear
(B) Postsurgical phase
Implant connection
Impression making
Fabrication of working cast
Design and fabrication of framework, a ball/stud attachment, a magnetic retention, or a bar/clip system can be used for orientation of the prosthesis
Sculpting the ear and modification
Three-piece stone mold fabrication for silicone casting
Intrinsic coloring and establishing an intrinsic formula
Casting of silicone ear prosthesis
Extrinsic coloring
Care of the prosthesis and tissue around the implant abutments
Regular follow-up

NEW DIRECTIONS FOR EAR RECONSTRUCTION PATIENTS

Tissue engineering

Autogenous ear reconstruction with cartilage remains a technical challenge for the plastic surgeon because of the intricate configuration of a human ear. Technology for tissue engineered generation of new bone, cartilage, and liver tissue offers great potential for improving the outcome of tissue regeneration.[103-105] Tissue engineering is defined as a science in which the material properties of synthetic compounds are manipulated to enable formation of new functional tissue. The Harvard-MIT research group brought significant attention in the biomedical area with recent accomplishments. The group seeded a high density of functional, dissociated cells onto synthetic biocompatible, biodegradable polymers of different chemical compositions and physical configurations and transplanted them into animals for the purpose of generating new functional tissue. The cell-polymer construct is config-

ured to allow gas exchange and nutrient diffusion until the successful engraftation of cells with new tissue formation can occur.[106] Their study produced tissue engineered cartilage in the shape of a human ear using chondrocytes seeded onto a synthetic polymer fashioned in the shape of a 3-year-old child's auricle.[107] The finding also concludes that after 8 weeks of implantation, new cartilage will mature and retain the same complex architecture with the application of a stent to resist the initial contraction forces of wound healing. Although this is an animal model, it opens many avenues of potential clinical applications of surgical treatment for patients with HFM.

Computer design

The conventional method of making the auricular prosthesis includes impression making, master cast fabrication, wax model sculpting, dewaxing, silicone packing, and coloration. It requires artistic skills and is time-consuming. Computer technology has become an important tool in modern medicine and dentistry. Three-dimensional imaging acquired from computer tomography scans or laser surface scanning have been combined with CAD/CAM technique for clinical fabrication of facial prostheses and craniofacial surgery.[108-111] The CAD/CAM technique has been well-developed in industries and useful in the fabrication of dental restorations, such as porcelain inlays and crowns.[112,113] Transformation of 3-dimensional image data to a CAD/CAM system for successive mathematical processing, design simulation, and model production can potentially minimize the time and skills required for sculpting an ear prosthesis for patients with HFM and provide new perspectives for future maxillofacial prosthodontics. However, sophisticated computing and machining need to be simplified and reduced in cost to be commonly used by clinicians.

CONCERNS OF CRANIOFACIAL IMPLANTS FOR CHILDREN WITH HFM

Long-term follow-up results of craniofacial titanium implants in the adult subjects without irradiation has been favorable.[114,115] Published pediatric data on the use of osseointegrated titanium implant systems are sparse. Children from ages 5 to 12 are considered at a higher risk for complications because of thinner and softer temporal bones and are at an increased risk for a disruptive accident injury. Tjellström[97,116] reported that during placement of implants for hearing aids, 12% were in contact with the dura mater, the wall of the sigmoid sinus was seen at the bottom of the implant site in another 12%, and mastoid air cells were seen in 25%. Children with severe craniofacial defects pose special problems relative to the implant site because of aberrant facial nerve course, low middle cranial fossa dura, and small mastoid.

The lack of information on the influence of craniofacial growth and development on the stability of osseointegrated craniofacial implants in pediatric populations is of great interest for clinicians. Placement of implants in areas of bony resorption may decrease the functional life of implants during active craniofacial growth in HFM patients. Moreover, long-term stability of implants may be further compromised at puberty when the mastoid air cells undergo their greatest development.

The psychologic impact of early placement of implants is another important issue. Studies of psychologic and social effects on adults with facial deformities reported that all the patients went through periods of emotional turmoil related to their appearance, and all experienced periods of marked depression that impaired emotional and social functioning.[117] Psychologic problems in children with craniofacial deformities have included lack of emotional attachment between parent and child, inadequate development of peer relationships, and the experience of shame related to a poor body image.[118] Children with facial deformities at school age of 5 years would start to show self-consciousness and receive pressure from their peers. Would early autogenous or alloplastic ear reconstructions for HFM children age 5 to 10 years minimize psychologic problems? What are the risks and benefits of these 2 methods in treating HFM children? These are important questions to be answered in future psychologic studies.

CONCLUSIONS

Hemifacial microsomia is the second most common craniofacial defect after cleft lip and palate; its cause is unresolved and complicated. Extreme variability of expression of the disease is characteristic of HFM. It was once thought that definitive treatment would not be successful until facial growth was complete, but now more clinicians believe that early diagnosis and treatment is beneficial.

Surgically reconstruction is better for HFM treatment rather than a lifetime of prosthetic management, namely, continual remakes of the prosthesis. With the introduction of craniofacial implants, an implant-retained auricular prosthesis has become a viable option to surgical reconstruction for microtia for patients who are not candidates for surgical reconstruction. Advantages and disadvantages of autogenous and alloplastic ear reconstructions are discussed. Both treatments can provide excellent cosmetic results in appropriately selected situations. New exciting research directions, such as tissue engineering and fabrication of auricular prosthesis by CAD/CAM, offer potentials to improve the treatment for HFM in the future.

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