Comprehensive Prosthetic Rehabilitation of A Case of Hemifacial Microsomia – Clinical Report

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Abstract
Maxillofacial defects can result from trauma, treatment of neoplasm or congenital malformations. Many a times due to size, location of the defect or because of the patient’s medical condition, surgical reconstruction may not be possible. In these cases, rehabilitation is indicated with the help of a maxillofacial prosthesis. Hemifacial microsomia is a congenital anomaly that affects development of mouth, face, eyes and ears. Such abnormalities or defects compromise form, function, esthetics, social acceptance and deeply affect the psychological status of an individual. This report describes the comprehensive prosthetic rehabilitation of a case of Hemifacial microsomia with congenital defect of ipsilateral orbit and ear. The individual was rehabilitated with a custom made orbital and auricular prosthesis using room temperature vulcanizing silicone. The prosthetic rehabilitation significantly improved aesthetics, phonetics and function as well as social and psychological status of the patient.

Keywords: Oral Potentially Malignant; Malignant Lesions; Diagnostic Aids
Introduction

Hemifacial microsomia is a congenital syndrome that cause asymmetrical anomalies of derivatives of the first and second brachial arches.[1-3] According to studies it is the second most common congenital craniofacial birth defect after cleft of the lip and palate.[1,5,6] However, recent finding of a higher figure of 1 in 3000 have been reported.[7,8] Predilection for males with a male-female ratio of 3:2 has been demonstrated by many investigators.[6]

Hemifacial Microsomia occurs sporadically with no positive family history of this deformity, hence there is strong consensus that it is genetic and not hereditary.[6,7] Phenotypic expressions of HFM depend on the extent of this hemorrhage and its effect on these two arches.[11-13] Therefore, there is a wide spectrum of presentation of this malformation varying from the mild to the severe spanning the skeletal, neural, muscular tissues and soft tissue. It affects the development of the lower half of the face, most commonly the ears, the mouth and the mandible.[9,13]

The more commonly affected structures include ear (external and middle which result in conduction defects between 30-50%), mandible [ascending ramus, condyle and temporomandibular joint (TMJ)], orbit, zygomatic arch and maxilla. Soft tissues majorly involved include facial nerve and muscles such as masseter and temporalis [11-13]. The positioning of the orbit might be altered (orbital dystopia) with presence of dermoids (epibulbar), retinal or choroidal coloboma, blepharoptosis, microphthalmia or anophthalmia and others.[13] Some patients could also present with absent ear (anotia), small ear (microtia), disorders of the middle ear and very bad cases with hearing loss. [6-9] Furthermore, the seventh (facial) cranial is frequently affected with different degrees of affectation of the upper or lower branches and in severe cases the fifth (trigeminal) and twelfth (hypoglossal) cranial nerves could also be vulnerable.[6] In addition, findings of abnormal teeth development and eruption such as dental hypoplasia, agenesis, microdontia, malocclusion and delayed teeth eruption have been demonstrated. Since hemifacial microsomia causes physical, psychological social impact on individuals, so intervention at early age gives the best results. A complete team effort is required to correct syndrome’s complex deformities which requires a multidisciplinary approach including Pediatrician, Prosthodontist, Reconstructive surgeon, Oral surgeon, Otolaryngologist and Speech therapist.

Case Report

A 23 year old male patient reported with the chief complaint of congenitally missing left ear and left eye. Also the patient could not hear from left ear neither had any vision from left eye. Medically history revealed a diagnosed case of hemifacial microsomia and multiple attempts have already been made for surgical reconstruction but none proved successful. The family history was non-contributory. On general physical examination, it was found that all the vital signs were within the normal limits. Extraoral examination revealed hypoplastic mandible. The patient had a small ear tag on left side (Figure 1) representing surgical reconstruction attempt was made. On examination it was noticed that the tissue tag would interfere with complete seating of prosthesis so pre prosthetic surgical excision of tissue tag was planned. Also on examination of left eye, there was complete paralysis of orbital muscles which didn’t allow placement of ocular prosthesis. (Figure 1) On examining the patient intraorally, all the teeth were present. There was increased overjet and overbite. Other findings included high arched palate. Radiographic investigations confirmed the condition as hemifacial microsomia.

Figure 1: Pre-op view showing tissue tags on left ear and paralysis of left orbital muscles
A comprehensive prosthetic rehabilitation was planned for the patient with custom made adhesive retained silicon orbital prosthesis for rehabilitation of ear defect and a custom made adhesive retained auricular prosthesis for orbital defect. Treatment was initiated with psychological counseling and reassured treatment outcome regularly during treatment.

Fabrication of orbital prosthesis began with marking the reference landmarks on face which included midline of face, inner canthus, outer canthus and mid pupilary lines. (Figure 2) This was done to achieve bilateral symmetry. The patient preparation was completed and a custom made special tray was prepared using impression compound (DPI, India). A complete face impression (Figure 3) was made using irreversible hydrocolloid (AlgiteX) and poured in dental stone (Kalabhai).

The iris was obtained from a closely matching stock eye and wax pattern fabrication was completed using modelling wax (Ruthenium wax products) following reference marks for bilateral symmetry. Room temperature vulcanizing silicone (Copsil T – 30 TN, COP) was used for fabrication of prosthesis. Silicone was mixed with intrinsic colours following the manufacturer’s instructions to closely match the shade of the patient. After packing of silicone, the complete flask assembly was closed and left at room temperature for 24 hours to complete curing. The prosthesis was retrieved and finished. Characterization of prosthesis was done by adding eye lashes, eyebrows using artificial hair and further life-like appearance was produced using extrinsic colours. (Fig 5) The prosthesis was inserted in situ and was evaluated for fit and aesthetics.
Fabrication of ear prosthesis: Impressions of ear was made using a two step impression technique (Algitex) [Figure 2], using elastomeric impression material (Aquasil Putty, Dentsply, Germany). A donor with same size of unaffected ear was selected, and impression was made with irreversible hydrocolloid (Algitex) for preparation of wax patterns. For the fabrication of the wax pattern, molten wax was poured in mould and retrieved after cooling. Wax patterns were adjusted on a Polymethyl methacrylate (DPI, India) substructure according to remaining ears (Fig 6).

The patterns were finalized according to location, size, symmetry, and characterization. Room temperature vulcanizing silicone (Copsil T – 30 TN, COP) was used for fabrication of prosthesis. Silicone was mixed with intrinsic colours following the manufacturer’s instructions to closely match the shade of the patient. After packing of silicone, the complete flask assembly was closed and left at room temperature for 24 hours to complete curing. The prosthesis was retrieved, finished, characterized using extrinsic colours. The prosthesis was inserted in situ and was evaluated for fit and aesthetics (Fig 7) Patient was instructed about the limitations, use, and maintenance of prosthesis. A comprehensive prosthetic rehabilitation not only improved form, function and aesthetics but also remarkably improved the confidence, psychological attitude and social acceptance of the patient.
Figure 6: Wax pattern for Auricular prosthesis

Figure 7: Post-op after insertion of prosthesis
Discussion

The clinical features of this anomaly vary considerably but commonest dominator is the facial asymmetry associated with mandibular hypoplasia and TMJ incongruity.\cite{8,9} This is majorly unilateral but occasionally can be bilateral. Maxillary/zygomatic hypoplasia, external/internal ear abnormalities/ataresia, coloboma, parotid hypoplasia and microphthalmia. \cite{10,11} There are also several dental derangements such as oligodontia, malocclusion, open bite and delay eruption. Other congenital anomalies that might be present include vertebral anomalies, cardiac defects, renal defects, mental retardation and host of other soft tissue disorders.\cite{13} Various prosthetic materials like acrylic resins, acrylic copolymers, vinyl polymers, polyurethane elastomers and silicone elastomers have been reported in literature. Silicone became popular over other materials as they have a range of good physical properties, low degree of toxicity, easier to manipulate, chemical inertness, high degree of thermal and oxidative stability. Further they can be stained intrinsically or extrinsically to give them more lifelike natural appearance.\cite{14} The follow-up visit should be 24 hours after prosthesis delivery. At that time, the condition of the prosthesis and the health of skin should be assessed. Subsequent visits are planned at one week and then every month to evaluate fit, color, function, retention of prosthesis and also to examine the health of underlying skin. Maintaining hygiene of the prosthesis is important for the health of soft tissue underneath the prosthesis and for preserving the prosthesis itself in a good condition. Silicone materials are more difficult to clean than resins as these materials are permeable so are more susceptible to microbial colonization.\cite{15} Water and neutral soap, together with gentle brushing using a soft nylon bristles have been recommended.\cite{16} The use of Chlorhexidine has been shown as an excellent method of disinfection, in this case prosthesis was cleaned using 4 percent chlorhexidine immersion for 1 minute everyday followed by rinse with water\cite{17} Multiple studies highlight that the use of disinfecting agents and rigorous cleaning adversely affects the physical properties of silicone material.\cite{18,19}

Conclusion

Hemifacial microsomia affects the patients physically, functionally, psychologically and socially. So one has to plan treatment taking multiple aspects into consideration. A team effort of multiple specialities is required to rehabilitate patient and improve quality of life of these patients.
References


