Case Report

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# Hemifacial Microsomia with mild Radial Hypoplasia and Hypodontia: A Case Report

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## ABSTRACT

**Background**: Hemifacial Microsomia (HFM) is an asymmetric craniofacial malformation which results in hypoplasia of the components of the first and second branchial arches. There can be various anomalies, which include conductive hearing loss which is caused by external and middle ear deformities. HFM is the second most common congenital facial anomaly, which is seen after cleft lip/palate. Here we present a unique case of 7years old boy with hemifacial microsomia associated with mild radial hypoplasia and hypodontia.

**Keywords:** Hemifacial Microsomia, Branchial arch syndrome, radial hypoplasia, hypodontia.

## INTRODUCTION

Hemifacial Microsomia (HFM) was first described by Carl Ferdinand Von Arlt in 1881. HFM involves first and second branchial arch derivatives with highly variable phenotypes. It is also known as a first and second branchial arch syndrome, otomandibular-facial dysmorphogenesis and lateral facial dysplasia.[1] HFM is primarily a syndrome of the first branchial arch, which involves underdevelopment of the temporomandibular joint, mandibular ramus, muscles of mastication and the ear. Abnormal development of the auricular hillocks leads to microtia or atresia of the pinna, and it is proportional in severity to the abnormal external auditory canal development[2].

HFM is the most common congenital facial anomaly, which is second only to facial clefts [2,3]. There is a definite male predilection (3:2), with the right side being more commonly affected (3;2) [4]. Hemifacial microsomia is estimated to occur in 1:3,000 to 1:5,000 live births [5]. The aim of this paper is to report the present case in literature with a unique association of mild radial hypoplasia and hypodontia.

## CASE REPORT

An 8 year old boy, reported in the department of Pedodontics and Preventive dentistry in Rishiraj college of Dental Sciences and Research Center for painful teeth in all quadrants and asymmetric facial appearance. Parental history was taken for familial recurrence, consanguinity marriage, presence of diabetes mellitus, gestational diabetes, use of assisted reproductive technology or use of any vasoactive medication during pregnancy. None of the asked possible etilogy found positive. Parents have 3 children. All are prematured with normal delivery around 8-months 10-12 days at home. First, two's are the girl with good health and built.

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Only, he was born with facial asymmetry and malformed ear with a birth weight of 2000 gm.

Developmental milestone was normal. The child was not suffering from cardiological, respiratory, urogenital, gastroenteric or skeletal abnormalities. On extraoral examination, facial asymmetry was observed on the right side of the face, with the right pinna being situated at a much lower level than the left one, and it was small and malformed. No preauricular tag or pit was present. The hearing was less with the right ear as compared to the left ear. The size of the right eye is the same but located at a higher level than the left one. There was no presence of any coloboma or strabismus or epibulbar dermoids. Mild radial hypoplasia is present on the right hand. Cant of occlusion was 6-15 degree higher on the right side. Minimal soft tissue deficiency was present on the right mandibular region with chin deviation

Intraoral examination reveals mild dysmorphism with no ankyloglossia. Teeth present were 16,55,54,53,11,21,62,63,64,65,26,36,75,74,73,32,31, 42,83,84,85,46. Clinically, 41 was missing. The anterior open bite was present. Deep occlusal caries was present in 55,65, 74, 84, 85 smooth surface caries in 53,63and 83. The patient was advised for Intraoral Periapical (IOPA) X-rays of 55,65,74,84,85 to assess carious involvement and succedaneous tooth development. Orthopantomogram (OPG) and lateral cephalogram to assess facial asymmetry and entire dentition status. OPG examination revealed mandibular right central incisor was not developed. Right mandibular ramus was short in length. Right mandibular condyle was not well developed as compared to the left side, grade-I (Pruzansky,1969). The inferior border of the right mandibular body was not well developed and short in height.

By assessment of IOPA X-rays, Pulpectomy was planned and done for 55,65, and 74. 84 and 85 were extracted as the pulpal infection caused severe root resorption. Since the mandibular functions were normal, slight deviation of the mandible to the right was not considered very important at this stage. Due to the lack of the right temporomandibular joint, vertical and sagittal mandibular development, causing the mandible to rotate bodily to the right. On correlating the clinical and radiologic findings, a diagnosis of HFM was made and the patient was referred to Oral Maxillofacial Surgery and Orthodontia Department for aesthetic rehabilitation of hypoplastic mandible and correction of unilateral crossbite and anterior open bite.

Fig 1: Facial asymmetry, right eye located on a higher level, deviated chin and lip toward the right

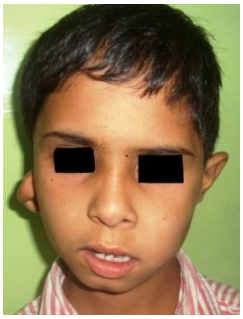


Fig 2: (i) severely malformed pinna of right ear (ii) left profile of patient

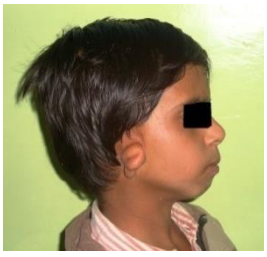


Fig 3: Mild radial hypoplasia in right hand

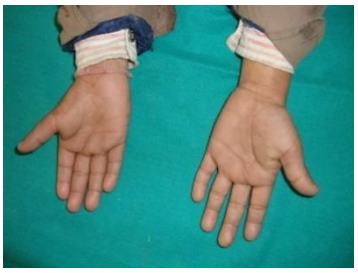


Fig 4: Anterior open bite, missing right mandibular central incisor.

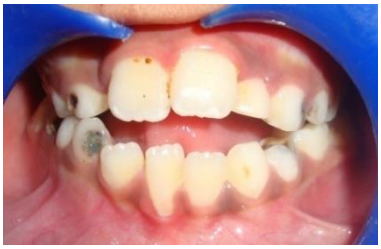


Fig 5: OPG shows absence right mandibular incisor, short right mandibular body, and the right condyle is also not well developed.

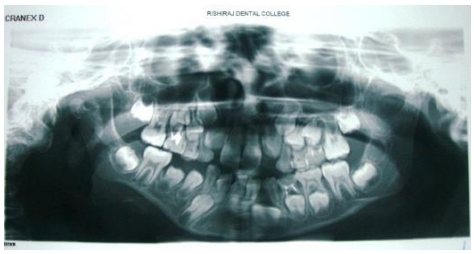
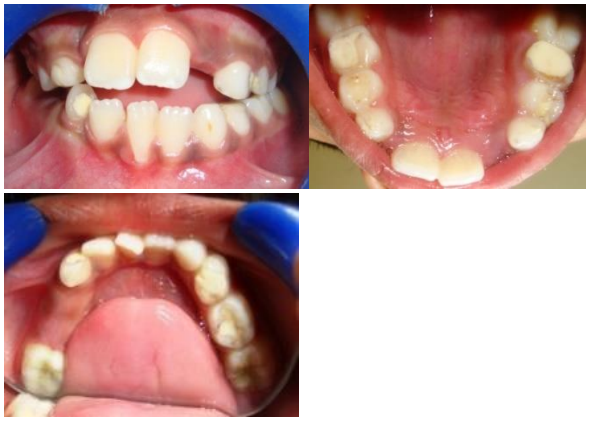


Fig 6 (i) After restoration of caries (front view), (ii)maxillary arch, (iii) mandibular arch



## DISCUSSION

The term hemifacial microsomia (HFM) was coined by Gorlin and Pindborg (1964).[6] There are many theories given for the HFM based on embryologic, clinical and laboratory studies, but the exact aetiology is yet not known, but pathogenically it is heterogeneous. The laboratory studies suggest that there are certain factors that are responsible for the clinical presentation of HFM, which can be due to early loss of neural crest cells [7,8]. The defective genes, teratogens and vascular anomalies are singly or collectively responsible for the disruption of normal development, which leads to anomalies seen in these patients[9].

Minimal diagnostic criteria for HFM include asymmetric hypoplasia of facial structures with preauricular tags or microtia with or without preauricular skin tags. A 30–50% of the patients have auditory problems as a result of malformations in the middle ear.[8] The OMENS classification is the most commonly used classification system[9]. The acronym OMENS designates each of the five major areas of involvement: O= orbital, M = mandibular, E = ear, N = facial nerve, and S = soft tissue. Accordingly, present case was classified as $O_3M_{2A}E_2N_0S_1$. Branchial arch syndromes are thought to result from retardation or failure of differentiation of maxillary mesoderm at and after the 50mm stage of the embryo[10]. The first visceral arch of the visceral mesoderm also advances secondarily to form the mandible, and again retardation occurs on the same basis[10]. The hypodontia or agenesis of mandibular central incisor, in this case, might be explained in relation to this failure or retardation. Mild radial hypoplasia defect is present in the present case in the right hand, which may be considered as a unique association in this spectrum. Radial limb deformity in the form of a hypoplastic or aplastic radius and/or thumb, or bifid or digitalized thumb, occur in about 10 % [11]. Patient with HFM must be distinguished from those with Goldener syndrome, Treacher Collins syndrome, and Parry Romberg syndrome.

Goldenhar's syndrome can be distinguished by varying degrees of vertebral, cardiac and sometimes structural kidney defects[12] which are not present in this case. Lack of vertebral malformation and epibulbar dermoids, which may be bilateral in 75% of Goldenhar's syndrome cases [13] eliminated the possibility of this syndrome.

Most of the features of Treacher Collins syndrome include hypoplasia of facial bones especially malar and mandibular bones, malformation of the external, middle and internal ear, macrostomia, high palatal arch which mimic HFM. However, the other features like antimongoloid palpebral fissures, deficiency of eyelashes, a blind fistula between angles of the ear and mouth with facial cleft and skeletal deformities with the characteristic appearance of the face are "birdlike or fishlike" [14] which were not seen in the present case. Parry-Romberg syndrome (PRS) also known as "progressive facial hemiatrophy" is characterized by progressive but self-limited atrophy of the skin and subcutaneous tissue on one side of the face[15], no hearing loss or ear deformity is involved. So, the present case excludes this diagnosis.

The management of HFM is a multidisciplinary approach, which includes surgery done during growth phase or after the growth phase is over, limited autogenous bone grafting of deficient

portions of the craniofacial skeleton, bilateral mandibular jaw advancement in patients with mild to moderate mandibular micrognathia with a combined Le Fort I osteotomy. The genioplasty and microvascular free flaps can be used for augmenting the soft tissue of the affected side of the face and costo-chondral grafts which can be used to provide a new growth centre for treating this anomaly[16].

## CONCLUSION

HFM is a developmental malformation in which there is a deficiency in both skeletal and soft tissues of the maxillofacial region on one side of the face. Hence, early diagnosis and multidisciplinary treatment approach are required for proper functioning and esthetics of the orofacial structures, for a good prognosis.

## CONFLICTS OF INTEREST

The authors declare they have no potential conflict of interests regarding this article.

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