Oto-Mandibular Dysostosis

* *Ramesh Chandra, M.S., M.S.,* †*R.N. Sharma, M.S., F.R.C.S.* & ‡*V.N.S. Yadava, M.S.*

Congenital unilateral (very rarely bilateral defects of the Ear, the musculo-skeletal tissue developing from the 1st and 2nd branchial arch and macrostomia are the classical features of cases included in the Oto-mandibular dysostosis first described by Francis and Haustrate (1954). The disorder is connected with the 1st branchial arch and is quite identical to the "Oral-Mandibular-Auricular syndrome" documented by Stark and Saunders (1962) and others. The earliest reference of association of deformities of these areas was probably made by Canton, Ogston & Mason (1860).

Aetiology

The exact aetiology of this syndrome (Otomandibular dysostosis) is not yet known. Spemann (1936) gave the theory of organisers. According to him, development depends upon the presence of normal organisers and if they are damaged faulty development of the affected area results. This is governed by the gene mutation process. The fundamental anomaly according to (Neehan 1942) is metabolic which may manifest itself in different ways.

Mc-Kenzie and Craig (1955) put forward the suggestion that the mandibulo-facial syndrome results from the defective stapedial artery during foetal development. Greer Walker (1962) however suggested Intra-uterine necrosis due to environmental hazards as the possible aetiological factor. Stark and Saunders (1962) suggest a deficiency of mesoderm in the branchial arch. Franceschetti and Klein (1949) suggested a genetic origin, and thought that heredity had no role in the genesis of otomandibular syndrome.

The present paper is an analysis of 14 patients admitted to our service during the five year period i.e. 1968 to 1973. Our centre provides for the reconstructive needs of the entire state of U.P. with a population of 8.8 crores and an area of 113.409 sq. miles (1971 census), besides scattered cases from adjoining states. Our surmise is that there are many unrecorded cases, and their families have accepted the deformity for want of information regarding treatment facilities or because of their poor socio-economic conditions.

Table 1 shows the age group of patients on admission and their sex ratio which is 2.5 : 1 suggesting male predominance in our series. Majority of the patients came during school going age and some when faced with marriage problems.

* Reader in Plastic Surgery. † Prof. & Head, Department of Plastic Surgery. ‡ Post-graduate student.
Department of Plastic Surgery, King George's Medical College Lucknow, India.

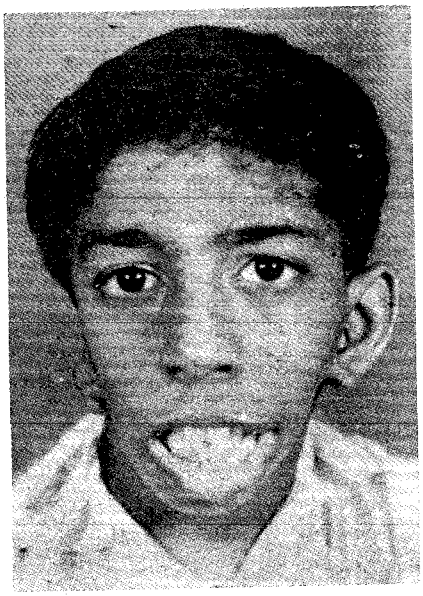
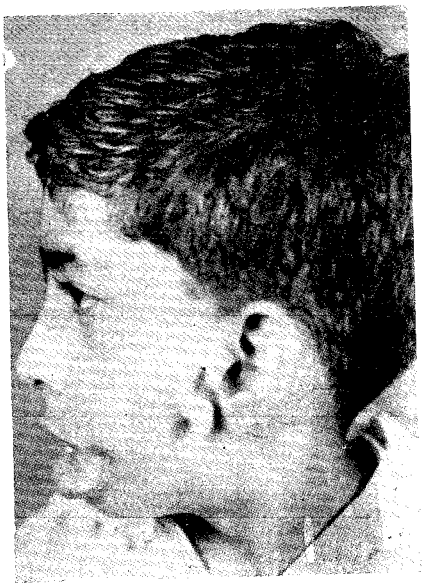
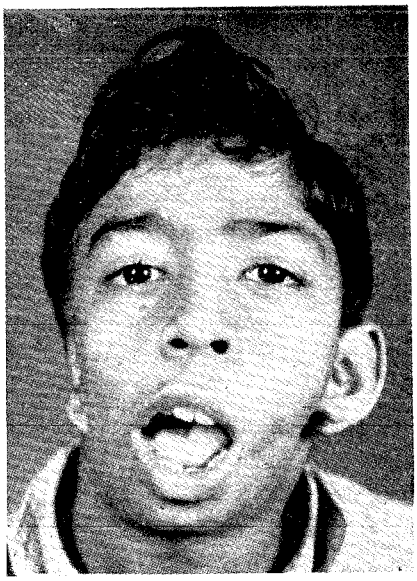


Fig. 1 a.—Front view showing bilateral involvement.



b.—Left lateral view.



c.—The post-operative appearance.

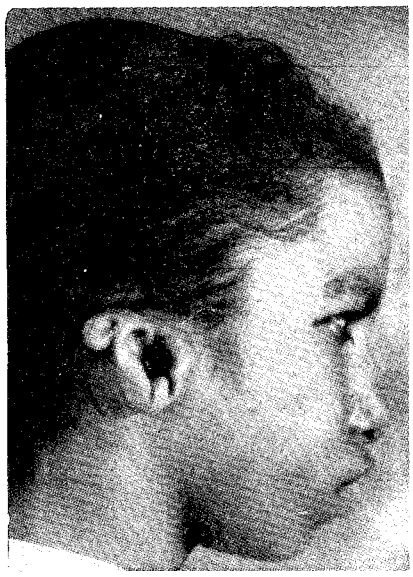


Fig. 2 a.-Right lateral view showing the abnormality of the ear.

b.—Front view showing bilateral involvement along with facial paralysis.

c.—Left lateral view showing the abnormality of the ear.

Table 1

Age in years	Male	Female	Total
Below 5	2	—	2
6 —10	1	2	3
11—15	4	1	5
16—20	1	1	2
21—25	2	—	2

Table 2 shows cases with bilateral involvement. Arvind the first case (Fig. 1) was a 12 years male child while Madhu the second case (Fig. 2) was a 11 years old female who had left sided facial palsy, a rare finding hitherto undocumented.

Table 2

Sl. No.	Side	Ear	Musculo-skeletal hypoplasia	Oral involvement
1.	Left	Preauricular tabs.	Mandibular	Macrostoma
	Right	Abnormal, low placed	Mandibular	Nil
2.	Left	Abnormal, low placed	Mandibular	Macrostoma, facial palsy
	Right	Abnormal, normally placed	Nil	Nil

Table 3 shows six cases with classical unilateral (left side) involvement. The associated abnormalities being disturbed occlusion in case No. 3 and bilateral epicanthal folds (Fig. 3 a, b, c, in case no. 6.)

Table 4 shows another group of six cases who did not have macrostoma. The associated abnormalities being facial paralysis (Fig 4 a, b, c,) in case no. 9, involvement of he 1st arch and 2nd arch tubercles in case no. 9. and 11 and right sided palatal palsy (Fig. 5) and nasality of the speech in case no. 12.

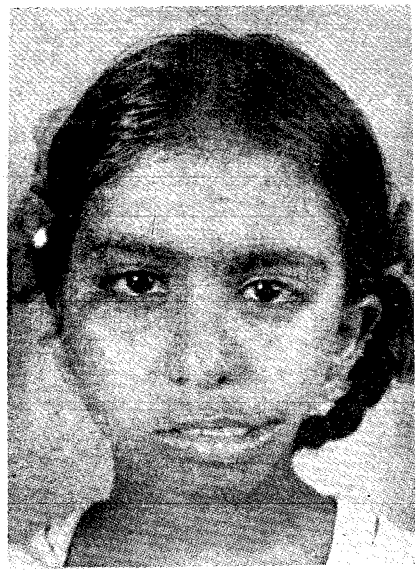
Treatment :

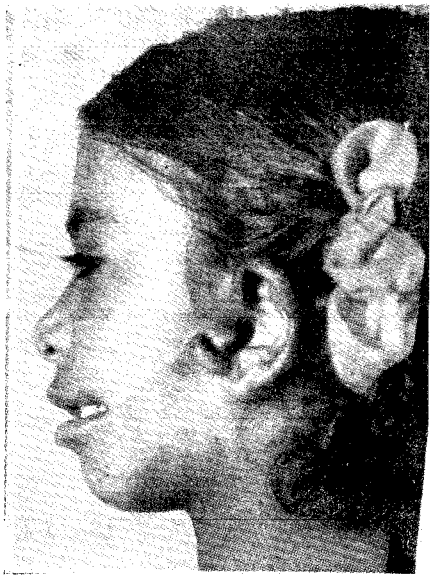
After necessary investigations the priorities in the treatment were assigned. The Macrostoma was managed either by a straight line closure and or by incorporation of a 'Z' plasty. The accessary tubercles, preauricular tabs or the abnormal tragus were set right by excision and local adjustment. In case of atresia of external auditory canal, meatoplasty was performed.

None of the patients agreed for the correction of the mandibular deformity. Those having facial muscle palsies were advised physiotherapy and electrical massage therapy beside supportive treatment.

Summary :

14 patients of otomandibular dysostosis a rare congenital anamoly of 1st branchial arch have been presented. Two patients had bilateral defects while there was Palatal paralysis, facial paralysis, disburbed occlusal pattern, bilateral epicanthal folds and involvement of the 2nd arch tubercles as associated abnormalities in other cases. The abnormality affected the males and the left side of the face predominantly. Patients were satisfied with correction of the musculoskeletal deformities associated with this syndrome.





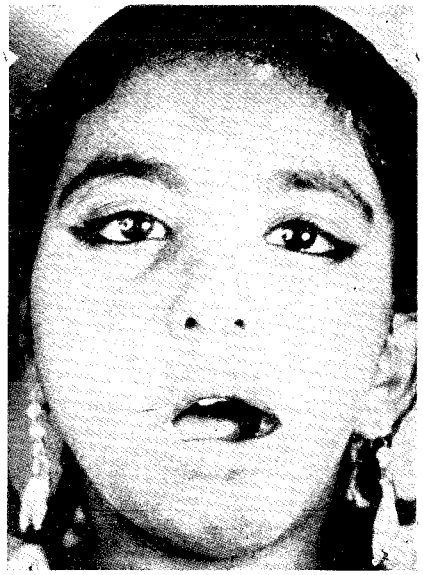
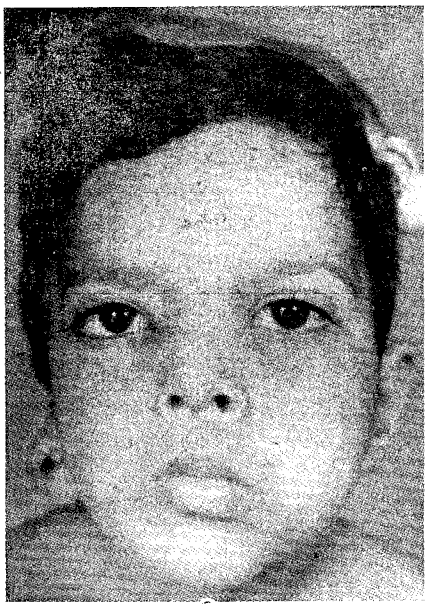
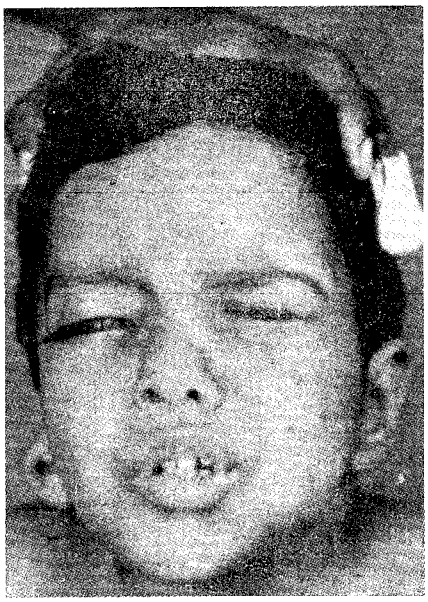


Fig. 3 a.—Front view showing the classical unilateral involvement.

b.—Left lateral view.

c.—Showing the post-operative appearance





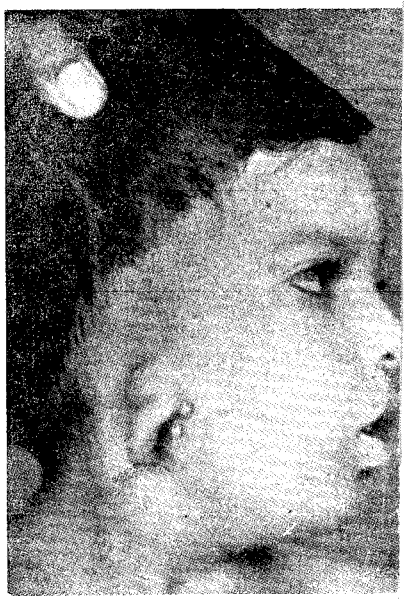


Fig. 4 a.—Front view showing the abnormality.

b.—Front view showing facial paralysis.

c.—Right lateral view showing abnormality of the ear.

Table 3

Sl. No.	Name, Age, Sex	Ear	Musculoskelatal hypoplasia	Oral involvement	Associated abnormalities
3	R. K. 3 Yrs Male	Abnormal tragus, Abnormal tabs.	Mandibular	Macrostoma	Disturbed occlusion
4	R K. 6 Yrs Female	Abnormal tragus, Accessary tabs near angle of mouth	Mandibular	Macrostoma	Nil
5	Vinod 7 Yrs Male	Abnormal tragus	Mandibular	Macrostoma	Nil
6	Baby 8 Yrs Female	Accessary tabs	Mandibular	Macrostoma	Bilateral epicanthal folds.
7	M.T. 13 Yrs. Male	Abnormal tragus, Accessary tabs	Mandibular	Macrostoma	Nil
8	M J. 21 Yrs Male	Abnormal tragus, Accessary tabs.	Mandibular	Macrostoma	Nil

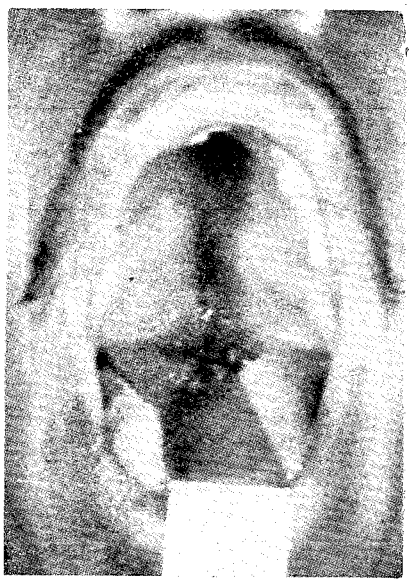


Fig. 5 - Showing Right sided palatal paralysis in case no. 12.

Table 4

Sr. No.	Name Age Sex	Side	Ear	Musculoskelatal hypoplasia	Oral involvement	Associated abnormalities
9	M. S. 4 Yrs Male	Right	Poor development of 1st and 2nd arch tubs Low placed ear	Mandibular Maxillary	Nil	Facial palsy (Rt side)
10	A.P.S. 11 Yrs Male	Left	Poor dev. of 1st arch tubercles	Mandibular	Nil	Nil
11	NNS 16 Yrs Male	Right	Poor dev of 1st arch tubercles	Mandibular	Nil	2nd arch tubercle also affected
12	M. T. 16 Yrs Male	Right	Accessary tabs, Atresia ext. audit. canal	Maxillary & Mandibular	Nil	Rt. side palatal palsy
13	A. K. 11 Yrs Female	Right	Abnormal tragus, Atresia ext. audit. canal	Mandibular	Nil	Nil
14	O.N.J. 15 Yrs Male	Right	Poor dev. of 1st arch tubercle	Mandibular	Nil	Nil

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