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# An Unusual Case of Otomandibular Dysplasia

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***Abstract:***
*Otomandibular dysplasia are characterised by the combination of anomalies of the ear and the mandible. Otomandibular dysplasia is a branchial arch deformity which needs correction.The objective of this study was to describe to illustrate the imaging aspects of otomandibular dysplasia through a case presented in D Y Patil Hospital, Navi Mumbai and to highlight the contribution of imaging in the preoperative assessment of the lesions of otomandibular dysplasia.*

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## I. Introduction:

Otomandibular Dysplasia is a condition in which the lower half of one side of face is underdeveloped and does not grow normally. It is also sometimes referred to as: hemifacial microsomia, first and second branchial arch syndrome, oral-mandibular-auricular syndrome, lateral facial syndrome, or otomandibular dystosis. Acral anomalies associated with facial dysostosis allow the distinction between Treacher-Collins syndrome and acrofacial dysostosis (Nager and Miller syndromes). Unilateral and bilateral asymmetrical anomalies, namely facioauriculovertebral syndrome, hemifacial microsomia, otomandibular dysostosis, no. 7 cleft, first branchial arch syndrome, Goldenhar syndrome were lumping together by Gorlin in 1990, who proposed to use the term "oculoauriculo vertebral spectrum".The syndrome varies in severity, but always includes the underdevelopment of the ear and mandible. This is the second most common facial birth defect after clefts.

## II. Case Study:

A 13 years old girl presented to the ENT OPD with need of operative correction of the deformed left ear (microtia) and ipsilateral left sided deviation of the chinsince birth. She had undergone HRCT temporal bone for preoperative assessment and further correction of the deformity.

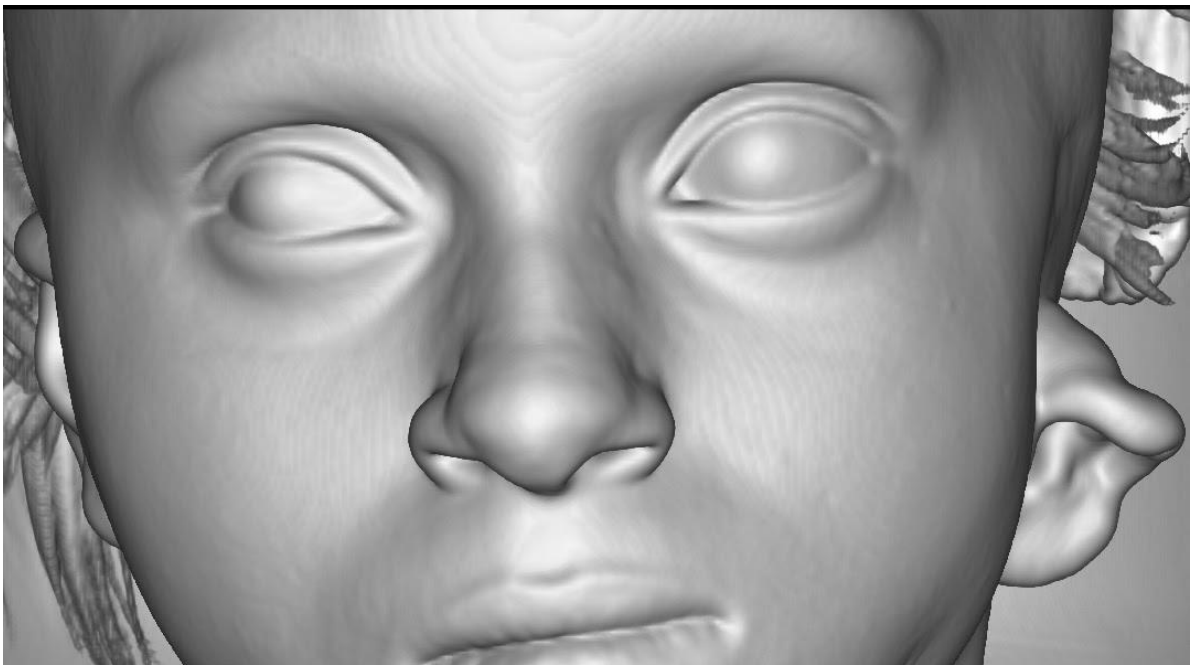


Figure: Deformed left ear (microtia) and ipsilateral left sided deviation of the chin.

**AURICULAR DEFECTS**

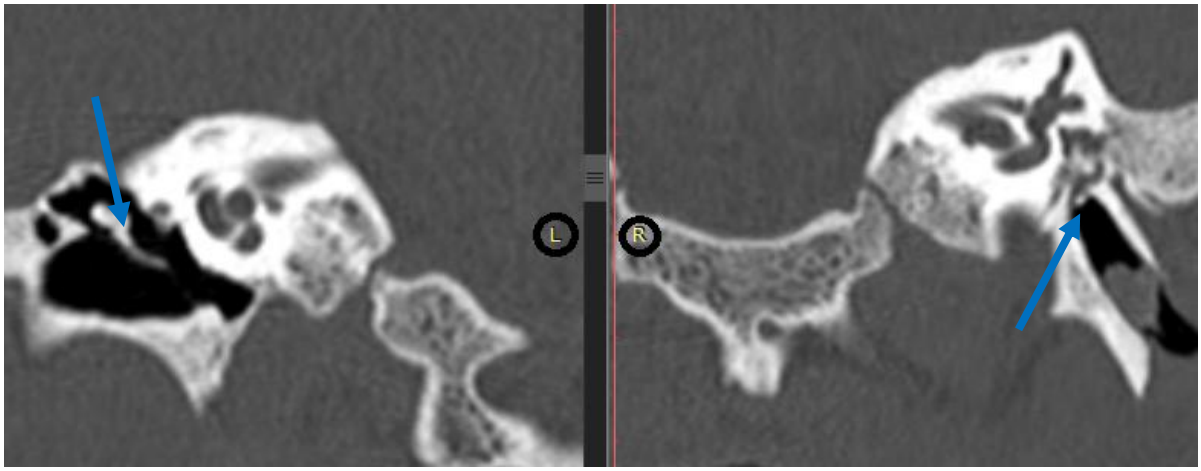



Figure: CT axial image showing small left middle ear cavity with few hypoplastic ossicular remnants.*(→)*

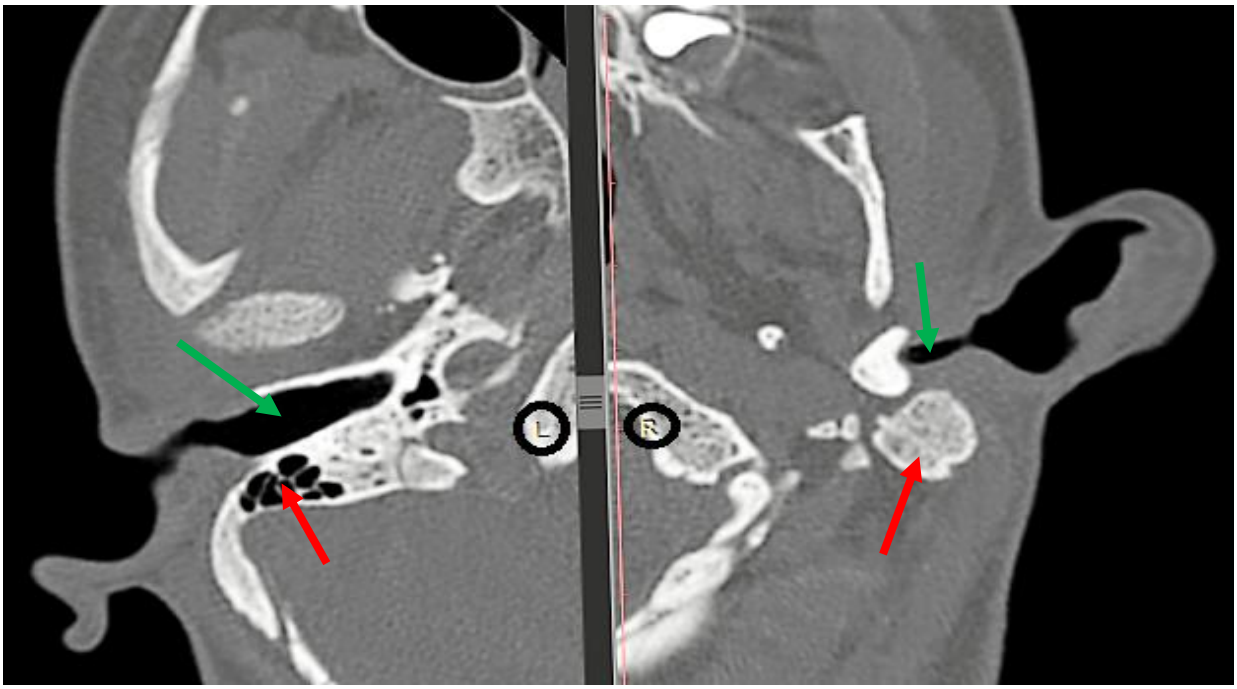



Figure: CT axial images showing left small external auditory meatus and external auditory canal as compared to right. (→).

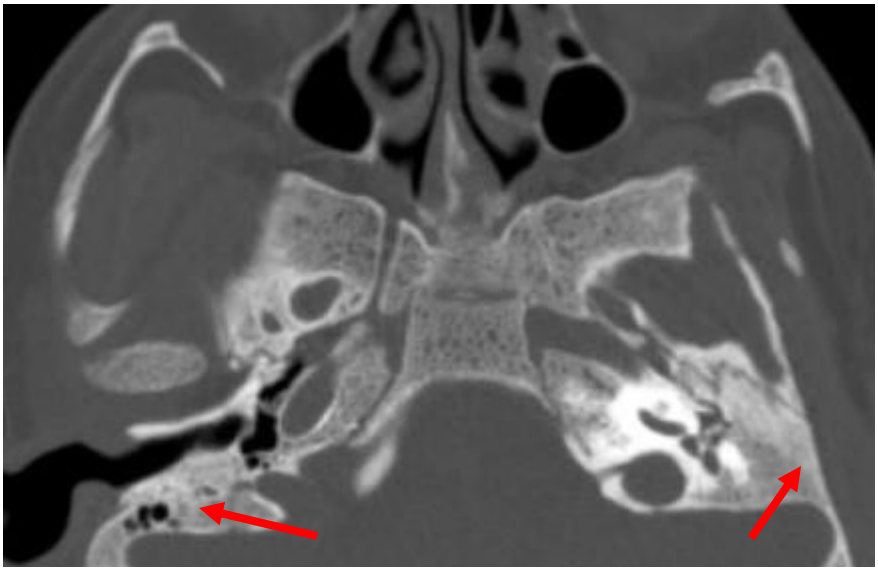


Figure: CT axial image showing hypo-pneumatization of the left mastoid air cells. ( →)

**MANDIBULAR DEFECTS**

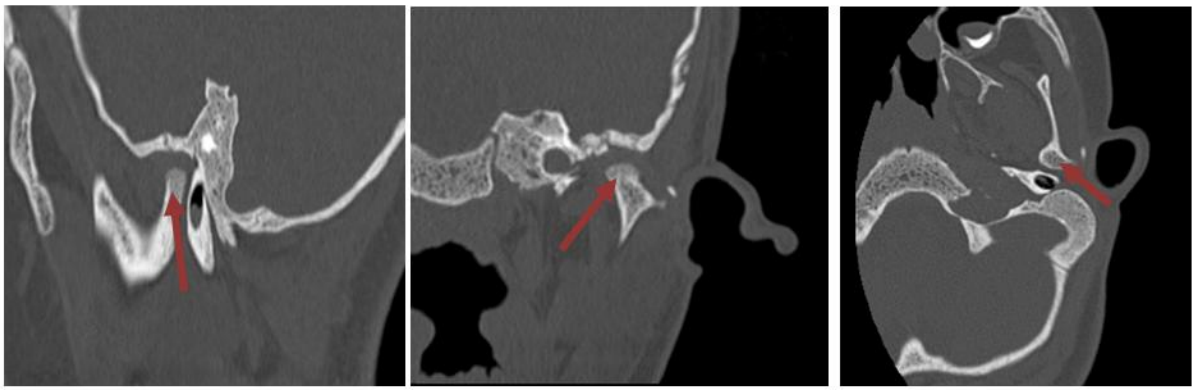


Figure: CT sagittal, coronal and axial images showing hypoplastic left mandibular condyle with foreshortened and underdeveloped right mandibular ramus. (➡)

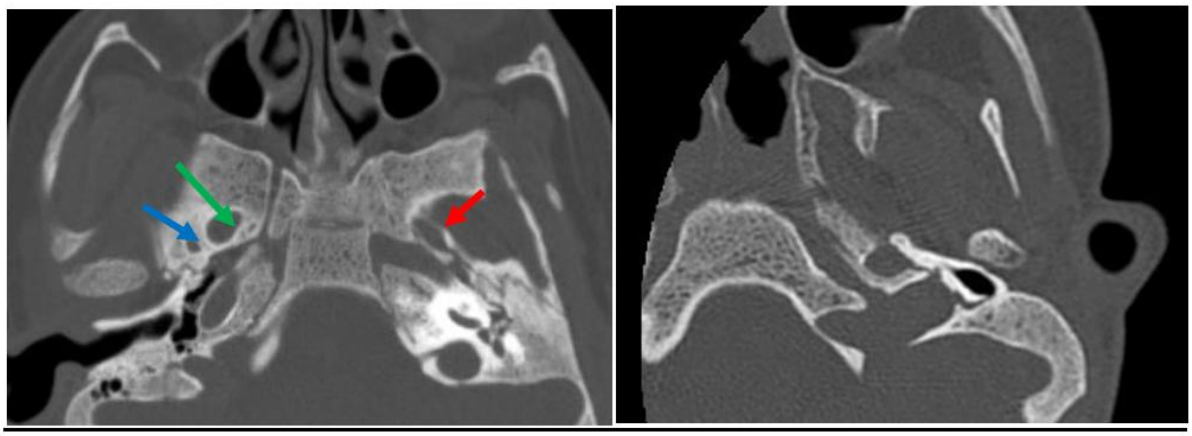
**PRUZANSKY CLASSIFICATION**

**Grade I:** minimal hypoplasia of the mandible

**Grade II:** functioning but deformed temporo-mandibular joint with anteriorly and medially displaced condyle

**Grade III**: absence of the ramus and glenoid fossa

**FORAMEN DEFORMITIES**



CT axial images showing hypoplastic left Vidian canal with poor visualization of foramen ovale and spinosum(➡).

Normal right foramen ovale (➡) and right foramen spinosum (➡).

**FACIAL NERVE DEFECTS**

The labyrinthine segment of the left facial nerve appears thin in calibre with absent 1st genu. Tympanic segment of the left facial nerve not well visualized with the variant anterior course of the labyrinthine segment of the facial nerve.

## III. Discussion:

It is primarily a syndrome of first and second branchial arch where there is deficiency in the amount of hard and soft tissue on one side of the face. It involves underdevelopment of the temporo-mandibular joint, mandibular ramus, masticatory muscles and ear.Usually unilateral and always asymmetrical if presents bilaterally.

**GOLDENHAR SYNDROME:** A variant of HFM in which vertebral anomalies and epibulbar dermoids were present.

## IV. Conclusion:

Computed tomography (CT) is the best modality to provide a three dimensional view of both hard and soft tissue. Information on comparative muscle development can be assessed through CT or MRI on a case by case basis.

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