

Reply

Dr. Nair has raised the issue of ventilation in 11 cases of transient tachypnea of the newborn (TTNB). We have elaborated upon this diagnosis in the text. This label was assigned to cases of respiratory distress in whom conditions such as hyaline membrane disease, pneumonitis, aspiration, malformation can be excluded. Thus, essentially, it was a diagnosis of exclusion. However, they all fulfilled the criteria for ventilation (*i.e.*, progressive respiratory distress with inability to maintain normal blood gases in oxygen hood at FiO₂ 0.6). In all of them, the possibilities such as HMD or pneumonia could not be excluded at the point of initiating ventilatory support. However, they had a benign course as

evidenced by need for only CPAP in all except two of them. The two who needed IPPV were preterm babies whose initial course resembled HMD, but unlike cases of HMD they recovered within 48-72 hours. All units which institute ventilation must be coming across cases of transient respiratory distress as encountered by us. We have used the term TTNB in a broad sense to label such cases.

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Rare Associations with Goldenhar Syndrome

A case of Goldenhar Syndrome with rare association of anophthalmos and calcification of falx cerebri is reported.

A 5-year-old boy was brought with the complaints of asymmetry of the face since birth. On examination the child was averagely built and nourished. He had hemifacial hypoplasia with the clinical signs of frontal bossing, plagiocephaly, microphthalmos (left side), microtia (left side), mandibular hypoplasia (left side) and mental subnormality. A detailed otorhinolaryngo-

logical examination revealed microtia of the left pinna which was displaced inferiorly along with atresia of left external auditory canal and conductive hearing loss. The right ear, nose and throat were normal. The left eye showed anophthalmos and blepharophimosis. The right eye was normal. Systemic examination was essentially normal. X-ray skull revealed plagiocephaly, flat occiput, micro-orbit (left side) hemimandible (left side) especially condylar hypoplasia, hypoplasia of left sided facial bones and calcification of falx cerebri (*Fig. 1*). X-ray cervical spine revealed congenital block vertebra CV2, CV3 and CV4 with spina bifida CV6 and CV7. X-ray chest revealed 13 ribs with rudimentary 1st ribs on both sides.

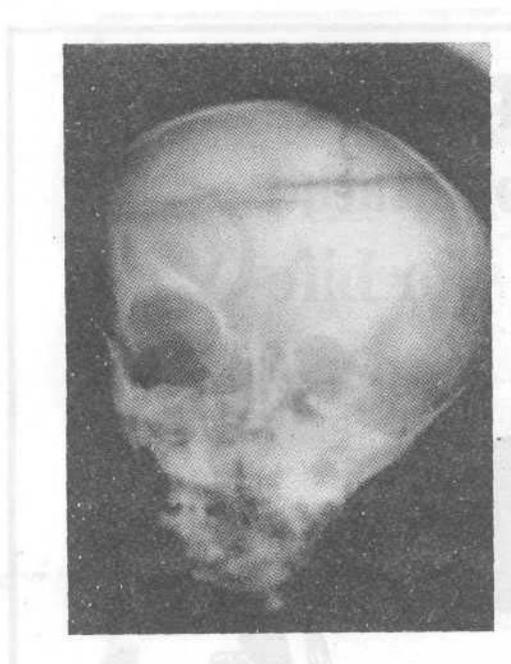


Fig. 1. X-ray skull AP view showing micro orbit (left side), hypoplasia of facial bones (left side), hypoplastic mandible (left side) and falx calcification

In 1952, Goldenhar gave the first description of a combination of epibulbar dermoids, auricular appendices and vertebral anomalies which goes with the name of Goldenhar syndrome. Since then about 150 cases have been traced in the available literature(1). Estimates of the incidence of the syndrome have ranged from 1 per 5,600 to 1 per 3,500 births. The cause of this malformation has focussed primarily on devel-

opmental defects involving derivatives of first and second branchial arches. Poswillo concluded that a hemorrhage or occlusion of the stapedia artery before its normal regression in embryonic life could lead to abnormalities in derivatives of the first and second branchial arches(2).

Our patient had all the features of Goldenhar Syndrome except epibulbar dermoid. The rare association of anophthalmos and blepharophimosis has not been reported earlier in the available literature. The finding of calcification of falx has been reported previously in one case only(3). The child is being managed with reconstructive procedures.

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