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Half Nose

by

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Summary

A very rare abnormality is reported of congenital absence of the right half of the nose in combination with microphthalmia, coloboma, epiphora and median cleft of the alveolus.

Case report

F. R. was born on 8 March 1982, and referred to ENT outpatients on the next day as only half the nose was present. The boy was born of a full-term pregnancy, his mother's second. There was no history of illness during pregnancy and he weighed 6½ lbs at birth. There was no family history in the mother's or the father's family of any congenital abnormality in two previous generations. Examination showed a healthy infant in his mother's lap breathing quietly through the left half of his nose. The right half of nose was completely absent and there was a depression in place of the right nasal bone.The right eye was smaller and coloboma was present. There was a swelling attached to the medial end of the upper eyelid like a small proboscis and mucus was discharging from the lower canaliculus (Fig. 1). Ocular pendular nystagmus was present. The nasal cavity, turbinates and nasal septum on the left side were normal; examination of the mouth revealed a midline cleft of the alveolus (Fig. 2). The hard palate looked normal and no submucosal cleft could be felt by digital palpation.

X-Ray examination showed normal maxillary sinuses but there was a bony defect in the hard palate. An Olympus fibreoptic nasopharyngoscope was passed under sedation and local anaesthetic and this revealed a normal left choana. On the right side the Eustachian opening was present but above it there was a blind pit, with the nasal septum pushed towards the right side, with slight widening of the left choana.

This boy has been observed for 19 months. He has had three attacks of dacryocystitis and there is constant epiphora which was noted on earlier examinations. Pus can be expressed on pressure over the lacrimal sac and the depression of the right nasal bone has increased with age.

Discussion

According to Birrell (1978), absence of one half of the nose is very rare. Cases have been reported by Fatin (1955), by Ogura and Onoda (1967) and by Kukreja (1973). Other cases have been reported in the German literature: Selenkoff (1884) reported, postmortem, complete absence of the right half of the nose with a high arched palate in an adult male who had died of alcohol and fever; Tiefenthal (1910) reported absence of the right half of the nose with deformity of the right half of the face in a 37-year-old female; and Wicke (1972) reported another case in a 3-year-old female child who was born prematurely and whose breathing was obstructed.

Schweckendiek and Hein (1973) also described one case and mentioned seven others. In their own case, unilateral absence of the nose was associated with microphthalmia, coloboma, ocular nystagmus and complete cleft palate, including a median cleft of the alveolus.

This appears to be the first case in which congenital absence of half the nose has been associated with ocular and palatal abnormalities. The present case was also associated with abnormalities of the eye and palate, the difference being that cleft palate was minimal in our own case, involving only the alveolus and a bony defect in the hard palate.

Handousa-Bey (1949), while discussing midline congenital malformations, suggested that these abnormalities could not be explained by conventionally described embryological development and put forward a theory of bilateral origin of the nose, both sides developing symmetrically to fuse together in the midline in early embryonic life. Badrawy (1967) suggested that the two nasal cavities come together, forming a front-nasal process.

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FIG. 1

The boy in his first week of life showing absence of right half of nose and a proboscis in the right eye.



FIG. 2

The upper lip has been retracted to show a midline cleft of the alveolus.

This is broad at first but later thins out by a process of folding of the nasal septum, which fuses in the midline, thus providing separate medial and lateral walls to each nasal cavity. This allows independent growth of each cavity, one half of the nose developing normally while the other half can be retarded.

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