Indian Journal of Plastic Surgery (1988), 21(1), pp. 56-58

COMPLETE CONGENITAL ABSENCE OF NOSE A CASE REPORT

M. H. KHAN, A. B. KHAN AND A. H. KHAN

SUMMARY

A case of complete congenital absence of the nose is being reported for its rarity along with the review of literature.

(*Key Words* : Nose, Nasal deformities)

Case Report

A six weeks old, male baby was referred to the plastic surgery unit of the J. N. Medical College, Aligarh from the Department of Paediatrics because the baby had complete congenital absence of the nose (Fig. 1). The patient was admitted for further investigations and evaluation.

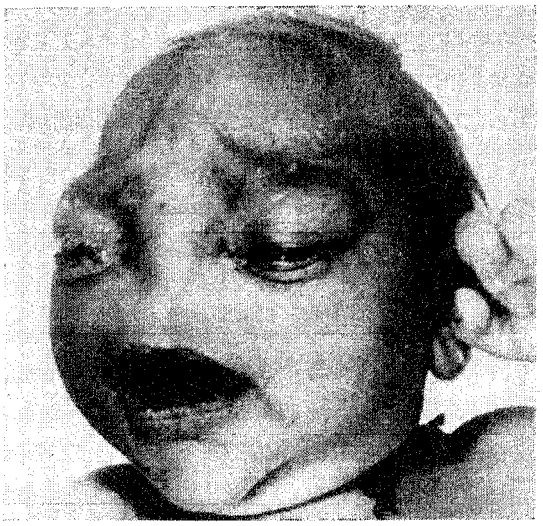

Fig. 1. Baby with complete absence of nose.

The baby was the result of a full term normal delivery. Family history of siblings having congenital anomalies was absent. There was no history of the mother having taken drugs or having been exposed to radiation during the pregnancy. Feeding was initiated soon after birth, with a cleft lip feeder. There were no signs of respiratory distress.

On examination, the face appeared large. There was bossing between the eyes and the lower part of the forehead. There was complete absence of the nose and nasal airway. The

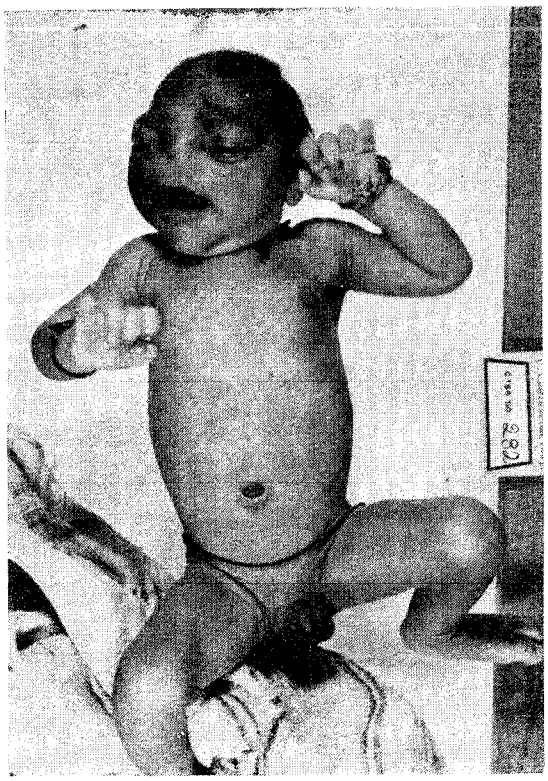

Fig. 2. Baby with complete absence of nose, no other anomaly.

upper lip was well formed with a normal philtral groove. The hard palate was high arched but the soft palate was normal and functioned normally. The eyes appeared large and widely placed. There was no evidence of any congenital heart disease. The patient was passing faeces and urine normally. There were no other associated congenital anomalies (Fig. 2).

The patient was discharged on request and the parents were instructed to bring him again at the age of 3 years for review.

Review of literature: Complete congenital absence of the nose is an extremely rare anomaly. Cases have been reported by Wakely (1904), Denis (1919) and Walker (1961). Gifford and McCullum (1972) reported two cases associated with complete absence of nose and nasopharynx. Wahby (1903) reported examination of a skull from the Cambridge University Museum in which the nasal bones were absent along with the premaxilla and there was hypotelorism. Davis (1963) mentioned this anomaly in his text book and suggested a V-shaped, midline, upper lip flap to shape the nostrils and the columella. Blair described a classical repair of such a case. Berger and Martin have reported a single case of this anomaly.

Discussion

The formation of the nose occurs between 2 to 28 days of the intra uterine life by the sinking in of the olfactory pits of ectoderm. This is associated with proliferation and thinning out of the surrounding mesoderm at 35 to 38th day of IUL or 10 mm stage of the embryo. The 2 layers of epithelum rupture leaving a primitive choanal opening posterior to the premaxilla. Now the horizontal process of the maxilla and the palatine bones develop from the masses of mesoderm lying laterally. This leads to the formation of the secondary palate. This mesoderm is also invaded by the olfactory pits which grow caudally to join the foregut growing anteriorly. At the meeting point the two epithelial surfaces rupture giving rise to the establishment of the posterior choanae.

In the case under review there was complete absence of the nasal openings and the external nose.

The external nose develops in the 2nd month of intra uterine life from the fusion of the fronto nasal processes. The Olfactory epithelum will not be formed in the absence of invagination of the nasal placodes. So such babies are expected to lack a sense of smell alongwith congenital absence of the nose.

The secondary palate in these cases will presumably be abnormally high because of the absence of the nasal cavity and the nasal septum.

Cases of complete congenital absence of the nose have been reported to progress well and reconstructive rhinoplasty has been suggested when the patient has attained adult size and dimensions. In the intervening period a nasal airway can be made at the site where the external nares should be and a silicone nasal prosthesis put in place. This procedure will have 2 advantages then—

1. The patient will be able to breathe while eating and so will not have to gulp down his food like a canine.
2. The prosthesis will boost up self confidence and make them socially acceptable.

REFERENCES

1. Davis, J.: Human Developmental Anatomy. Ronald Press Company, New York. 1963.
2. Gifford, G. H. and McCullum, D. W.: In: Paediatric Otolaryngology. C. F. Ferguson and E. L. Kendig Jr. (eds.), W. B. Saunders, Philadelphia, 1972; p. 932.
3. Scott-Browns: Diseases of the Ear, Nose and Throat. 4th edition. 1979; p. 73-81.
4. Sprinkle, P. M. and Sporck, F. T.: Congenital malformations of the nose and paranasal sinuses. In: Paediatric Otolaryngology. C. H. Bluestone and S. E. Stool (eds.), W. B. Saunders, Philadelphia, 1983; p. 769-780.

5. WAHBY, B. : Congenital absence of the Nose and premaxilla J. Anat. 1903; 4 : 38-49.
6. WALKER, D. G. : Malformations of the Face, E. and S. Livingstone Ltd., Edinburgh, 1961; p. 191.

The Authors

PROF. MASOOD H. KHAN, *Professor*, Deptt. of Surgery, J.N.M.C., A.M.U. Aligarh.
DR. ARIF BASIR KHAN, *Postgraduate*, Deptt. of Surgery, J.N.M.C., A.M.U. Aligarh.
DR. ARSHAD H. KHAN, *Clinical Registrar*, Deptt. of Surgery, J.N.M.C., A.M.U. Aligarh.

Request for Reprints

PROF. M. H. KHAN, M.S., M.S., *Professor & Head* of the Plastic Surgery, J. N. Medical College, Aligarh.