Double nose

By P. Ghosh, Mrs. K. Dada, Mrs. S. Gupta, N. Jamwal and R. Daljit
(New Delhi)

Congenital deformities of the nose which is largely responsible for beauty and personality of a person is not rare, but the incidence is less than that of aural deformities. Presence of two complete noses, complete absence of nose and single median nose as in cyclops are extremely rare. The first case of double nose was probably reported by Muecke and Souttar (1923) and the second one by Erich (1962). Anomalies, e.g. bifid nose, proboscis lateralis, choanal atresia (unilateral or bilateral), atresia of anterior nares, rudimentary nasal appendages, etc., are seen more often. There may be associated lip and/or palate deformity, as median cleft of the upper lip is quite often an accompaniment of bifid nose. Association of hypertelorism is not uncommon, though it is more apparent than real in most of the cases. These all give a preternatural and demoniacal look to the baby, apart from the functional disabilities that are quite often concomitant. This is fraught with the risk of the parents being outcast by the superstitious society. Moreover, when the child grows with the deformity he is likely to develop serious psychological problems. Bilateral choanal atresia may lead to inanition and retardation of growth and speech defect. And that is why an early correction is mandatory for cosmetic improvement and functional rehabilitation. The following is the report of a case having two complete noses without any other congenital deformity elsewhere in the body.

Case report

Baby G, 22 days old, was brought to the hospital with the following complaints.

1. Double nose with widely separated eyes since birth.
2. Inability to breathe through nose. Breathes through mouth only and gets exhausted and restless periodically.
3. Inability to suck mother's breast effectively and continuously. In between it has to stop sucking for breathing. Failure in putting on weight.
4. Excessive nasal discharge (purulent) from all the four nostrils.

Perinatal History was non-contributory. Respiration in the neonatal period was not unduly alarming. No history of cyanosis or jaundice was present.

Mother is not an elderly primae, does not have diabetes mellitus and was all right during pregnancy. Father is hale and hearty. There was no positive family history of anybody having congenital or hereditary disease. Parents did not have any major surgery under general anaesthesia, never had sunstroke and did not undergo any radiological procedures which might be responsible for bringing about genetic mutation.

On examination: An emaciated small baby, periodically struggling for air followed each time by opening the mouth wide and inspiring noisily. There

https://doi.org/10.1017/S0022215100074284 Published online by Cambridge University Press

was slight tugging in of the suprasternal notch. There was no other congenital anomaly elsewhere in the body. Systemic examination revealed no abnormality.

Local examination: There were two complete noses with a discharging sinus in the upper part of the internasal space which, on probing, was found to be communicating with the left medial nasal fossa. The medial nostrils were smaller than the lateral ones. The nasal fossae were full of mucopus. After sucking out, the mucosa was found to be moderately congested. There were small turbinals (only inferior ones could be seen) in the lateral nasal fossae, but none in the medial ones. Rubber catheter could not be passed into the nasopharynx through any of the anterior nares. With eustachian catheter the choanal atretic partitions could be felt to be quite hard and bony. Palate and the lips were normal. No other deformity in the pharynx or ears was detected. There was marked hypertelorism and a dermoid cyst on the glabella region. No bony defect was felt there. The hearing was normal.

Nasograms—injection of the dye into the nasal fossae did not yield a good skiagram. Small cotton pledgets, soaked in dinosil, were inserted into the nasal fossae and a satisfactory skiagram obtained which showed that the ends of the

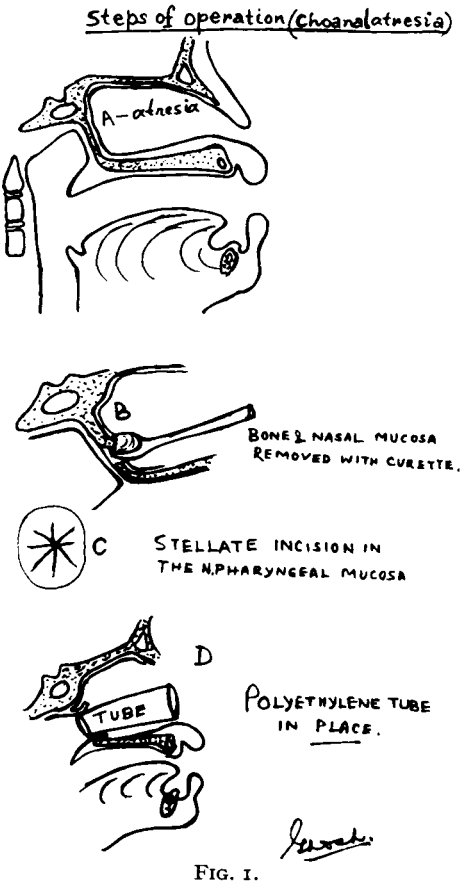


FIG. 1.

https://doi.org/10.1017/S0022215100074284 Published online by Cambridge University Press

pledgets stopped short at the atretic plates which were bony. Air shadow of the nasopharynx did not communicate with that of nasal cavity.

X-ray spines—no bony lesion seen. No basilar impression was present.

Sphenoid bone shows a vertical disposition with the hypophysial fossa looking directly backwards. The sphenoid body is solid (Fig. 1).

Other sinuses—normal (Fig. 1a).

Interorbital space is wider than usual. Skull shows patchy ossification, and sutures and fontanelles are too wide. Clavicles and shoulder joints were normal.

Heart—normal in size and shape. No situs inversus was discovered.

There were broncho-pneumonic changes in the lungs which cleared up after a course of antibiotics.

Blood and urine examinations did not reveal any abnormality.

Treatment: Under general anaesthesia through endotracheal tube the atretic plates closing the lateral nasal fossae were broken in the following manner (Fig. 1). The medial fossae were left untouched because, (1) these were very small and rudimentary and (2) for performing rhinoplasty in future, since it has been tentatively planned that the medial fossae, which are lacking in functional tissues (viz. turbinals), would be removed followed by anastomosis of the lateral halves with each other to make one complete nose.

After perforating the atretic plates two polyethylene tubes of proper size were brought through the nose from behind with the help of a rubber catheter passed through the anterior nares bringing out through the mouth. The posterior ends of the tubes were at the level of the nasopharynx just behind the planes of atresia, this being ensured by palpation, inserting a fine polyethylene suction catheter into the nasal tubes and by the presence of easy breathing with mouth closed after the endotracheal tube was removed.

The nasal tubes were fixed by a safety pin and held in place by means of adhesive straps. The tubes were kept *in situ* for 3 weeks with, of course, weekly changing (Figs. 2 and 3). After removal of the tubes the patient could breathe normally through the nose and by about 4 weeks it had gained 4 lbs. There is no nasal discharge now. (Fig. 4).

At a subsequent stage it is proposed that the following surgical measures would be undertaken: (1) fistulectomy, (2) removal of the dermoid cyst and (3) correction of the deformity offering the patient a single functioning nose.

Discussion and Comments

The development of the nose is complex indeed. The development of face during the second month of gestation is dominated by the formation of the nose so that the embryo assumes a human shape. The prospective homosapiens thus attempts at successfully emerging into a definitive human form from a shapeless embryo by the predominant development of the snout.

The frontonasal process is the most important component in the development of the nose. Olfactory placodes deepen to form the olfactory pits which mature into primitive nasal fossae. The deeper portion of the lower part of the frontonasal process goes to form the premaxilla and the primitive nasal septum, and the superficial part is incorporated in the upper lip. The head broadens and the median portion of the frontonasal process is compressed with apposition

https://doi.org/10.1017/S0022215100074284 Published online by Cambridge University Press



FIG. 2.



FIG. 3.

https://doi.org/10.1017/S0022215100074284 Published online by Cambridge University Press

of the nostrils and nasal fossae to each other with the result that septum and the dorsum of the nose are formed. From either side two tectoseptal processes arise from the maxillary swellings of the first arch and extend medially to meet each other and project downwards in the midline as a bilaminar plate to form the secondary nasal septum. This fuses with the posterior border of the primitive septum to form the definitive septum thereby dividing the nasal cavity into two halves. Along with this process the bucconasal membrane which

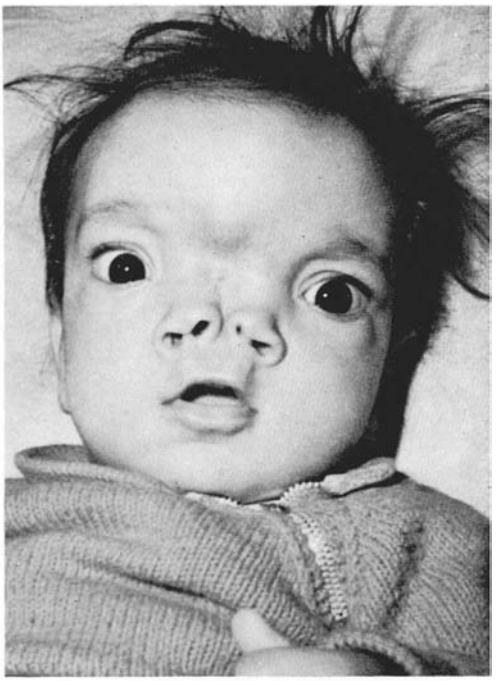


FIG. 4.

was separating the nasal fossae from the oral cavity migrates posteriorly and ruptures during the second month. Persistence of this is the cause of choanal atresia. At times an epithelial plug may be present in the choanal regions which may fail to get canalized giving rise to atresia.

The embryological explanation for double nose would not be very convincing. Muecke and Souttar (1923) thought that one of the noses was that of an included twin. This is very unlikely. Dichotomy by atavism leading to reduplication of nostrils was considered by Lindsay (1906), but does not seem plausible (Erich, 1962). Bifid nose or broad nose may be explained on the basis that the middle part of the frontonasal process has not been compressed adequately secondary to broadening of the head leading to failure of fusion of the 'nasal laminae' (Erich, 1962), that had grown backwards from the globular processes, along with failure of the tectoseptal processes to come in apposition to each other and fuse. Perhaps, the influence of the primary organizer (head organizer) through the elaboration of its evocator is responsible for duplication of an organ.

https://doi.org/10.1017/S0022215100074284 Published online by Cambridge University Press

This again depends on the competence of the substrate tissue. If, for any reason, the inductive stimulus of the organizer is separated, in space (Fig. 5a), into two or more components, the organ concerned may be subdivided or reduplicated (supernumerary organs, e.g. hyperdactylism, double ureter and kidney(Hamilton, Boyd and Mossman, 1964). Each stimulus will give rise to two nasal placodes

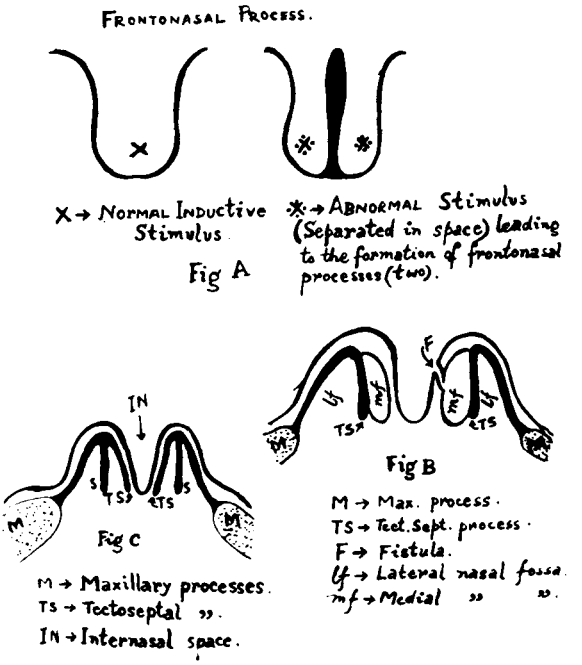


Fig. 5.

on each frontonasal process, two processes having been formed under the influence of double inductive stimuli. Presence of dermoid in the midline above the nasal bridge is more suggestive of the presence of two frontonasal processes which when uniting with each other in the upper part sequestrated some ectodermal tissue, the intermediate part having failed to unite. That the lower parts united is evidenced by the absence of palate or lip defect. The primitive nasal fossae and the septum developed in each frontonasal process as usual giving rise to two complete noses anteriorly. In this case there has been a failure of the tectoseptal processes to unite with each other in the midline accounting for the presence of the median cleft between the two noses (Fig. 5b). Under the influence of the separated inductive stimuli it is likely that there was a downgrowth (Fig. 5c) from the concavity of the tectoseptal process on each side to form the definitive septum.

The fistulous track was communicating with the medial nasal fossa of the left nose. If the medial crus of the tectoseptal process had extended medial to the medial nasal fossa, perhaps, the track would not have had the chance to communicate with the medial fossa. Cartilage is resistant to pressure and it is

https://doi.org/10.1017/S0022215100074284 Published online by Cambridge University Press

difficult for the track to pass through the cartilage (Littlewood, 1961). This tends to evince that the tectoseptal process turned downwards contributing towards the formation of the septum and consequently the medial fossa did not have any component of this process medially (Fig. 5b).

Because of this lack of mesodermal support, probably, functional tissue has not grown in the medial fossae. Moreover, as Erich (1962) postulates, some irregularity in the evolution of the two olfactory placodes causes them to bring forth four olfactory pits, rather than two and such an anomaly would alter the developmental pattern of the medial nasal processes; hence lateral nostrils are larger and more normal in size than the medial ones, since the lateral processes are not involved in the developmental defect. In all probability, identical development took place on the opposite side. In both the cases the vomer is expected to be unilaminar instead of a normal bilaminar one which will be verified at reconstructive surgery.

Summary

A case of double nose is reported. Possible explanations (embryological) have been given.

Acknowledgements

We are grateful to Professor S. Achaya, Head of the Department of Anatomy and Principal, Lady Hardinge Medical College, for her advice on the embryological aspect and kindly permitting us to use the case records. We also thank Professor A. Paul, Head of the Department of Paediatrics, for her kind help in preparing the article.

REFERENCES

Erich, J. B. (1962) *Plastic and Reconstructive Surgery*, **29,** 159.

Hamilton, W. J., Boyd, J. D., and Mossman, H. W. (1964) *Human Embryology*, Cambridge.

Littlewood, A. H. M. (1961) *British Journal of Plastic Surgery*, **14,** 169.

Muecke, F. F., and Souttar, H. S. (1923) *Proceedings of the Royal Society of Medicine* (*Section of Laryngology*), **17,** 8.

Department of Otolaryngology,
Lady Hardinge Medical College and Hospital,
New Delhi,
India.

https://doi.org/10.1017/S0022215100074284 Published online by Cambridge University Press

https://doi.org/10.1017/S0022215100074284 Published online by Cambridge University Press