Lateral nasal proboscis

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THE lateral nasal proboscis is a rare condition. Only nine references could be found, the most recent being Bishop (1964) and the earliest Keith (1923). Blair and Brown (1931), Young (1949), Smith (1950), MacLaren (1955), Bremar (1956) and Rao (1963) all describe one case. Biber (1949) gives a review of the literature and credits Selenkoff (1884) with the first description and describes one case. Rosen and Gitlin (1959) describe a case of bilateral proboscis and review the literature.

Case Report

History

The child, a boy, was examined at age 18 days. No family or antenatal history was available as he had been referred to the care of an Institution shortly after birth.

Clinical examination

The main abnormality was the tubular fleshy proboscis arising above the right inner canthus, approximately 3 cm. in length, and from $\frac{3}{4}$ cm. in breadth at its base to 1 cm. at its free end. The mass rested on the cheek; from an aperture on its free end a blind track extended along the length of the proboscis.

Beneath the proboscis a small lower lid cleft was hidden from view. The right side of the nose was undeveloped and an associated atresia of the right anterior naris was present. On the lower right surface of the nasal septum a blind hair-lined track passed into the septum (Fig. 1). A submucous cleft of the soft palate with bifid uvula was an additional finding. The upper lacrimal punctum was absent. The lower punctum was present and led downwards and inwards, ending with no evidence of a lacrimal sac. With growth of the face a mild degree of hypertelorism and slight hypoplasia of the right maxilla became apparent. Radiographic examination of the skull showed that the right nasal passages were hypoplastic.

Treatment

On the initial admission to hospital 2 years after birth, a palatal repair and a Hynes pharyngoplasty were performed.

Treatment of the nasal anomaly commenced at age 4. At the preliminary operation a hair-lined track passing into the nasal septum was excised. The right nasal cavity was exposed from the front by removal of bone. This prepared nasal cavity was lined with two local skin flaps. Four months later the proboscis was split vertically along its posterior surface for approximately 2 cm. spread, and its free borders inserted into the cheek and bridge of the existing nose to

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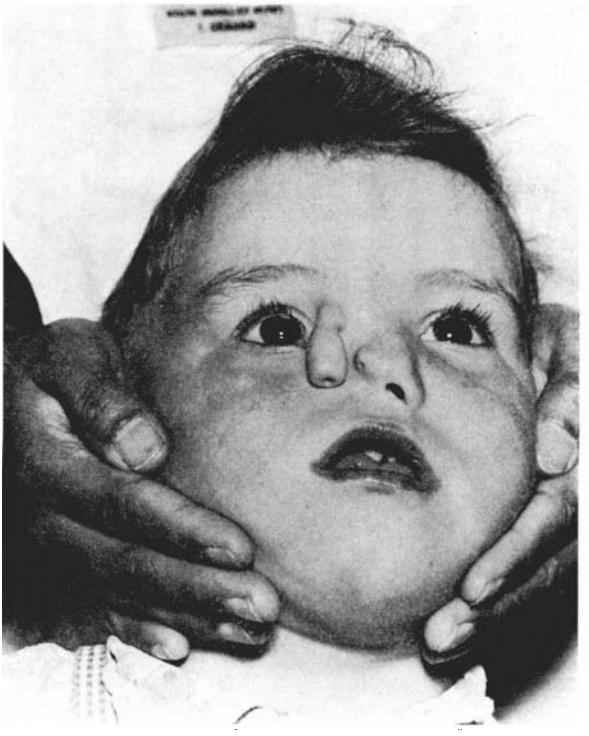


FIG. 1.

Child of 4 years of age before two-stage reconstruction of nose.
Note: Hair-lined track passing into nasal septum, aperture at free end of proboscis and lower coloboma palpebralum.

form a right ala (Fig. 2). The proboscis was divided 3 weeks later and the ala base inset. Post-operatively, pus discharged from a sinus at the base of the proboscis. The area was explored and the sinus lining and an associated bony spur removed. No communication with the meninges was found.

Assessment of the cosmetic result was made when the child was 6 years old and minor adjustments were made to the right side of the nose. At the same operation, a number of eyelashes on the inner surface of the right lower lid were excised and the lower lid cleft was closed in two layers. In the last 4 years growth of the normal tissue of the nose has outpaced that of the repaired side, and although the result is acceptable, further adjustment will be made (Fig. 3).

Comment

On inspection of the literature on such anomalies, the reviewer's attention is immediately attracted by their similarity.

The proboscis is typically described as a pendulous tubular mass arising superior to the inner canthus. It is normally between 2 and 4 cm.

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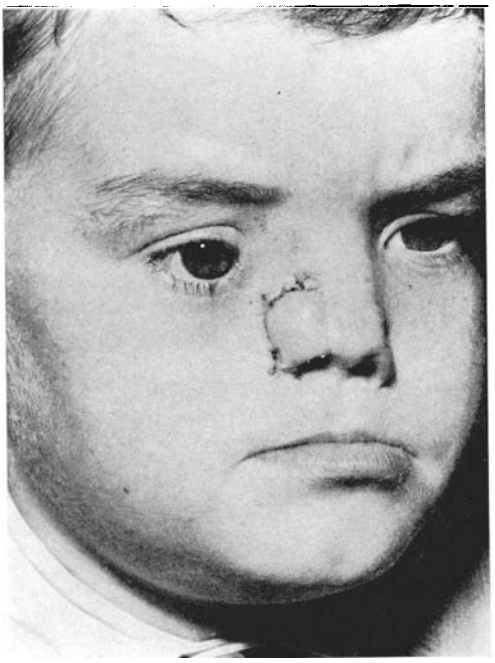


FIG. 2.
Child after reconstruction of the right side of the nose.

in length and ½ to 1 cm. in breadth, with a blind track of variable length leading from an orifice on its free end. In two cases, however, namely those of Young (1949) and Rosen and Gitlin (1959) the probosces were solid.

Biber in his review states: 'The histological examination of the removed proboscis in cases by Kirchmayer (1906), Selenkoff, and Zacherd (1920) and Seefelder (1910) showed invariably the same conditions; the covering skin has sebaceous glands. The connective tissue beneath it contains striated muscle fibres of the same kind as normally found in the facio-nasal muscles and some cartilagenous elements corresponding to the normal cartilage of the lateral nasal wall. The mucous lining of the central canal contains stratified columnar epithelium.' Bishop (1964) and MacLaren (1955) have described comparable histological findings.

The proboscis may be associated with nasal, ophthalmic, oral or intracranial abnormalities. No specific combination is apparent; no other generalized associated body deformities have been described.

The most common facial malformation is hypoplasia of a variable degree of the iso-lateral surface of the nose, as in the cases of Bishop (1964), Young (1949), Blair and Brown (1931) and Rosen and Gitlin (1959). MacLaren (1955) had, however, sufficient nostril and ala nasi present for reconstruction of the nose.

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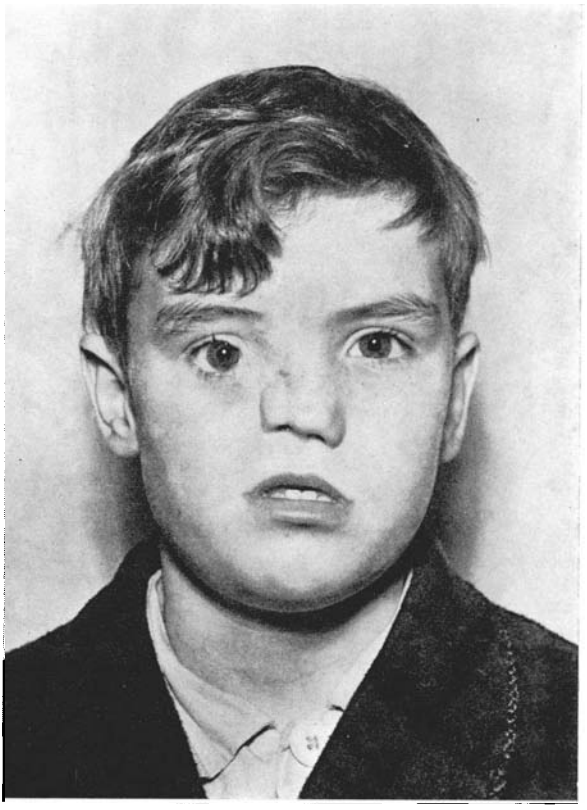


FIG. 3.
Child at 10 years of age. Note that the growth of the normal side has outpaced that of the repaired side.

Coloboma of the iris and clefts of upper and lower lid are the next most common anomaly. Cleft of lip and palate, absence of paranasal sinuses, absence of incisor teeth, have all been described. In a post-mortem examination in the cases of Seefelder (1910), Selenkoff (1920) and also in the bilateral case of Rosen and Gitlin (1959), the olfactory bulb and cribriform plates were absent on the side of the proboscis.

Discussion

Origin

Speculation as to the origin and character of the proboscis has resulted in three theories:

(1) It represents the naso-lacrimal duct.
(2) It is formed from a variable part of the lateral nasal process.
(3) It is formed from the lateral and medial nasal processes.

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Blair and Brown (1931) support the first view and comment on tears that appear to drain through the tube. Biber (1949) and Smith (1950) describe a similar picture of tears appearing at the orifice when the child cried. Smith (1950), by careful X-ray study of the tube after injection of lipiodol, demonstrated, in his case, that the proboscis was connected by a tear duct to the lacrimal sac.

The second view is supported by Biber (1949) who considers that where the nose is normal or slightly hypoplastic, the proboscis is a 'dislocated appendage' formed from a variable portion of the lateral nasal process. Rosen and Gitlin (1955) agree with this view and add that where there is absence of one side of the nose, the third hypothesis is true. Bremar (1956) believes the proboscis is formed from the lateral and medial nasal processes, and explains that the position of the philtrum may vary according to the degree of compensatory growth of the globular process of the normal side. Should the vacant globular space remain unfilled, a cleft lip will occur. This is seen in the cases of Keith (1923) and MacLaren (1955). I agree with the third hypothesis and present the following points as further evidence.

(a) There is atresia of the anterior nares.

(b) In those cases where post-mortem has been performed, the cribriform plate was missing and the olfactory bulb was either rudimentary or absent. Rosen and Gitlin (1959), Selenkoff (1920) and Seefelder (1910).

According to Hamilton *et al.* (1962) the olfactory nerve fibres develop from the primitive olfactory epithelium of the blind sac and grow towards the roof of the cartilaginous nasal capsule.

It is likely that there has been an abortive development of the nasal placode from the start with no development of the blind sac and its lining cells, allowing maxillary mesoderm to come into contact with the frontonasal process and in some manner isolating the tissue of the lateral and medial processes. Where the proboscis is present in conjunction with a normal or slightly hypoplastic nose, the possibility of the existence of two placodes on the same side must be considered.

Surgery

The ideal operation would remove the proboscis, replace the missing tissue and restore function. The proboscis is the obvious choice of tissue for reconstruction—'a natural tube pedicle'—it is identical in colour and texture to the normal side and readily accessible. Operations performed by Blair and Brown (1931), Biber (1949), Bishop (1964), Young (1949), Smith (1950) and the operation in the case described, all utilize the substance of the proboscis with slight variations in operative technique.

Biber (1949) operated on his case at age 6 months and considers that for the best cosmetic results, the operation should be performed at

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maturity, as there would be less danger of the normal side out-growing the operated side. Bishop (1964) was presented with his case when the child was 7, and felt that the results were reasonable at the time when the patient was last seen. In the case presented, the operation was performed at 4 years of age and since that time the normal side has out-grown the operated side.

A report from the association who looks after our patient, remarks 'he is not a very happy little boy, being aggressive and jealous of other children'. They have tried, without success, to find a foster home where there would not be the same competition. Biber's remarks would appear valid from a purely surgical point of view, but one wonders how long a child should be allowed to suffer such a deformity and be subjected to the mental distress which such an anomaly must incur. On taking these factors into consideration, an early repair is considered better.

Limited success has been achieved by previous authors in maintaining a patent nasal cavity. In the case presented there was failure. Smith (1950) considers the exposure of the nasal cavity unnecessary, as the skin lining required to maintain it would not duplicate the physiological function of ciliated columnar epithelium with mucous cells. Bishop (1964) designed an operation to utilize the epithelial lining of the proboscis, to line the cavity, but such a refined operation only seems justified where the nasal cavity is normal.

In conclusion, a cosmetic operation is suggested, providing an early reconstruction of the ala at 4 years before the child goes to school—over-correcting the repair in anticipation that the growth of the normal side will be greater than that of the repair, hence alleviating the necessity of regular adjustments.

Summary

A case of a lateral nasal proboscis is described. A review of the available literature is given.

The origin and surgery of these anomalies are discussed. An early cosmetic operation over-correcting the repair is suggested.

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REFERENCES

BIBER, J. J. (1949) *Journal of Laryngology and Otology*, **63,** 734.
BISHOP, B. W. (1964) *British Journal of Plastic Surgery*, **17,** 18.
BLAIR, V. P., and BROWN, J. B. (1931) *Surgery, Gynecology and Obstetrics*, **53,** 797.
BREMAR, J. L. (1956) *Congenital Anomalies of the Viscera: their Embryological Basis*. Cambridge, Mass.
HAMILTON, W. J., BOYD, J. D., and MOSSMAN, H. W. (1962) *Human Embryology*. Third edition, Cambridge.
KEITH, A. (1923) *Human Embryology and Morphology*. Fourth edition, London.
MACLAREN, L. R. (1955) *British Journal Plastic Surgery*, **8,** 57.
RAO, P. B. (1963) *Journal of Laryngology and Otology*, **77,** 1028.
ROSEN, Z., and GITLIN, G. (1959) *Archives of Otolaryngology*, **70,** 545.
SMITH, F. (1950) *Plastic and Reconstructive Surgery a Manual of Management*. Philadelphia.
YOUNG, F. (1949) *Plastic Reconstructive Surgery*, **4,** 59.

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