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Some clinical considerations on congenital anomalies of the nose

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SUMMARY

The diagnosis and treatment of nasal cysts and fistulas are reviewed on the basis of a material of 16 cases. Definite evidence of cyst size is often unobtainable. Surgery of nasal cysts and fistulas should be done as early as possible to assure unimpaired nasal growth. In secondary fistulas re-operations may be quite problematic.

Bifid noses vary substantially in terms of severity. Severity, in fact, will dictate the surgical techniques to be employed, i.e. decortication or external incisions. Saddling, which is a consistent finding, can be corrected by preserved cartilage. The results thus obtainable are satistactory by follow-up evidence.

Congenital flat nose is a rare anomaly. Eight cases were encountered in the past 10 years. The techniques of columellar lengthening and strutting of the nasal tip are reviewed.

Congenital anomalies of the nose are rarely seen in hospitals. This may explain, why they are frequently misdiagnosed and why the treatment ultimately instituted is inadequate, producing what often is an unsatisfactory result.

Disregarding the etiology and pathogenesis of nasal anomalies, this paper will primarily focus on the diagnosis and treatment, as practiced by the authors in nasal cysts and fistulas, bifid noses and congenital flat noses.

NASAL CYSTS AND FISTULAS

In the past years reports on nasal cysts and fistulas were published by Bruck and Kittinger, 1963; Crawford and Webster, 1952; Felleti and Guccione, 1971; Hoshaw and Walike, 1971; Hoskino et al., 1971; Legler, 1977; Little-

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wood, 1961; Masing, 1972; Müsebeck, 1968; Partheniades, 1965; Skolnik et al., 1971; Szalay and Bledsoe, 1972; Weisman and Johnson, 1964; Wayoff, 1968.

In our center 20 pertinent cases were seen and treated in the last 15 years. When compared with the total number of patients undergoing surgery of the nose, this is equivalent to a ratio of about 1 : 1000. These nasal cysts and fistulas are not very likely to be encountered at minor rhinologic services.

DIAGNOSIS

Roundish bulges along the nasal dorsum in infants should always be considered as suggestive of nasal cysts. In their presence the skin overlying the nasal dorsum should be carefully examined for fistulas. Often enough the small mouth of a fistula, which may manifest itself as a shallow depression in the nasal skin, will go unnoticed, unless examinations are done with a magnifying glass. We have repeatedly seen cutaneous fistulas, which invariably localize along the midline, to the altogether missed. The mouths of fistulas may be encountered anywhere along the nasal dorsum from the root of the nose down to the columella (Crawford and Webster (1952); Legler

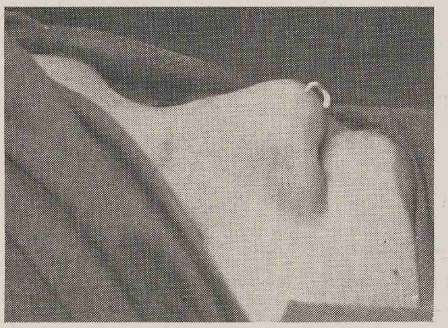


Figure 1a. Fistula on the nasal tip expelling dermoid mash by gentle pressure of the dorsum.

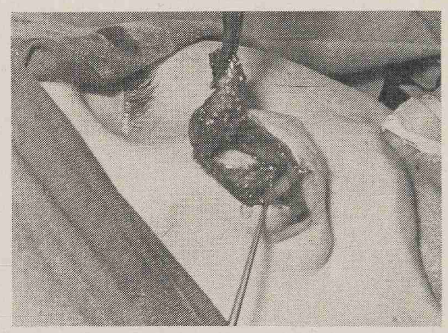


Figure 1b. Same case. The fistula is exposed and followed upward 25 mm to the end.

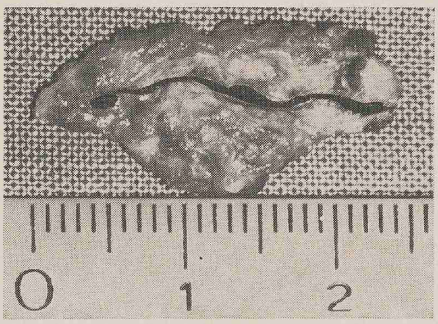
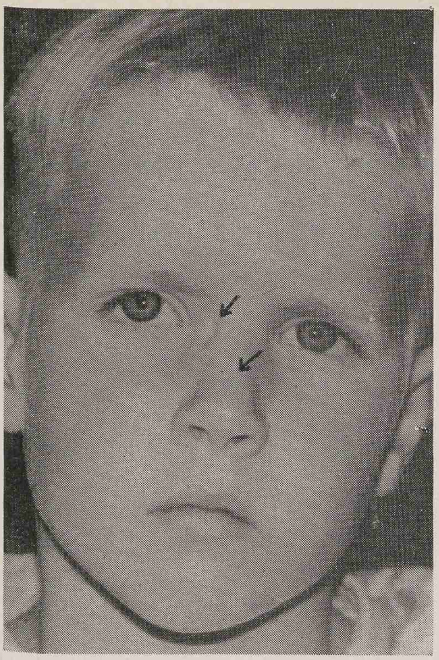
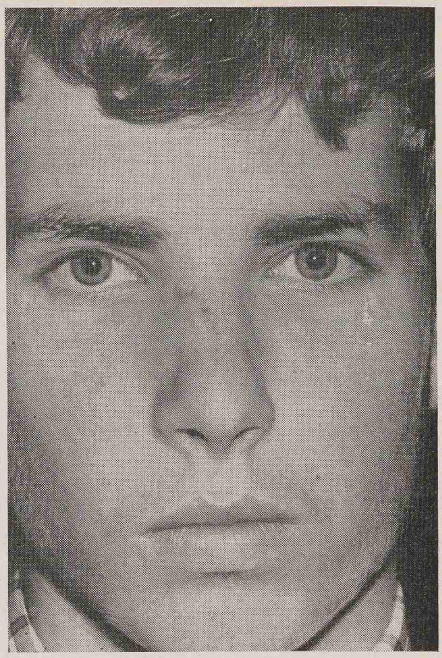
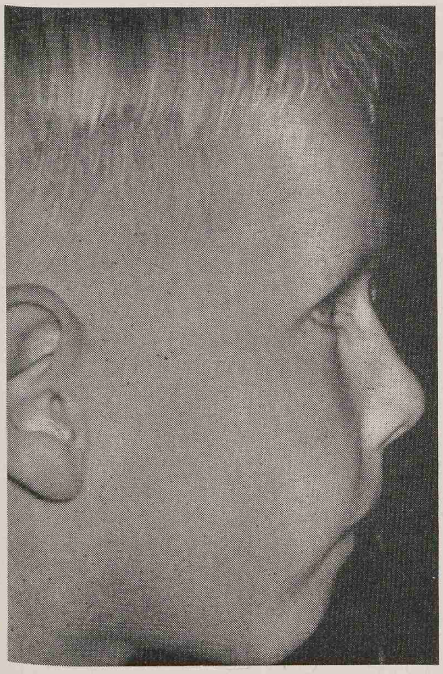


Figure 1c. Excised fistula opened.







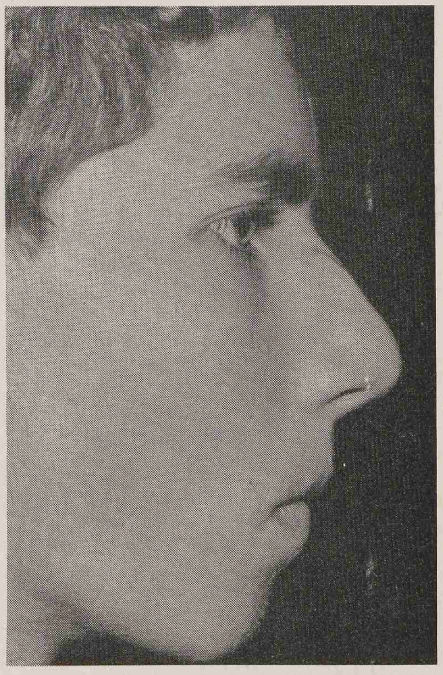


Figure 2a. 5 years old boy showing a hardly visible fistula on the dorsum (arrow) and a scar on the right nasal root after repeated abscesses.

Figure 2b. Follow up examination 12 years later after radical dissection of the fistula and reconstruction of the bony dorsum.

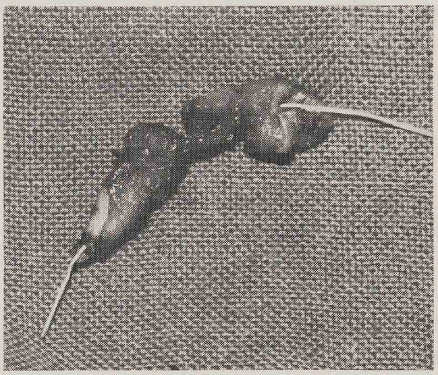


Figure 2c. Surgical specimen of the fistula, length 23 mm.

(1977); Masing (1972); Müsebeck (1968); Walter (1973)), (Figure 1). Müsebeck, in fact, distinguished between high, central and low cysts. Cysts may be superficial or lie deep to the nasal surface (Walter, 1973). Secondary cysts of an inflammatory origin which, as a rule, manifest themselves in terms of dacryocystitis at the medial canthus constitute a particular diagnostic problem (Masing). In the differential diagnosis of nasal cysts, gliomas (Hoshaw and Walike, 1971) and encephalomeningoceles should be ruled out. This is generally no problem, as they are usually combined with major central midface deformities.

Plain X-ray films are unlikely to furnish any definite evidence of the size of fistulas or cysts (Weisman and Johnson, 1964). Felletti consequently recommended to instill radiopaque substances in the fistulous tract. We have, however, found the tracts to be poorly visualized with this technique, so that no unequivocal information can be obtained their length and extension.

Probing of the fistulous tract is inconclusive, since the original situation may have been changed substantially by recurrent inflammatory conditions and previous surgery. Surgery had, in fact, been repeatedly done in one third of our cases, which suggests that the true extension of the fistula has been misjudged.

Methylene blue has been found to be best suited for visualizing the fistulous tract. It will generally also stain secondary fistulas (Denecke and Meyer, 1964; Masing, 1972).

Suction biopsies constitute an alternative procedure. The presence of dermoid material clinches the diagnosis.

TIMING OF SURGERY

In our view surgery for nasal fistulas and cysts should be done as early as possible. The youngest patient with a nasal cyst we ever operated was aged 14 months at the time of surgery. The presence of a cyst is bound to affect

the nasal infrastructure so that the nose tends to become increasingly broad and deformed. In addition, surgery will be appreciably complicated by repeated previous infections with extensive scarring. Both cysts and fistulas can be optimally visualized under the microscope or magnifying glass (Denecke and Meyer, 1964). There is, consequently, no reason to postpone surgery even the growth of the nose seems to be not influenced as shown in Figures 2a, b and c.

SURGICAL TECHNIQUE

The scope of this contribution precludes a review of the technique in all its details. These have, at any rate, been discussed extensively in pertinent publications and surgical manuals (Crawford and Webster, 1952; Denecke and Meyer, 1964; Masing, 1972; Walter, 1973). There is general agreement among workers in this field that, for the success of surgery to be permanent, cysts and fistulas should be radically removed.

In terms of priority, radicality thus overrides any cosmetic considerations. Incisions should be designed for an optimum visualization of the cyst in its entire extension (Crawford and Webster, 1952; Müsebeck, 1968). While nasal cysts and fistulas localize either above or below the embryonic cartilaginous capsule of the nose, i.e. the septo-dorsal cartilage, they never enter the capsule proper. Dissection is thus best done along the cartilage.

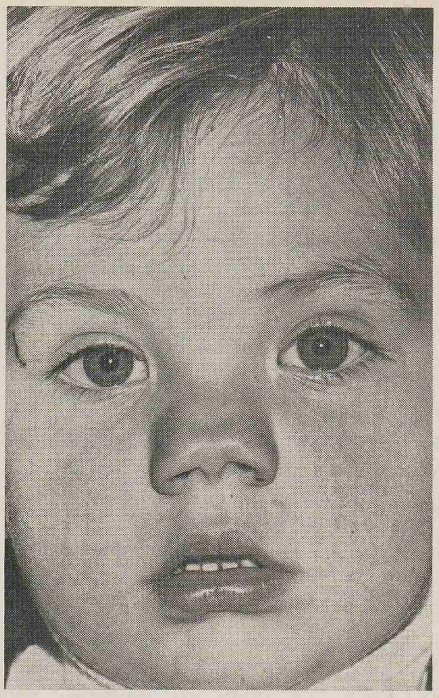
In fistulas we have come to use a midline approach throughout, the mouth of the fistula being circumcised in an ovoid pattern. Fistulas which continue below the nasal bones necessitate bone resection for exposure of the cartilaginous capsule. In older children this can be done by cutting, removing the nasal bones either in toto or in 2 fragments and reimplanting them at the end of surgery. In infants the nasal bones are often completely absent.

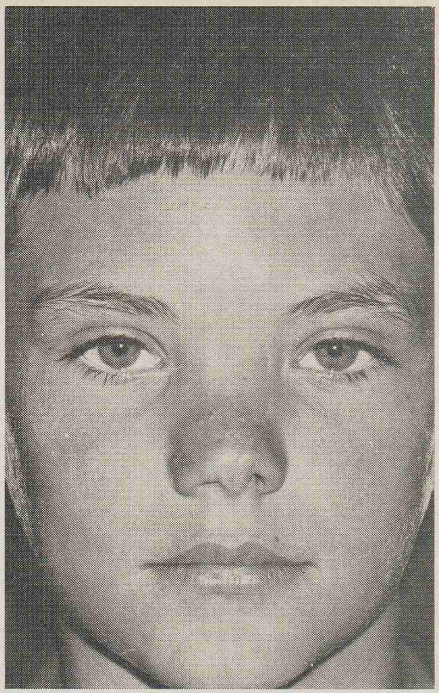
If a strictly subperichondral route is followed on dissection, rupture of the fistulous tract is unlikely. In most cases the tract ends in a cul-de-sac at the base of the skull. If there is no bone at this site, as in 2 of our cases, it will lead up to the dura. C.S.F. leakage in this area necessitates closure of the defect with a fascial flap.

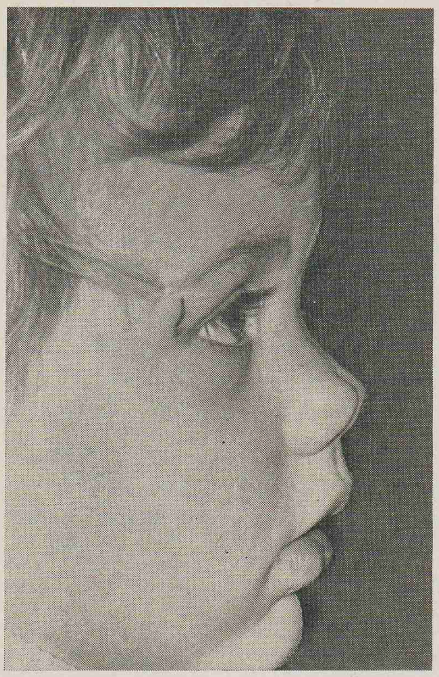
Once the tract has been excised, the defect is packed with cartilaginous, bony or fascial material. We have come to use preserved cartilage for this purpose and found it to heal unproblematically.

BIFID NOSE

Bifid nose is characterized by a median furrow which substantially extends the width of the nose. In the literature (Denecke and Meyer, 1964, Webster and Deming, 1950) a distiction has been made between minor, moderate and major cleft noses on the basis of the severity of the malformation. Severe







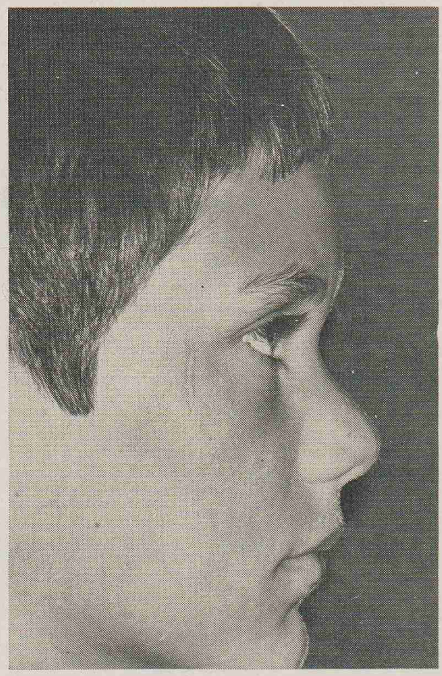


Figure 3a. 5 years old boy with bifid nose.

Figure 3b. Same case 5 years after surgical correction. Notice the tiny scar on the dorsum.

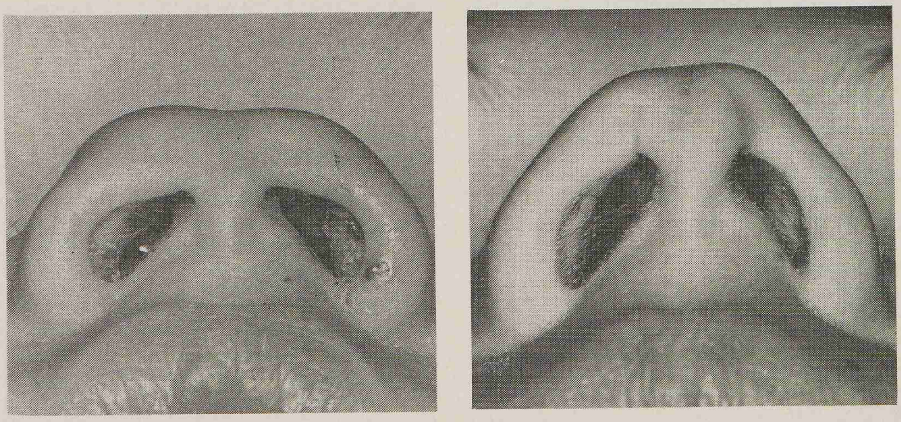


Figure 3c. Same case base view before (c) and after (d) surgery.

bifid noses are invariably associated with central midface deformities. Depending on the degree of severity true or pseudohypertelorism may be an additional finding.
In the past 15 years we have seen 12 pertinent cases, of which 3 had associated true hypertelorism.

DIAGNOSIS

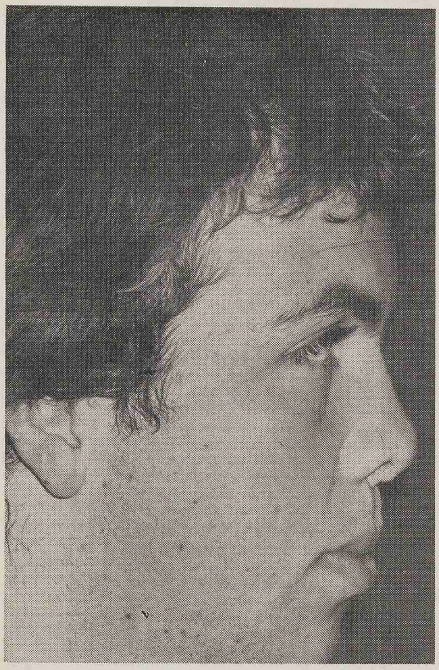
Normally the diagnosis does not present any problems. But one of our patients, a 17-year-old male, with severe bifid nose had undergone submucous septal resection for obstructed nasal breathing, while he was a child. This necessitated reconstruction of both the cartilaginous and the bony nose in terms of rhinoplasty.

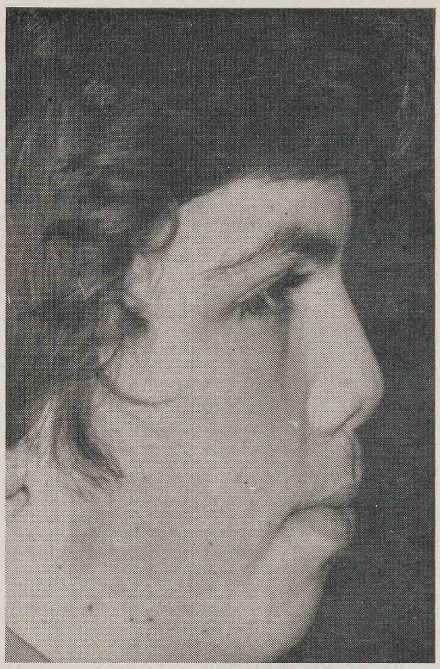
TIMING OF SURGERY

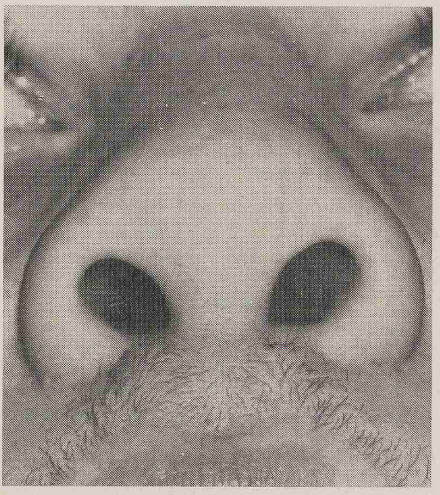
Bifid noses are readily amenable to corrective surgery during childhood. In none of the patients we followed up did we find any conspicuous growth anomalies of the nasal bone to occur postoperatively.

SURGICAL TECHNIQUE

In bifid noses of minor severity with or without alar deformities surgery can be done by using the decortication technique. When extending the incisions up to the root of the nose (Walter, 1973), excellent vision is obtained. Dissection is strictly extramucous. The double arch formed by the septodorsal cartilage is exposed and scarified for medial displacement. Often the bony pyramide is not fully established. Timing of the nose for correcting nasal width by lateral and transversal osteotomies requires prior resection of the







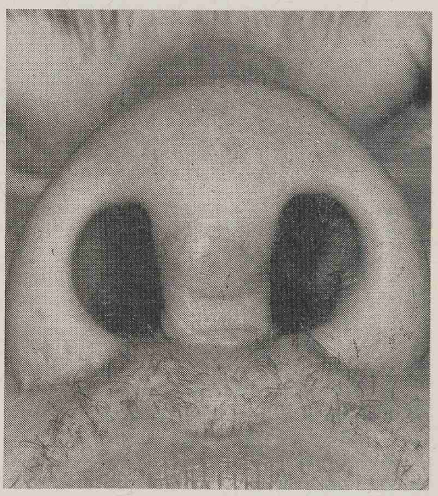


Figure 4a. 18 years old male patient with a congenital dysplasia of the cartilaginous vault and absence of the nasal spine with short columella.

Figure 4b. Same case 4 years later after lengthening of the columella by using composite graft and a preserved cartilaginous strut.

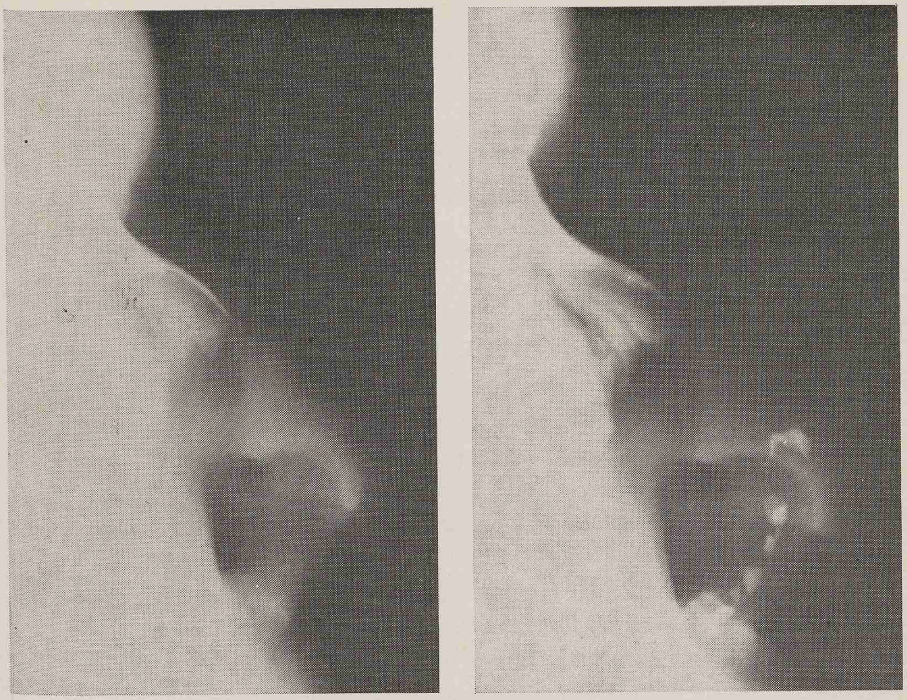


Figure 4c and d. Same case. X-ray control immediatly after implantation of the cartilage (c) and 4 years later (d). The implant shows a good calcification which ensures an adequate projection of the nasal tip.

frontal spine. Wiring the lateral bony walls after mobilization with full-thickness mattrass sutures proved to give satisfactory results in terms of substantially reducing the pseudotelorism.

Severe bifid noses with a wide root are best corrected with a direct approach through external incision (Converse; Denecke and Meyer, 1964; Webster and Deming, 1950). We prefer a curved incision from the root to the tip of the nose, leaving a barely visible scar on the nasal dorsum (Figures 3a, b, c and d). If pronounced hypertelorism is present, satisfactory results can only be obtained by craniofacial orbital osteotomies (Edgerton at al., 1964).

CONGENITAL FLAT NOSE

Congenital flat nose is characterized by predominantly cartilaginous nasal dysplasia. The anterior nasal spine is, however, consistently absent in these cases so that the projection of the cartilaginous nose is abnormal. The nasal base is typically flat and the columella short. The cartilaginous nasal skeleton is dysplastic and the septum poorly developed.

Meyer and Fleming (1969) reported 7 pertinent cases in 1969. Our experi-

ence with congenital flat nose extends to 8 cases operated in the past 10 years. If the anomaly is diagnosed during childhood, the procedure to be adopted is analogous to that in septal abscesses: implantation of a septal chip will elevate the cartilaginous dorsum thus stimulating soft tissues and cartilage growth. Once growth is completed, the nose is reconstructed by rhinoplasty. In most cases, however, the columella will need lengthening. This is done either by local flaps from the upper lip or by composite grafting. (Figures 4a and b).

ZUSAMMENFASSUNG

Diagnose und Therapie der Nasencysten und -fisteln werden anhand von 20 beobachteten Fällen besprochen. Über die Ausdehnung der Cysten läßt sich häufig keine sichere Aussage machen. Die Cysten und -fisteln der Nase sollten möglichst frühzeitig operiert werden, um das Wachstum der Nase nicht zu beeinträchtigen. Bei Vorliegen der Sekundärfistel können Nachoperationen besonders schwierig sein.

Die mediane Spaltnase zeigt einen sehr verschiedenen graduellen Unterschied der Mißbildung. Je nach Schweregrad kann diese Mißbildung mit Hilfe der Dekortikation oder gebogener äußerer Schnittführung korrigiert werden. Der vorliegende Sattel läßt sich mit konserviertem Knorpel aufbauen. Nachkontrollen zeigten ein befriedigendes Resultat.

Die congenitale Flachnase ist eine seltene Mißbildung, die in den letzten 10 Jahren in 8 Fällen beobachtet wurde. Die Technik der Verlängerung der Columella der Abstützung der Nasenspitze wird erläutert.

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