Pathology and Treatment of Nasoschizis

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A craniofacial malformation is caused by a developmental arrest that results in focal fetal dysplasia. The severity, location, and number of these dysplasias make a wide spectrum of abnormalities possible. In this spectrum nasal dysplasia and, more particularly, the malformations characterized by a cleft in the lateral part of the nose play a prominent role, but in past years they have received only scant attention. In Tessier’s classification this nasoschisis is the Number 1 cleft. We present here our observation in 2 patients with nasoschisis and the procedure used to correct it. Both patients were operated on at the Rambam Medical Center in Haifa, Israel.

The term “nasoschisis” relates to clefts of the lateral part of the nose. The severity of these defects ranges from a simple notch in the alar rim to a defect or cleft involving part or all of the nasal half. Clefts of this nature should be distinguished from a small group of malformations characterized by dislocation and dysplasia of the alar base. In Tessier’s classification [17], these malformations are referred to as “Number 2 clefts,” though Tessier himself is not convinced they form a distinct entity and submits that they represent a transitional form between his Number 1 and his Number 3 cleft [3]. Mazzola [8] named these malformations “unilateral nasal hypoplasias.”

In the patients with nasoschisis seen by the first author the nasal defect was commonly associated with a “widow’s peak.” Monolateral as well as bilateral cases occur and combinations with other manifestations of focal fetal dysplasia, such as defects in or between the frontal bones [3], teleorbitism and unilateral microphthalmia [14], bilateral microphthalmia [1], cleft lip [6], or even cleft lip and microphthalmia [8, 16], have been observed. Combinations of severe monolateral nasoschisis and deficiencies in growth of the nasal septum (internasal dysplasia) have been reported by Kindler [5], Rosasco et al [15], and Van der Meulen [19]. An example of a combination of severe bilateral nasoschisis and internasal dysplasia was recently observed by Das Gupta [2]. In his patient, as in many other patients with nasoschisis, the malformation was associated with choanal atresia. Patients with a combination of nasoschisis (monolateral as well as bilateral), internasal dysplasia, and a medium cleft lip have been described by De Meyer et al [11], Millard et al [12], and Iregbulum [4].

Pathomorphogenesis

Early development of the nose should be considered as that of two organs. Each nasal placode is transformed by an outgrowth of three facial swellings, the maxillary process, the lateronasal process, and the medianonasal process via the nasal groove into the nasal tube. Both medianonasal processes develop in the area between the nasal placodes—the interplacodal area (20, 21). They are separated by the internasal groove, which gradually disappears by outgrowth of the septum in a frontocaudal direction.

If the nasal septum does not grow, the internasal groove will persist and the two nasal halves will re-
main separated. Internasal dysplasia (medium cleft nose, or bифid nose) will be the result [21].

Nasoschizis, on the other hand, is produced when outgrowth of the medianal and lateronasal processes is disturbed and, consequently, a malformation of one nasal half is produced. The combination of a nasal defect and a widow’s peak can be explained by the discrepancy in growth that results when development is arrested in one area while it continues normally in the surrounding tissues. An hourglass deformity will be produced with the transition in the middle as the original area of focal fetal dysplasia and the defects at both ends, in this particular malformation the nasal defect and a widow’s peak representing the areas that have not developed as a result of the arrest. The direction of the hourglass deformity will correspond with the direction of growth [20].

Case Reports

Patient 1

After the birth of our first patient, a 1-year-old girl, neighbors advised the parents to kill the girl because she looked like a pig—which indeed she did.

She was born with a protruding mass of soft tissue on the right forehead, a widow’s peak, extreme hypertelorism, complete unilateral nasoschizis (Fig 1), a wide right nasal cavity with a pulsating mucosa-lined roof, a nostril at the medial canthal level, minor internasal dysplasia, and a median cleft lip. Motor and psychological development was normal at 1 year.

Computed tomography revealed a large frontal bone defect (Fig 2A) extending into the floor of the anterior cranial fossa over the nasal cavity, an interorbital distance of 4 cm (Fig 2B), a large lipoma above the corpus callosum (Fig 2C), and wide brain ventricles. Ophthalmic examination showed alternating exotropia and free movements of both eyes. Fundus examination was normal, apart from a posterior staphyloma on the left eye.

An attempt was made to correct as many of the described malformations as possible. Surgical exposure was through coronal and lower palpebral incisions. The medial canthal tendons were not detached. After dissection of the borders of the frontal bone defect, the floor of the anterior cranial fossa was exposed by the neurosurgeons. A forehead flap was then raised from the area overlying the frontal mass with the intention of using it for reconstruction of the right nasal half (Fig 3). The mucosa lining
Fig. 2. Computerized tomography scan of Patient 1 showing (A) large frontal bone defect; (B) a 4-cm hypertelorism; and (C) large lipoma above the corpus callosum.

Fig. 3. Intraoperative photograph (Patient 1) and diagram of the raised forehead flap.
the roof of the right nasal half was dissected from the dura and moved interiorly with the same purpose. A solid undifferentiated mass of cartilaginous tissue was removed from the glabellar area and classic four-wall osteotomies [18] then made it possible to close most of the defects in the frontal bone and to reduce the interorbital distance to 2.0 cm. The midline osteosynthesis was performed under some tension, due to residual soft tissue traction which could not be released. The median cleft in the lip and the right half of the nose were finally reconstructed in the manner shown in Figure 4. Postoperatively, the child made an uneventful recovery. Her appearance after this operation is shown in Figure 5.

A year later the child was reoperated upon for correction of the canthal drift and further modeling of the nose. Extracranial exposure showed excessive fibroosseous thickening of the right medial canthal region. The canthal tendon was detached. The nasal bridge was narrowed, producing an interorbital distance of 2.0 cm and a canthopexy was performed. The surplus of skin in the glabellar area and over the left half of the nose was removed and the contour of the right nose was improved.

The child's appearance nine months after the second operation is shown in Figure 6. Some canthal drift has recurred on the right side but the mother of the child is satisfied and not interested in corrective surgery.

Fig 4. The various stages in nasal reconstruction in Patient 1. (A) Preoperative appearance of patient. (B) Intraoperative marking of surgical incisions.
Fig 4 (Continued). (C) Enlargement illustrating three flaps, a, b, and c, which were designed for reconstruction of the right nostril. (D) Raised flaps a and b, as well as elevation and approximation of nasal lining. (E) Partial overlapping of flap a over flap b. (F) The relationship of the three flaps, a, b, and c, to each other and their respective contribution to the nasal reconstruction.
Fig 5. Patient 1, two weeks postoperatively.
Patient 2

The patient, 1 year old at the time of consultation, had been born with a first-degree hypertelorism, severe right-sided nasoschisis, a right upper eyelid coloboma, internasal dysplasia, a double cleft lip, and complete cleft palate (Fig 7), a combination of malformations which, to our knowledge, has never been described before.

Functional development was normal and computed tomography revealed no other craniofacial anomalies.

The double cleft lip was closed first (Fig 8). Planning of the nasal reconstruction six months later was complicated by the absence of septum and columella. However, the following procedures were performed (Figs 9–12): the right nostril defect was reconstructed in a manner similar to the one used in our first patient. A forked flap was raised in the lip using a modification of the procedure originally described by Marcks et al [7]. The septum was reconstructed with a thin triangular bone graft, and the columella was made by suturing the flaps as in the form of a Z-plasty.

The present state of the patient is shown in Figure 13. Further correction of the nose and scars of the lip is being considered.
Fig 9. (A) Patient 2. Outline of flaps a, b, and c, designed to restore the right side of nose. Flaps d and e were used for columella reconstruction. (B) Enlargement of flaps a, b, c, and d.
Fig 10. Operative procedure in Patient 2. (A) Elevation of flaps a, b, and c. (B) Elevation of flaps d and e prior to insertion of bone graft beneath them. (C) Tailoring and suturing of flaps.
Fig 11. (A) Patient 2, front view, showing bone graft in position. (B) Side view.

Fig 12. Patient 2 postoperatively.
Fig 13. Patient 2, one year postoperatively.
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