

Pediatric Facial Plastic and Reconstructive Surgery

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Chapter 3: Nasal Surgery for Congenital and Acquired Disease

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The topic of nasal surgery in children has been debated for decades. As in all other aspects of pediatrics, it is imperative that we first do no harm while attempting to correct debilitating conditions. The controversy revolves around whether or not surgical manipulation of the nose will influence it and surrounding structures in a manner that would be aesthetically or functionally beneficial. It must be emphasized that surgery should only be performed after intensive medical therapy has failed to relieve severe obstruction, persistent rhinorrhea, recurrent sinusitis, or upper respiratory illness. Obviously, immediate procedures need to be performed for septal hematoma or abscess or traumatic or congenital deformity causing airway compromise.

Surgeons interested in performing nasal surgery for acquired or congenital disease need to understand the development of the nose and the many surgical options available. Today, innovative instrumentation and techniques allow rapid, accurate diagnosis and give surgeons the opportunity to correct debilitating problems without unreasonable disrupting future aesthetic or function.

Development

Development of the nose begins at approximately 4 weeks of fetal life. At this time the neural tube is closing and neural crest cells begin their migration. In the head and neck neural crest cells form the connective tissues of the face. They are induced by closely associated tissues to differentiate into various types of connective tissue.

Collections of these neural crest cells undergo proliferation, forming the nasal placodes. Other collections proliferate in areas that become the medial and lateral nasal swellings and the maxillary swelling. The medial and lateral swelling soon grow together, forming the two nasal pits, the floor of which is the oronasal membrane. This soon ruptures and then is closed by fusion of the medial portions of the maxillary swellings, thus forming the hard palate. The nasal pits, meanwhile, continue to burrow deep in the mesenchyme until only the nasobuccal membrane remains. This soon ruptures and by week 10, the nose communicates with the choana.

While this is occurring, the cartilaginous framework of the nose is developing. Three paired condensations of mesenchyme in the medial and lateral nasal swellings initially are formed: the trabecular cranii, in the primitive nasalseptum; the tectal condensations, located dorsally around the primitive nares; and the paranasal condensation, located further posteriorly in the lateral nasal swellings. This cartilage model will ossify over the next 4 weeks, becoming the posterior septum, ethmoid complex, and nasal bone.

Postnatal growth and development is predictable and sporadic. The infantile nose is proportionally smaller and more broad with visible nares. Growth is very rapid initially and then slows in early childhood. Another burts occurs during the pubertal growth spurt, after which activity declines. Many factors influence postnatal development of the nose such as occlusion of the teeth, tongue placement, facial musculature, and development of the sinuses. Whether or not the nasal septum is the main determinant of postnatal development is debatable.

Effects of Surgical and Nonsurgical Management on Subsequent Development

The goal of nasal surgery in the pediatric patient is to restore function without deleteriously altering development. It seems logical then to avoid areas of the septum that are "growth centers" and to avoid resection of any tissue if possible. Vetter and his group helped to delineate the metabolically active area of the septum by analyzing small cartilaginous strips obtained at septoplasty from children and adults. They incubated these strips in ³⁵S-labeled NaSO₄, and measured the uptake of this material and showed age-related differences in metabolic activity. In prepuberty the suprapremaxillary area and anterior septum showed the greatest activity, whereas in pubertal children and adults only the anterior end of the septum showed high activity. The conclusion from this study was that these areas appear important metabolically and should be avoided during surgery.

Verwoerd and Verwoerd-Verhoef used a rabbit model to investigate the effects of future development after surgery on various parts of the septum. They found that elevation of the mucoperichondrium unilaterally or bilaterally had no effect on future development and that a small (1 mm) submucous resection resulted in no foreshortening of the nose or maxilla. These rabbits did develop a septal deviation at the point of resection. However, removing a basal strip of the septum resulted in foreshortening of the nose and midface.

Verwoerd and Verwoerd-Verhoef have reviewed several other studies using models that have shown other areas of the septum, along with the premaxilla and the anterior septum, to be crucial in development, including (a) the upper lateral cartilages, whose integrity ensures straight outgrowth of the nose; and (b) the integrity of the T-bar configuration of the septal and upper lateral cartilages. Trauma or disruption of this unit affects the total development of the dorsoseptal cartilage, which in turn causes maldevelopment of the nasal bones and vomer.

The potential effects of large resections as discussed above are discouraging. However, expectant management of a pediatric patient with nasal obstruction may be debilitating also. The dental literature is replete with reports that describe facial maldevelopment attributed to "mouth breathing". Bresolin et al examined 30 allergic children who appeared to breathe through the mouth and compared them to 15 nonallergic children who appeared to breathe predominantly through the nose. All were subject to intraoral and cephalometric analysis. They found that the allergic children had longer faces with narrower maxillae and retruded jaws. However, they readily admit that they had not objectively determined that their patients indeed were breathing through the suspected route. A Canadian group performed a careful study and showed that there was no difference in nasal airflow between "mouth breathers" and normal children. This obscurity then shifts the debate to a question of whether the abnormal anatomy causes the appearance of mouth breathing or is caused by mouth breathing. This

discussion is presented because there are a large number of professionals who believe nasal obstruction and mouth breathing adversely affect development.

A question then arises, do deformities that cause obstruction, specifically, septal deformities, correct themselves over time? Gray examined the septa of 145 babies at birth and at 8 years. He found that all untreated deviations were present or had increased in severity during subsequent development. He correlated this with a high proportion of otitis media. Only surgically corrected septa were straight at follow-up.

This evidence supports surgical manipulation when clinically indicated. The question then becomes, Can surgery be performed with acceptable aesthetic and functional results? Ortiz-Monasterio and Olmedo presented a series of 44 rhinoplasties, which included septoplasties, all done on patients between 8 and 14 years of age. Follow-up consisted of all patients being seen at least 5 years later with results comparable to adult patients. Pirsig presents his more than 15 years of conservative rhinoplasty experience showing poor to excellent results in children with traumatic nasal injuries. Verwoerd et al, despite showing deleterious results of resections of specific portions of the septum, also mention that their findings do not contraindicate a conservative procedure that avoids critical areas of the septum. Healy presents ten children, all with severe obstructing deviations, who had their obstructions relieved via a sublabial route with no disturbance in facial development 10 to 60 months later. Jugo's total septal reconstructive technique also provides good evidence that septal surgery is safe in the younger patient.

Optimal Timing of Nasal Surgery in Children

Wide experience, as demonstrated above, has not shown hard evidence to absolutely contraindicate nasal surgery in the younger patient if performed in a conservative manner for appropriate indications. Obviously, in a child with septal hematoma or septal abscess, immediate drainage needs to be performed. A newborn with a dramatically deviated nose may need immediate correction; however, most correct spontaneously. A young child with symptomatic obstruction should have surgery within the guidelines listed below. It may be prudent to perform surgery to relieve obstructing lesions before the pubertal growth spurt, if indeed nasal obstruction causes midface maldevelopment. If a nonobstructing lesion is problematic, surgery should probably be delayed until after the nose is mature. Buck and Brown, in a longitudinal study of nasal growth of children age 6 to 18, showed that 87% of nasal growth is obtained by age 12 in girls and age 15 in boys. Thus, if surgery can be delayed until after the midteens, it should. However, Ortiz-Monasterio and Olmedo reported several children of pubertal age who had rhinoplasty without deleterious results.

Psychological Aspects

Surgery in the pediatric population is universally stressful. Parents do not wish to see their children in discomfort and are often distraught themselves when dealing with an operative situation. Children often have great anxiety about parental separation, general anesthesia, postoperative pain, and other aspects of surgery. But at the same time, children, particularly adolescents, may suffer psychological stress over the unacceptable appearance of the nose. The positive benefits and negative factors must be considered on an individual basis. Many reluctant children in whom corrective surgery would be clearly beneficial will often, over a

period of time, accept surgery with proper explanation and guidance on the part of the physician, parents, and others involved. The procedure should be performed for precise clinical indications with special emphasis on restoring function and reasonably relieving aesthetic deformities that can be emotionally devastating. Proper preoperative preparation of the parents as well as the child is crucial. Every effort should be made to allow the child as much control as possible without being detrimental to his/her well-being. A perioperative team trained to work with children in circumstances that all find comfortable is most beneficial. When the above considerations are given proper attention, the surgery will often be a surprisingly smooth experience for the child. If there is any question on the part of the surgeon or any member of the team, including the parents, professional psychological evaluation and treatment, if necessary, should precede surgery.

Technical Challenges

The technical challenges are (a) the small operative area, (b) the delicacy of the tissue and the need to always be as conservative as possible, (c) avoiding "growth centers," (d) gaining adequate exposure, (e) aesthetic acceptability, and (f) the underlying pathology causing the deformity. Jugo and Healy use open procedures to perform septoplasty and have obtained excellent results. The new pediatric, rigid nasal endoscopes may dramatically affect surgery by making diagnosis more precise and will allow superior visualization intranasally. More experience with the endoscopic techniques may make open procedures less desirable.

Congenital Disorders

The nose is deformed in a multitude of genetic disorders, many of which involve the orbit, midface, palate, or cranial vault. There are also tumors such as dermoids, gliomas, and encephaloceles that present as nasal deformities. These will be covered in other chapters in this book.

Choanal Atresia

The newborn with bilateral choanal atresia is usually brought to the otolaryngologist's attention immediately after birth when the patient develops respiratory distress. After obtaining an adequate oral airway the diagnosis can be made by attempting to pass a suction catheter through the nostrils (Table 1A and B). If time and conditions permit, a 2.7 mm rigid endoscope can be used to directly visualize the atretic plate after properly decongesting the nasal mucus membranes. Computed tomography (CT) scanning gives the definitive diagnosis. However, its primary use is to identify the composition of the atretic plate prior to surgery.

Table 1A. *Choanal atresia*

- 1 to 7.000 live births
- 2 females to 1 male
- 2 unilaterals to bilateral
- 90 percent bony/10 percent membranous
- 50 percent associated with other congenital anomalies

The clinicians must keep in mind that many patients (20% to 50%) have associated anomalies, ie, the CHARGE association (coloboma, heart defects, atresia, retarded growth and central nervous system (CNS) abnormalities, genital hypoplasia, eardrum deformities). Choanal atresia occurs more often in females (2:1), most often unilaterally, and in 90% the atretic plate is bony.

Table 1B. *Choanal atresia - Diagnosis*

Bilateral - Birth - Respiratory distress relieved by oral airway

Unilateral - Later - Unilateral rhinorrhea

- CAT Scan Finding:
1. Narrow nasal cavities
 2. Lateral bony obstruction
 3. Medial (vomer) bony obstruction
 4. Membranous obstruction
 5. Bony choanal plate.

The definitive cause for choanal atresia is unknown. There are four basic theories: (a) persistence of the buccopharyngeal membrane, (b) persistence of the nasobuccal membrane of Hochstetter, (c) the abnormal persistence or location of mesoderm, or (d) abnormal neural crest migration.

Table 2A. *Choanal atresia - Treatment*

Bilateral - Early

- Transnasal:
1. Blind puncture
 2. Visually assisted - microscope and/or endoscope

Transpalatal - More widely used

Unilateral - Elective

The treatment is surgical (Table 2A and B). Various methods have been employed: tanspalatal, transseptal, and transnasal puncture. Richardson and Osguthorpe found that 83% of transpalatal procedures were successful and Ferguson and Neel also reported excellent results in a small number of patients. We prefer to use the brilliant illumination of the rigid fiberoptic endoscope to perform this procedure intranasally in those cases in which the stenosis is membranous or when the bony plate is not thick and when the nasopharynx does not have a lateral bony constriction. In the older child, the endoscopic approach is also preferable as a larger size of structures will allow this surgery to be more easily performed. However, in the infant with bony atresia who does not respond to medical management, the transpalatal approach is quite appropriate. In using the transpalatal approach, we prefer the inverted horseshoe approach rather than midline splitting as we feel this is functionally superior and has fewer complications.

Table 2B. *Choanal atresia - Key Surgical Aspects*

1. Conserve periosteum and mucosa
2. In addition to atresial plate, remove posterior septum and lateral bony buttress.

Nasal Duplication

Fortunately, this entity is exceedingly rare (only three reported cases) as the obvious deformity; if uncorrected, it would be psychologically devastating. The patients present with two nearly complete external noses and hypertelorism with a nonfunctional nasal airway.

The origin of this deformity is unknown. The abnormality obviously arises before the nasal placodes develop. Interestingly, there is a report of "tripe nostrils" in which the author also postulates that an extra nasal placode is theoretically possible. However, a recognizable nose or pseudonose structure does not develop.

Treatment is initiated early for functional and cosmetic reasons. The first goal is to provide an adequate nasal airway using procedures to perforate the atretic plates. The second portion of the procedure is to remove the medial portion of each "nose" and one septum is removed. The remaining lateral portions are approximated to give a more normal appearance.

Proboscis Lateralis

This is another rare deformity that presents as hypertelorism with a nose-like structure located at the inner canthus and oriented in a plane perpendicular to the normal nose. Midfacial and nasal development may be affected. The embryology is also unknown but probably involves a similar mechanism of abnormal nasal placode development. Whether Hengerer and Oas' theory of abnormal neural crest migration, as they mentioned in their theory of choanal atresia, is involved in some or all of these nasal deformities, has not been proven.

Treatment is surgical, first to ensure an adequate nasal airway, and second to establish a more acceptable countenance.

"Cyrano de Bergerac" Hemangiomas

This deformity is caused by a capillary or capillary-cavernous hemangioma primarily involving the nasal tip, giving rise to the "Cyrano" appearance. Multiple modalities have been used to treat hemangiomas of the head and neck, such as excision, steroids, and expectant treatment. Spectacular results due to spontaneous involution and without active medical management frequently occur.

This conservative mode of thinking may be the best choice for the Cyrano lesions also. Thompspon and Lanigan retrospectively reviewed their experience and found that although at time difficult, the conservative treatment, at least initially, is probably the best. In selected patients, surgical treatment may be the more conservative approach. For example, with persistence and growth, there are selected cases in which the appearance is psychologically disturbing or in which bleeding is a significant problem. Although carbon dioxide freezing and laser surgical excision have been found to be appealing, in our experience the straightforward surgical excision utilizing a modified external rhinoplasty approach is quite successful. Bleeding, surprisingly, is easily controlled by the usual methods.

Birth Trauma

Deformities of the nose, independent of the genetic or embryologic effects, are commonly caused by trauma in utero or during delivery. It is common to see nasal flattening at birth; however, this normally corrects itself. Closed digital manipulation or selected surgical instrumentation of severely displaced structures is appropriate. However, open approaches are seldom, if ever, indicated. It is surprising that, even in those infants in whom the external nose appears markedly displaced, self-correction is usually the rule. Jazbi studied 100 consecutive cases from 7,129 live births and found a frequency of occurrence of less than 1%. The deformities were generally caused by septal dislocation. In older children whose deviations were not corrected at birth, septoplasty may need to be performed by any of the various acceptable approaches, considering each patient and the deformity on an individual basis. Healy has obtained excellent exposure with minimal morbidity using a sublial approach.

Septoplasty

If the projection of the nose and midfacial development in general is dependent upon the nasal septum, then it is logical to strive to attain an acceptable nasal airway prior to the "point of no return". As previously mentioned, Gray showed that watchful waiting did not allow septal deviations to correct themselves. If a patient has an obstruction, then a minimal, closed procedure is our usual approach. The sublial approach of Healy certainly seems reasonable. We have not yet used the total septal reconstruction approach of Jugo, though his experience would indicate that this is a reasonable approach.

Septorhinoplasty in Older Children and Adolescents

In children whose deformity will not be sufficiently corrected by septoplasty alone, consideration of reconstructive septorhinoplasty is appropriate. These are primarily children in whom the nasal pyramid as well as the septum are deviated and there is significant nasal obstruction. There has been no firm evidence to show that simple osteotomies to reposition the bony pyramid has resulted in growth deformities. On the other hand, the temptation to perform complete reconstructive septorhinoplasty, including resection of the dorsal tissue, should usually be resisted. However, dorsal augmentation should not be unduly delayed. It has been our philosophy to correct severe deviations by repositioning and augmentation at an age when these problems become physiologically or psychologically important, with the full understanding that further correction may be desired after full growth has been obtained.

Cosmetic septorhinoplasty in older adolescents is quite rewarding; however, the psychological aspects demand careful consideration. In our experience involving both adults and adolescents, there are a greater proportion of psychological pitfalls in the adolescents; however, these are usually more easily managed than when present in the adult. To manage them, they must first be identified and if the surgeon is not fully capable in dealing with these problems, psychological consultation should be obtained. However, this has been surprisingly infrequent. The surgeon who is comfortable in dealing with adolescents will find the adolescent among his most satisfied and satisfying patients.

Proper, informed consent is especially important in these patients, with specific attention to the possible effects on future growth. The younger the adolescent, the more likely there will be further growth changes, changing the appearance over time. In addition to the age guidelines given by Buck and Brown concerning growth, each patient must be considered on an individual basis with regard to size and with reference to parents and siblings, with regard to whether the patient has completed pubertal growth, and particularly, with regard to whether there had been growth in the last 6 months.

Even when all these factors are considered, there may be further growth. A slight alteration in structure will not significantly affect the appearance; however, if there is significant change, it will usually be as a result of increased dorsal projection, which can later be managed with minor revisional surgery. There is increasing evidence that careful conservative septorhinoplasty in younger patients can be safely performed. Fedor has recently reported satisfactory results in 22 patients where conservative surgical guidelines were carefully followed.

The guiding surgical principle of septorhinoplasty in children is to never overoperate - *less is better*. Attempts are made to achieve the desired result by repositioning tissues whenever possible. Incising, reshaping, and resecting of tissue is only done as a last resort with resection of tissue kept to an absolute minimum. It should be remembered that it is much easier to later correct errors of omission than those of commission.

Table of Guidelines

1. Do as little and as few surgical maneuvers as possible to achieve satisfactory results. Repositioning is preferable to reshaping and incising. Reshaping is preferable to resecting.

2. Use only lateral osteotomies and avoid medial osteotomies unless absolutely necessary. Unless absolutely necessary, do not separate the upper lateral cartilage from the septum.

3. Use sharp dissection to modify the cartilaginous profile. The bony profile in the adolescent can be generally remodified with a rasp, as the tissue is immature and responds readily to rasping. This aids in avoiding overresection.

4. Do not do extensive tip surgery in the younger patient, and in the adolescent use the most conservative tip modification approach to produce the desired result. Particularly avoid overresection of the lateral crus.

5. Augmentation measures are generally satisfactory in the adolescent.

Nasal Trauma

All children sustaining significant trauma to the nose should receive a thorough, detailed examination, whether treatment is apparently indicated or not. (For a discussion of soft tissue injuries, see the chapter by Farrior and Clark.) Of particular importance, it should be noted that soft tissue defects should have early repair, and that scars of the nose in children

and adolescents are erythematous for an extended period of time before "fading". Nasal fractures are a common event throughout childhood. Trauma in this area may influence further development of the premaxillary and maxillary septal and nasal bony elements. Unfortunately, on occasion, even after accurate diagnosis and adequate treatment, further growth and development may be affected. An important anatomical feature to remember in assessing and treating nasal injuries is that the smaller the child, the larger the cartilaginous proportion of the external pyramid. The nasal bones are formed on the surface of the cartilaginous nasal capsule and are initially separated in the midline by an open suture. There is a large overlap of the nasal bones on the upper lateral cartilages. If these are detached, upper lateral cartilage can be prolapsed into the nasal cavity, creating an obstruction. Fractures of the septal cartilage may occur with or without nasal, bony fractures. An untreated greenstick fracture of the septum may result in progressive deformity.

Septal hematoma occurs more frequently in children due to the loose attachment and mobility of various anatomical components. If untreated, this frequently leads to septal abscess with cartilage resorption and subsequent "saddle nose" deformity. The hematoma should be evacuated as early as possible and the septal components sutured together with a mattress type suture, following which the nose is packed. If an abscess has occurred, immediate evacuation with thorough irrigation should be undertaken. Close inspection of the cartilage for evidence of absorption should be done at the time of surgery. A drain is left in place, the nose is packed, and appropriate antibiotics are administered for at least 2 weeks. If the cartilage has absorbed, the patient must then be observed closely after healing and should dorsal depression become evident, early grafting (usually with conchal cartilage) will prevent progressive "saddling".

Nasal Obstruction Due to Other Causes

Nasal obstruction is not always secondary to septal deformities; consequently, an accurate etiologic diagnosis is required. Allergy and other metabolic conditions such as cystic fibrosis require proper management. Other causes that may require surgical management include nasal polyposis, chronic hypertrophic sinusitis, turbinate hypertrophy (especially pneumatization of the middle turbinate, ie, concha bullosa). It is our practice to include a coronal computed tomography (CT) scan as part of the diagnostic evaluation in all children with airway obstruction not obviously due to septal nasal deformity or obvious medical causes. Many of these patients will have an excellent response to functional sinus surgery and/or turbinoplasty. (The details of evaluation and surgery are beyond the scope of this chapter.)