Congenital and Acquired Deformities of the Nose

Fernando D. Burstein • Bruce S. Bauer

As the central and prominent feature of the face, the nose commands front billing, and even minor nasal abnormalities and discrepancies draw immediate attention. There has always been an inherent desire in people to look like their fellow humans and not appear peculiar or different.¹ The complex sequence of events that shapes the frontonasal structures and functional airway is vulnerable to atypical cellular processes and deformational forces that may cause minor abnormalities in nasal shape at one end of the spectrum, or near-total absence of nasal formation at the other end. Once the normal nose has formed, its prominent position makes it subject to the forces of trauma, particularly in healthy, active children. Regardless of whether the deformities result from abnormal developmental processes or an errant bat during Little League, there is a need either to create a more normal appearance or restore the structure so pediatric patients can interact comfortably and successfully with their peers. Although other chapters address nasal deformity resulting from cleft lip and palate, the range of nasal clefts in atypical facial clefts (see Chapter 26), and primary rhinoplasty for adolescents with indications and techniques (see Chapter 34), this chapter reviews diagnosis and treatment of some of the more common conditions that may affect the development and/or appearance of the nasal structures. These conditions include nasal hypoplasia, atypical nasal clefts, congenital nasal masses, vascular lesions, cutaneous lesions, and traumatic nasal deformities.

NOSE EMBRYOLOGY

During the fourth week of gestation, neural crest cells migrate to the developing face from the lower forebrain, the midbrain, and rhombomeres 1 and 2 of the upper hindbrain. These migratory neural crest cells are the predominant source of facial connective tissue, including cartilage,
bone, and ligaments. Because neural crest cells migrate to the face as cohorts of cells from different portions of the brain, they carry with them different developmental programs. Mutations arising in the premigratory or early migratory neural crest cells may affect a specific clone of cells, which then carries that mutation to a predestined site in the face.²-⁶

At week 4, five identifiable primordia surround the stomodeum (Fig. 33-1). The single, unpaired frontonasal prominence lies in the midline just superior to the stomodeum.² By the end of the fourth week, even before the neural folds close, paired thickenings of ectoderm appear on the surface of the frontonasal prominence just superolateral to the stomodeum. These oval nasal placodes, located at the 1 o'clock and 11 o'clock positions, give rise to the future nose and nasal cavities. Development of the nasal placodes (and the lens placodes) requires the paired box gene Pax 6. In the absence of Pax 6, neither the nasal placodes nor the lens placode develop.² During the fifth week of gestation, mesenchyme in the margins of the nasal placodes proliferates to form horseshoe-shaped elevations—the nasomedial and nasolateral processes. The nasomedial processes are longer than the nasolateral processes.³ The tissue surrounding the placodes thickens and elevates so the nasal placodes appear recessed within depressions in the surrounding tissue, which are designated the nasal pits. The nasal pits are the primordia of the anterior nares (the future nostrils) and the nasal cavities.³

From the fifth week of gestation, the nasal pits gradually deepen toward the oral cavity, forming substantial depressions. By 6.5 weeks' gestation only a thin oronasal membrane separates the oral cavity from the nasal cavities. This oronasal membrane then breaks down so that the oral cavity communicates with the nasal cavity through openings posterior to the primary palate.² These openings are designated the nasal choanae. Fusion of the two palatal shelves lengthens the nasal cavity and carries the communication posteriorly to the upper pharynx. The nasal septum grows down from the frontonasal prominence to the level of the palatal shelves when the shelves fuse to form the definitive secondary palate. The fusion of the secondary palate with the secondary palate is marked by the incisive foramen.⁷

From the sixth to the eighth week of gestation, the cheeks and corners of the mouth form by the maxillary and mandibular processes merging. The upper lip is completed in the latter part of this period.² Simultaneously the expanding nasomedial processes merge with the superficial region of the maxillary processes on both sides along epithelial seams called the nasal fins.² Mesenchyme penetrates the nasal fins and forms continuity between the nasomedial and maxillary processes. The fusion of the two nasomedial processes displaces the frontonasal prominence posteriorly. Therefore the frontonasal prominence does not contribute significantly to the definitive upper lip, jaw, or nasal tip, despite the fact that it formed a prominent portion of the stomodeal border earlier. The fusion of the paired nasomedial processes forms the nasal tip, the crest of the nose, and a portion of the nasal septum.²

Facial and limb malformations are known to result from deficient or excess molecular signaling (sonic hedgehog, fibroblast growth factor, and retinoic acid signaling among others). Similar phenotypes, such as clefting, may result from either deficiency of the appropriate midline tissue or such excess of other midline tissue that the appropriate processes cannot meet to fuse. In experimental animals, reduced retinoic acid signaling diminishes the expression of sonic hedgehog and fibroblast growth factor 2 in the mesenchyme, increases apoptosis locally, and decreases proliferation of tissue in the forebrain and frontonasal process. These animals show holoprosencephalic phenotypes with hypoplastic forebrains, fused eyes, and absence of structures derived from the frontonasal process.² ⁴⁻⁷⁻⁸ Conversely, excess sonic hedgehog stimulates frontonasal growth and widens the frontonasal process (an average of 48%), resulting in cleft palate, and more severe phenotypes show ectopic midfacial structures with duplication of the nasal bones²⁻⁹ (Fig. 33-2).
Fig. 33-1 Embryogenesis of the face from 4 to 10 weeks' gestation demonstrates the contribution of the frontonasal process and the developing nasomedial and nasolateral processes to the formation of the nose as the structure of the lip forms around the developing stomodeum and the nasal pit deepens.
THE FACIAL SKELETON AND NASAL STRUCTURES

The cartilage of the nasal capsule is the foundation of the upper part of the face.\textsuperscript{10} The bony elements of the facial skeleton appear around it and replace it in part. The lateral masses of the ethmoid form by endochondral ossification of the nasal capsule. The frontal processes of the maxillary bones, the premaxillary bone, the nasal bones, the lacrimal bones, and the palatine bones all form in membrane in close relationship with the roof and lateral walls of the cartilaginous nasal capsule.\textsuperscript{10} The vomer develops in membrane in relation to the perichondrium of the septal process. Eventually nearly all of the nasal capsule becomes ossified or atrophied. All that remains of the nasal capsule cartilage in adults is the anterior part of the nasal septum and the alar cartilages that surround the nostrils.\textsuperscript{10}

Specifically, the midline septal cartilage is continuous with the cartilaginous skull base. At birth, the skull base has three major ossification centers, and the septal cartilage has not yet ossified. The lateral masses of the ethmoid have ossified, forming the paramedian bones, but the cribriform plate is still cartilaginous or fibrous. At birth, therefore, the entire midline of the face may be a lucent strip of cartilage situated between the paired lateral ossification centers. This lucent midline can simulate a midline cleft from the nares to the presphenoid bone.\textsuperscript{10} Because the appearance of the nasal septum varies with the patient's age, one must interpret imaging "evidence" of midline defects and sinus tracts carefully. Of patients less than one year of age, 14% have no midline ossification of the anterior fossa or septum.\textsuperscript{10-13}
OVERVIEW OF CONGENITAL NASAL ANOMALIES

Losee et al.\textsuperscript{13} presented a classification scheme for congenital nasal anomalies based on a review of 261 patients seen at the craniofacial center at the Children's Hospital of Philadelphia over a 22-year period. They separated anomalies into four types: Type I, hypoplasia and atrophy; type II, hyperplasia and duplications; type III, clefts; and type IV, neoplasms and vascular anomalies. These types represented 62\%, 1\%, 16\%, and 20\% of cases, respectively. The high association of both type I and type III anomalies with craniofacial syndromes may give way to the somewhat skewed prevalence of these groups of anomalies in this chapter.

Type I anomalies represent paucity, atrophy, or underdevelopment of skin, subcutaneous tissue, muscle, cartilage, and bone in varying degrees. Although arhinia and unilateral nostril agenesis are quite rare, lesser forms of nasal hypoplasia are quite common as part of many craniofacial syndromes. Type II anomalies involve an excess of tissue and include the spectrum of deformities from bifid septum and columella associated with dermoid cysts and sinuses, to the rare proboscis lateralis. Type III anomalies include Tessier nasal clefts (Tessier 0, 1, 2, 3) and their cranial extensions (Tessier 11, 12, 13, 14). These clefts usually result in hard and soft tissue deficiencies and are also frequently associated with other craniofacial syndromes. Type IV anomalies include congenital masses such as the spectrum of dermoid neoplasms, glioma, encephalocele, vascular malformations, and benign neoplasms.

Although we have taken a slightly different path with the basic sections of this chapter, the reader may find it helpful to keep Losee types in mind when considering the spectrum of congenital nasal lesions.

**Congenital Absence of the Nose**

Reviewing the previous relatively brief discussion of the very complex processes that occur in development of the nose much seems to rest on the effect of molecular signaling and tissue patterning in the face. Sonic hedgehog may be the morphogenic organizer, whereas fibroblast growth factors may serve as the stimuli for mesenchymal outgrowth. Insufficiency of the frontonasal and nasomedial processes may result in hypoplasia or absence of the nose and intermaxillary segment (see Fig. 33-2, A). If there is decreased proliferation in the forebrain and frontonasal processes, then the plumping of the nasomedial and nasolateral processes do not progress, or progress incompletely, and the ectodermal sculpting that should progress to create the nares and the gradual breakdown to communicate with the pharynx may either not progress at all or progress incompletely (Fig. 33-3). Lesser distortions of this process may result in isolated nasal deformities with incomplete canalization of the nares and residual soft tissue and cartilage masses within the nasal airway. Not surprisingly, given the intimate relationship between the developing brain and the central facial structures, the most severe forms of nasal hypoplasia may be associated with brain anomalies (often in the holoprosencephaly spectrum)\textsuperscript{14,15} (see Fig. 33-2, B).

Given the fact that newborn infants are obligate nose breathers, children born with either incomplete formation of the nose or total absence of the nose face the same emergent problem as children born with complete choanal atresia, and after immediate intubation these infants need a tracheostomy. Subsequent reconstruction of an absent nose is complicated by hypoplasia of the supporting tissues (both nasal and maxillary, cartilaginous and bony), soft tissue deficiencies, and the desire to create a functional nasal airway and maintain its patency, if possible. These
procedures may need to be staged over many years and are not typically completed until the affected child reaches skeletal maturity (Fig. 33-4). A combination of forehead flaps, nasolabial flaps, and local tissue advancements are necessary for soft tissue coverage.\(^{16,17}\) Cranial bone grafts, rib grafts, and conchal grafts can be used for skeletal reconstruction. Microsurgical tissue transfer may be necessary in the most severe cases to provide enough tissue for internal lining and external covering.

---

**Fig. 33-3** This infant has a rare combination of nostril agenesis and complete bilateral cleft lip. The border of the nostril is evident on the left side, but the nasal pit failed to deepen and break through the buccopharyngeal membrane. Consequently there is a solid mass of tissue in the airway to the level of the piriform margin where the posterior airway is patent.

---

**Fig. 33-4** A. This child was born with severe nasal hypoplasia, absence of the nasal airway on the right side, a hypoplastic nasal passage on the left side, mild hypotelorism, and a midline cleft of the lip. Additional anomalies were noted in the heart and gastrointestinal tract. She showed no central nervous system anomalies. During her first week of life, she underwent a tracheostomy. B, Three-dimensional CT scan showing frontonasal hypoplasia, medialization of the orbits, absence of a nasal passage on the right side, and presence of a nasal passage at the level of the piriform margin larger than what is seen at the skin level. C, After an initial failed attempt to reconstruct the external nose with expanded regional flaps when this patient was 5 years of age, formal reconstruction with expansion of the forehead was begun when she was 9 years of age.
Fig. 33-4, cont'd  D, Two weeks before forehead flap reconstruction, regional flaps were elevated around the nostril pit on the left side, bone was drilled out to open the airway to the point where the local surface flaps could be sutured to lining tissue behind the piriform aperture. E, Tissue expander in place and flaps healed around the reconstructed left nasal passage. F-H, Incisions were made at the borders of the planned external nasal reconstruction, and the border flaps were turned toward one another to provide lining tissue for the left nasal vault and coverage for the combined cranial bone and cartilage grafts used for support beneath the forehead flap. A pseudonosrill was created on the right side. I-K, Results at 14 and 19 years of age.
Nasal Hypoplasia

Nasal hypoplasia can present in a number of ways and with varying degrees of deformity. The severity of the deformity may be a function of how early in development the normal processes are interrupted and whether the tissues required to form the individual nasal parts are not developed, partially developed, or present but displaced. It can be associated with dwarfism and other genetic conditions11 (Fig. 33-5; see also Fig. 33-3). Trauma at an early age can also result in lack of nasal growth. The nasal dorsum may be very low because of a lack of growth of the nasal bones and seprum.8,9 Less frequently the tip cartilages themselves may be hypoplastic with resultant aesthetic and functional sequelae. The severity of the condition may not become fully apparent until skeletal maturation.

Having established a functional airway early, it is generally advisable to allow time for growth of the nasal structures. When possible, initial corrective efforts are directed at placing structures as aesthetically as possible before the child reaches school age. Typically bone grafts that are placed early will need to be augmented in later years. Our experience has demonstrated growth of these grafts, but not sufficient enough to obviate the need for later augmentation. Early grafts may still have the benefit of stretching the overlying skin envelope (see Fig. 33-5). Definitive reconstruction may be delayed until the child is close to skeletal maturity. Reconstruction through an external rhinoplasty approach can yield gratifying results. Autologous cartilage grafts from the septum, conchae, and rib serve as a nasal reconstruction framework. Wide undermining of the nasal skin to the frontal bone superiorly and the maxilla laterally usually yields enough soft tissue to cover the framework.

---

Fig. 33-5  A, Patient with congenital nasal deformity characterized primarily by hypoplasia of the underlying bone.
Fig. 33-5, cont'd  B, Intraoperative view of costal cartilage L-strut being carved. Note removal of perichondrium to prevent warping. A small K-wire is also inserted along the longitudinal axis of each strut to prevent warping. C, Stacked cartilage grafts can be carved from the nasal septum, conchal cartilage, or costal cartilage. This technique is useful in teenagers with mild-to-moderate hypoplasia of the dorsum. D, Patient after placement of the L-shaped cartilage strut to reconstruct the hypoplastic dorsum.
Facial Clefts

Deranged development of the frontonasal process, and/or failure of adjacent processes to merge successfully, results in a coherent series of malformations.\textsuperscript{14,15} The insufficiencies of frontonasal and nasomaxillary processes mentioned earlier result in some of the common manifestations of holoprosencephaly.\textsuperscript{8,9,11} Failure of the two nasomaxillary processes to merge in the midline produces the more rare, true midline cleft lip and palate with hypertelorism. These defects and the full spectrum of typical and atypical clefts have varied effects on the overall structure of the nose (Figs. 33-6 through 33-9). The discussion of the typical cleft lip nasal deformity and a review of the classification and features of the atypical nasal clefts appear elsewhere. Several different anatomic classifications for rare craniofacial clefts have been reported. In 1976 Tessier\textsuperscript{18} proposed a new system of classifying facial and craniofacial clefts based on the orbit as a reference structure. He recognized that there was a soft tissue as well as a bony component to these clefts. His system attempts to offer a common terminology for the description and treatment of these rare and complex clefts. As noted earlier and described in detail in Chapter 26, the Tessier nasal clefts include clefts 0, 1, 2, and 3, with their cranial extensions being Tessier clefts 11, 12, 13, and 14. Kawamoto\textsuperscript{19} reported the incidence of these atypical clefts as 1.43 to 4.85 per 100,000 births, making them far rarer than typical cleft lip and palate malformations. David et al\textsuperscript{10} studied these rare clefts with CT and three-dimensional reconstructions, improving our comprehension of the anatomic anomalies and reconstructive challenges. As with the design and planning of a typical cleft lip repair, the repair of nasal clefts in almost all cases requires rearrangement of the cartilaginous structures (with or without additional cartilage grafts, and one or more Z-plasty-type lengthening procedures) to correct the vertical deficiencies inherent in all the of these cases\textsuperscript{21} (see Figs. 33-7 and 33-8). For more complex clefts, repair often requires simultaneous correction of the eyelid(s) and lip (see Fig. 33-9), although other clefts may be confined entirely to the nose. Not surprisingly, most need staged revisions as the child grows.\textsuperscript{22,23} Often the defect in these cases has a bony component that involves the maxilla and skull. Reconstructive planning in these complex cases requires bony and soft tissue reconstruction, which is staged as the patient develops. Complex reconstructive techniques, including soft tissue expansion, bone grafting, recon-

![Fig. 33-6](image-url)

This infant was born with bilateral Tessier number 1-13 clefts and asymmetrical number 3 clefts without clefting of the lip, associated with a small encephalocele.
**Fig. 33-7**  A, This 5-year-old child was born with a Tessier number 2 cleft on the left that was not associated with any bony anomaly. B-E, Outline of the nasal flaps, including both skin and cartilage, used for the cleft repair. F and G, Result 2½ years after the first surgery and 6 months after a revision procedure to modify a limited alar web.
Fig. 33-8  A, Child with Fraser syndrome, a rare autosomal recessive disorder, demonstrates the features of cryptophthalmos and nasal anomalies including a broad nose with a midline groove, depressed nasal bridge, and hypoplastic nares with Tessier number 3 clefts. B and C, Outline of Z-plasty to reposition parts of the disrupted alar margins carried out when the child was 4 years of age. D, Appearance 2 years after repair. E and F, Appearance at 12 years of age.
struction of the lacrimal system, and on occasion craniosynostosis surgery, may be required. Often, if minor, these complex clefts can be resolved with local flaps. If major lateral nasal clefts are present, a combination of a forehead flap with tissue expansion and cartilaginous reconstruction in staged fashion may be required (see Fig. 33-9). Occasionally, composite grafts, usually from the concha, can be used to reconstruct the lateral ala with excellent color match and function. Clefts limited to the nose can vary in severity and include the upper and lower lateral cartilages as well as bones in severe cases. Surgical techniques are individualized according to the severity of the cleft. Recognizing the deficiency of soft tissues, and often bone in three dimensions, can facilitate surgical planning and reconstruction.

Fig. 33-9  A, This child illustrates the more “typical” Tessier-type clefts in combination with atypical clefts and hypoplasia resulting from amniotic band syndrome. B, Markings for surgery when the child was 13 months old, after tissue expansion, showing the initially proposed combination of advancement flaps, Z-plasties, and forehead flaps, in addition to planned repair of the lip and palate clefts (earlier surgery was delayed because of neurologic issues and delayed scalp healing). C, Result at 1½ years after the outlined single surgery, along with conformer expansion of the reconstructed eye socket and placement of an ocular prosthesis.
Nasal Masses

Nasal masses in the pediatric population have various causes, and proper diagnosis is the key to optimal treatment. The differential diagnosis includes nasal dermoids, gliomas, encephaloceles, and vascular lesions (hemangioma and vascular malformation). The embryologic development of the frontonasal region helps explain how the former three groups of lesions represent a continuum in the disrupted process.11,26-30 Early in the developmental process there is a potential space between the developing frontonasal bony structures and the nasal capsule, which develop the nasal cartilages. This prenasal space represents an embryonic “fault line” from the foramen cecum to the skin surface (Fig. 33-10), as clearly demonstrated in the classic paper by Sessions et al.31

Physical examination can usually, but not always, narrow the differential diagnosis. An MRI with contrast and often a complementary CT scan may be necessary to determine the extent of a lesion. MRI has the advantage of not irradiating the radiosensitive tissues of the eye and is therefore the preferred diagnostic method. The embryologic development of the frontonal nasal region, which has been previously discussed, helps explain the basis for gliomas, dermoids, and encephaloceles11,26,31 (Fig. 33-11; see also Fig. 33-10).

Nasal Dermoids

The presence of a frontonasal dermoid may be noted at birth or in early infancy as a midline pore, possibly with protruding hair and possibly with associated widening of the nasal dorsum (Figs. 33-12 through 33-15). Nasal dermoids represent the most common congenital nasal mass. Some children may initially present with an infection in the cyst, with swelling and erythema anywhere from the glabella to the nasal tip (see Figs. 33-14 and 33-15). On rare occasions, a child may present with a brain abscess without previous recognition of the cyst or sinus opening from which the infection arose22-34 (see Fig. 33-13).

Because the extent of soft tissue tracts may not be recognized beforehand, a CT scan or MRI is essential before surgery. The preference for one study over the other is mostly an indi-
Fig. 33-11  Spectrum of nasal cysts and masses. Classification depends on location and what tissue remnants are left behind. A, Dermal sinus with intracranial extension; B, dermal sinus tapering to fibrous tract that extends through the foramen cecum; C, dermal cyst alone; D, glioma; and E, encephalocele.

individual one, and some lesions may require both studies to determine the continuity of the sinus tract through the foramen cecum and any presence of an intracranial component to the lesion. An MRI scan can provide valuable information regarding the soft tissue tracts associated with dermoids and help differentiate normal anatomic variants of the anterior cranial fossa from sinus tracts penetrating intracranially (see Fig. 33-14, B and C). CT scans can provide the best picture of the bony anatomy of the nose and cranial base (see Figs. 33-13, B and C, and 33-15, B). CT scan data can even be translated into a life-sized anatomic replica of the patient's skull. These three-dimensional models can be a valuable planning and clinical adjunct if bony resection and/or reconstruction are necessary. Lesions with intracranial extension either course beneath the nasal bones through a widened septum, then through the foramen cecum, or through a widened frontonasal suture, then the foramen cecum. In both cases a bifid crista galli is usually noted on CT scan, and an intracranial portion of the dermoid may lie between the leaves of the falk. Excision
Fig. 33-12  A, This 2-year-old child had obvious swelling and a palpable cyst on the nasal dorsum with an overlying sinus opening. B, The tract was cannulated with a lacrimal probe at the time of excision. C, The cyst and sinus before final resection, and fully excised overlying the lacrimal probe.

Fig. 33-13  A, This 6-year-old child was first seen with an intracranial abscess. On close inspection of his nose, a sinus opening was detected at the nasal tip.
Fig. 33-13, cont'd  B and C, Coronal and axial views demonstrate the defect at the level of the foramen cecum. D and E, Anatomy of the cyst in both sagittal and three-dimensional views of the cup-shaped foramen cecum, widened by the presence of the intracranial portion of the dermoid cyst. F, The full length of the extracranial tract is shown. G and H, Intraoperative intracranial view of the dermoid cyst as resected from between the leaves of the falk cerebri and the cup-shaped bony defect illustrated in E.
Fig. 33-14  A. This unusual dermal cyst and sinus were found to extend not only intracranially but intraparenchymally (into the frontal lobe). A small hair can be seen protruding from the sinus opening. B, MRI showing cup-shaped defect in the crista galli. C, Intraparenchymal location of the cyst in another axial view. D, The keyhole frontal craniotomy is outlined along with the planned osteotomy line for removing the frontonasal complex en bloc. E and F, Excellent views of the area between the falx cerebri, which provides access for resecting the portion of the dermoid within the frontal lobe as well. G, The frontonasal bony complex and frontal bone are replaced and fixed with absorbable plates.
Fig. 33-15  A, This infant had intermittent drainage from a sinus opening in the area of the glabella. Despite recommendations to proceed with resection, the family did not return for 4 years. B, Series of axial views through the area of the cyst, which had eroded the surrounding bone. The films were interpreted as demonstrating chronic osteomyelitis. C and D, At the time of resection, the ragged edges of the bony defect were clearly visible.
of these more complex lesions is facilitated by a cooperative effort between the plastic surgeon
and the neurosurgeon using a craniofacial approach.35,36

The surgical approach to "simple" cysts or sinus and localized cyst alone varies depending on
the exact location on the nose, or glabella. As previously mentioned, the sinus opening at the base
of the columella extends to the nasal spine only, and, if associated with a cyst, the sinus is circum-
scribed for excision, and the remaining dissection of the cyst is accomplished through the labial
sulcus. For the remaining sinuses and cysts from nasal tip to radix, one should choose the tech-
ique that allows the best view of the tract and its final point of attachment, with every effort to
minimize the final scar (see Fig. 33-12). Some prefer excision of the sinus opening in combination
with an open rhinoplasty approach37; however, if the tract extends for any distance beneath
the nasal bones, it may be very difficult to see. A poor field of vision during the first surgery may
result in the problem recurring, so every effort should be made to remove the deep component
at that time. If the cyst or sinus is intimately attached to the skin, the tract may be difficult to
obliterate if it is not excised directly, which may lead to recurrence. Sinuses and cysts of the
glabellar area (fronticus nasofrontalis origin) without intracranial extension may be approached
through a curvilinear incision within the shadow of the lateral nose alone or in combination with
excision of the sinus opening. This yields a barely visible scar and gives an excellent view of the
area. A transnasal, endoscopically assisted approach has been reported for resection of lesions
that do not have an intracranial component.

For lesions with suspected or confirmed intracranial extension (see Figs. 33-13 and 33-14),
excision of the sinus opening begins from the external approach combined with a frontal cra-
niotomy, with delivery of the tract through to the fully exposed frontonasal area following it
through to its intracranial terminus.38 For tracts that seem to terminate in a foramen in the bone,
a biopsy will confirm the presence or absence of epidermal elements in the tract. If confirmed,
a limited craniotomy is performed for access. For more complex lesions and those with con-
formed intracranial extension, the above is combined with an en bloc removal of the frontonasal
bone with direct visualization of the tract, crista galli, and falx through a “keyhole” osteotomy39,40
(see Fig. 33-14). Immediate reconstruction of the nasal and cranial defects is preferred. For cases
in which a significant amount of the bony and cartilaginous skeleton of the nose have been re-
sected, the nasal dorsum should be reconstructed with a combination of cartilage and bone grafts.
If the cartilaginous defect is small, a conchal graft may suffice, but if the defect is larger a costal
cartilage graft is preferred. Most defects resulting from a keyhole resection involve the frontal
bone and anterior base of the skull. Cranial bone grafts are optimal for reconstruction and are
usually obtained from the parietal bone. Preserving a large anteriorly based pericranial flap allows
its interposition between the dura and the reconstructed anterior fossa, separating the nasal con-
tents from the dura. Donor defects are created if the skull is too thin to split. These defects are
behind the hairline and can be repaired satisfactorily using allograft combined with demineral-
ized bone matrix paste. Because these lesions typically occur in the infant or pediatric age group,
nasal development and frontal sinus development may be affected. Often further nasal recon-
struction will be necessary in the teen years. This is done using an external approach for optimal
visibility using autologous cartilage grafts, which can often be obtained from the septum. Frontal
sinus hypoplasia can occur after keyhole resection if the frontal sinus origins are disturbed, but
this usually does not manifest until adolescence. The external manifestation is a visible depres-
sion occurring just above the nasofrontal juncture. This can be corrected with gratifying aes-
thetic results by using hydroxyapatite cement and a bicoronal approach.
Nasal Gliomas

Nasal gliomas represent heterotrophic neural tissue secondary to deficient regression of neurologic tissue in embryonic development. Gliomas in an extranasal location may still penetrate the bone in the frontonasal suture area and are often associated with both broadening of the nasal root and increased intercanthal distance (Figs. 33-16 and 33-17). Continued extension through the widened foramen cecum and attachment to the dura in the area of the falk should be ruled out by MRI scan. An MRI scan can distinguish the interface between cartilage, bone, and brain in the region of the frontonasal area. Dural continuity can be ruled out by metrizamide cisternography if an MRI scan is not definitive. Gliomas presenting intranasally are usually noted first during physical examination in association with complaints of nasal airway obstruction or evidence of nasal bone distortion secondary to an expanding intranasal lesion (Fig. 33-18). Most often gliomas are confused with hemangiomas. Once the diagnosis is made and the extent of the lesion determined, excision can be carried out. The surgical approach is again tailored to allow viewing and excising the entire lesion. Lesions having an intracranial communication may re-

Fig. 33-16  Extranasal glioma with normal overlying skin. A, At birth this infant had a firm, noncompressible, nonfluctuating mass typical of an extranasal glioma. B, The dural connection was pinched off and the glial tissue remained. C and D, The mass was resected, and the relationship to the underlying bony septum and nasal mucosa is shown. E, Patient at 1½ years after resection.
**Fig. 33-17** Extranasal glioma involving the overlying skin. **A,** This lesion masqueraded as a vascular lesion, but it was not compressible and did not fluctuate with breathing or a Valsalva maneuver. **B,** Defect in the nasal bone and underlying dorsal septum at the time of resection.

**Fig. 33-18** Intranasal glioma. **A,** Three-year-old child with a gradually enlarging mass in the left nasal cavity that caused splaying of the nasal bone. **B,** Anatomy of the intranasal glioma and its relationship to the overlying nasal bone.
require a frontal craniotomy for complete excision. Extranasal lesions may require excision of the overlying skin with flap reconstruction when the skin is involved. Endoscopically assisted excision, using a modified external rhinoplasty approach, may be possible for lesions with a subcutaneous presentation and no intracranial involvement. Histological examination reveals glial tissue, primarily astrocytes and fibrous tissue with occasional neurons and ependymal cells being found.\textsuperscript{37}

**Frontonasal Encephaloceles**

Encephaloceles or meningoceles can also present as midline or paramedian nasal masses, usually at the nasofrontal area. A midline or paramedian cystic mass presents at birth and can attain enormous proportions (Fig. 33-19). Size fluctuation may be noted during crying or straining. In general, much of the dural sac is filled with cerebrospinal fluid, but it may contain herniated frontal lobe along with amorphous glial tissue. Again, the most common path of herniation is through the open cranial base at the site of the previous foramen cecum and the widely patent frontonasal suture.\textsuperscript{14,15} Herniated tissue may extend well into the ethmoid air cells with an intranasal component being present. Encephaloceles may be associated with a widened intracranial distance or true hypertelorism. MRI and MRA can give a detailed multiplanar image of the extent of the lesion, the vascular anatomy, and the presence of neural tissue in the herniated sac.\textsuperscript{21} Arteriography is often beneficial for determining the surgical approach and the presence or
absence of functional brain tissue. Significant defects of the nasofrontal area and the anterior base of the skull may be present. A CT scan with three-dimensional reconstruction can give a complete picture of the extent and shape of the defect. A three-dimensional acrylic model, which may be sterilized for operative use, can provide valuable information for planning and executing the bony reconstruction. Treatment is best planned as a cooperative neurosurgical and plastic surgical effort. Histologically, the sac is lined by arachnoid with possible heterotrophic glial tissue. These are accompanied by underlying bony and dural defects. Repair of both the dura and the underlying bony defect, usually with cranial bone grafts, is recommended. Bone grafts can be cut and shaped using the three-dimensional representation. Frontal encephaloceles can often result in hypertelorism because of their extension into the ethmoid air cells. Such a residual deformity may require secondary correction depending on the final aesthetic and functional outcome of the primary repair.

Fig. 33-19 A, Frontonasal encephalocele in a newborn infant. B, Midsagittal CT view shows a defect in the area of the previous fonsiculus nasofrontalis; the continuity of the dura is also demonstrated. C, The defect is shown on a three-dimensional CT scan. D, Intraoperative view of the encephalocele once it is fully exposed and before resection.
Fig. 33-19, cont'd  
E, A cranial bone graft was used to reconstruct both the anterior cranial base and the nasal dorsum. F, Six months after repair. G, At 4 years of age the child demonstrated a trigonocephalic deformity with a prominent midfrontal ridge and antimongoloid slant to the orbits. This CT scan was obtained before craniofacial reconstruction with frontoorbital repositioning and frontal cranioplasty. H-J, One year after reconstruction.
Nasal Hemangiomas

Nasal hemangiomas are one of the most common pediatric nasal masses. They may present as an external red or purple mass, but occasionally they can be entirely subcutaneous, presenting as a swollen nasal tip (Figs. 33-20 through 33-24). They may have an intranasal component that can compromise nasal airway function. Although some spontaneous involution may eventually occur, nearly all hemangiomas of the nose require treatment to minimize long-term sequelae. Physical examination is often sufficient to differentiate hemangiomas from arteriovenous malformations. These malformations are usually present at birth, may have a thrill when auscultated, and are high flow. Purely venous malformations are typically darker than hemangiomas and can be compressed. If a question persists regarding the exact nature of a lesion, an MRA scan can resolve it in almost all cases. Strategic planning for approaching nasal hemangiomas should include minimizing external skin involvement, preserving function, and minimizing long-term distortion of the cartilaginous nasal skeleton. A number of approaches can be used to treat nasal hemangiomas successfully, including serial excisions, steroid injections, external laser treatment, intralesional laser treatments, and resection using an external rhinoplasty approach. We use a combination of these therapeutic options based on each patient’s individual needs (see Figs. 33-20 through 33-22). If a lesion is entirely subcutaneous with normal skin, significant shrinkage can be achieved using intralesional KTP or YAG lasers. This treatment is carried out every 4 weeks for a total of 3 to 6 treatments. A small amount (0.5 ml) of Kenalog 5% is injected into the deepest part of the lesion during each treatment. This small amount has not been found to cause subcutaneous thinning, and it helps decrease proliferation of the hemangioma and decrease scar tissue as the hemangioma becomes more fibrotic, which significantly decreases the vascularity and size of the lesion. If any excess bulk remains 6 to 9 months after the final laser treatment, and there has been no gross regrowth of the hemangioma, an external rhinoplasty approach can be used to remove any residual mass effect. A gull-wing incision that does not violate the nasal

![Fig. 33-20](image)

A, This infant had a nasal hemangioma with an internal component causing partial airway obstruction. B, Appearance after serial treatments with KTP laser, Candela laser (Candela Corp., Wayland, MA), and steroid injections.
tip subunit allows excellent exposure to debulk the residual fibrofatty tissue and address the associated cartilaginous deformity. It also allows for limited skin reduction, again without violating the subunits. Often the medial crura of the lower lateral cartilages are splayed in a bifid appearance by the pressure of the hemangioma (see Fig. 33-23). This can be reconstructed during an external rhinoplasty by simply suturing from the footplates to the domes, thus reversing the pressure changes of the hemangioma. If the hemangioma has significant skin involvement, or if the nasal tip skin has ulcerated during the most active proliferative stage leaving permanent tip

---

**Fig. 33-21**  A, This infant had a nasal tip hemangioma with external and internal components. B, Appearance after sequential KTP laser, Candela laser, and steroid injections, and a subsequent external rhinoplasty approach. The residual hemangiomas have been debulked and the nasal tip has been reconstructed.

---

**Fig. 33-22**  A, Large nasal dorsal hemangioma interfering with the right medial visual field. B, After initial treatment with KTP laser, Candela laser, and steroid injections to decrease the size of the lesion, the residual lesion was treated surgically with debulking and local flap reconstruction.
Fig. 33-23  A and B, This child, age 2 years 4 months, who had previously received steroid injections and laser treatment for hemangioma of the nasal tip, demonstrates the typical Cyrano nasal tip deformity that follows a bulky hemangioma confined to the nasal tip. Excess fibrofatty and vascular tissue remains, causing the domes of the alar cartilages to splay apart. C, The initial resection is carried out through a "gull-wing" incision. The upper border of the resection line is estimated, but is refined once the fibrofatty vascular mass is resected, and the skin is redraped. D, Result 1½ years postoperatively with residual vascular changes in the skin. E, Patient at 6 years of age, 4 years after resection.
Fig. 33-24  A, This infant had an extensive facial hemangioma with numerous ulcerations and significant deformation of the underlying alar cartilages. B, At 4 years of age this patient underwent extensive debulking, partial skin resection, and repositioning of the distorted alar cartilages. C, Patient 3 months after a second debulking procedure and 2 years after the excision shown in B. D and E, Although the residual lesion involuted, the scars matured further, and the nose was allowed to grow. The child is shown at 7 and 16 years of age after the initial resection. F and G, The patient is shown 8 months after revision of the alar margin with the addition of a small conchal cartilage graft.
scarring, an external skin excision is necessary and may be preferable (see Fig. 33-24). Often significant shrinkage of the lesion, as well as salvage of affected skin, can be achieved through a combination of external Candela laser treatment and internal KTP laser treatment. These treatments can be combined with local steroid injection to halt growth, minimize cutaneous involvement, and greatly facilitate the process of serial excision. Once maximal shrinkage of the lesion has been achieved, an external excision using a serial approach is advocated. Wide undermining of the normal skin can be achieved, and this skin can be used to replace the involved hemangioma skin over a period of years. Serial excisions are combined with reapproximation of the medial crura, as previously described. In addition, reconstruction may require conchal cartilage grafts when cartilage deficiency is also present to give the best overall aesthetic result (see Fig. 33-24).

**Cutaneous Lesions**

The range of both congenital and acquired cutaneous lesions in the pediatric population is covered in Chapter 6 and is not reviewed here. The smaller (half centimeter or less) acquired lesions present the same issues and concerns as acquired lesions in the adult population, but congenital nevi, both melanocytic and sebaceous, may create particular concerns, both in their extent and distribution, that warrant discussion here (Figs. 33-25 through 33-28). Unlike acquired malignant lesions in adults, reconstruction is typically confined to reconstructing the skin even for extensive lesions, because the underlying supporting cartilage and lining tissues are either uninvolved entirely, or, with the latter, may be left intact (balancing concerns about potential functional disturbance and the likely low risk of later malignant degeneration).

**Congenital Melanocytic Nevi**

Congenital nevi of the nose are uncommon and may present as confined to the nose alone or as part of a more extensive nevus of the periorbital region, hemiface, or bilateral face. Although the risk of malignant transformation of giant congenital nevi is well established, the risk of malignant transformation of congenital facial nevi appears to be less common; however, the psychological impact on a child and the child’s family from a pigmented facial nevus can be profound. Additionally, some nasal nevi can produce a variety of functional problems from marked thickening of the subcutaneous tissues creating a bulk and weight that may compress the ala and secondarily narrow the nostril. For these reasons we advocate excision and reconstruction of these nevi. Although some surgeons elect to delay the treatment of these very visible lesions until later ages, when nasal growth is more complete and some procedures may be easier, treatment delays may have a significant long-term impact on a child trying to deal with constant peer ridicule.

**Staged Excision (With or Without Adjacent Tissue Expansion)**

Although one must always design an excision and reconstruction with the aesthetic nasal subunits in mind, and give consideration to recruiting distant forehead tissue to provide coverage, excision of some of the nasal tip subunit, alar margin, and middle dorsum may be treated quite effectively using staged, serial excision, with or without local nasal tissue expansion (see Figs. 33-25 and 33-26). As long as underlying cartilage is left intact, even a limited reduction in nostril size (following alar margin excisions) may yield excellent long-term outcomes, with only minor scar revisions of balancing procedures necessary near completion of nasal growth.

**Excision With Full-Thickness Skin Graft Coverage**

Although nevus excision only involves excision of skin and subcutaneous fat to the perichondrial or fascial level, full-thickness skin grafts typically do not provide an excellent long-term outcome. However, they may be beneficial for removing deeply pigmented, very visible lesions
in children younger than some surgeons may be comfortable with. Full-thickness skin graft coverage as part of a larger expanded full-thickness skin graft covering the periorbital region, canthus, and lateral nose as a single unit, expanded, full-thickness skin graft may also provide excellent long-term results.\textsuperscript{51}

**Expanded and Nonexpanded Forehead Flaps**

Although there are differences of opinion regarding the benefits of expanding the forehead before using a forehead flap for nasal reconstruction, our extensive experience in treating large and giant congenital pigmented nevi of the face has demonstrated unique benefits of the expanded

---

**Fig. 33-25**  
A, This 2-year-old child had a small congenital pigmented nevus of the nasal tip and portion of the dorsum. She was treated with three serial excisions over a 1\(\frac{1}{2}\)-year period. B, Six months after completion of the excisions. C and D, She returned 12 years later to discuss correction of residual nasal asymmetry, which primarily consisted of a dog-ear of extra skin along the alar margin and just within the nares. This could have been addressed many years earlier, but the family had moved out of the country in the interim.
Fig. 33-26  A, Five-year-old girl with a bulky congenital pigmented nevus of the nasal tip and dorsum. Although excision and reconstruction with a forehead flap was an option, the decision was made to excise the lesion with expansion of adjacent normal skin. B, Close-up view of the nevus before placing two 3 cc custom expanders through an incision within the nevus that also allowed a conservative partial resection of the nevus. C, Appearance of the expanders just before resection of the remaining nevus. D, Patient 8 months after resection. E, Same patient at 10 years of age.
forehead flap (see Fig. 33-27). When advancing the expanded central forehead laterally to excise nevi of the lateral forehead and temporal region, there is a relative "redundancy" of skin in the glabella and the ipsilateral side of the nevus excision. This tissue can be used based on the same supratrochlear vascular pedicle as a standard forehead flap, as a small flap to cover the nasal dorsum alone, or for total nasal coverage, with the added benefit that the donor scar will lie along the brow, rather than centrally in the forehead (see Fig. 33-27, C and D). Our experience with expanded forehead flaps in the early stages of treating large and giant nevi is still limited (in the 18- to 24-month age range), but our results clearly support its role as a powerful reconstructive option.

Fig. 33-27 Early stages of reconstruction in a child with a congenital giant pigmented nevus of the face. A, Patient at 6 months of age. B, After successful expansion that began at 1 year of age, with expanders in both the forehead and neck, the forehead flap is oriented so that the donor scar lies along the brow and continues out into the zygomatic temporal region. C, Appearance a few days after resection of the nasal nevus and expanded flap reconstruction. D, Six months after the procedure shown in B, an expander was placed in the child's cheek. After an additional 3 months of expansion, the remaining nevus of the cheek was resected and the forehead flap was lifted from the nose, thinned, and replaced. Patient as she appeared 10 days postoperatively.
Linear Sebaceous Nevi

Linear sebaceous nevi may present in isolation as a lesion or a series of striped lesions on the nose, or as part of a larger more extensive facial sebaceous nevus associated with sebaceous nevus syndrome.32

Although some physicians have elected to treat linear sebaceous nevi with lasers alone, any remaining portions of a lesion require an extended series of treatments, and it still may be visible enough to create an unsightly deformity. Of interest is the fact that the skin between stripes

Fig. 33-28 This infant was born with extensive sebaceous nevi, without associated central nervous system involvement. A, Larger lesions covered large segments of the scalp in combination with multiple linear lesions in the central and lateral face. This type of lesion is approached using a combination of tissue expansion and staged excisions of some of the narrower linear lesions. These excisions may be timed to allow partial excision at the time of expander placement and repeat excisions later at the time of more extensive excision and flap reconstruction. B and C, The first set of expanders at the completion of expansion. D, Patient's appearance 5 years after the start of staged excision and reconstruction with the greater part of the nevus on the nose, forehead, and scalp excised but residual lesions in the nasolabial region, lip, and lateral eyebrow just before further excision.
of lesion is normal, and serial linear excisions can preserve this normal tissue and allow excellent long-term outcomes. With an extensive facial nevus, staged linear excisions of the nasal lesion may be carried out simultaneously with procedures for either placing tissue expanders, or removing them and undertaking an extensive excision and flap reconstruction (see Fig. 33-28). Because the later procedures are typically carried out with 11 to 12 weeks of expansion, with a 4- to 6-month interval between each group of procedures, this allows multiple opportunities to serially excise nasal lesions.52

ACQUIRED NASAL DEFORMITIES

Acquired nasal deformities are most commonly caused by trauma. Traumatic deformities can be divided into those with an open skin component and those of a closed nature. When either open or closed nasal injuries occur, they can result in permanent nasal deformities. If the injury occurs at a young age, these deformities may not become apparent until the child has grown, at which point they may present with a saddle nose deformity or even complete nasal hypoplasia. Closed nasal trauma injuries are common, are associated with sports, or occasionally fisticuffs, and may involve injury to both the cartilaginous and bony components of the nose (Fig. 33-29). The initial evaluation should include a thorough history to reconstruct the mechanism of injury and approximate the force of impact. Standard radiographs may not reveal the extent of injuries in a developing nose, because there is a tendency for the nasal bones to undergo greenstick fracturing rather than shatter. Plain radiographs are not a substitute for a thorough examination. Computerized tomography of the maxilla and nose in both the axial and coronal planes can provide useful clinical information. An intranasal examination is necessary to determine whether a septal hematoma is present, which can lead to late pressure necrosis of the septum. A hematoma often presents as a unilateral purple swelling that can completely obstruct the nares. Patients with a septal hematoma often have pain that is disproportionate to the visible extent of their external

![Fig. 33-29](image_url) A, Eight-year-old girl with a bony and cartilaginous deformity of the nose that occurred when she fell off a horse. B, Same patient after closed nasal reduction.
nasal injury. Treating a closed nasal fracture consists of closed reduction and external splinting. If a septal hematoma is present it should be drained, and any loose fragments of septum should be debridged.\textsuperscript{51,52} Plain gut is used in a continuous mattress stitch to approximate the septal mucoperichondrial flaps and prevent recurrence of hematoma (Fig. 33-30). An external thermoplastic splint is applied, and no intranasal packing is used. If a septal hematoma is present a short course of oral antibiotics is prescribed. After 1 week the splint is removed. Contact sports are not allowed for 4 weeks. Open surgery for acute treatment of a closed nasal fracture is not indicated, because the skin and periosteal attachments holding the comminuted nasal bones can be disrupted, making stabilization challenging. Sequelae of untreated nasal fractures with or without septal hematomas include airway obstruction, aesthetic deformity, and saddle nose deformity. Even if optimal treatment is administered there may be a residual deformity that may become more apparent as facial growth progresses. Parents should be made aware of this possibility during the initial discussion of the injury and required treatment.\textsuperscript{51}

Open injuries often occur secondary to motor vehicle accidents, falls, animal bites, and sports injuries. Isolated nasal trauma requires thorough intranasal and extranasal examinations. Plain radiographs and CT both can provide valuable information. As with closed nasal trauma, it is imperative to determine whether there is a septal hematoma with open trauma. Open nasal fractures require reduction, inspection for septal hematomas, and prompt repair of the skin lacerations. CT scanning is recommended to rule out underlying maxillary injuries. If maxillary fractures are also present (and are more significant than nondisplaced greenstick fractures), they are treated with open reduction and resorbable fixation followed by closed reduction of the nasal bone fractures once the skin has been repaired. Wide undermining of the nasal skin should be avoided, because this further destabilizes bony fragments. Only obviously devitalized skin should be debrided, because even macerated traumatized skin is often viable. External splinting is used,
and occasionally resorbable nasal packing is used for very unstable fractures. Patients are kept on antibiotics and analgesics for 5 to 7 days. A conservative approach is recommended for the initial treatment of most open nasal fractures, followed by at least 12 months of observation. Approximately 40% of children who suffer significant nasal fractures exhibit a residual external or intranasal deformity when they reach skeletal maturity. Obstructive symptoms infrequently warrant intervention before skeletal maturity. A conservative approach is also recommended in these cases, with septal scoring and splinting, and turbinate reduction to improve the nasal airway if intervention is necessary before adolescence.51-53 If there is a symptomatic residual deformity at skeletal maturity, it requires an open rhinoplasty approach, often with cartilaginous reconstruction, septal reconstruction, and fracture reposition of the nasal bones. Autologous cartilage is advocated if augmentation is required, especially for the nasal dorsum. Septal cartilage can be useful for small defects, but costal cartilage seems to be more effective for optimal long-term results with larger dorsal defects.

Animal bites are most commonly from canines. Young children are particularly vulnerable, because they are at face height to many dogs. Teenagers may tease or roughhouse with dogs, putting their noses at risk of bite injury. Generally, the initial puncture by a dog’s teeth is followed by the victim pulling back, which often results in partial or complete tissue avulsion (Fig. 33-31). Initial treatment should include debridement, updating tetanus prophylaxis, and oral and topical antibiotics. The first steps of treatment are inspection of wounds with removal of all definitively devitalized tissue and prevention of secondary infection. Given the excellent blood supply in the area, initial debridement should be conservative. After the tissues have demarcated their viability the residual defect can be evaluated. If there is no significant tissue loss, scar revision may be considered 9 to 12 months after the initial injury, allowing time for scar maturation. If a true tissue avulsion is present, plans for a staged reconstruction should be discussed with the

Fig. 33-31 A, This teenager sustained a nasal avulsion injury after she was bitten by a dog. Note the loss of the lower lateral cartilage and caudal upper lateral cartilage with extensive skin loss.
Fig. 33-31, cont'd  B, Technique for harvesting and applying conchal graft for nasal reconstruction. Note that a graft can also be harvested with overlying skin as a composite graft. C, Same patient after staged reconstruction using conchal grafts for cartilaginous nasal vault reconstruction and a left nasolabial flap for external and internal soft tissue reconstruction.
patient and the family. Depending on the defect, local rotation flaps, forehead flaps, or composite grafts may be necessary for repair.\textsuperscript{54} Alar defects can be treated with local rotation flaps combined with conchal grafts or composite grafts for alar airway support. Larger defects of the skin and cartilage may require a forehead flap or nasolabial flaps, depending on the extent of tissue loss.\textsuperscript{54} Tissue expansion may be valuable for obtaining sufficient soft tissue for reconstruction. If adequate soft tissue coverage is available, the underlying nasal framework can be reconstructed at the same time. Significant defects involving the tip, dorsum, and lateral nasal sidewall cartilage are best treated using rib cartilage to construct a framework for supporting the overlying soft tissue flaps. Once the initial framework and soft tissue reconstruction have been accomplished and healed, several refining procedures can be considered. These include scar reduction and debulking procedures, aesthetic tip plasty, adding conchal cartilage grafts to minor defects, laser treatment of scars, and other modifications. Defects of the columella may require adjacent flaps, composite auricular grafts, or combinations of these procedures (Figs. 33-32 and 33-33). The patient, the patient’s family, and the treating plastic surgeon must all understand that reconstruction for extensive nasal defects will take multiple stages over a period of years. Significant improvement can be expected, but there will be residual donor and recipient site scars, and these will be permanent. It is important that the animal involved in a bite trauma be checked for rabies and reported to animal control authorities. Because lawsuits often result from dog bites, careful documentation is mandatory, along with a conservative estimate of the final aesthetic and functional outcome. It is impossible for the surgeon to estimate the total number of procedures and final outcome after the initial intervention, although many insurers and attorneys demand this information.

\textbf{Fig. 33-32} A and B, This 4-year-old boy was seen for reconstruction of columellar and tip defects resulting from partial necrosis of the columella caused by the pressure of the nasal cannulas that were used while he was in the neonatal intensive care.

\textit{Continued}
Fig. 33-32, cont'd  C, The patient returned at age 16 after undergoing three operations, including an attempted columnellar advancement according to the method of Cronin, two small composite grafts, and a scar revision. These procedures left him with considerable scarring and, according to the patient and his family, a more noticeable deformity. D, Lateral view before further reconstruction shows shortening of the columnella, blunting of the nasal tip, and notching of the tip. E, Patient at 17 years of age after reconstruction using an open rhinoplasty approach. After osteotomy and infracture of the nasal bones, septrhaphy and septal cartilage graft to the columnella, and flap reconstruction using portions of the previous composite grafts for additional tip support, the shape of the columnella and the nasal tip was restored. F, Lateral view 1 week after surgery shows improved tip projection and smoother contour of the columnella. G and H, Two months after reconstruction. This patient was seen on one other occasion to discuss minor scar revision, but he elected to postpone this.
Fig. 33-33 Forehead flap reconstruction of an extensive defect that occurred after tissue destruction resulting from a hemangioma. A and B, This 14-year-old girl is shown after destruction of the soft tissues and cartilaginous support of the nasal tip and columella subsequent to an involuting hemangioma. The skin over the entire nose is scarred and atrophic. The columella and right rim had been repaired by means of local tissue rearrangement when she was a young child. C, The skin of the nasal units is marked for excision. D-H, The angle of the septal cartilage and the entire alar cartilage complex are absent. Templates were made to guide exact replacement of the missing nasal skin and absent septal angle, tip cartilages, and alar support grafts. A triangle of septal cartilage was fixed to the residual septum with lateral spreader grafts to re-create the septal angle. Septal cartilage strips were then fixed to the distal septum and hinged laterally to re-create the medial, middle, and lateral crura to restore support and contour to the distal nose. (Case courtesy Frederick J. Menick, MD.)

Continued
Fig. 33-33, cont'd  I and J, The nose was resurfaced with a full-thickness forehead flap. K, At 3 weeks, the forehead flap is healed to the recipient site. The forehead donor site, which could not be closed under the hairline, was allowed to heal by secondary intention. L-N, Three weeks after transfer, the forehead skin was reelevated with up to 3 mm of subcutaneous fat. In this case it was maintained as a bipedicle flap hanging from its origin at the brow and from the distal columella inset. In most instances, the flap is completely elevated and temporarily positioned to the side of the face. The underlying excess of subcutaneous fat and frontalis is exposed. This is excised with a scalpel to re-create the subtle three-dimensional contours of the nasal surface. The flap is resutured to the recipient site with quilting and peripheral sutures.
Fig. 33-33, cont'd  O. Three weeks later the flap was divided, and its proximal aspect was returned to the forehead as a small inverted "V" at the brow. The forehead scar was subsequently revised. P-R, Postoperative result at 19 years of age. The excess fullness of the left alar base could be improved by minor revision.
KEY POINTS

- The definitive nasal structures, both external and internal, develop during the fourth to eighth weeks of gestation concomitantly with the developing primary and secondary palate.

- Facial and limb malformations are known to result from deficiency or excess of molecular signaling (sonic hedgehog, fibroblast growth factor, and retinoic acid signaling, among others). Similar phenotypes, such as clefting, may result from either deficiency of the appropriate midline tissue or excess of other midline tissue so that the appropriate processes cannot meet to fuse in the midline.

- Because the appearance of the nasal septum varies in radiologic examination depending on a patient's age, one must interpret imaging evidence of midline defects and sinus tracts carefully. In fact, of patients less than one year of age, 14% have no midline ossification of the anterior fossa or septum.

- One can best understand the spectrum of congenital frontonasal masses (dermoid cysts and sinuses, gliomas, and encephaloceles) with a clear understanding of the embryology of frontonasal development and the paths of potential embryonic "faults" through the foramen cecum and frontonasal suture.

- The choice of radiologic technique for evaluating frontonasal cysts and sinuses varies partly on surgeon preference. Some surgeons think that the bone anatomy and area of the crista galli is better delineated with CT scan, whereas others think that the sinus tract is better delineated with MRI. Complex cases with intracranial extension may best be studied using both.

- Dermal cysts and sinuses with obvious intracranial extension require a craniofacial approach typically through a "keyhole" frontal osteotomy with initial resection of the intracranial dermoid. For cases that have indefinite extension, the tract is followed to its tapered stalk, and a biopsy is taken to determine whether dermal elements exist. If they are present, the intracranial portion is then addressed.

- For lesions having an intracranial component, a follow-up MRI is recommended yearly for 3 years to rule out recurrent growth of any dermal remnants that were missed during the initial surgery.

- If a portion of the cartilaginous septum is removed surgically when following the sinus tract, it should be repaired during the same procedure to prevent a saddle deformity. If the defect is small, conchal grafts can suffice; if the defect is large, a costal graft may be required.
If there is a large defect in the frontal bone, the occipital bone can be switched to the frontal position to give a smooth contour to the forehead. The frontal bone defect is repaired with allograft and placed in the occipital position.

Nasal hemangiomas can be confused with venous malformations or arteriovenous malformations. If physical examination does not clarify the diagnosis, MRA with venous phase views can be helpful.

If a patient has a history of nosebleeds without ulceration of the external component, a significant intranasal component may be present. An MRA of the nose and face can reveal the internal extent of the lesion.

When treating nasal hemangiomas with laser therapy, more treatments with less intensity decreases ulcerations and burns. Power settings should be increased incrementally over several treatments until the minimal effective power settings are determined.

The Cyrano nasal tip deformity is a common sequela of hemangiomas of the nasal tip, with the involuting lesion leaving excess fibrofatty tissue with splayed alar cartilage domes.

Costal grafts are notorious for warping, but steps can be taken to prevent this. All of the perichondrium is removed, and a 0.4 mm Kirschner wire is inserted and cut just below the surface of the cartilage. Alternatively the rib cartilage can be cut longitudinally into thin layers that can be “bruised” with a cartilage crusher. These cartilage sheets can be stacked and sutured together to achieve the desired dorsum height.

Standard radiographs do not always accurately reveal nasal fractures in pediatric patients because bones can undergo greenstick fracture rather than fracture outright. In addition, the cartilaginous septum cannot be seen on plain radiographs. Accurate initial clinical assessment for significant bony and cartilaginous deviation as well as septal hematoma is crucial. If swelling prevents satisfactory assessment, ask the patient to return in 5 days with an old anteroposterior photograph for comparison.

The septum can be splinted with sterile x-ray film cut to fit and sutured with the septum in the middle. This is removed after 7 days.

If secondary surgical correction is necessary after initial closed reduction in adolescents, allow 9 to 12 months for complete healing before rhinoplasty. This interval can reveal areas of dorsal cartilage loss or deviation that may not be apparent initially.
REFERENCES—With Key References Annotated


The role of sonic hedgehog is particularly critical to the development of the frontonasal structures, and this paper provides an excellent early overview of its role as a basis for understanding much of the newer work and study of frontonasal development.


This chapter provides one of the most complete overviews of both head and neck embryology, with discussion of pathogenesis of the full spectrum of anomalies of the head and neck and special emphasis on radiological findings.

This article outlines a simple yet comprehensive classification scheme dedicated to congenital nasal anomalies, dividing cases into the following four subtypes: type I, hypoplasia and atrophy; type II, hypoplasia and duplications; type III, clefts (Tesier numbered); and type IV, neoplasms and vascular malformations.

The confusing spectrum of rare clefts is clarified. The bony and soft tissue components and reconstruction are discussed in this important paper.

Application of computerized imaging to rare craniofacial clefts further refines the three-dimensional understanding of these complex deformities.
   Sequelae of untreated septal hematoma are discussed in this article. Abscess formation can result in destruction of the dorsal septum and long-term deformity.