

# Congenital Anomalies of the Nose

Jamie L. Funamura, MD<sup>1</sup> Travis T. Tollefson, MD, MPH, FACS<sup>2</sup>

<sup>1</sup>Department of Otolaryngology and Communication Enhancement, Children's Hospital Boston, Boston, Massachusetts

<sup>2</sup>Department of Otolaryngology, University of California, Davis, Sacramento, California

Address for correspondence Travis T. Tollefson, MD, MPH, FACS, Facial Plastic and Reconstructive Surgery, Department of Otolaryngology-Head and Neck Surgery, University of California, Davis, 2521 Stockton Blvd., Suite 7200, Sacramento, CA 95817 (e-mail: tttollefson@ucdavis.edu).

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## Abstract

Congenital anomalies of the nose range from complete aplasia of the nose to duplications and nasal masses. Nasal development is the result of a complex embryologic patterning and fusion of multiple primordial structures. Loss of signaling proteins or failure of migration or proliferation can result in structural anomalies with significant cosmetic and functional consequences. Congenital anomalies of the nose can be categorized into four broad categories: (1) aplastic or hypoplastic, (2) hyperplastic or duplications, (3) clefts, and (4) nasal masses. Our knowledge of the embryologic origin of these anomalies helps dictate subsequent work-up for associated conditions, and the appropriate treatment or surgical approach to manage newborns and children with these anomalies.

## Keywords

- ▶ nasal deformities
- ▶ nasal dermoid
- ▶ Tessier cleft
- ▶ nasal cleft
- ▶ nasal hemangioma

Congenital anomalies of the nose are thought to be relatively rare, affecting approximately 1 in every 20,000 to 40,000 live births.<sup>1</sup> The exact incidence is difficult to quantify, as minor anomalies may be overlooked or left untreated, while experience with more severe presentations tends to cluster at specialized medical centers. Our knowledge of congenital anomalies is largely based on the detailed observation of the vast range of patients and also case reports and small case series. For the individual practitioner confronted with a patient with a rare congenital anomaly of the nose, a basic understanding of nasal embryology, classification system, and associated conditions and clinical considerations will be important to appropriately manage these uncommon conditions.

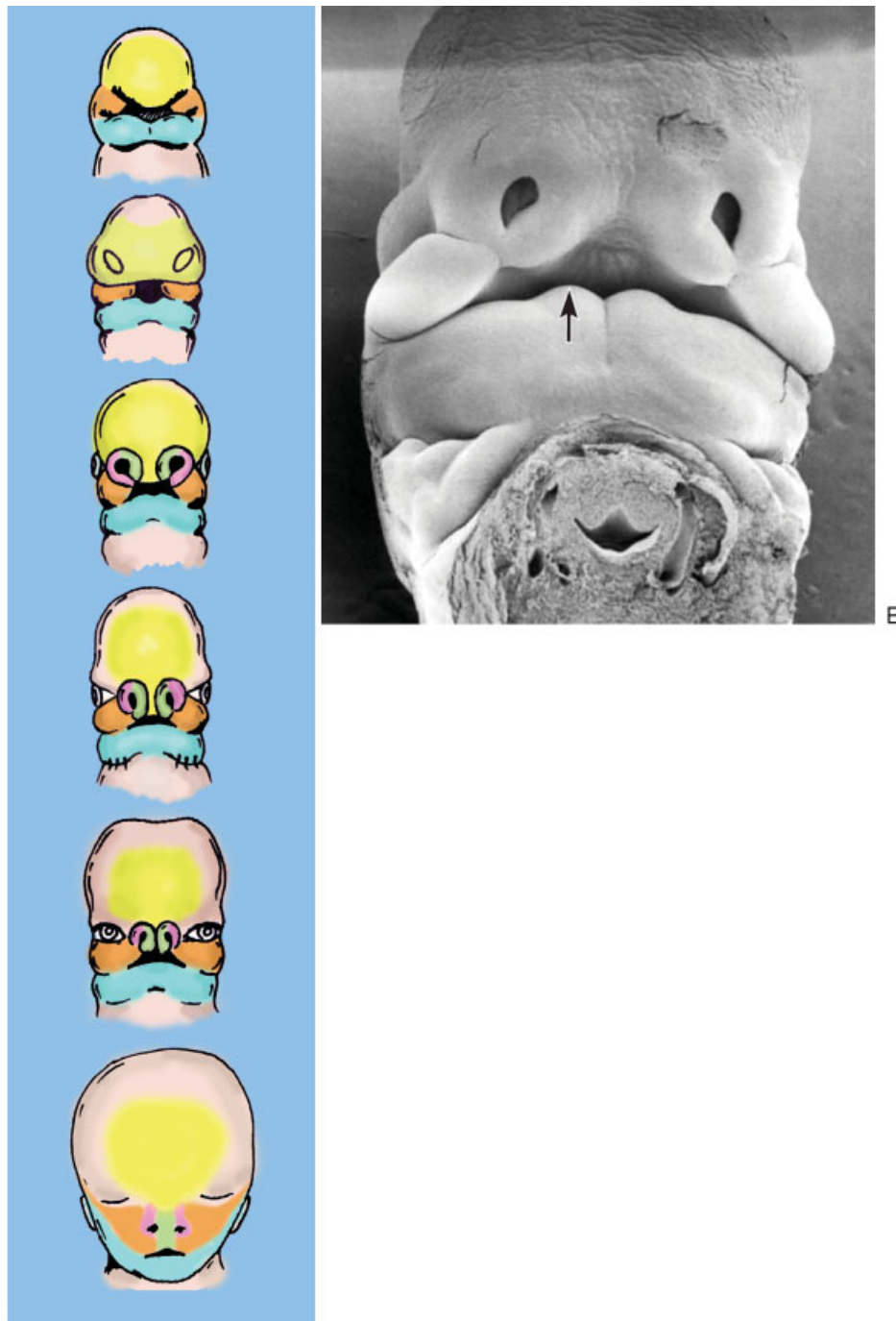
## Embryology of the Nose

Nasal development begins in the fourth week of gestation and is mostly complete by the eighth week. Of the five facial primordia, the frontonasal prominence is the primary structure responsible for nasal development. Neural crest cells migrate into the frontonasal prominence and form the olfactory (nasal) placodes which deepen into nasal pits.<sup>1,2</sup> These nasal pits are surrounded by mesenchymal cells that proliferate, developing into the horseshoe-shaped medial and lateral nasal processes on each

side<sup>1-4</sup> (▶ **Fig. 1A, B**). The medial processes will ultimately fuse, contributing to the nasal septum and the medial crura of the lower lateral cartilages. The lateral processes develop into the nasal bones, upper lateral cartilages, ala, and lateral crura of the lower lateral cartilages. The nasal dorsum and glabella are derived directly from the frontonasal prominence.<sup>3,4</sup> The other four facial primordia—the paired maxillary and mandibular processes of the first branchial arch—will ultimately fuse with the medial and lateral processes, completing facial formation by the 14th week of gestation.<sup>2-4</sup>

## Classification Systems

The most recent and comprehensive classification system for congenital nasal anomalies was developed by Losee and colleagues in 2004.<sup>1</sup> Noting that previous classification systems addressed nasal anomalies in conjunction with craniofacial syndromes, a 22-year retrospective review of craniofacial center patients from the Children's Hospital of Philadelphia was conducted to create a broad classification scheme of congenital nasal deformities based on hypoplasia, hyperplasia, or clefting of nasal structures as well as congenital nasal masses (▶ **Table 1**). In keeping with this broad classification system, the most well-studied congenital nasal



**Fig. 1** (A) Illustrations of the human craniofacial development with embryos shown at 4, 5, 5.5, and 6 weeks and term infant from top to bottom. The major facial prominences are color coded: *blue*, mandibular; *orange*, maxillary; *pink*, lateral nasal; *green*, medial nasal; and *yellow*, frontal. (Artwork courtesy: Amir Rafii, MD.) (B) Scanning electron microscopic view of an embryo early in development of the merging medial nasal prominences. In this *Macaca fascicularis* embryo, all the lateral nasal, medial nasal, maxillary, frontonasal, and mandibular prominences are seen. (Used with permission from Senders CW, Peterson EC, Hendrickx AG, Cukierski MA. Development of the upper lip. Arch Facial Plast Surg 2003;5(1):16–25.)

anomalies will be reviewed here in terms of the absence, excess, clefting, or mass lesions of the nose.

### Aplastic and Hypoplastic Anomalies

Congenital anomalies of the nose in which there is a paucity or underdevelopment of nasal structures ranges from complete aplasia—arhinia—to a subtle hypoplasia of a part of the

nose. Aplastic and hypoplastic anomalies are thought to represent the most common class of congenital nasal anomalies (→ Fig. 2).

Total arhinia or complete nasal agenesis, by definition, is the complete absence of the external nose, nasal cavities, and olfactory apparatus (→ Fig. 3). It is an extremely rare condition and the etiology is unknown, although associated anomalies of chromosome 9, 13, and 21 have been reported. Embryological

**Table 1** Classification scheme for congenital nasal anomalies

	Category	Description
Type I	Hypoplasia and atrophy	Paucity, atrophy, or underdevelopment of skin, subcutaneous tissue, muscle, cartilage, and/or bone
Type II	Hyperplasia and duplications	Excess tissue, ranging from duplication of parts or complete multiples
Type III	Clefts	The Tessier classification of craniofacial clefts is applied
Type IV	Neoplasms and vascular anomalies	Benign and malignant neoplasms, vascular anomalies

Modified from Losee et al.<sup>1</sup>

theories of origin include failure of nasal placode invagination.<sup>1,5</sup> Coexistent midline anomalies such as holoprosencephaly, bilateral choanal atresia, hypotelorism, meningocele, encephalocele, choanal atresia, and orofacial clefting are common and therefore should be evaluated for prior to any elective interventions. Certain coexistent conditions may have a poor life expectancy. For example, lobar holoprosencephaly (the severest presentation of holoprosencephaly) is rarely compatible with life beyond 1 year of age.<sup>5</sup>

Heminasal aplasia, in which unilateral nostril agenesis is present, has been reported in isolation as well in combination with anomalies affecting the ipsilateral face. Radiographic studies have demonstrated an associated absence of the cribriform plate, which is thought to represent a loss of the ipsilateral nasal placode during development.<sup>6</sup>

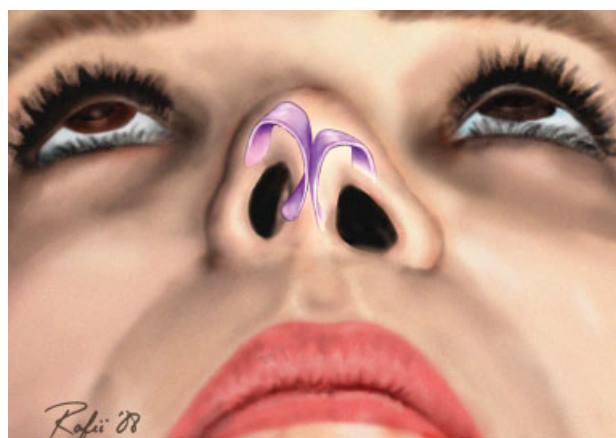
Hypoplasia or absent portions of the nose without associated orofacial clefting are rare. Columellar agenesis (missing medial crura of the lower lateral cartilages and soft-tissue covering), with normal septal development, and complete or partial nasal bone agenesis have been reported.<sup>7-9</sup> It has been suggested that isolated hypoplastic anomalies of the nose may in fact represent a carrier state of orofacial clefting.<sup>10</sup>

Several craniofacial syndromes are known to have associated nasal hypoplasia. Binder syndrome, also called nasomaxillary hypoplasia, is characterized by hypoplasia of the

anterior nasal spine and columella with midface hypoplasia. In Fraser syndrome, an underdeveloped nasal dorsum and hypoplastic nares present with cryptophthalmos.<sup>1</sup> Apert and Crouzon syndromes also present with characteristic midface hypoplasia with retrusion of the nasal dorsum as well as nasal cavity stenosis and maxillary sinus hypoplasia.<sup>11</sup> Unilateral hypoplasia of the nasal ala can be seen with hemifacial microsomia.<sup>1</sup> Bosma arhinia microphthalmia syndrome illustrates the common embryologic origin of nasal, ocular, and pituitary structures, presenting with severe nasal hypoplasia or arhinia, microphthalmia, anosmia, and hypogonadotropic cryptorchidism.<sup>12</sup>

Atresia of the anterior, middle, or posterior nasal cavity can occur. Atresia of the anterior nasal cavity in the form of pyriform aperture stenosis will be discussed here, as it affects the external nasal structure. However, posterior nasal cavity obstruction—choanal atresia—and stenosis of the nasal cavity itself, although rare, should also be considered in the work-up of a newborn presenting with symptoms of nasal obstruction.

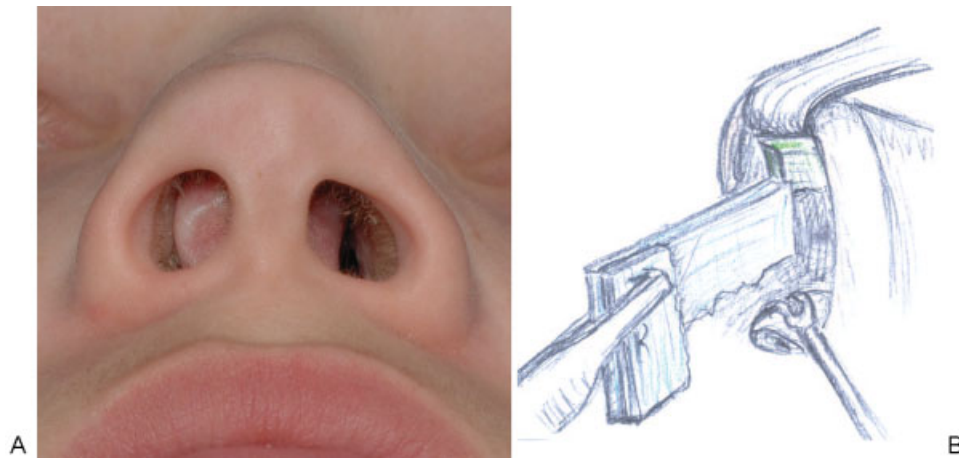
Pyriform aperture stenosis is characterized by a narrowing of the premaxillary pyriform aperture (1–2 mm), resulting in an anterior nasal obstruction. As the pyriform aperture is the narrowest part of the nasal airway in a newborn, this can result in varying degrees of respiratory distress. Associated congenital anomalies include other midline hypoplastic anomalies such as holoprosencephaly, submucous cleft palate, central megaincisor, and hypothalamic-pituitary axis anomalies.<sup>5,13</sup> These associated conditions have led some to suggest that pyriform aperture stenosis is a microform of holoprosencephaly.<sup>14</sup> Mild cases may be managed with nasal hygiene (suctioning, humidification) and topical steroid



**Fig. 2** Illustration of a base view of a patient with a hypoplastic left lower lateral cartilage and dysmorphic nasal ala (without signs of cleft lip). (Used with permission from Tollefson TT, Humphrey CD, Larrabee WF Jr, Adelson RT, Karimi K, Kriet JD. The spectrum of isolated congenital nasal deformities resembling the cleft lip nasal morphology. *Arch Facial Plast Surg* 2011;13(3):152–160.)



**Fig. 3** Child with complete absence of the nose and nasal apertures, termed arhinia.



**Fig. 4** (A) Photograph in base view of congenital nasal septal deviation with tip and lower lateral cartilage asymmetry. (B) illustration of extracorporeal septoplasty approach to reconstruct the L-strut (shown in *blue*) by securing to the "keystone" of the ethmoid perpendicular plate/dorsal quadrangular cartilage (shown in *green*).

drops. Severe cases may require surgical intervention, usually via a sublial approach with a drill-out of the stenosed aperture. Care should be taken to avoid injury to the tooth buds and nasolacrimal duct when drilling inferiorly and posterolaterally, respectively.<sup>5,15</sup>

Congenital deformations of the nose such as neonatal septal deviation or nasal tip deviation have also been reported (→ **Fig. 4A, B**). Although not truly hypoplastic or aplastic congenital anomalies, these nasal deformities may nevertheless have potential cosmetic or functional implications. Increased risk of neonatal septal deviation is noted with prolonged labor, intrauterine pressure, and primiparous delivery. Many of these deformities have been observed to resolve without intervention or late sequelae. The degree of deviation and presence of airway obstruction determines the need for intervention. Often, treatment can be limited to a simple closed reduction shortly after diagnosis if severe, although early hemitransfixion approaches have also been described.<sup>16–18</sup>

### Hyperplastic and Duplication Anomalies

Hyperplasia or duplication anomalies of the nose have been reported ranging from supernumerary nostrils to duplicate columella to true duplication of the entire nose, or polyrhinia (→ **Fig. 5A, B**). It is thought that an accessory olfactory pit or duplication of the olfactory placode is the embryologic origin of these anomalies.<sup>1,2</sup> The lateral nasal process then develops normally but a duplication of the medial nasal process results in polyrhinia. A milder, unilateral version of this process is represented by the supernumerary nostril.<sup>2,5</sup> Of note, duplicate or bifid structures may also represent a midline craniofacial cleft.<sup>1</sup> Important associated anomalies include pyriform aperture stenosis and choanal atresia.<sup>1,17</sup> In general, as hyperplastic and duplication anomalies are often associated with underlying bony anomalies, computed tomography (CT) and/or magnetic resonance imaging (MRI) should be used to assist with timely management of nasal obstruction and for preoperative planning.



**Fig. 5** An infant with right polyrhinia, right cleft lip and alveolus, and intact palate is shown in (A) frontal and (B) base/intraoral views. Frontonasal malformation is noted with hypertelorism and a broad nasal root.

The rare but classically described congenital nasal deformity is proboscis lateralis, which typically presents as a tubular tract emanating from the lateral nose or medial canthus. The structure is characterized by a squamous and ciliated respiratory epithelium lining, and associated with ipsilateral sinonasal hypoplasia or aplasia. The sinonasal hypoplasia may include a heminasal aplasia as well as absent turbinates, nasolacrimal duct, and/or frontal, ethmoid, and maxillary sinuses.<sup>3,17,19</sup> As proboscis lateralis presents with this ipsilateral sinonasal hypoplasia or aplasia, it could be argued that it belongs to the hyperplastic category of congenital anomalies of the nose. The embryologic cause is thought to be a lesion or absence of the olfactory placode early in development, as proboscis is also associated with an absent olfactory nerve, olfactory lobe, and cribriform plate on the affected side. The degree of contribution from the medial or lateral nasal process determines the site of attachment of the proboscis, ranging from midline to a more lateral position at the medial canthus of the eye. Proboscis lateralis can interfere with normal eye and eyelid development. The result can be hypertelorism and/or coloboma of the eyelid, iris, retina, or optic nerve.<sup>3</sup> Oftentimes, the skin and soft tissue of the proboscis can be used in the reconstructive effort in conjunction with bone and cartilage grafts, with possible dacryocystorhinostomy.<sup>1,3,19</sup>

## Nasal Clefts

Clefting of the nose results from the failure of the frontal processes to develop appropriately, and may present as either medial or lateral clefts. Nasal clefts range from the well-known cleft of the nasal floor associated with cleft lip and palate deformities to lateral involvement of the frontal bone or orbit. The most commonly used classification system is that developed by Tessier in the 1970s.<sup>1,20</sup> The Tessier classification system uses the orbit as the primary structure of reference, and localizes clefts of the soft tissue and bone of the face and cranial vault.<sup>20</sup> The craniofacial clefts that apply to the nose are the facial Tessier No. 0, 1, 2, and 3 clefts, and their cranial extensions: Tessier No. 11, 12, 13, and 14.<sup>1</sup>

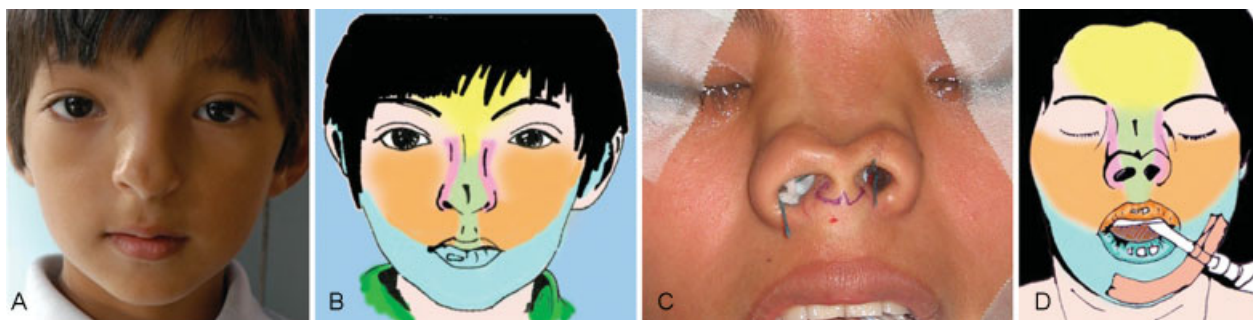
Median facial clefting, also known as frontonasal dysplasia (malformation), represents the Tessier No. 0/14 cleft. The most severe presentation of this cleft is a frontonasal en-

phalocele with hypertelorism and a broad nasal dorsum, with the appearance of failure of closure of the anterior neuropore. The forehead is typically v shaped (→Fig. 6).<sup>1,20</sup> Frontorhiny is thought to be an intermediate presentation, characterized by hypertelorism, a wide nasal dorsum, bifid nasal tip, and broad columella with widely separated narrow nares.<sup>21</sup> At the far end of the spectrum, mild nasal bifidity (see →Fig. 6A) may represent the microform condition of this cleft. Of note, given the association of midline masses and clefting anomalies, a concurrent nasal dermoid cyst or encephalocele must be ruled out in a midline cleft.<sup>5</sup>

The Tessier No. 1/13 cleft involves a soft-tissue cleft of Cupid's bow and the nasal ala, extending through the upper lateral cartilage and medial aspect of the brow, displacing the medial canthus laterally. The bony cleft involves the alveolus, pyriform aperture, nasal bone, and frontal process of the maxilla.<sup>1</sup>

Cleft lip and nasal deformity, the most common and well-defined cleft nasal deformity, is part of the spectrum of Tessier No. 2/12 clefts. A complete Tessier No. 2/12 cleft is a rare occurrence, presenting with a soft-tissue defect of the lip extending to the alar rim, with involvement of the upper lateral cartilage, and cranial extension through the frontal process of the maxilla.<sup>1</sup> Failure of the fusion of the medial and lateral nasal processes with the maxillary process is thought to be the underlying cause of the common cleft lip nasal deformity.<sup>3</sup> Tessier thought it likely that heminasal aplasia, supernumerary nostrils, and proboscis lateralis are part of the spectrum of this type 2 cleft.<sup>20</sup>

The Tessier cleft that involves both the nose and the orbit is the No. 3/11 cleft. The soft-tissue defect of the lip is similar to the common unilateral cleft lip, but with involvement of the medial canthus (a feature not present in the other clefts involving the nose). The bony cleft affects the nasolacrimal system, resulting in duct obstruction and recurrent infections. Tessier No.4 clefts occur in a spectrum of cleft lip, decreased distance between the medial canthus and mouth, orbital dystopia, and possible cleft palate or colobomas of the eyelids (→Fig. 7). Colobomas of the lower lid, medial to the punctum, with microphthalmia or anophthalmia are often common. The medial upper lid, brow, and forehead are also affected. The bony cleft involves the floor of the orbit or results in orbital dystopia.<sup>1</sup> The embryologic origin of these lateral nasal clefts is thought to be disorganized mesenchymal



**Fig. 6** Color-coded illustration of the spectrum of midfacial clefts showing the embryologic derivation of the facial structures. The major facial prominences are color coded as: *blue*, mandibular; *orange*, maxillary; *pink*, lateral nasal; *green*, medial nasal; and *yellow*, frontal. (A) Minimal orbital hypertelorbitism and bifid nasal tip. (B) Color-coded illustration. (C) Intraoperative base view of median nasal cleft with tip and dorsum bifidity. (D) Illustration. (Artwork courtesy: Amir Rafii, MD.)



**Fig. 7** Photograph of child with a right Tessier No. 4 atypical cleft, extending up from the lip, coursing lateral to the alar base and nasolacrimal system, and distorting the medial canthus.

flow between the medial and lateral nasal processes.<sup>5</sup> Tessier observed that this cleft was “the most vicious cleft to repair,” due to the absence of the frontal process of the maxilla, absent septation between the nasal cavity and maxillary sinus, and shortness of the nose.<sup>20</sup>

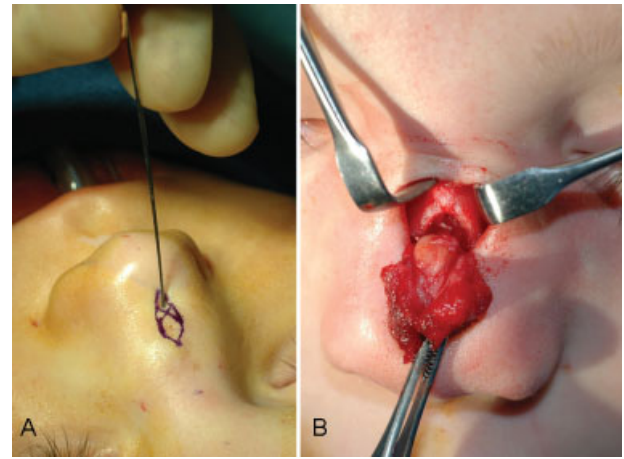
### Congenital Nasal Masses

Congenital nasal masses are rare lesions that can present in the external nose as well as within the nasal cavity, paranasal sinuses, nasopharynx, oral cavity, and orbit.<sup>22</sup> We focus here on the lesions that affect the external nasal structures affecting the form as well as function of the nose. The differential diagnosis of a congenital nasal mass includes encephalocele, meningocele, glioma, dermoid cyst, vascular malformations, and, less commonly, malignant and benign neoplasms.<sup>23</sup>

Dermoid cysts, encephaloceles, and gliomas are the three classically described midline congenital nasal masses. Despite their distinct clinical and pathologic characteristics, they are thought to be embryologically related to developmental anomalies of the frontonasal region. Faulty closure of the anterior neuropore resulting in a persistence of an anterior cranial defect is thought to be the responsible mechanism.<sup>24,25</sup>

#### Nasal Dermoid Cysts and Sinuses

Nasal dermoid lesions may present as cystic masses, sinus tracts, or a combination of the two. Although the most common location is the lower third of the nasal dorsum, dermoid cysts may occur from the glabella to the nasal tip or columella.<sup>24,26</sup> Clinically, nasal dermoid cysts present as firm, slow-growing masses that (1) are noncompressible, (2) do not transilluminate, and (3) often have a nasal dermal pit (→ **Fig. 8A, B**). These masses do not enlarge with crying or straining. Importantly, intracranial extension occurs in 20 to 45% of cases. Imaging characteristics suggestive of intracranial extension include a bifid crista galli and enlargement of the foramen cecum on CT.<sup>27</sup> Given the incomplete ossification of the ethmoid bone and crista galli at the typical age of presentation, CT imaging alone may produce false positives. The presence of these findings on CT has been suggested to be an indication for subsequent MRI, while a normal foramen cecum and crista galli can rule out intracranial



**Fig. 8** Intraoperative photograph of child with a nasal dermoid. (A) Lacrimal probe is inserted into the dermal puncta to identify the tract for elliptical excision and tracing into the nose. Preoperative MRI did not demonstrate a foramen cecum deformity in the skull base. (An open rhinoplasty approach can be added to improve visualization.) (B) Dissection of the dermoid from the underlying nasal bone and upper lateral defect is shown.

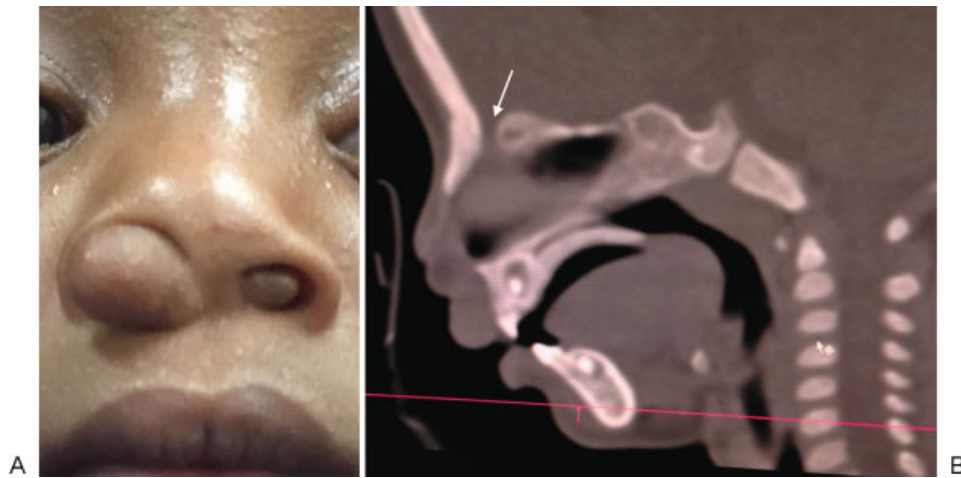
involvement.<sup>28,29</sup> If left untreated, dermoid cysts and sinuses may lead to local inflammation or abscess formation. If an intracranial connection is present, these may ultimately lead to cerebrospinal fluid (CSF) leak, meningitis, and cavernous sinus thrombosis. The growth or presence alone of a dermoid cyst may also cause a cosmetic issue that worsens with time with deformational effect on the nasal bones and/or cartilages.

#### Nasal Encephaloceles

Congenital nasal encephaloceles represent an extracranial herniation of meninges and brain tissue. Encephaloceles are classified by the site of herniation, with sincipital (nasofrontal, nasoethmoidal, and naso-orbital) encephaloceles presenting at the forehead, nasal dorsum, or orbit, respectively. Although the external mass will present at one of the aforementioned locations, the internal skull defect is located in the midline.<sup>22,30</sup> Encephaloceles are soft compressible masses that transilluminate; intranasally, they can be mistaken for polyps (→ **Fig. 9A, B**). Patients with encephaloceles demonstrate a positive *Furstenberg test*, in which the mass enlarges with increased intracranial pressure caused by crying or straining. On MRI, encephaloceles will demonstrate CSF that is in continuity with the intracranial space. Encephaloceles carry the risk of CSF leak, meningitis, and intracranial abscess if left untreated.<sup>22</sup>

#### Nasal Gliomas

Nasal gliomas similarly represent brain tissue that has persisted through an anterior cranial defect; however, unlike encephaloceles, the meningeal connection has been lost. The term glioma implies a true neoplasm and is thus a misnomer; terms such as encephaloma, nasal cerebral heterotopia, or neuroglial heterotopia have been proposed to more accurately reflect the nature of this lesion.<sup>31</sup> Nasal gliomas are typically firm, noncompressible masses, and can present from



**Fig. 9** (A) Encephalocele shown protruding from both nostrils. (B) The sacs are fluid filled and extend up to a skull base defect shown on CT lateral view of the defect (*white arrow*) at the foremen anterior skull base connecting to the midline nasal mass. (Artwork courtesy: Dr. David Shaye.)

the glabella to the nasal tip. An estimated 60% are extranasal, 30% intranasal, and 10% a combination of the two.<sup>22</sup> Intranasal gliomas can be mistaken for nasal polyposis, but are typically less translucent. A pedicle of glial tissue with a dural connection is found in 15 to 20% of cases, but the absence of extracranial continuity of CSF flow should be apparent on MRI.<sup>25</sup> MRI alone is thought to provide sufficient information for preoperative planning for these lesions without the need for concurrent CT imaging.<sup>31</sup> While at a lower risk than encephaloceles for intracranial complications given the lack of meningeal continuity, similar to dermoid cysts, gliomas may become infected and can result in deformation of the septum or nasal bones.

### Vascular Anomalies of the Nose

The most common vascular anomaly of the nose is nasal hemangioma. Infantile hemangioma are designated a benign vascular tumor of endothelial cell origin, which appear during the first several weeks of life. The natural history is then characterized by a rapid proliferation phase during the first year of life followed by a quiescent and then involutonal period (~18 months of age) that can last several years. Classically, treatment of lesion was reserved for lesions that caused functional issues such as nasal obstruction or visual impairment, or lesions that ulcerated and bled. Since the discovery of propranolol as an effective treatment for accelerating involution with rare side effects, medical treatment has become a common practice.<sup>32</sup> Topical  $\beta$ -blockers have also been shown to have good effect with an even lower risk of side effects of treatment. Other treatment options include intralesional steroid injections, pulse dye laser therapy, and surgical excision. The authors, in line with other surgeons, advocate a subunit approach for surgical resection. The paradigm has shifted toward earlier resection (→ **Fig. 10A–E**). Intervention is dictated by the surgeon's experience and the nature of the lesion, although there has been an increasing trend toward

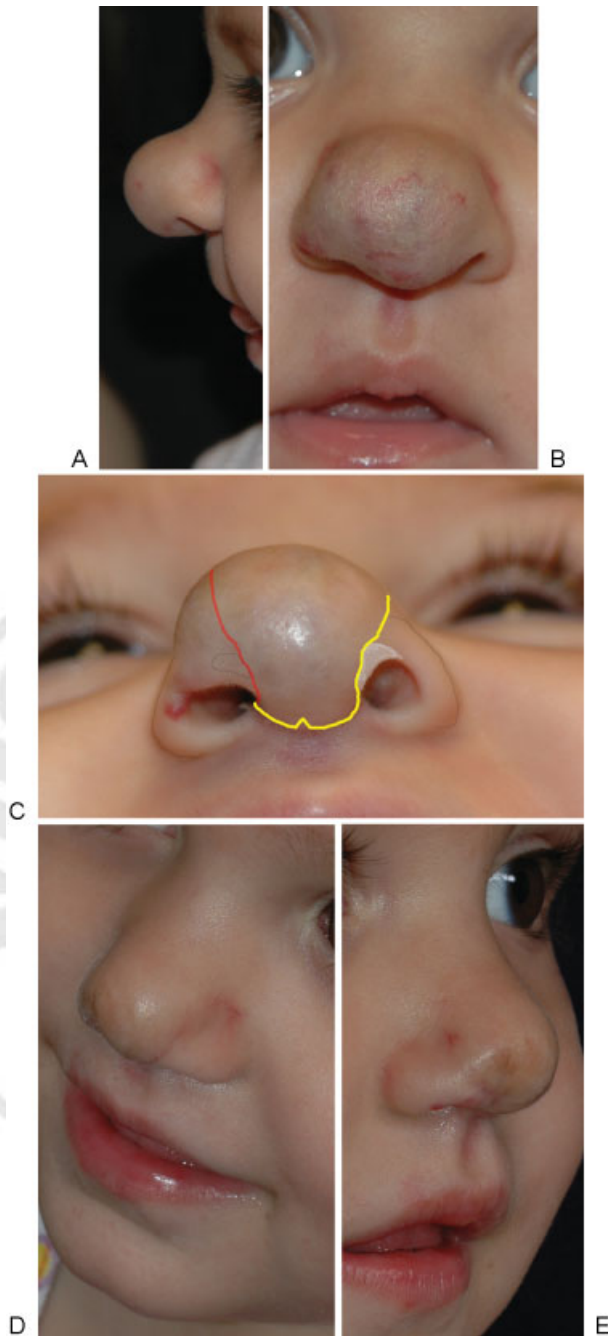
early surgical excision to prevent the deformational effects of the lesion.<sup>33,34</sup>

### Neoplasms

Neoplastic lesions of the nose that have been previously reported include pilomatrixoma, lipoma, neurofibroma, neuroblastoma, rhabdomyosarcoma, and teratoma.<sup>1,35</sup> Of these, teratomas are the most well-described congenital lesions and have the greatest potential for causing life-threatening airway obstruction at birth. The incidence is thought to be 1 in 20,000 to 40,000 of live births. Head and neck teratomas account for only 5% of neonatal teratomas, sacrococcygeal lesions being more common. These neoplasms, composed of tissue from all three germ layers, can often be diagnosed on prenatal ultrasound and confirmed with fetal MRI. Although cervical teratomas are the most common, the nasopharynx is the second most common site. Resection is usually undertaken in the early newborn period after the airway has been secured.<sup>36,37</sup>

### Clinical Considerations

The timing of surgical intervention in patients with congenital anomalies of the nose is largely dependent on the specific pathology. However, there are several unique characteristics of the nose that warrant special consideration. Infants are obligate nasal breathers for at least the first 6 weeks of life and up to the first 6 months.<sup>38</sup> Therefore, any lesion that causes bilateral nasal obstruction or has the potential to if infection were to occur should be closely monitored and consideration given for early surgical intervention. Parents and providers should be alert to signs of respiratory distress as well as difficulty feeding and failure to thrive that may signify concerning nasal obstruction. Mild cases or transient periods of worsening nasal congestion due to upper respiratory infections may be temporized by nasal saline or steroid drops and assiduous nasal hygiene. Of note, newborns with congenital



**Fig. 10** Infant with infantile hemangioma presented at birth with only a small red dot on nose. (A) Lateral and (B) frontal view at 6 months of age. This was prior to the use of propranolol, so steroid injections and pulse dye laser were used until the child was 16 months of age. (C) A subunit approach was designed for resection. (D) Left oblique and (E) right oblique postoperative views. Vascular laser treatment with a 585-nm pulse dye laser treatment was used on the residual hemangiomatous skin.

nasal anomalies may also have coexistent congenital cardiac or neurologic anomalies that can contribute to cyanosis and poor respiratory effort.

Psychosocial reasons for addressing deforming facial lesions have been frequently cited as justification for surgical intervention of nasal deformities in early childhood. The development of self-awareness between the ages of 2 and

3 years and the socially defining time point of school matriculation at the age of 5 years are two important developmental landmarks for young children.<sup>33</sup>

The potential for deformational effect of a nasal mass in the growing infant nose should also be considered. As has been well documented, infants have a greater cartilage-to-bone ratio of the nose than do adults, with the newborn septal cartilage extending from the nasal tip to the skull base. A young child has an absent perpendicular plate and rudimentary vomer, with gradual ossification of the cartilaginous septum and regression of the upper lateral cartilages with age. The perpendicular plate, which merges with the vomer, is thought to be fully formed between 6 and 8 years of age.<sup>39,40</sup> Clinical evidence of traumatic inhibition of the development of the nasal skeleton and maxilla has been demonstrated in comparative observational studies of monozygotic twins.<sup>41,42</sup> Studies have also shown two specific windows of accelerated growth of the nose: the first 2 years of life and during puberty.<sup>43</sup> A significant deformational effect can therefore result from a delay in addressing a congenital nasal mass that displaces nasal structures. Conversely, a cosmetic result that may appear acceptable in early childhood could drastically change during adolescence. These clinical observations on the patterns of growth of the nose form the basis of delaying definitive rhinoplasty until after puberty.

## Conclusion

Congenital anomalies of the nose are rare occurrences that can be divided into four broad categories. The first, ranging from hypoplasia of parts to complete aplasia of the nose, may present with associated oculocephalic deformities. Complete radiographic work-up and management of any neonatal respiratory distress that may result from absent or hypoplastic nasal structures should be performed expeditiously. The second category, hyperplasia or duplication anomalies, can range from a supernumerary nostril to a complete nasal duplication. Underlying bony anomalies are prevalent as well, with associated pyriform aperture stenosis, choanal atresia, or clefting that warrant imaging prior to reconstructive efforts. The third category, nasal clefts, is thought to belong to the spectrum of craniofacial clefts as classified by Tessier. Isolated nasal clefts are extremely rare, and thought to be microform presentations of the broader craniofacial clefts. Nasal masses comprise the fourth category of congenital nasal anomalies. Dermoid cyst and sinuses, gliomas, and encephaloceles are the classic nasal masses that result from similar midline fusion anomalies during embryogenesis. These typically require early surgical management to prevent infectious and deformational complications. Management of vascular malformations such as hemangioma is more lesion-specific due to the natural history of proliferation and involution of this entity. Of significance, newborns are obligate nasal breathers for up to the first 6 months of life, and also undergo a significant period of nasal growth within the first 2 years. This is in addition to the development of self-awareness and social interactions that might impact a child with a nasal deformity during the early school-aged years. Timing of surgical intervention, therefore, is a balance of



functional, cosmetic, and psychosocial considerations, which is often further complicated by associated conditions that often present with congenital anomalies of the nose.

## References

- Losee JE, Kirschner RE, Whitaker LA, Bartlett SP. Congenital nasal anomalies: a classification scheme. *Plast Reconstr Surg* 2004; 113(2):676–689
- Williams A, Pizzuto M, Brodsky L, Perry R. Supernumerary nostril: a rare congenital deformity. *Int J Pediatr Otorhinolaryngol* 1998; 44(2):161–167
- Rontal M, Duritz G. Proboscis lateralis: case report and embryologic analysis. *Laryngoscope* 1977;87(6):996–1006
- Sadler TW. Head and neck. In: Langman's Medical Embryology. 11th ed. Philadelphia, PA: Lippincott, Williams & Wilkins; 2009: 265–291
- Otteson TD. Congenital malformations of the nose and paranasal sinuses. In: Bluestone CD, Simons JP, Healy GB, eds. *Bluestone and Stool's Pediatric Otolaryngology*. Vol. 1. Shelton, CT: PMPH; 2014: 1017–1035
- van Kempen AA, Nabben FA, Hamel BC. Heminasal aplasia: a case report and review of the literature of the last 25 years. *Clin Dysmorphol* 1997;6(2):147–152
- Lewin ML. Congenital absence of the nasal columella. *Cleft Palate J* 1988;25(1):58–63
- Guerrissi JO. Congenital absence of nasal bones. *Ann Plast Surg* 1993;30(3):260–263
- Manning KP, Singh SD. Hypoplasia of the nasal bones. *J Laryngol Otol* 1977;91(12):1085–1091
- Tollefson TT, Humphrey CD, Larrabee WF Jr, Adelson RT, Karimi K, Kriet JD. The spectrum of isolated congenital nasal deformities resembling the cleft lip nasal morphology. *Arch Facial Plast Surg* 2011;13(3):152–160
- Ginat DT, Robson CD. CT and MRI of congenital nasal lesions in syndromic conditions. *Pediatr Radiol* 2015;45(7):1056–1065
- Graham JM Jr, Lee J. Bosma arhinia microphthalmia syndrome. *Am J Med Genet A* 2006;140(2):189–193
- Shikowitz MJ. Congenital nasal pyriform aperture stenosis: diagnosis and treatment. *Int J Pediatr Otorhinolaryngol* 2003;67(6):635–639
- Johnson PJ, Rydlund K, Hollins RR. Congenital nasal pyriform aperture stenosis. *Plast Reconstr Surg* 1999;103(6):1696–1699
- Visvanathan V, Wynne DM. Congenital nasal pyriform aperture stenosis: a report of 10 cases and literature review. *Int J Pediatr Otorhinolaryngol* 2012;76(1):28–30
- Podoshin L, Gertner R, Fradis M, Berger A. Incidence and treatment of deviation of nasal septum in newborns. *Ear Nose Throat J* 1991; 70(8):485–487
- Leung MK, Krakovitz PR, Koltai PJ. Congenital sinonasal disorders. In: Kennedy DW, Hwang PH, eds. *Rhinology: Diseases of the Nose, Sinuses, and Skull Base*. New York, NY: Thieme; 2012:381–393
- Emami AJ, Brodsky L, Pizzuto M. Neonatal septoplasty: case report and review of the literature. *Int J Pediatr Otorhinolaryngol* 1996; 35(3):271–275
- Abou-Elhamd KE. Proboscis lateralis: a report of two cases. *Int J Pediatr Otorhinolaryngol* 2004;68(4):503–505
- Tessier P. Anatomical classification facial, cranio-facial and latero-facial clefts. *J Maxillofac Surg* 1976;4(2):69–92
- Pham NS, Rafii A, Liu J, Boyadjiev SA, Tollefson TT. Clinical and genetic characterization of frontorhiny: report of 3 novel cases and discussion of the surgical management. *Arch Facial Plast Surg* 2011;13(6):415–420
- Rahbar R, Resto VA, Robson CD, et al. Nasal glioma and encephalocele: diagnosis and management. *Laryngoscope* 2003;113(12): 2069–2077
- Bradley PJ, Singh SD. Congenital nasal masses: diagnosis and management. *Clin Otolaryngol Allied Sci* 1982;7(2):87–97
- Hughes GB, Sharpino G, Hunt W, Tucker HM. Management of the congenital midline nasal mass: a review. *Head Neck Surg* 1980; 2(3):222–233
- Hedlund G. Congenital frontonasal masses: developmental anatomy, malformations, and MR imaging. *Pediatr Radiol* 2006;36(7): 647–662, quiz 726–727
- Pratt LW. Midline cysts of nasal dorsum: embryologic origin and treatment. *Laryngoscope* 1965;75:968–980
- Posnick JC, Bortoluzzi P, Armstrong DC, Drake JM. Intracranial nasal dermoid sinus cysts: computed tomographic scan findings and surgical results. *Plast Reconstr Surg* 1994;93(4):745–754, discussion 755–756
- Pensler JM, Bauer BS, Naidich TP. Craniofacial dermoids. *Plast Reconstr Surg* 1988;82(6):953–958
- Rahbar R, Shah P, Mulliken JB, et al. The presentation and management of nasal dermoid: a 30-year experience. *Arch Otolaryngol Head Neck Surg* 2003;129(4):464–471
- Hoving EW. Nasal encephaloceles. *Childs Nerv Syst* 2000; 16(10–11):702–706
- Adil E, Robson C, Perez-Atayde A, et al. Congenital nasal neuroglial heterotopia and encephaloceles: an update on current evaluation and management. *Laryngoscope* 2016 (e-pub ahead of print). doi:10.1002/lary.25864
- Hogeling M, Adams S, Wargon O. A randomized controlled trial of propranolol for infantile hemangiomas. *Pediatrics* 2011;128(2): e259–e266
- Hochman M, Mascareno A. Management of nasal hemangiomas. *Arch Facial Plast Surg* 2005;7(5):295–300
- McCarthy JG, Borud LJ, Schreiber JS. Hemangiomas of the nasal tip. *Plast Reconstr Surg* 2002;109(1):31–40
- Zeitouni AG, Shapiro RS. Congenital anomalies of the nose and anterior skull base. In: Tewfik TL, Der Kaloustian VM, eds. *Congenital Anomalies of the Ear, Nose, and Throat*. New York, NY: Oxford University Press; 1997:189–200
- April MM, Ward RF, Garelick JM. Diagnosis, management, and follow-up of congenital head and neck teratomas. *Laryngoscope* 1998;108(9):1398–1401
- Azizkhan RG, Haase GM, Applebaum H, et al. Diagnosis, management, and outcome of cervicofacial teratomas in neonates: a Childrens Cancer Group study. *J Pediatr Surg* 1995;30(2): 312–316
- Valdez TA, Ainsworth T. Neonatal nasal obstruction. In: Schoem SR, Darrow DH, eds. *Pediatric Otolaryngology*. Elk Grove Village, IL: American Academy of Pediatrics; 2012:109–123
- van Loosen J, Verwoerd-Verhoef HL, Verwoerd CD. The nasal septal cartilage in the newborn. *Rhinology* 1988;26(3):161–165
- Poublon RM, Verwoerd CD, Verwoerd-Verhoef HL. Anatomy of the upper lateral cartilages in the human newborn. *Rhinology* 1990; 28(1):41–45
- Grymer LF, Bosch C. The nasal septum and the development of the midface. A longitudinal study of a pair of monozygotic twins. *Rhinology* 1997;35(1):6–10
- Grymer LF, Pallisgaard C, Melsen B. The nasal septum in relation to the development of the nasomaxillary complex: a study in identical twins. *Laryngoscope* 1991;101(8):863–868
- Nolst Trenité GJ. Rhinoplasty in children. In: Papel ID, ed. *Facial Plastic and Reconstructive Surgery*. 3rd ed. New York, NY: Thieme; 2009:605–617