Chapter 39: Congenital Anomalies of the Nose

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The human nose has evolved as an organ that maintains a constant, direct contact with the environment. A person's future nose is visible as a midfacial unit as 3.5 to 4 weeks into gestation, when interaction occurs between it and the other facial components. A teratogenic influence may occur anytime up to birth of the child; the nose is therefore at risk of alteration in its normal development for much of the individual's developing life and is susceptible to involvement in many other facial anomalies. The reader is referred to embryologic tests (see the Suggested Readings section, which closes this chapter) and to Chapter 18 for a detailed examination of the timing of these influences.

Nasal anomalies may arise as primary embryologic defects, which in some cases involve environmental expressions (for instance, fetal alcohol syndrome), and also as secondary to defects in other facial units (such as cleft lip and palate). In addition a wide range of anomalies occurs from the rare to the very common; in some cases, the clinical presentation and management of nasal pathologies overlap.

Using a team that includes an otolaryngologist and individuals with craniofacial surgery expertise is often the best approach in complex cases.

Uncommon Nasal Anomaly

Many rare nasal anomalies are associated with distinct genetic syndromes, and some follow known exposures of the fetus to teratogenic drugs or environmental conditions at critical times. The result is a failure or delay in mesodermal reinforcement of tissues derived from the neural crest. Multiple migrational pathways are possible for this process, but a preordained pattern must occur if an anomaly is to be prevented.

Arrhinia (nasal aplasia)

Arrhinia, the complete absence of the external nose and nasal airway, may occur as an isolated defect or with other anomalies. A unilateral form has been reported. In all reported cases, the nasal chambers are incomplete or absent, and the palate is highly arched and hypoplastic. The facial appearance is varied although usually distinctive, consisting of a "dish-face" deformity with hypertelorism or hypotelorism and normal labial development (often including normal philtral columns). Anophthalmia or hypoplasia of the eye and orbits are common (Fig. 39-1).

The initial management of arrhinia is the same as for choanal atresia: one awaits acquisition of reflex mouth breathing and the coordination of swallowing and breathing during feeding. In two reported cases (Gifford et al, 1972) and one case I have seen, satisfactory coordination was possible within 2 to 3 days of life. Reconstructive attempts (if needed) are best deferred until midfacial development progresses (at 5 to 6 years of age), when mucosal and skin-

lined flaps from external and transpalatal areas can be rotated. A long-term approach to correcting this deformity is usually required. A nasal prosthesis without creation of a nasal passage is recommended (Fig. 39-2).

Proboscis deformity

In proboscis deformity, the medial and lateral processes and the globular processes are absent. The maxillary process on the affected side fuses with the opposite nasal and globular process, creating nasal closure with absence of the nasal cavity, choanae, and ethmoid and nasal bones. A trunklike appendage covered in skin and dimpled on the outer end droops from the medial canthus area. A bilateral expression of this anomaly is reported (Hengerer and Oas, 1980). Nose anomalies may be associated with central nervous system (CNS) anomalies.

Excision of the appendage improves facial appearance. Construction of nasal chambers can be performed using a transpalatal and external combined approach, although long-term stenting is often required and prosthetic restoration may be a suitable alternative.

Polyrrhinia

Polyrrhinia, or double nose, has been reported three times as an isolated defect (Ghosh et al, 1971). Hypertelorism is usually present. A combined craniofacial-neurosurgical-otolaryngological team approach is best. A median excision of the nasal duplication is deferred until the associated choanal atresia or stenosis is corrected by a transpalatal approach. Following this, a craniofacial restoration may be performed.

Median nasal cleft

Median nasal clefts very from a simple furrow on the nasal dorsum usually associated with some widening of the nasal bones to an appearance similar to polyrrhinia. The incidence of other associated regional defects increases with the severity of deformity. These are hypertension, facial cleft, coloboma of the lower eyelid, iris, or retina, encephalocoele, and choanal atresia.

Age may improve the facial appearance by soft tissue growth in patients with minor furrows; these, therefore should be watched. Severe deformities require surgical assistance, the timing of which needs to be coordinated with a craniofacial group and with the emotional development of the child.

Computed tomographic (CT) scanning may be required, because a coincidental dermoid or encephalocele in the median septum is distinctly possible.

Lateral nasal cleft

A lateral nasal cleft is a rare anomaly, varying from a notching of the lateral ala nasi to a severe lateral nasofacial cleft with an open defect extending through to the lacrimal system and medial canthus of the eye.

Minor defects can be managed by local flaps or composite free grafts (usually from the auricle). Major defects have a high incidence of other anomalies, particularly in the craniofacial and cardiac areas. A team approach that includes the parents is advised for this type of patient.

The frontonasal syndrome combines some aspects of medial and lateral nasal clefts. Usually lateral alar notching and deformity of the nasal cartilage and nasal bones occurs on the affected side. Hypertelorism and frontal bone defects of the skull also often exist. The incidence of anterior encephalocele, hemangioma of the septum, and cerebral tumor is high in these patients.

A craniofacial group approach is best, using rotational-flap techniques and cranial adjustment.

Common Congenital Anomalies of the Nose

Cleft lip and palate

The nasal deformity associated with a cleft of lip is present to some degree in all cleft-lip patients, even those with an incomplete cleft. The more severe the unilateral cleft, the more severe is the nasal deformity. A bilateral lip cleft has a double-sided nasal deformity, reflecting the degree of anomaly present on each side (Fig. 39-3).

The nasal deformity of a unilateral cleft lip is characterized by the following:

1. Deviation of the caudal septum and nasal spine to the noncleft side by the unrestricted muscular pull of the normal side.

2. Displacement of the alar base in three directions: laterally by the free drift of the ala and unopposed pull of lateral facial muscles; inferiorly by the same and by the circumoral muscles; and retrodisplacement by the relative hypoplasia of the maxilla on that side.

3. The nasal dome is lower on the cleft side, and the ala is flattened, which produces horizontal nares.

4. The medial crus of the lower lateral cartilage is longer, flattened, and pulled out over an increased distance. This thinning produces physiologic weakness of the cartilage, and, together with adherent vestibular skin, the cartilage is drawn into a hooded S-shaped fold (Fig. 39-4).

5. In a complete cleft, the nasal floor is absent, and in an incomplete cleft it may be notched.

6. The nasal bone on the cleft side is affected by growth and muscular pull. The tendency is for an unrestricted noncleft pull to deviate the cleft side medially and to orient that bone more vertically.

The deformity of a bilateral cleft simply duplicates these deformities and, in addition, adds the effect of an unstable, possibly overprotuberant premaxilla and accentuates the lack of columella and definitive philtral columns.

The deformity in a cleft nose continues as a dynamic anomaly altered by nasofacial growth. Theoretically, therefore, the deformity is best corrected once growth ceases (at 14 in females to 18 years of age in males). One must, however, balance the impact of facial appearance on the emotional development of that individual child; a team approach including a psychologist is thus worthwhile. The team should also include an orthodontist who can advise as to the dentofacial relationship, particularly anteriorly in the maxilla, because the maxilla is the foundation of the nasal pyramid.

Since children are subject to peer pressure and the indirect pressures of parents who desire that their deformed child be made as normal as possible immediately, the best approach when the deformity is severe may be to correct the lower nasal deformity (tip and septum) at around 6 to 8 years of age, leaving the bony and upper lateral cartilage deformity until the teenage years approach.

The best approach for a cleft nose deformity is to incorporate in the lip repair *some* correction of the nasal deformity, while avoiding excessive dissection, which is likely to create scars and may even restrict nasal development (as does dissection over the face of the maxilla or excessive dissection of the lower lateral cartilages).

The correction of the deformity associated with cleft lip and palate is considered in four parts as follows, proceeding stepwise with age:

1. Correct the nasal deformity as a part of the lip repair without lower lateral cartilage dissection or incision (soft tissue dissection to allow a dome to realign is acceptable). For example, the results of the Millard-type lip repair speak well for this conservatism by lengthening the columella and placing the alar base in an anatomically correct position in relation to lateral and inferior misplacement.

2. Create a stable, symmetric nasal base by aligning the alveolar segments and sometimes by the use of onlay grafts placed on the maxilla beneath the alar base to correct the third mismatch of alar position (hypoplasia of the maxilla); then, "minimally" correct (with cartilage preservation) septal deflections only when severe nasal obstruction exists.

3. Correct the lower lateral cartilage and columellar deformity, including the septal deflection, usually with an external rhinoplastic approach. This correction allows excellent symmetric positioning of the lower lateral cartilages and the creation of equal domes together with columella lengthening by V-Y advancement. Occasionally, this step can be combined with the final step.

4. Create a symmetric skeleton with lateral and medial osteotomies, often combined with labial scar modification, sometimes with dermabrasion of scars.

In general no step should be missed, although steps may be combined or additional modifications incorporated. (For example, during external rhinoplastic correction of the lower lateral deformity, Wehr's incisions and replacement of the ala base may be required.)

Nasal dermoid cysts

Although for terminologic reasons, nasal dermoid cysts include epidermal inclusion cysts with only epidermal elements, the true dermoid cyst or dermoid sinus contains skin appendages - hair follicles, sweat glands, and sebaceous glands. There is an important difference between a dermoid cyst and a dermoid sinus with a cyst. A true dermoid cyst may occur alone subcutaneously on the nasal dorsum superficial to the nasal bones without a cutaneous opening. Simple dermoid cysts therefore appear as slowly enlarging masses under the skin, which over time may simply deform the underlying structures. Histologically and surgically this lesion is easily removed by excision.

The dermoid sinus with or without a cyst is an extensive lesion extending into nasal cartilage and bone, usually passing deeply at the level of the osteocartilaginous junction on the dorsum of the nose.

Dermoid cysts with sinus are probably more common than the approximately 150 reported cases would indicate, and as a measure of anatomic prevalence, nasal presentations occupy about 13% of dermoid sinuses with cysts in the head and neck. The floor of the mouth, submental, and submandibular areas contribute a further 24%, and 50% of dermoid cysts with sinuses occur in the orbital area.

Their creation is an embryologic fault best delineated in detail by embryologic tests. Between the developing nasal bones and cartilaginous capsule behind, a potential space - the prenasal space - exists. A normal herniation of dura passes through a gap in the vault - the foramen cecum. The herniation is continuous with connective tissue, which forms the inner periosteal lining of the nasal bones, and this skin maintains a connection to underlying fibrous tissue or the cartilaginous nasal capsule. Epithelial elements may be pulled inward to form a sinus, which usually has hair protruding from the opening. The sinus may adhere to the dura by its extension through the foramen cecum. The sinus may also extend through the gap between the frontal bones superiorly and the nasal bones inferiorly (the so-called fonticulus nasofrontalis), appearing as a bulging mass at the glabella.

The following variant theory has been proposed to explain the clinical variant in which the dermoid is encountered deep in the septum of the nose or in the pericranial region, often without fistulous connection. The midlayer of the fetal trilaminar septum is a ventral extension of the dura (and therefore and ectodermal derivative). As the posterior septum chondrifies and then ossifies, this layer degenerates; a failure of degeneration would leave ectodermal elements to develop a potential dermoid cyst.

Incidence and presentation

Dermoid sinuses and cysts are slightly more prevalent in males than in females, and familiar cases occasionally have been reported.

The dermoid cyst with sinus is usually visible at birth. The opening is usually at the osteocartilaginous junction of the dorsum. The occasional discharge of purulent material and the presence of a group of hairs at the opening are diagnostic.

Although a sinus without cyst can occur, the tract is often narrowed, suggesting cystic formation proximal to the narrowing. As already explained briefly by embryologic theory, these lesions may extend deeply to the basisphenoid and into the septum or beyond the cribriform plate to the dura.

Deep extension may be predicted when the nasal bones are splayed or true hypertelorism is present. (Deep extension beyond the nasal bones reportedly occurs in 45% of cases.)

Investigation

Radiologic evaluation by standard sinus films or tomographic methods supplemented occasionally by dye contrast fistulography has been preempted by CT scanning, particularly in infants. Standard radiographic methods are reported in one series as having failed to show deep cribriform involvement in two cases (Weisman and Johnson, 1964).

The major CT findings include fusiform enlargement of the septum, a bifid appearance of the septum, widening or erosion of the nasal vault, destructive changes in the glabella or nasal bones, or a defect in the cribriform area. All of these help to distinguish nasal dermoid cysts. Unfortunately, a wide fusiform septum superiorly and broad nasal vault may also be normal findings in infants.

Management

Complete nasal excision is required for cure. Although cauterization, injection of sclerosing agents, and limited dissections have been tried, a high recurrence rate resulted. Preoperative injection of methylene blue into the sinus is an aid.

Occasionally, a secondary infection is the presenting problem. An incision and drainage with antimicrobial treatment resolves the problem acutely, but a fistulous tract with drainage usually persists and requires removal.

Although a horizontal elliptic incision for removal of the sinus opening is cosmetically desirable, a wide exposure with medial nasal osteotomies may be required, and a Y-incision with the vertical limb midline down the nasal dorsum may be needed. An alternative is an H type of incision with both vertical limbs as the incision sites for an external ethmoidectomy. At the point where a dural connection is expected, a combined subfrontal otolaryngologic-neurosurgical approach is advisable.

The nasal deformity caused by the lesion or the extensive surgical approaches may require secondary correction, although incorporating some cosmetic correction is usually possible at the time of the primary excision.

The timing of removal has been debated; one must weigh the risk of infection or infectious complications with the progressive deformity associated with waiting. Growth inhibition within the anterior facial skeleton can occur from extensive dissection, resulting in trauma to growth centers. However, with conservative dissection the former inhibitions about surgical incisions in the bones of the midface have to a great extent been dispelled by the experience of the craniofacial surgery currently undertaken in very small infants.

Nasal gliomas

Nasal gliomas and encephaloceles are rare lesions and are usually described together because of their similar appearance and embryogenesis. Gliomas are deposits of glial tissue in an extradural site, which have not maintained an attachment to the central nervous system or subarachnoid space. Approximately 15% of gliomas have a fibrous connection to the subarachnoid space but no cerebrospinal fluid connection. These are technically gliomas but may have originally been encephaloceles.

Embryogenesis

The following three theories of glioma embryogenesis have been proposed.

1. Gliomas are heterotopic nervous tissue from the olfactory apparatus; the tissue is cut off from the olfactory bulb as the cribriform plate closes or as the frontal and ethmoid bones fuse.

2. Gliomas are developed from neuroglial tissue within the nasal mucosa associated with the olfactory area.

3. The blastomatous theory maintains that a cell designated to become neural tissue is displaced and becomes ectopic neural tissue.

The intermediate lesions, which maintain a fibrous connection to the central nervous system, may be indicative of a common development etiology for glioma and encephalocele.

Clinical presentation. Gliomas contain a mixed stroma of fibrous and neural elements with no true capsule. Mitosis is rarely noted. No metastasis or aggressive local infiltration is recorded, although local recurrence is reported, usually attributed to incomplete local excision. Gliomas are therefore not true neoplasms; the term itself is a misnomer. *Glial heterotopia* would perhaps be preferable. Most are noticed at birth or early childhood. No sex predilection exists, and heredity does not appear to be a factor. About 60% of gliomas are extranasal, 30% are intranasal, and 10% are a combination.

Extranasal gliomas. Extranasal gliomas are smooth, firm noncompressible masses that usually occur along the nasomaxillary suture or near the glabella, but are occasionally found in the midline. The overlying skin may be discolored, telangiectatic, or violaceous, but it is not tethered to the mass. If the lacrimal apparatus is compressed, epiphora may exist.

Intranasal gliomas. Intranasal gliomas create a nasal obstruction, sometimes manifested as difficulty with feeding in infants (as commonly noted with choanal atresia). Large gliomas create septal distortion or may protrude from the anterior or posterior nares like a choanal polyp. They appear as pink-red polypoid masses that are firm and noncompressible and do not distend with crying or jugular vein compression (Furstenberg's sign). Widening of the nasal bony skeleton and hypertelorism is possible with large gliomas. Occasionally, combined dumbbell-shaped masses are encountered with both external and internal components.

Dural connection. Overall, approximately 15% of gliomas have fibroglial connections to the dura, with external gliomas less frequently connected than internal gliomas (9% versus 35%). The dural connection in extranasal gliomas is located close to the nasofrontal suture, whereas in intranasal gliomas, the connection is through a cribriform plate defect. Rarely, a spontaneous cerebrospinal fluid (CSF) leak (rhinorrhea) or meningitis is the presenting sign.

Differential diagnosis. The most common lesion to differentiate from a glioma or encephalocele is a dermoid cyst. It is of particular importance to recognize the potential for a central nervous system connection and to carefully search for evidence of such a connection before initiating treatment.

An extranasal glioma and an encephalocele are rarely confused. Encephaloceles are soft and compressible, and a bony defect is always present on radiographs. Extranasal gliomas rarely show bony dehiscence and are firm and non-compressible. Should bony dehiscence exist when a glioma is suspected, an approach like that for an encephalocele is prudent.

An intranasal glioma and an encephalocele are more easily confused. Both may appear as polypoid masses; nevertheless the same distinctions of softness, compressibility, and enlargement on crying apply to these encephaloceles. Intranasal encephaloceles usually protrude through a bony defect in the cribriform area, whereas this protrusion is less common in intranasal

gliomas. Needle aspiration of the mass has been suggested to be useful, but false positive results have been reported for cystic gliomas, and a risk of introducing infection into the subarachnoid space exists. Preoperative biopsy is not recommended for similar reasons.

Plain radiographic films in three planes of the anterior skull (posteroanterior, lateral, base), along with either polytomography or CT scanning, are therefore recommended for both gliomas *and* encephaloceles.

Management

Extranasal gliomas. For external gliomas an approach identical to that for a dermoid cyst with a complete excision is appropriate: a horizontal ellipse or a Y incision may be needed, occasionally with an H type of incision over the nasal dorsum. If a small unanticipated CSF leak is encountered, a subfrontal transethmoid approach may be used to repair the defect; the incision is extended into a Sewall incision on the side of the leak. If profuse leakage is encountered, a bifrontal craniotomy approach may be needed.

Intranasal gliomas. Some authors have advocated routine craniotomy to sever any connection between the intranasal mass and the subarachnoid space. This approach may occur via a craniotomy or an external ethmoidectomy. Since the bifrontal approach itself carries a significant morbidity, it would seem unreasonable to advocate such an affront routinely; and individualization should occur. Certainly a combined neurosurgical-otolaryngologic opinion should be sought.

A primary exploratory craniotomy should be reserved for patients with (1) a history of meningitis, (2) a compressible nasal mass, (3) a positive Furstenberg sign, (4) radiographic evidence of an osseous defect, or (5) CSF rhinorrhea. The craniotomy can, with good neurosurgical support, be performed synchronously with excision of the intranasal mass through an extended lateral rhinotomy approach, exposing the cribriform plate and controlling any moderate CSF leak with a facial graft slid in from below (extradurally) and supported with both a septal flap swung medially and intranasal packing.

Encephaloceles

Encephaloceles maintain a connection to the central nervous system but are nevertheless histologically identical to gliomas.

Embryology. An encephalocele is an extracranial herniation of the meninges and brain, so a cranial defect is always present. Technically meningoceles are identical, except that no brain tissue is herniated. A meningohydroencephalocele contains a communication with the ventricular system. In common usage, these terms are often used interchangeably: *encephalocele* is used to encompass all of the following.

An encephalocele may be congenital or acquired in trauma. Some congenital encephaloceles are secondary to a generalized dysplastic process or mesodermal migrational failure, for instance, neurofibromas and Ehlers-Danlos syndrome.

The following three main theories of encephalocele embryogenesis exist:

➤ In the first theory, the mechanical origin theory, an incomplete separation of epithelial tissue occurs in the anterior neuropore as the neural tube closes. This error leads to the attaching of brain tissue to skin, and as mesodermal elements attempt to migrate inward to produce bone, a mechanical block exists. A bony defect is produced, and brain appears through the defect. The anatomic site of the anterior neuropore evolves to become the frontoethmoid junction and the posterior neuropore becomes the lumbosacral area. Encephaloceles certainly occur in these areas, but this theory unfortunately does not adequately explain occipital encephaloceles.

➤ In a second theory, a primary defect of mesodermal reinforcement migration occurs (too little or too late), allowing herniation of brain through the defect. Encephaloceles are known to be associated with neurofibromatosis and Ehlers-Danlos syndrome, which seem to support this theory.

➤ The third theory postulates that the anterior cranial vault is derived from cells of neural crest origin. Since migration of these crest cells occurs predominantly anteriorly, a failure of migration into the dorsal head region leaves it devoid of mesodermal elements. The anterior and occipital areas of the skull base require the farthest migration of mesodermal cells, so any delay in migration would result in both anterior and occipital encephaloceles. The association of encephaloceles with other midline fusion defects, for instance, cleft lip or palate, is common and supports this theory. Partial closure or late closure of the defect could theoretically pinch off neural tissue, creating a glioma.

Epidemiology and incidence. Lumbosacral spina bifida is five times more common than a cranial encephalocele is; the latter is an uncommon occurrence. Most encephaloceles occur in the occiput (75%), and a further 10% occur in the parietal areas. Only 15% occur in the frontal region, making a nasal encephalocele very common.

Some geographic variations occur. There is an encephalocele incidence of 1 in 6000 live births in southeast Asia and Russia, and 1 in 35.000 in western Europe, the USA, Australia, Japan, China, and Indica. There is no sex predilection, and only one pair of identical twins has been reported with encephaloceles.

If one excludes the massive brain herniations that occur with hydrocephaly and microcephaly (with encephalocele), then an encephalocele is compatible with normal brain development and intelligence. A high incidence of other CNS anomalies exists with encephalocele, and a CT scan of the brain to look for other anomalies - such as agenesis of the corpus callosum, ventricular dilatation, and so forth - is worthwhile (Fig. 39-5).

Presentation and investigation. Nasal (or anterior) encephaloceles are anatomically divided into (1) *sincipital* - those associated with a visible external mass and occurring around the nasal dorsum, the orbits, or the forehead; and (2) *basal* - those occurring with no nasal mass and occurring in the nasopharynx, or posterior orbit.

Sincipital encephaloceles. The bony defect occurs between the frontal and ethmoid bones, corresponding to the foramen cecum ossis frontalis in front of the crista galli. The defect is midline in one-half of the cases and bilateral in one fourth. A single unilateral defect occurs in the remaining one fourth.

The following three anatomic subgroups are recognized:

1. *Nasofrontal.* The defect is between the medial orbital walls, which become displaced laterally. Although the nasal bones, nasal cartilages, and the frontal processes of the maxilla remain in a normal relationship, they are pushed *inferiorly* by the mass that expands between the frontal and nasal bones.

2. *Nasoethmoid.* The nasal bones and frontal processes of the maxilla articulate with the frontal bone but are pushed *superiorly* by the mass expanding between the nasal bones and the nasal cartilages anteriorly. The nasal cartilages and septum remain attached to the ethmoid complexes above.

3. *Nasoorbital.* The frontal and nasal bones are in a normal position, and the mass pushes down behind the nasal bones and then extends *laterally* through a defect in the frontal process of the maxilla at the medial orbital wall. The lacrimal bone and lamina papyracea form the posterior limit of the opening.

Sincipital encephaloceles are most often midline at the glabella; a protrusion to the side is less common. The size varies from barely perceptible to a mass equal to the child's head. The degree of hypertelorism varies directly with the size of the protrusion.

Various changes in the overlying skin occur, such as hyperpigmentation, wrinkling, and thinning out of thickening; rarely, CSF is observed to leak spontaneously. Transmitted pulsation may be seen, although most encephaloceles are described as firm but somewhat compressible.

Conventional radiology usually demonstrates a bony defect that is best delineated by CT scanning with and without enhancement. A full head scan should be requested in addition because of the correlation with hydrocephaly. Linear tomography, arteriography, and pneumoencephalography are usually reserved for basal encephaloceles, although very large sincipital masses may require this additional information.

Management. Enlargement of the brain herniation occurs over time with increasing traction on the optic nerves, ventricular system, and cerebral vasculature. A real risk of meningitis exists, and early elective surgical intervention is best. If hydrocephaly exists, a shunt should first

be inserted, and then an approach used to expose the defect so that a CSF-proof closure can be achieved.

The anatomic-clinical distinctions of each subgroup become important here. The nasofrontal subgroup has a short anterior stalk amenable to an extracranial repair, whereas the nasoethmoid and nasoorbital types have longer, wide stalks that require intracranial separation and dural closure followed by a nasal removal, usually staged.

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In the very young child (under 3 years), usually no craniofacial adjustment is recommended other than excision of excess soft tissue and appropriate cosmetic closure of incisions as growth tends to realign displaced nasofacial bones. In an older child a craniofacial interdisciplinary approach is best.

Basal encephaloceles. Basal encephaloceles are less frequently encountered than the sincipital type. The defect lies in the anterior cranial fossa between the anterior border of the cribriform plate and either the superior orbital or the posterior clinoid fissure. Therefore the encephalocele herniates posteriorly into the nose, nasopharynx, or orbit.

The following four subgroups are recognized:

1. *Transethmoidal.* An intranasal mass emerges from a defect in the cribriform plate.

2. *Sphenoethmoidal.* A defect exists in the posterior cribriform or posterior ethmoid area.

3. *Transsphenoidal.* A frequently large defect extends from posterior cribriform to posterior clinoid, and a nasopharyngeal mass exists.

4. *Sphenoorbital.* A mass extends through the superior orbital fissure and protrudes into the posterior orbit.

Both sphenoorbital and transsphenoidal encephaloceles are very rare. The sphenoorbital type appears as unilateral exophthalmos and both neurosurgical and ophthalmic referrals are appropriate. The transsphenoidal type appears as a nasopharyngeal mass, occasionally large enough to cause feeding or nasal respiratory dysfunction. Hypertelorism and an increase in bitemporal diameter are usual. A quarter of the reported cases have had a cleft palate as well.

The transethmoidal type is more common than the sphenoethmoidal type, although both appear as intranasal masses that are typically unilateral. Unilateral rhinorrhea, feeding difficulty, and an examination-revealed polypoid mass that may be pulsatile or expansile on crying or

jugular compression are typical. Some broadening of the bony nasal vault and hypertelorism are not uncommon.

Plain-film radiography initially may *not* demonstrate the defect, unless the rare transsphenoidal type is present. Polytomography may miss the defect if thick cuts are used, and the radiation dosage to the lens of the eye should be borne in mind. CT scanning therefore becomes the initial investigation of choice. If a strong clinical suspicion exists, arteriography may demonstrate displacement of the anterior inferior frontal artery to a point below the level of the cribriform plate.

Small basal encephaloceles may remain undetected until later in life than the normal presentation, which is before the age of 5 years. Some authors have suggested that post-traumatic acquisition of a basal encephalocele is possible.

Management. A CSF-proof closure of the dural defect is almost impossible to achieve through an extracranial approach in a basal encephalocele. A staged or combined approach severing the intracranial communication is used first, occluding the bony defect and then approaching from below to reinforce this.

Transsphenoidal encephalocele poses special technical problems - a larger defect and a difficult (high-mortality) intracranial approach; a very difficult transseptal reduction of the mass is required.

Choanal atresia (Fig. 39-6). Bilateral or unilateral choanal atresia occurs as an isolated congenital anomaly, and 50% are associated with other defects (for instance, Treacher Collins' syndrome, branchial arch anomalies, and cardiac or gut abnormalities).

Incidence

Choanal atresia is not a common anomaly, nor is it rare. It occurs in 1 of every 5000 to 7000 live births and is twice as common in females as in males. It is unilateral more often than bilateral, and it occurs twice as often on the right side as the left side. About 90% are bony, and 10% are membranous. An incomplete atresia is termed a *choanal stenosis.*

Clinical presentation

A unilateral atresia is interesting but requires no immediate attention. A bilateral atresia is a medical, not a surgical, emergency. Newborn infants are obligate nasal breathers; mouth-breathing is a learned reflex that can take from hours to days to acquire.

Until opening of the mouth is learned, a typical cyclical respiratory obstruction occurs. As the child falls asleep, the mouth closes, and a progressive obstruction occurs, starting with stridor and progressing to increased effort and cyanosis. If an observer open the mouth, the obstruction disappears and normal skin color reappears as cyanosis fades.

Normally the initial alerting event is the first feeding of a newborn infant. As the child startles, a progressive obstruction of the airway occurs with cyanosis and hypoxia, to the point of aspiration of the milk and choking. This initial event can mimic the presentation of a tracheoesophageal fistula. If the obstruction is relieved or if gavage feeding is substituted, no cyanotic episodes are seen.

Diagnosis

Once the child has been observed while feeding - to confirm the cyclic nature of the problem - catheters (No. 6 Fr.) should be passed into the nasopharynx. The nose should be examined for edema if many attempts have previously been made to pass catheters, and mucus should be suctioned from the nose. A solution of 0.25% Neo-Synephrine with a little gentian violet can be instilled, and the dye looked for in the pharynx.

Confirmation of the diagnosis can be made by instilling two or three drops of iodized oil (Lipiodol) and taking a cross-table lateral radiograph of the face (Fig. 39-7).

Differential diagnosis

If the cry is abnormal in tone and/or volume, one should suspect a pharyngeal abnormality or congenital subglottic stenosis or subglottic hemangioma.

If the cry is normal and respiratory distress is not relieved by crying, then laryngomalacia should be considered. If the obstruction is relieved by crying and nasal bubbles are observed or a No. 6 Fr. catheter passes the nasopharynx bilaterally, then either a tracheoesophageal fistula, macroglossia, or glossoptosis with a small mandible exists. If catheters fail to pass on one or both sides, the diagnosis is either catheter trauma and edema or choanal atresia on the side through which the catheter fails to pass.

Initial management

Airway maintenance is the first concern. A large nipple can be modified by having its end cut off. This is called a McGovern nipple. Ties are then attached to the nipple and are placed around the occiput, or tape can be used to secure the nipple in the mouth. The nipple acts as an oropharyngeal airway through which the baby can breathe.

Feeding is the next concern. Children with choanal atresia tend to swallow air while feeding and require "breathing breaks" and burping. Sometimes gavage feeding is initially required until mouth-breathing is learned.

During this period the parents need encouragement, and the child needs skilled nursing care and careful observation. Once mouth-breathing is learned, the period of danger is over and the timing of elective repair can be discussed.

If an adequate airway cannot be maintained with an oropharyngeal airway during this critical learning period, surgical intervention will be required. A transnasal opening of the atresia is usually successful in providing an adequate airway, at least temporarily.

Later management

Two surgical approaches can be successfully used: transnasal and transpalatal. The anatomy of the atretic plate is such that it lies obliquely across the nasal airway and the lateral nasal wall is tractioned inward. The surgeon is therefore dealing with a small nasopharynx and an oblique plate. The plate is bony in 90% of cases and membranous in 10%, and may be complete or partial. The latter is termed *choanal stenosis* and does not always require a surgical procedure. Waiting may have some benefit for a stenosis, since the opening enlarges with growth.

Transpalatal approach. The transpalatal approach achieves better visualization and preservation of nasal lining, but the incisions are identical to those for a cleft palate repair and may have a restricting effect (by scar) on lateral maxillary growth. Most surgeons prefer to wait with this approach until at least 16 teeth have erupted (usually by about 12 to 18 months).

Transnasal approach. In the older child with a unilateral atresia, or the infant in whom there is difficulty in maintaining an adequate airway, a transnasal approach may be preferred. In the very young child, ear instruments, aural specula, and a microscope (sometimes with the laser attached) are used. Attempts are made to preserve the lining, since scars form where denuded bone is left. With a rotating burr, currettes, or the laser, the bone in the area of occlusion is removed, with care being taken not to injure the basisphenoid posteriorly as the atretic plane is removed.

A soft stent to hold the flaps in place is always required with all techniques, and most surgeons remove the posterior margin of the vomer to create one large choana, which is less likely to stenose than two smaller ones.

Interim management

In the interim, stasis of secretions in the nose, unless frequently suctioned by parents, leads to infection, with consequent excoriation of the nasofacial skin. Antibiotics, suctioning, and cleansing may be needed.

Feeding can also pose difficulties, particularly if prolonged gavage feeding is required and a failure to gain weight or problems with care of the nose would lead to a need for surgical intervention earlier than the transpalatal approach allows.

Choanal atresia is an easily recognized and diagnosable medical emergency, which requires simple logical steps to manage. The early treatment is conservative, with the timing of surgery dictated by the infant's progress and the parents' ability to manage his difficulties with the assistance of skilled personnel.