Disorders of the Midface

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Embryology

The face and nasal structures develop from cells of ectodermal, neural crest and mesodermal origin, which occurs during the fourth through eighth week of gestation. At 4 weeks gestation, the first sign of facial development begins with the stomodeum, a depression below the developing brain. Surrounding the stomodeum are five structures arising from neural crest cells migrating into the first pharyngeal arches. These structures, the frontonasal prominence, the paired maxillary prominences, and paired mandibular prominences, will guide the development of midface structures.

The frontonasal prominence forms during the fourth gestational week and consists of a frontal and a nasal component. The frontal part of the frontonasal prominence is the precursor to the nasal dorsum and forehead. Near the end of the fourth week, paired ectodermal thickenings on the nasal part of the frontonasal prominence called nasal placodes form. During the fifth week, mesenchymal thickenings form on the margin of the nasal placodes. These are known as the lateral nasal prominences and medial nasal prominences. This proliferation creates a central depression in the placodes called nasal pits. From the fifth week, the nasal pits deepen toward the oral cavity forming the nares and nasal cavity. This progresses until only a thin oronasal membrane separates the oral cavity from nasal cavity. This membrane is an epithelial plug that normally resorbs during the third trimester resulting in posterior choanae.

During the sixth and seventh weeks, the maxillary prominences extend medially toward each other and push the medial nasal prominences medially. The upper lip, nasal tip, columella, philtrum, primary palate, and columella are formed as fusion occurs at the junction of these processes. The nasal septum grows inferiorly from the frontonasal prominence. As the maxillary processes fuse, they form the lateral upper lip and secondary palate.

At the end of the sixth week of gestation, the lateral nasal processes fuse with the maxillary processes to form the lateral borders of the nostril. The nasolacrimal grooves form at the junction of the lateral nasal and maxillary processes. Surface ectoderm migrates from the naso-optic fissure within the nasolacrimal grooves to form epithelial cords, which canalize by the 6th month to form the nasolacrimal ducts and sacks.

The lateral nasal wall begins to take shape at 7–8 weeks gestation when the cartilaginous capsule that surrounds the nasal cavity extends from the chondrocranium of the skull bases. Pre-turbinates form as protrusions from this capsule. Between 9 and 10 weeks gestation, cartilage penetrates the pre-turbinates and the cartilaginous precursor to the uncinate process forms.

Superiorly, the nasal and frontal bones are separated by the fonticulus frontalis. As the frontal and nasal bones grow, they obliterate this space, forming the frontonasal suture. At the same time, the nasal bones and cartilaginous nasal capsule framework are separated by a prenasal space. A dural extension extends from the anterior cranial fossa through the foramen cecum where the frontal bone articulates with the ethmoid bone (maybe define this space) into the transient prenasal space where it contacts the tip of the nose prior to receding. As the nasal bones grow, they obliterate this prenasal space.

History, Physical and Imaging

In the setting of a neonate with a severely compromised airway, history taking will be secondary to assessment and stabilization. However, once the child is stabilized, a focused history should be taken (Fig. 1). It is of significance to note

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History	Physical
Onset of symptoms (immediate vs delayed) Constant vs intermittent Exacerbating/remitting factors History of adjunctive procedures required Gestational age Route of delivery Instrumentation during delivery Prenatal complications Known nutritional deficiencies (folate) Exposures (drugs, alcohol, toxins, radiation) Family history of craniofacial syndromes Any history of prenatal screening Prenatal ultrasound results	Evidence of respiratory distress: • Cyanosis • Loud stridor/stertor • Retractions, alar flaring • Hypoxemia/hypercarbia Facial dysmorphisms Midface hypoplasia Bubbling from the nares Anterior rhinoscopy Mirror exam to look for fogging Passage of suction catheter Flexible fiberoptic nasopharyngoscopy

Fig. 1 Midfacial Disorders: History and Physical Examination

when the respiratory distress started (immediately at birth or delayed onset) and whether it is constant. Exacerbating or remitting factors such as crying, feeding, or positioning should be noted. There should be inquiry into which adjunctive airway measures were used (positive pressure, jaw thrust, nasal trumpet). One should know the gestational age of the child, whether there were any significant peripartum events, the route of delivery (Cesarean section or vaginal) and whether there was instrumentation of the child during delivery. Parental questioning should include the presence of maternal pregnancy complications, risk factors for nutritional deficiency (specifically folate), exposures to drugs, alcohol, toxins or radiation and family history of craniofacial or other syndromes. One should know whether any prenatal genetic screening was done and whether there were any abnormalities on prenatal ultrasonography.

Initial assessment of an infant with respiratory distress should focus on the ability of the child to maintain ventilation and oxygenation. Signs that may indicate the need for rapid intervention include cyanosis, loud stridor or stertor, the presence of retractions, hypoxemia or hypercarbia.

In the more stable neonate, or in the neonate with a secure airway, a more thorough physical examination can take place. With regards to midface abnormalities, one must pay attention to stigmata of craniofacial syndromes such as gross dysmorphisms or midface hypoplasia. Anterior rhinoscopy can demonstrate mass lesions obstructing nasal airflow. Signs of bubbling of nasal secretions suggest patency of the nasal airway. A mirror placed under the nares will fog if nasal airflow is present and a flexible suction catheter can be passed to assess patency of the nasal airway. Flexible fiberoptic nasopharyngoscopy and laryngoscopy allows a rapid evaluation of nasal passages as well as the nasopharynx, oropharynx, and larynx.

Imaging studies may be useful in the setting of a suspected mass or neoplasm, or if there are concerns for osseous or soft tissue obstruction of the nasal airway. Computed tomography (CT) is the imaging modality of choice to evaluate bony narrowing and osseous lesions but should be used judiciously given the concerns regarding ionizing radiation exposure in infants. Magnetic resonance imaging (MRI) may be more useful when soft tissue evaluation is needed.

Syndromic Midfacial Obstruction

Crouzon Syndrome

Etiology

Crouzon Syndrome was first described by French physician Octave Crouzon in 1912 who noted the features in a mother and daughter. It is one of the craniosynostosis syndromes that result from a mutation of the fibroblast growth factor receptor gene. It is classically associated with bicoronal synostosis, midface hypoplasia, proptosis, and normal intellect.

Epidemiology

Crouzon Syndrome is a rare disease, affecting only approximately 1.6 in every 100,000 births [1].

Pathogenesis

Crouzon syndrome is a genetic syndrome with autosomal dominant inheritance and typically complete penetrance. The majority of cases are associated with various mutations of the FGFR2 genes although one variant, Crouzon with acanthosis nigricans is associated with mutations of the FGFR3 gene [1]. Typically these are missense mutations [1].

Clinical Presentation

Patients with Crouzon's typically present at birth with classic phenotypic abnormalities that should raise the clinician's suspicions for one of the craniosynostosis disorders. These findings include craniosynostosis (i.e., bilateral coronal suture synostosis, pansynostosis, or clover leaf skull), hypertelorism, beaked nose and midface hypoplasia. There may also be limb involvement, however, the absence of syndac-

tyly and broad thumbs can help differentiate the syndrome clinically from others such as Pfeiffer and Apert's (see below). Also, unlike patients with Pfeiffer and Apert's, patients with Crouzon's do not typically have neurocognitive impairment. Due to the presence of craniosynostosis, these patients are at high risk for development of increased intracranial pressure and should be monitored closely for signs of such.

Diagnosis

The diagnosis of Crouzon's is typically suspected based on clinical findings as noted above, however, ultimately diagnosis typically requires molecular testing and identification of mutations within the FGFR2 gene.

Management

As with all patients with midface disorders, the first step in management of these patients is the establishment of a stable airway. Midface retrusion, choanal atresia, nasopharyngeal narrowing, and tracheal/laryngeal abnormalities may all contribute to airway obstruction. Stabilization may require nasopharyngeal airway placement, intubation, and/or tracheotomy. Shallow orbits and severe proptosis require aggressive management to prevent exposure keratitis and ulcerations. Temporizing tarsorrhaphies may ultimately be required until midface reconstruction can be completed.

In patients with elevated intracranial pressures, decompression procedures are warranted on an urgent basis. Of the craniosynostosis disorders, Crouzon's has the highest risk of significant intracranial hypertension [1]. However, in the absence of these findings, cranial vault expansion has traditionally been delayed until around 6–12 months of age [2]. With the advent of endoscopic approaches however, the age to intervene is trending toward a younger age with some institutions performing elective strip craniectomies as early as 3 months [3]. Midface procedures are typically delayed until around 5 years of age [4]. Due to the underlying biology of these syndromes, high rates of reoperation have been reported [4].

Multidisciplinary Considerations

As with patients with other craniosynostosis syndromes, children with Crouzon's are typically best served at large medical institutions with multidisciplinary craniofacial teams that include pediatricians, otolaryngologists, oral maxillofacial surgeons, geneticists, and others. All patients should also be evaluated with developmental screens and management as indicated.

Apert Syndrome

Etiology

Eugene Apert, a French pediatrician, first described nine people in 1906 with the similar findings of craniosynostosis, midface hypoplasia, and syndactyly of hands and feet [5, 6]. Most cases (>98 %) are due to the mutation of chromosome bands 10q25–q26 [7].

Epidemiology

The prevalence of Apert Syndrome is roughly 1 in 65,000 newborns. It is the cause of 4.5 % of cases of craniosynostosis and is equally distributed between male and females [8].

Pathogenesis

Apert syndrome is most commonly caused by a missense substitution mutation in chromosome bands 10q25–q26. The mutation affects downstream production of adjacent amino acids (i.e., Ser252Trp, Ser252Phe, Pro253Arg) in the linker between the second and third extracellular immunoglobulin domains of fibroblast growth factor receptor 2 (FGFR2). The inheritance pattern is thought to be autosomal dominant [6].

Clinical Presentation

The typical findings of Apert syndrome are those of craniosynostosis (coronal, sagittal, metopic), midface hypoplasia, and syndactyly of the hands and/or feet. The skull appearance is a flat elongated forehead with bitemporal widening and occipital flattening. The skull may appear like a "cloverleaf" depending on the position of the temporal bones. The midface is hypoplastic with a flat nose and bulbous tip. The palate is arched with swelling of the palatine processes creating a "pseudocleft" in the midline. Soft palate clefting is found in 30 % of cases. Dentition tends to be crowded with an anterior open bite. Hand anomalies consist of variable syndactyly of the second through fourth fingers, ranging from webbing to complete fusion. Equal variability is seen between the second and fourth digits of the feet. The combination may be referred to as "mitten hand" and "sock foot" [8, 9].

Diagnosis

The majority of cases are diagnosed based on physical exam findings in keeping with Apert Syndrome. Imaging in the form of plain films and computed tomography will be required for diagnostic and therapeutic purposes. Genetic evaluation may be performed to confirm the diagnosis [8, 9].

Management

Each of the three typical components of Apert Syndrome will typically require surgical intervention. Craniotomy will be required in the first year of life for associated craniosynostoses: coronal, sagittal, and/or metopic. Syndactyly repair will be carried out soon thereafter for functional gain. Midface and frontoorbital advancement is typically performed later for cosmetic improvement while orthodontic treatment will be carried out as soon as possible to improve teeth alignment [8, 9].

Multidisciplinary Considerations

Given the cranial and extracranial manifestations of Apert syndrome, multidisciplinary care of patients is required. Ideally, in a collaborative center, surgical intervention will be staged based on functional and cosmetic goals.

Pfeiffer Syndrome

Etiology

Pfeiffer syndrome was first described in 1964 as a rare craniosynostosis syndrome associated with craniosynostosis, midface hypoplasia, broad thumbs, great toes, and variable syndactyly of the hands and feet [10]. It is caused by mutations in the fibroblast growth factor genes [10–13]. These mutations can be transmitted in an autosomal dominant fashion or arise de novo. Interestingly, the spontaneous mutation is thought to be related to advanced paternal age [13].

Epidemiology

Pfeiffer syndrome affects an estimated one in 100,000 live births [14]. Men and women are affected equally [15].

Pathogenesis

Pfeiffer syndrome is associated with more than 25 mutations on one of the two FGFR genes. Five percent of the patients have a mutation on FGFR1. These individuals are likely to present with the less severe phenotype (Type 1). The majority of patients, however, present with a mutation on the FGFR2 gene (Type 2 and 3).

Clinical Presentation

Abnormal development of structures derived from preformed cartilage appears to be at the root of many of the abnormalities seen with Pfeiffer's Syndrome [15]. These structures, which include the skull, trachea, spine, fingers, and ribs can be affected to various degrees depending on the severity of the phenotype. Type 1 "classic" Pfeiffer syndrome involves mild manifestations including brachycephaly, midface hypoplasia, and short, broad thumbs and great toes. These individuals generally have normal intelligence and good long-term prognosis. Type 2 Pfeiffer syndrome is generally associated with the classic "cloverleaf skull," extreme proptosis, finger and toe abnormalities, elbow ankylosis or synostosis, developmental delay and neurological complications. Type 3 is similar to type 2 but without a cloverleaf skull. Infants born with any craniofacial dysostosis may have moderate to severe midface hypoplasia. This may significantly narrow the nasal and nasopharyngeal airway potentially causing severe airway obstruction.

Diagnosis

The majority of cases of Pfeiffer syndrome are diagnosed clinically based on classic phenotypic findings to include craniosynostosis with a wide head and flat occiput, midface hypoplasia, ocular proptosis, short broad thumbs and great toes with deviation away from other digits, various degrees of syndactyly [16]. Molecular diagnosis may also play a role particularly in suspected, but not classic cases of Pfeiffer's. Rarely, prenatal diagnosis is possible via ultrasound. Ultrasound findings of craniosynostosis and broad thumbs and toes should raise a suspicion for Pfeiffer syndrome.

Management

Infants are obligate nasal breathers and therefore any condition affecting the midface can potentially affect their airway. Airway management is therefore a cornerstone of treatment for all neonates born with midface disorders. For infants with severe obstruction, nasopharyngeal airway placement or intubation may be necessary. Ultimately many of these children will require tracheostomy tube placement. Once the patient's airway is secure, long-term plans for reconstructive surgery can be made. Temporizing procedures such as tarsorrhaphy are often necessary due to the severe proptosis which may inhibit complete eye closure. Ultimately the synostotic sutures require release in order to decompress the brain. This procedure may take place as early as 3 months of age [16]. Subsequent surgeries typically include midface distraction osteogenesis to both improve airway dimensions as well as orbital volumes. These procedures may involve external or internal devices depending largely on the patient's age and surgeon preferences [17].

Multidisciplinary Considerations

The complexity of these patients mandates that a multidisciplinary approach be taken to their long-term management. These children are typically best served at large medical institutions with multidisciplinary craniofacial teams that include pediatricians, otolaryngologists, oral maxillofacial surgeons, geneticists, and others.

Inflammatory and Traumatic Disorders

Neonates are obligate nasal breathers so nasal obstruction may lead to respiratory distress, feeding difficulties, cyanotic episodes, and even death. Respiratory distress that improves with crying is the classic clinical scenario for nasal airway obstruction.

Neonatal Rhinitis (Rhinitis Neonatorum)

Etiology/Epidemiology

Rhinitis of infancy is a clinical entity that is seen commonly in pediatric otolaryngology practice; however, a paucity of literature exists regarding this problem. The condition is the most common cause of neonatal nasal obstruction.

The etiology is unclear although there appears to be a seasonal component with most cases presenting in the fall or winter months [25].

Clinical Presentation/Diagnosis

Presenting signs can include stertor, mucoid nasal discharge, mucosal edema, difficulty feeding and intermittent apneas. Thought to be an under-recognized problem [25], it has even been implicated in the sudden infant death syndrome [28].

There have been some questions as to whether there is an atopic component to rhinitis of infancy. Nearly 10 % of children will display symptoms of allergic rhinitis by 18 months and there seems to be an association with parental history of allergic rhinitis [24]. However, immunologic mechanisms seem unlikely to play a major role in the first weeks of life given the mechanism of allergy as it is currently understood.

Rarely, primary ciliary dyskinesia (PCD) can manifest as neonatal rhinitis causing respiratory distress [20]. Suspicion is raised when plain films demonstrate dextrocardia. Diagnosis is by electron microscopy studies demonstrating morphological abnormalities in cilia obtained by bronchial or nasal mucosal brush biopsies. Treatment aims at improving pulmonary toilet [20]

Management

Nasal saline and bulb suctioning should be utilized to clear the mucoid discharge. For severe cases, a short course of topical decongestant such as 0.125 % neosynepherine alone or in combination with topical corticosteroids such as 0.1 % dexamethasone ophthalmic drops may be considered. Dexamethasone drops can be cadministered for up to 1 month and then tapered [25]. Rarely does this condition require further intervention and most infants will respond within 12 weeks [25].

Multidisciplinary Considerations

Neonatal rhinitis can usually be managed by the primary care physician or otolaryngologist. When indicated, allergy/immunology consultation should be considered.

Nasal Septal Deviation

Etiology/Epidemiology

It has been recognized since as early as 1936 that the forces on the neonatal face encountered during the birth process may impact the morphology of the nose and face [18]. The incidence of nasal septal deviation in the newborn is described at between 1.25 and 25 % [26, 27]. Incidence may be related to intrauterine positioning of the fetus with a breech position being associated with the highest incidence [18].

Clinical Presentation/Diagnosis

Morphologically, neonatal septal deviation can take the form of anterior dislocation off of the maxillary crest or anterior/posterior septal deformity [21]. On occasion, neonatal nasal septal deviation can be so severe as to cause obstructive symptoms, with cases severe enough to present with apneas and cyanotic episodes while awake [21].

Management

Closed reduction of the anterior septum can be performed in the first days of life with good results [21]. In severe cases, formal septoplasty can be performed on infants as young as 8 days [21]. This can be done through either a transnasal or sublabial approach, either directly or with endoscopic visualization [21, 22].

Multidisciplinary Considerations

Neonatal septal deviation can usually be managed by the otolaryngologist. In cases of suspected trauma, additional consultations may be necessary.

Septal Hematoma

Septal hematoma can present either as a result of birth trauma or as the consequence of non-accidental trauma. It may be misdiagnosed as a nasal mass. Treatment is transnasal incision and drainage [19].

Congenital Nasal Masses

Intranasal Infantile Hemangiomas

Etiology/Epidemiology

Infantile hemangiomas (IH) are the most common vascular tumors of infancy and are known to present very early in life. One review article published in the NEJM in 1999 reported that in neonates with infantile hemangiomas 55 % are present at birth and the remainder develops within the first few weeks of life [31]. Older studies suggest an incidence as high as 10 %, [30] however, more recent reviews of the existing literature highlight the general lack of methodologically standardized studies and place the presumed incidence more towards 4–5 % [46].

Clinical Presentation/Diagnosis

Infantile hemangiomas are unique in that they are characterized by a rapid proliferative phase followed by a spontaneous slow involution phase [35]. Warner and colleagues [32] highlighted their propensity for growth along embryological fusion planes, which can result in nasal distortion and obstruction in the neonate. Clinically, appearance of these

lesions varies based on depth, location, and stage of involution [35]. In neonates, however, they often appear as relatively pale, soft masses covered with telangectasias until they begin the proliferative phase [35]. They then may begin to appear like the classic soft, red, elevated hemangioma that we often think of. Diagnosis of IH is made based on the clinical appearance of the lesion and, when indicated, imaging of the facial soft tissues. Surveillance for additional skin or internal hemangiomas may be warranted.

Management

The primary goal of treatment, as with any nasal obstruction in the neonate, is focused around airway support. Once this is established, more definitive management of the lesion itself can be entertained. Historically, laser treatments, systemic corticosteroids, intralesional corticosteroids, and surgical resection have been the mainstay for treatment of infantile hemangiomas. However, with the introduction of propranolol as a treatment modality in 2008, there has been a dramatic shift in the management of these lesions. While the exact mechanism by which propranolol treats IH remains unknown, proposed mechanisms include induction of endothelial cell apoptosis [33], vasoconstriction, and blocking of proangiogenic signals [34]. A general paucity of high quality, prospective studies have limited recommendations on propranolol, however, the recent consensus conference on initiation and use of propranolol for IH has led to some guidelines which may help guide clinicians in their use of this medication. Currently the consensus group recommends an initiation dose of 1–3 mg/kg/day with most members advocating 2 mg/kg/day divided TID [35].

Multidisciplinary Considerations

Management of IH often requires a team approach, including input from the dermatologist, plastic surgeon, and ophthalmologist, when indicated.

Nasopharyngeal Germline Malformations

Etiology/Epidemiology

Teratomas of the head and neck account for less than 5 % of all teratomas and reportedly occur in 1 in 20,000 to 1 in 40,000 live births with a female predominance of 5–6:1 reported in the literature [36, 40]. The cervical neck is reported to be the most common head and neck site involved with the nasopharynx second [36]. Like all teratomas, they are composed of all three embryologic germ layers (ectoderm, endoderm, and mesoderm).

Clinical Presentation/Diagnosis

While the literature on nasopharyngeal teratomas is somewhat limited, clinically these lesions seem to have associations with central nervous system abnormalities [37], cleft palates [38], and cardiac abnormalities [40]. A review of 113 cases of germline malformations of the nasopharynx by Chaudhry et al. [39] reported that true teratomas are often sessile lesions while dermoids are more often pedunculated. Both most frequently present with respiratory distress, however, teratomas are associated with a higher incidence of preterm birth, delivery via Cesarean section, and neonatal distress [40].

Nasopharyngeal germline malformations may be identified on prenatal US for maternal polyhydramnios and/or elevated alpha fetoprotein. A literature review by Coppit et al. found that while polyhydramnios is identified in 18 % of patients with cervical neck lesions, it is seen much less frequently in patients with NP malformations.

Management/Multidisciplinary Considerations

When teratomas are identified prenatally, a multidisciplinary approach to perinatal management is warranted. Considerations for airway management include endotracheal intubation, tracheotomy, and even consideration for an EXIT procedure (ex utero intrapartum treatment). Once the airway is secured, treatment of both teratomas and dermoids is focused on surgical resection, which typically occurs via a transnasal approach. Recurrence rates are dependent on completeness of the surgical resection with a higher rate of recurrence in teratomas thought to be secondary to the more difficult resection of a broad-based lesion in the neonate.

Nasal Dermoid Cysts

Etiology/Epidemiology

The nasal dermoid cyst is the most common congenital midline nasal lesion and represents approximately 4–12 % of head and neck dermoids [50]. It is composed embryologically of mesoderm and ectoderm due to failed separation of dural diverticulum and the overlying ectoderm [47]. Contrary to nasopharyngeal teratomas, dermoids present with a slight male predominance.

Clinical Presentation/Diagnosis

These lesions can be located anywhere from the columella to the anterior cranial fossa and can be intranasal, extranasal, or a combination of the two [49]. Clinically, these lesions present as a noncompressible mass with a sinus tract that drains sebaceous material and can occasionally cause recurrent local infections [49]. Hair protruding through a cutaneous punctum is pathognomonic for a nasal dermoid [49]. In a retrospective chart review performed by Wardinsky et al., nasal dermoids were associated with other anomalies in 41 % of cases and with intracranial extension in 45 % of cases.

While the exact percentage of intracranial extension varies in the literature, it highlights the importance of preoperative

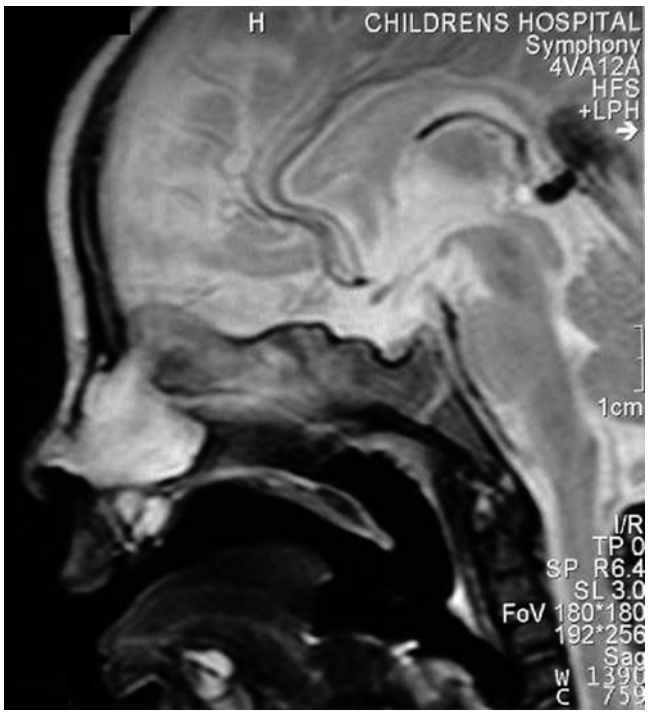

Fig. 2 Sagittal MRI of Nasal Glioma

imaging in patients with suspected dermoid cysts. Both MRI and CT scan may provide complimentary information crucial to the perioperative planning period although MRI avoids potential radiation risks. Normal anatomic variations in the pediatric patient can be easily mistaken for intracranial extension. For instance, a midline gap between the paired nasal bones, non-ossification of the cribiform plate, and the presence of the foramen cecum can all lead to false-positives (Zapata). Widening of the foramen cecum (up to 10 mm) is normal, a bony defect in the crista galli, and a bifid or dystrophic crista galli suggest intracranial extension (Posnick). Dermoids appear hyperintense on T1 and T2 images (Saettele). Contrast may be useful in helping to differentiate dermoids, which are non-enhancing, from other enhancing structures such as infantile hemangiomas, teratomas, and even normal nasal mucosa.

Management/Multidisciplinary Considerations

Treatment of nasal dermoids revolves around complete surgical excision with a high recurrence rate associated with incomplete excision. While multiple surgical approaches have been described, the open rhinoplasty remains the most widely used (Zapata). This approach allows for a single-staged intracranial–extracranial resection if indicated (Zapata). Neurosurgical consultation and involvement in the perioperative and intraoperative management is crucial if intracranial extension is suspected since craniotomy may be required for complete extirpation (Zapata).

Epidermoid

Nasal epidermoids are similar to dermoids in their embryological development but differ in the fact that they contain ectodermal tissue only and therefore never form communication with the central nervous system. Imaging is often required to differentiate epidermoid and dermoid lesions. Epidermoids are characteristically hypointense on T1 and T2 MRI imaging with restricted diffusion. CT scan shows fluid attenuation (Saettele).

Nasal Cerebral Heterotopia/Glioma

Etiology/Epidemiology

Nasal cerebral heterotopias, previously known as nasal glioma, is the least common of the midline nasal masses (Saettele). Embryologically, nasal cerebral heterotopias are similar to dermoids with the addition of rests of neural glial tissue (Saettele). The lack of subarachnoid communication helps distinguish these lesions from anterior encephaloceles (Saettele). While not in direct communication with the subarachnoid space, approximately 15 % of nasal cerebral heterotopias do maintain a stalk connection to the dura [48].

Clinical Presentation/Diagnosis

Clinically, these lesions present as firm, noncompressible masses with overlying skin telangectasias. They grow in proportion with the child and they do not transilluminate nor enlarge with crying (Saettele). They are typically isolated lesions with the most common sites for presentation being the glabella, nasomaxillary suture, and intranasally [48]. On MRI, these lesions are hypointense with T1 signal and can be hyper or hypointense on T2 depending on the degree of gliosis (Fig. 2). They appear isointense to brain matter on CT scan and rarely enhance.

Management/Multidisciplinary Considerations

Surgical resection of heterotopias is generally curative so long as the stalk is removed as well if present (Saettele). Surgery is usually performed using a transnasal approach. When indicated, neurosurgical consultation may be necessary.

Encephalocele

Etiology/Epidemiology

A bony defect in the skull with resulting protrusion or herniation of varying degrees of meninges and brain parenchyma is termed an encephalocele. In contrast to gliomas, encephaloceles maintain a patent communication with the subarachnoid space, which plays an important role in the diagnostic and therapeutic approaches to these lesions.

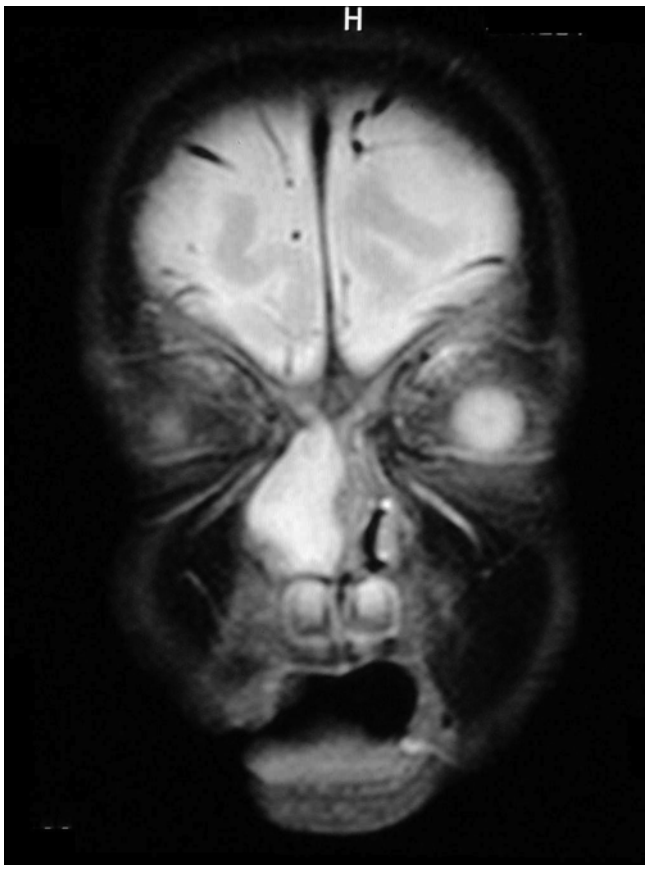

Fig. 3 Coronal MRI of Transethmoidal encephalocele

The majority of encephaloceles are posteriorly based; however, the 25 % that arise anteriorly tend to present much greater diagnostic and therapeutic challenges due to their associated functional, anatomic, and cosmetic affects. Potential complications include but are not limited to: nasal obstruction and impaired nasal function, facial disfigurement, impaired binocular vision, and risk of CNS infection [42]. These anterior, or sincipital, encephaloceles are relatively uncommon in the western hemisphere however are noted to have a high predilection for Southeast Asia with an incidence of 1:6,000 live births [44].

In the Sunwanela [45] classification was published and remains one of the more commonly used classification systems for sincipital encephaloceles. This system classified sincipital encephaloceles based on their location of skull base herniation: frontoethmoidal, interfrontal, and those associated with other craniofacial clefts. Under the umbrella of frontoethmoidal clefts, encephaloceles are further subdivided into: nasofrontal, nasoethmoidal, and nasoorbital.

In addition to their location, encephaloceles are also named based on the tissue that has herniated through the bony defect. Specifically, herniation of meningeal tissue only is referred to as a meningocele and herniation of both meningeal tissue and brain parenchyma is referred to as a meningocephalocele. In extreme cases, a portion of the ventricular system may also be protruding through the bony defect in which case it is referred to as a hydroencephalomeningocele [44].

Theories surrounding the development of encephaloceles are debated within the literature with some believing that failure of bony fusion results in prolapsed tissue whereas others believe that the preexistence of the prolapsed tissue and the resulting stalk prevents normal bony fusion [41]. Regardless of the underlying cause, the resulting prolapsed tissue creates cephalic displacement of the frontal bones, caudal displacement of the nasal bones, and anterolateral displacement of the medial orbital walls [41].

Clinical Presentation/Diagnosis

On examination, an encephalocele will present as a midline nasal mass with a bluish coloration. The mass is soft, compressible and due to its subarachnoid communication, may be visibly pulsatile [44]. Crying, Valsalva, and internal jugular vein compression will lead to enlargement of the mass, again secondary to its intracranial connection. This is referred to as a positive Furstenberg sign. Other characteristic exam findings include a long, flat, and widened nose along with the universal presence of telecanthus. Depending on the severity of these anatomic malformations, this diagnosis may be made in utero during a routine prenatal screening US or much later in life. If identified perinatally, alpha-fetal protein and acetylcholinesterase levels are typically elevated given that it is considered a neural tube defect [43].

MRI is the imaging modality of choice if an encephalocele is suspected (Fig. 3). MRI allows identification of CSF within the malformation, extent of herniated cerebral tissue, and the presence of hydrocephalus [43].

Management/Multidisciplinary Considerations

The process of herniation through the bony defect tends to be a progressive one with enlargement of the mass overtime. As a result, surgical excision is the treatment of choice. Options for surgical excision include combined intracranial/extracranial, fully extracranial, and endoscopic resections, often times in conjunction with a neurosurgical specialist. Regardless of the surgical approach, the general concepts remain the same: resect the mass, repair the skull base defect, and attempt to reconstruct the midline structures. It is important to note that the herniated neurologic tissue is unviable and can be safely removed without compromising neurologic function.

Nasolacrimal Duct Cyst

Nasolacrimal duct cysts (NLDC) arise from incomplete canalization of the epithelial cord that is the precursor to the

nasolacrimal drainage system. Obstruction often occurs distally at Hasner's valve resulting in epiphora, crusting, and ocular irritation. This occurs in up to 6 % of infants. Dacrocystoceles result from cystic swellings of the lacrimal sac when both the valve of Rosenmuller and Hasner's valves are obstructed. If large enough, NLDC can cause nasal obstruction. While NLDCs are most often unilateral, bilateral lesions have been described and can lead to respiratory distress [23] In emergent cases, a simple puncture of the cyst can be performed at the bedside to relieve obstruction; however, definitive management requires marsupialization of the cyst itself. This can be done in a number of ways to include at the bedside under rigid endoscopic guidance. The mucosa is decongested with lidocaine and phenylepherine and then alligator forceps are used to strip the mucosa from the mucocele. Other techniques have been described including the use of powered instrumentation [29] and either endoscopic or external dacrocystorhinostomy for long segment occlusion of the nasolacrimal system. If the intervention is performed under general anesthesia, then the lacrimal system can be probed with nasolacrimal ducts to ensure patency of the system.

Conclusions

Midface physiologic and developmental abnormalities can cause potentially life threatening airway obstruction. Appropriate recognition of airway threats, diagnosis of the correct problem, and directed treatment can mitigate the potential impact.

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