Diagnostic and Therapeutic Sinonasal Endoscopy in Pediatric Patients

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1. Introduction
Fifty years ago, the extracorporeal cold light and its transmission by glass fibers, along with the Hopkins rod lens system, were introduced. The development and application of these new technologies to upper airways allowed studying, understanding, and improving knowledge of the anatomy, physiology, and diseases of the nasal cavity and sinuses. In particular, some fundamental concepts of modern rhinology are based on endoscopic nasal findings and Messerklinger’s investigations of the pathophysiology of sinus mucosa. These studies radically changed traditional understanding of sinus inflammation and revolutionized its treatment using endoscopic conservative surgical management (Messerklinger, 1966, 1967, 1978). In the 1980s, Kennedy (Kennedy, 1985) first utilized this surgical technique in the United States and termed it functional endoscopic sinus surgery (FESS). At the beginning, the technique was performed only for treatment of rhinosinusitis in adult patients. In following years, the surgical indications were extended to selected malignant neoplasms (Kennedy & Senior, 1997; Lund, 1997; Nicolai et al., 2009, 2011). Due to the good results observed by FESS, in 1990s the development of smaller endoscopes and instrumentation adapted for pediatric patients was encouraged. For the treatment of recurrent or chronic rhinosinusitis in children, favorable results were obtained with endoscopic surgery (Lusk & Muntz, 1990; Wolf et al., 1995). During subsequent years, other diseases of sinuses were treated successfully with a nasal endoscopic surgical approach (Triglia & Nicollas, 1997: Berlucchi et al., 2003, 2010; Woodworth et al., 2004; Nicollas et al., 2006; Durmaz et al., 2008; Al-Mazrou et al., 2009; Presutti et al., 2009; Nicolai et al., 2010). In this chapter, a description of endonasal diagnostic techniques and a brief report of sinonasal disorders that may be effectively treated by FESS in pediatric patients are presented. Finally, fundamental surgical steps and their relation between pediatric endoscopic sinus surgery (PESS) and facial growth is briefly discussed.

2. Diagnostic nasal endoscopic procedures
The availability of adequate equipment such as flexible and rigid nasal endoscopes of various degrees and sizes (Fig. 1,2,3) is fundamental to achieve accurate endonasal diagnoses.
Fig. 1. Nasal rigid endoscopes.

Fig. 2. Flexible endoscope.

Fig. 3. Tips of the rigid nasal endoscopes of various degrees.
The choice of nasal endoscope is related to the age and compliance of the pediatric patient. In compliant children and in those older than 8 years, 4-mm and/or 2.7-mm rigid nasal endoscopes are usually well tolerated and provide good endoscopic nasal views. Because of possible traumatic complications, in non-compliant children and in those younger than 8 years, 3.5-mm and/or 2.5 mm flexible endoscopes must be utilized even if they provide an endonasal vision that is qualitatively inferior compared to rigid endoscopes. Before performing nasal endoscopy, cottons soaked with decongestant and local anesthetic are placed in the nasal cavities for about 10 minutes. This allows simultaneously augmenting the space of nasal fossae and obtaining a topical anesthetic effect. This may be easily performed in adolescents, whereas in toddlers and non-compliant children a local anesthetic is preferable sprayed in the nasal cavities. In infants and neonates, topical drugs are not generally utilized. During rhinoscopy, the child is placed in either a sitting position or kept in the arms of a nurse in relation to age and compliance. Nasal endoscopy must be performed correctly, meticulously, and accurately to avoid traumatic lesions of endonasal structures. Before starting endoscopic evaluation, whenever possible it is important to explain the diagnostic procedure to the child in the attempt to obtain full collaboration. After removal of cottonoids and treatment of the endoscopic lens with a thin film of anti-fog solution, the endoscope is inserted slowly and delicately in the nasal fossa. First, the floor of the nose and nasal septum, inferior nasal turbinate and its meatus are examined (Fig. 4).

![Fig. 4. Endoscopic view of the left nasal cavity: inferior turbinate (IT), inferior meatus (IM), nasal septum (NS), and nasal floor (NF).](www.intechopen.com)
Advancing posteriorly, the entire nasopharynx, Eustachian tube orifices, and torus tubarius can be assessed (Fig. 5).

![Image of nasal anatomy](image_url)

Fig. 5. Nasopharynx and left Eustachian tube orifice (arrow) at endoscopic evaluation.

Afterwards, coming back and turning the endoscope superiorly, the middle nasal turbinate and its meatus are explored (Fig. 6).

When the endoscope moves toward the uncinate process area, fontanellae, accessory maxillary sinus ostia, and sphenethmoidal recess can be assessed. By rotating the endoscope superiorly when it is located anteriorly to the head of middle turbinate, it is possible to observe the anterior olfactory region. In addition to evaluation of nasal anatomy, rhinoscopy allows assessment of mucosa status, the presence and type of endonasal secretions (i.e., serous, mucous, or purulent discharge) and their suspicious origin, associated disorders, and their relationships with surrounding structures. Furthermore, rhinoscopy allows monitoring sinonasal diseases such as rhinosinusitis and adenoid hypertrophy, as well as postsurgical follow-up of nasal sinuses. It can also evaluate response to medical treatment, ease cavity debridement in the post-operative period to favor healing of the sinuses, and identify persistent or early recurrences of sinonasal lesions.
Fig. 6. Endoscopic examination of the head of the middle turbinate (MT) and middle meatus (MM).

3. Sinonasal disorders treated by endoscopic sinonasal surgery

Numerous sinonasal diseases can be successfully treated by endoscopic sinus surgery. Extensive surgical experience is mandatory to treat some sinonasal lesions and to obtain good results. Several sinonasal pathologies will be briefly discussed.

3.1 Inferior turbinate hypertrophy
Inferior turbinate hypertrophy can be either congenital or acquired. The former is rare, whereas the latter is usually due to septal deviation, allergic rhinitis, or gastroesophageal reflux disease (Kwok et al., 2007; Cingi et al., 2010). The primary presenting symptom is nasal obstruction occasionally associated with seromucosal rhinorrhea, itching, and sneezing. Moreover, chronic nasal obstruction may modify the normal function of the Eustachian tube causing effusion in the middle ear (Pelikan, 2009). Diagnosis is made by nasal endoscopy. Rhinomanometry in basal conditions and after decongestion can be added in selected cases.

3.2 Adenoid hypertrophy
Adenoid hypertrophy is probably the most frequent pathology in the pediatric population. This disorder manifests usually between 3 and 6 years of age in both sexes. Children
complain of bilateral nasal obstruction associated with snoring, rhinorrhea, mouth breathing, hyponasal speech, and cough (Berlucchi et al., 2007). In some cases, obstructive sleep apnea syndrome can also be observed. The pathology may lead to cardiorespiratory syndromes such as cor pulmonale in extreme cases. Furthermore, adenoid hypertrophy may favor other illnesses such as recurrent and effusive otitis media and recurrent/chronic rhinosinusitis. Nasal endoscopy is the gold standard diagnostic technique to evaluate adenoid size, inflammatory and infectious status, and its anatomical relationship with the nasopharyngeal orifice of Eustachian tubes. Moreover, it allows checking changes in adenoid size after medical therapy (Cassano et al., 2003; Berlucchi et al., 2007). At endoscopic assessment, adenoids appear as a single pyramid-shaped aggregation of lymphoid tissue with the apex pointed toward the nasal septum and the base at the level of the superior and posterior wall of the nasopharynx. The adenoid pad appears as a lobulated and pinkish mass, partially or totally occupying the nasopharynx (Fig. 7).

Fig. 7. Adenoid hypertrophy (asterisk) totally obstructing the right nasal fossa.

3.3 Sinonasal polyposis

Sinosal polyposis is an uncommon pathology in pediatric subjects (Triglia & Nicollas, 1997). In the 1990s, the disorder was classified in 5 subtypes: antrochoanal polyps (this
lesion will be described separately due to its peculiar characteristics), choanal polyps, polyps associated with chronic rhinosinusitis (non-eosinophil dominated), polyps associated with chronic rhinosinusitis (eosinophil dominated), and polyps associated with specific illnesses such as cystic fibrosis, Kartagener’s Syndrome, and asthma (Stammberger, 1999). Even though the etiology of sinonasal polyposis is unknown, some predisposing factors have been identified. The lesions affect both sexes and can be either monolateral or bilateral. Clinically, children complain of nasal obstruction, rhinorrhea, reduction of the sense of smell or anosmia, headache, and facial pain (Triglia & Nicollas, 1997). At nasal endoscopy, polyps show a characteristic edematous and translucent appearance (Fig. 8).

Fig. 8. Nasal polyps (asterisks) associated with mucous secretion (arrows) in a patient with cystic fibrosis.

They can fill partially or totally the nasal cavity and may be associated with a broad or narrow pedicle. Imaging is the diagnostic technique of choice, and CT of the sinuses is the gold standard procedure as it shows exact extension of disease and presence of anatomic anomalies, which may favor sinonasal polyps and/or influence surgical strategy (Triglia & Nicollas, 1997).
3.4 Antrochoanal polyp

First described by Paefyn in 1753 (Paefyn, 1753), antrochoanal polyp (ACP) or Killian’s polyp is a benign, solitary, nasal polypoid lesion. It represents 4-6% of all nasal polyps in the general population (Yaman et al., 2010). It is also prevalent in the pediatric age, and ACP is found in about one-third of pediatric cases with polyps (Schramm & Effron, 1980; Basak et al., 1998; Ozdek et al., 2002; Yaman et al., 2010). The mass originates inside the maxillary sinus and as it grows it extends from the accessory or natural ostium of maxillary sinus to the middle meatus (Fig. 9), finally protruding toward the choana in the nasopharynx.

Fig. 9. Left antrochoanal polyp (asterisk) that come out from natural ostium (white arrow) of maxillary sinus.

From an etiological point of view, ACP develops from intramural Tornwaldt’s cyst in the wall of the maxillary sinus. This particular origin reflects the presence of cysts in the antral portion of the polyp (Berg et al., 1988; Skladzięń, 2001). Chronic sinus inflammation and allergy are other factors favoring formation of ACP (Skladzięń et al., 2001). The disorder, whose etiology is still unknown, is usually unilateral and more frequent in males (M:F=2.1:1). ACP is composed of cystic and solid portions. The former occupies the maxillary sinus, while the latter, which generally emerges through an enlarged maxillary
accessory ostium, is found in the nasal fossa. The most common symptoms are unilateral nasal obstruction, rhinorrhea, bleeding, headache, snoring, and foreign body sensation (Orvidas et al., 2001; Aydil et al., 2008). Moreover, in 20-25% of cases nasal obstruction may be bilateral, in relation to complete blockage of the nasopharynx. Moreover, some reports have described dysphagia and dyspnea correlated with mouth extension. Nasal endoscopy and CT are the gold standard diagnostic procedures. At endoscopic examination, the lesion appears as a white and bright mass located in the middle meatus. This mass juts out the maxillary sinus and occupies the nasal fossa (Frosini et al., 2009). At imaging, ACP fills the maxillary sinus growing through the accessory or natural ostium into the middle meatus to the choana (Pruna et al., 2000). By MR, the lesion reveals hypointense T1 and enhanced T2 signals, and the cystic part is enhanced in the peripheral area after intravenous gadolinium administration (De Vuysere et al., 2001).

3.5 CSF leak
Cerebrospinal fluid (CSF) leak occurs when there is abnormal communication between the space containing CSF around the brain (subarachnoid space) and the sinonasal tract and/or the ear (middle ear/mastoid system) (Pianta et al., 2005). It implies a breach of the underlying dura mater and adherent pia-arachnoid mater resulting in a pathological communication between the intracranial cavity and either the nasal or middle ear cavity (Lloyd et al., 2008; Presutti et al., 2009). According to Ommaya’s classification (Ommaya, 1976), CSF leaks can be divided into non-traumatic (with high or normal CSF pressure) and traumatic (accidental or iatrogenic lesion). About 80% and 16% of CSF leaks are due to head trauma and sinuses or skull base surgery, respectively (Beckhardt et al., 1991). Spontaneous fistulae, which are more frequent in obese females in the fourth decade of life (Pianta et al., 2005), represent 3-4% of cases (Beckhardt et al., 1991; Yerkes et al., 1992; Nachtigal et al., 1999; Schlosser & Bolger, 2002). Moreover, skull base tumors or other congenital lesions (such as untreated aqueductal stenosis) may cause CSF leaks directly through erosion of the skull base or indirectly through the development of hydrocephalus. Other congenital causes of CSF leak are the developmental of skull base defects with associated meningoceles, meningoencephaloceles, large arachnoid granulations or cysts, or congenital inner ear anomalies (Lloyd et al., 2008). If the pathogenesis of traumatic fistula is intuitive, spontaneous leaks may have a multifactorial origin. Among these, intracranial pressure, brain pulsation, cranial base pneumatization, and arachnoid pits are thought to play a major role (Pianta et al., 2005). Spontaneous CSF fistula occurs commonly at the ethmoid roof, cribriform plate, perisella of sphenoid sinus, or inferolateral or pterygoid recess (Lloyd et al., 2008). Patients with CSF leak suffer from unilateral or bilateral watery persistent or intermittent rhinorrhea with positive history for a previous head trauma or surgery of the sinonasal tract, middle ear/mastoid, or skull base. Increase of postnasal drip in the supine position may be reported. Moreover, patients can complain of a salty or sweet taste in the mouth. Recurrent meningitis should alert the physician to a diagnosis of CSF leak (Pianta et al., 2005). Intermittent clear nasal discharge may be exacerbated by the Valsalva maneuver and/or compression of both internal jugular veins (Pianta et al., 2005). When the lesion is located in the temporal bone, CSF reaches the nasopharynx via the Eustachian tube and becomes evident in most cases as bilateral clear rhinorrhea (Pianta et al., 2005). Patients with intermittent CSF leak complain frequently of headache, which appears whenever rhinorrhea stops and the CSF pressure increases (Beckhardt et al., 1991). Finally, signs and symptoms
such as headache, vomit, or edema of the papilla are suggestive for intracranial hypertension (Pianta et al., 2005). “Reservoir sign” is a feature suggestive for the presence of a CSF fistula at the sphenoid, and is due to accumulation of CSF in the sphenoid sinus when the patient is recumbent. It remains in the sinus until the patient assumes an erect position and the head is leaned forward. At that moment, fluid exits from sphenoid ostium and sudden profuse rhinorrhea becomes evident (Nuss & Costantino, 1995). Diagnosis of CSF leak includes laboratory testing, imaging, and fluorescein test. The former includes dosage of several proteins (i.e., beta-2 transferrin or beta-trace protein) on the watery fluid collected from the nose. These are polypeptides produced in the brain, leptomeninges, or choroid plexus that may be identified in nasal mucus when CSF leak is present (Bachmann et al., 2000; Lloyd et al., 2008). Radiological procedures such as CT and MR are used to localize and characterize the involved site, to evaluate for an underlying cause, and to exclude an associated meningocele or meningoencephalocele (Lloyd et al., 2008). Finally, fluorescein test is performed by intra- or peri-operative intrathecal injection of dye solution diluted with 10 ml of CSF (Pianta et al., 2005). This can allow localization of the site of leak and ensure successful closure during surgical intervention.

3.6 Rhinosinusitis

As rhinitis and sinusitis are usually simultaneous, the use of the term rhinosinusitis is medically correct. This disorder is a common upper airway infection in the pediatric age. It is an inflammation of nasal cavity and sinuses and is characterized by two or more symptoms of which should be either nasal obstruction or nasal discharge associated or no with facial pain/pressure and reduction or loss of smell. Based on duration of symptomatology, rhinosinusitis can be divided into: 1) acute rhinosinusitis, when total resolution of aforementioned symptoms may take up to 12 weeks; 2) chronic rhinosinusitis, when clinical picture persists for more than 12 weeks; and 3) recurrent acute rhinosinusitis, when multiple acute rhinosinusitis occurs with total resolution of each acute episode (Fokkens et al., 2007). Several predisposing factors such as allergy, adenoid mass, gastroesophageal reflux disease, sinonasal anomalies (i.e., septal deviation, concha bullosa, Haller cell, choanal atresia, and paradoxical middle turbinate), immunological disorders, primary ciliary dyskinesia, cystic fibrosis, exposure to tobacco, and daycare attendance have been noted to favor rhinosinusitis (Lusk, 1992, 1997; Clement 2008). The classical triad, which is generally responsible for upper respiratory infections (i.e., Streptococcus pneumoniae, Haemophilus influenzae, and Moraxella catarrhalis), has been shown to be involved in most acute rhinosinusitis as well. Staphylococcus aureus and anaerobes can be occasionally found (Lieser & Derkay 2005). Clinically, rhinosinusitis is characterized by rhinorrhea, nasal obstruction, cough, headache, and facial pain. Purulent rhinorrhea, periorbital edema, and high fever may be observed in severe form. Signs and symptoms of chronic rhinosinusitis are those of the non-severe acute form, but they persist for more than 12 weeks. At rhinoscopy, diffuse mucosal inflammation associated with turbinate congestion is the typical endonasal endoscopic appearance of acute rhinosinusitis (Fig. 10). Mucopurulent secretions can be also present and, in relation to their site, it is possible to suspect which sinuses may be affected. Purulent secretions located at middle meatus or sphenoidal recess are a sign of involvement of maxillary, ethmoid, and/or frontal sinus and sphenoid sinus, respectively. Under endoscopic control, cultures can be taken directly
from the involved meatus. Polypoid changes around the middle turbinate insertion is indicative of inflammation of the frontal sinus, whereas the presence of polyps suggests chronic rhinosinusitis (Joe et al., 2001). Moreover, nasal endoscopy allows monitoring inflammation and objectively evaluating the response to treatment. For this reason, serial endoscopic nasal examinations are mandatory to individualize therapy and, eventually, to modify antibiotic administration when no improvement is observed. Diagnosis is based on careful assessment of the patient’s history and clinical picture. In dubious cases, endoscopy of nasal cavity can confirm clinical suspicion. Microbiological cultures are not routinely necessary, but when sinus infection does not improve using antibiotic therapy within 48-72 hours, occurs in an immunocompromised patient, the child is toxic or extremely ill, suppurative complications are evident, or when infectious sinonasal illness recurs 1-2 weeks after the end of medical therapy, microbiological evaluation is mandatory (Lusk & Stankiewicz, 1997; Clement, 2008). Imaging is not indicated to confirm a diagnosis of rhinosinusitis. CT is performed after failure of medical therapy and, therefore, in the planning of surgery or when surgical treatment may be considered as in the aforementioned pathological situations (Lusk & Stankiewicz, 1997). Furthermore, examinations for allergy, cystic fibrosis, immunological disease, gastroesophageal reflux, and primary ciliary dyskinesia can be performed as necessary.
3.7 Choanal atresia

Choanal atresia (CA) is a rare, congenital disease characterized by complete obstruction of the posterior nasal passages. Its incidence is 1:5000-8000 live births (Teissier et al., 2008).

Fig. 11. Endoscopic view of the left choanal atresia (asterisk). The inferior turbinate (black arrow) and middle turbinate (white arrow) are also evident.

There is a female predominance with a F/M ratio of 5/1 among Caucasians. The lesion may be unilateral (60%) or bilateral (40%) and can subdivided into bony (90%) or membranous (10%) types (Vatansever et al., 2005). The genetic aspect of CA remains unclear and is likely multifactorial. In 50% of patients, CA is associated with other anomalies as in the CHARGE syndrome (coloboma, heart abnormalities, CA, retarded growth and development of central nervous system, genitourinary anomalies, ear defects) (Jyonouchi et al., 2009). Several theories such as persistence of the buccopharyngeal membranes, failure of the oronasal membrane to rupture either the nasobuccal membrane of Hochstetter or buccopharyngeal membrane of the foregut, incomplete resorption of nasopharyngeal mesoderm, or locally misdirected mesodermal flow have been proposed to explain the occurrence of CA, but
none have been universally accepted. This process occurs between the 4th and 11th fetal week (Dunham & Miller, 1992; Keller & Kacker, 2000; Samadi et al., 2003). Since neonates are obligate nasal breathing, at birth bilateral choanal atresia can manifest with dyspnea, cyanosis, severe hypoxia, and sucking difficulties, whereas the unilateral form presents monolateral rhinorrhea. Its diagnosis can be late and, often, occasional. Endoscopic examination with flexible nasal endoscope is mandatory when CA is suspected. Endonasal evaluation shows complete closure of involved choana that may be associated with inflammation of nasal mucosa and mucous stagnation (Fig. 11).

Imaging is the next, fundamental diagnostic procedure. CT performed in axial and coronal projections provides a thorough assessment of CA, reveals the bony or membranous nature of the disease, and shows the narrowing of posterior nasal cavity and the thickening of the vomer (Schweinfurth, 2002).

3.8 Mucocele
Mucocele is a benign, cyst-like, locally expansile paranasal sinus mass. The pathology consists of accumulation of secretion products, aseptic slimy mucus, desquamation, and inflammation lined by the respiratory mucosa (Marks et al., 1997; Busaba & Salman, 1999), developing within a paranasal sinus associated with expansion of its bony walls as a consequence of ostium blockage. A mucocele grows slowly and expands by eroding the surrounding bony walls. The obstruction can result from congenital anomalies, chronic rhinosinusitis, previous radiotherapy and/or surgical treatment, trauma, and sinonasal neoplasms (Johnson & Ferguson, 1998; Maroldi et al., 2005). Moreover, congenital illnesses such as cystic fibrosis and primary ciliary dyskinesia are considered predisposing factors for occurrence of mucoceles (Guttenplan & Wetmore, 1989; Thomé et al., 2000; Nicollas et al., 2006; Olze et al., 2006; Berlucchi et al., 2010). Mucoceles occur more frequently in the fourth and fifth decade of life, with a similar distribution in both sexes. Paranasal sinuses mucoceles are extremely rare in a pediatric age and most cases described have been associated with cystic fibrosis (Olze et al., 2006). The frontal sinus is involved in 60% of cases, followed by the ethmoid labyrinth and maxillary sinus with 30% and less than 10% of cases, respectively. Few cases are localized in the sphenoid sinus (Som & Brandwein, 1996; Arruè et al., 1998; Lloyd et al., 2000; Caylakli et al., 2006). The higher incidence of mucoceles in the frontal sinus seems to be related to anatomical variations of the frontal recess (Arruè et al., 1998; Martin et al., 2000). Mucoceles are usually monolateral, whereas bilateral mucoceles are infrequently observed (Varghese et al., 2004). The clinical picture, which varies in relation to the sinus involved, includes nasal obstruction, rhinorrhea, headache, cheek pressure or pain associated with or without check swelling, maxillary nerve hyperesthesia, infra-orbital anesthesia, dental pain, loosening of teeth, periorbital pain, proptosis, blurred vision, alteration of visual acuity, diplopia, and sudden loss of vision (Avery et al., 1983; Hayasaka et al., 1991; Moriyama et al., 1992; Curtin & Rabinov, 1998; Busaba & Salman 1999; Maroldi et al., 2005; Tseng et al., 2005). Whenever erosion of the anterior or posterior wall of the frontal sinus is present, a Pott’s puffy tumor or neurological symptoms may be evident (Maroldi et al., 2005). At nasal endoscopy, the appearance varies according to the site of the mucocele and the phase of growth. During the intrasinusal phase, no alterations are generally visible. The subsequent expansion of the mucocele may alter the paranasal sinus bony walls. In a maxillary localization, medialization of the middle turbinate, anterior dislocation of the uncinate process (Fig. 12),
Fig. 12. Anterior displacement of the left uncinate process (asterisk) due to mucocele of the maxillary sinus at endoscopic examination. Mucous secretion (black arrow) and middle turbinate (white arrow) are also observed.

and bulging of the agger nasi cells or the infundibular area can be observed, whereas submucosal remodeling or bulging of the sphenoethmoid recess or posterior ethmoid can be evident in sphenoidal mucocles. In frontal mucocles, endoscopic examination is usually negative (Maroldi et al., 2005) since the lesion has expanded inferiorly to involve the agger nasi. Diagnosis is based on signs and symptoms, nasal endoscopic evaluation, and imaging. By CT, the disease appears as a homogenous lesion that completely occupies the involved sinus with smooth clear-cut margins of bone erosion of its walls (Han et al., 1995; Busaba & Salman, 1999). Moreover, CT shows the site and extension of the disease, remolded cortex, bony erosion entity, anatomical variants, and hyperostotic changes, (Maroldi et al., 2005). MR is usually performed when mucocele formation is secondary to sinonasal soft tissue tumors in which the lining membrane of the mucocele will enhance after intravenous contrast (Jayaraj et al., 1999).
3.9 Meningoencephalocele

Cephalocele or encephalocele (EC) is an extracranial extension of any intracranial structure through a congenital or acquired defect of the skull base (Pianta et al., 2005). Such herniation may be represented by the leptomeninges associated with cerebrospinal fluid or it can also include the brain. The former is defined meningocoele (MC), whereas the latter is termed meningoencephalocele (MEC) (Naidich et al., 1992). The incidence of EC ranges from 1 case/5,000 live births in Thailand to about 1 case/40,000 live births in western countries (Mahapatra & Suri, 2002). The disorder may be divided into occipital, parietal, basal, and syncephalial types (Mc Carty et al., 1990). The latter group is subdivided into fronto-ethmoidal and interfrontal subtypes, and those associated with craniofacial clefts (C. Suwanwela & N. Suwanwela, 1972). The fronto-ethmoidal form, which accounts for about 10% of all meningoceles, includes: 1) naso-ethmoidal form that is the herniation of meninges with or without brain tissue through the anterior cranial base at the level of the foramen caecum between nasal bone and nasal cartilage; 2) naso-frontal form that occurs between nasal and frontal bones; and 3) naso-orbital form that develops between the maxilla and lacrimal bones. MEC is located at the occipital region in 75% of cases, followed in order of frequency by the frontoethmoidal and parietal area in about 15% and 10% of patients, respectively. (Hoving, 2000; Mahapatra & Agrawal, 2006). The neural tissue in MEC was initially considered dysplastic and non-functioning, but since functioning brain has been found in some occipital and trans-sphenoidal MEC, this concept has been recently revisited (Pianta et al., 2005). MEC may cause nasal obstruction and CSF rhinorrhea. This latter symptom can be unilateral or bilateral, persistent or intermittent, and it increases or may be elicited by maneuvers elevating CSF pressure such as compression of the internal jugular veins or the Valsalva maneuver (Pianta et al., 2005). Moreover, MEC can promote alterations and distortions of surrounding facial structures such as displacement of the medial orbital wall, orbit, telecanthus, broad nasal bridge, nasal and/or glabellar swelling, and hypertelorism. Ocular and lacrimal signs and symptoms (i.e., decrease of visual acuity, strabismus, epiphora and/or dacryocystitis) can be observed (Lello et al., 1989; Morris et al., 1989). At nasal endoscopic evaluation, the lesion may appear as a smooth, isolated, pulsatile polypoid mass arising from the olfactory fossa or sphenoid sinus (Samii & Draf 1989; Pianta et al., 2005). The site of the lesion may increase upon jugular vein compression (Furstenberg sign). In addition to evaluation of the clinical picture and nasal endoscopy, diagnostic work-up of MEC must include imaging. CT can show bony defects of the craniofacial junction and the sclerotic margins of the bone defect (Pianta et al., 2005), whereas MR may reveal the relationship with brain.

3.10 Lacrimal duct stenosis

With an incidence ranging from 6 to 84%, congenital lacrimal duct obstruction is a common disorder at birth. Fortunately, most cases resolve spontaneously within the first months of life. The remaining patients will require conservative procedures (lacrimal probing and intubation) and, if symptomatology persists, non-conservative management (dacryocystorhinostomy) will be performed (Berlucchi et al., 2003). The pathology is due to lack of canalization of the lacrimal system that generally intervenes at the distal end (Hasner’s valve). Epiphora and recurrent dacryocystitis represent the typical clinical picture observed. Rarely, some patients present bulging of the medial canthus that corresponds to dacryocystocele. This cystic lesion of the lacrimal sac is due to both proximal (Rosenmuller’s valve) and distal (Hasner’s valve) obstruction. When the lesion expands in the nasal fossa at the level of inferior meatus (Fig. 13), the patient may also complain of different degrees of nasal obstruction in relation to its size (Wong & VanderVeen, 2008); respiratory distress can also be observed in bilateral localization.
At nasal endoscopy, the nasal cavity can be completely normal or, in some cases, a nasolacrimal duct cyst can be identified in the inferior meatus. Ophthalmologic and otorhinolaryngologic evaluation, dacryocystography, and CT of sinuses are the diagnostic procedures indicated or required (Berlucchi et al., 2003).

3.11 Lobular capillary hemangioma

Also known as pyogenic granuloma, telangiectatic granuloma, granuloma pedunculatum, and infected granuloma, lobular capillary hemangioma (LCH) is a benign, rapidly growing, painless, easily-bleeding, solitary lesion, which occurs in the skin and mucous membranes (Maroldi et al., 2005). Although several factors (i.e., nasal trauma, hormonal influences, viral oncogenes, underlying microscopic arteriovenous malformations, and the production of angiogenic growth factors) have been advocated to favor this disorder, its etiopathogenesis remains unknown (Puxeddu et al., 2006). In the head and neck area, the lesion commonly occurs in the oral cavity (gingiva, lips, tongue, and buccal mucosa), whereas involvement of the nasal cavity is rare (Simo et al., 1998; Ozcan et al., 2004). Sinonasal localization ranges
from 7% to 29%, and the lesion more frequently involves the anterior portion of the nasal septum and the tip of the turbinates (Maroldi et al., 2005). The disease most often occurs in the third decade of life, with a female predominance (El-Sayed & al-Serhani, 1997; Maroldi et al., 2005), whereas its occurrence in pediatric populations has been only rarely reported (Berlucchi et al., 2010). The most common symptoms of LCH of the nasal cavity are recurrent unilateral epistaxis, nasal obstruction, and nasal discharge; facial pain, hyposmia and alteration of smell, and headache are rarely present (Ozcan et al., 2004; Puxeddu et al. 2006). At nasal endoscopy, the lesion usually appears as a single reddish hypervascularized polypoid mass that bleeds easily (Fig. 14).

Fig. 14. Lobular capillary hemangioma (asterisk) completely occluding left nasal cavity.

When a nasal LCH is small, diagnosis is not difficult, while problems occur when the mass is relatively large and its macroscopic appearance is unclear. In these situations, imaging is mandatory (Berlucchi et al., 2010) as it reveals important features of the lesion such as size, probable site of origin, and vascularization pattern. CT shows a soft-tissue density nasal
lesion with lobulated contours. MR reveals masses with an intermediate to hyperintense signal on T2-weighted images and a hypointense signal on T1-weighted images. Enhancement after contrast administration can be helpful (Berlucchi et al., 2010).

3.12 Nasal glioma
Nasal glioma (NG), also known as nasal glial heterotopias, brain-like heterotopia, glial hamartoma, heterotopic neuroglial tissue, nasal cerebral heterotopias, cephalic brain-like heterotopias, and nasal heterotopic brain tissue (Rahbar et al., 2003; Pakkasjärvi et al., 2008), is a rare benign developmental abnormality of neurogenic origin. The peak of occurrence is between 5 and 10 years of age, with a male-to-female ratio of 3:2 (Puppala et al., 1990; Vuckovic et al., 2006). The disorder represents 0.25% of all nasal tumors and accounts for approximately 5% of all congenital nasal swellings (Dabholkar et al., 2004, Vuckovic et al., 2006). The most widely accepted etiopathogenetic theory is that NG represents an encephalocele that becomes sequestered from the brain early in gestation. This is probably due to an abnormal closure of the nasal and frontal bone (fonticulus frontalis) that can lead to an ectopic remnant of glial tissue that remains extracranially (Ma & Keung, 2006). Since it is not a true neoplasm, the term NG is actually not correct. The lesion consists of ectopic/heterotopic neural tissue with neuroglial elements and glial cells in a matrix of connective tissue with or without a fibrous connection to the subarachnoid space or dura. It can grow within the nasal region and is covered by skin or respiratory mucosa (Lowe et al., 2000, Vuckovic 2006). Moreover, 90% of NG do not contain neurons and its benign nature is demonstrated by a low proliferative activity (Dimov et al., 2001). NG can be extranasal (60% of cases), lying external to the nasal bones and cavities; intranasal (30%), lying within the nasal cavity (Fig. 15), mouth, or pterygopalatine fossa; or mixed (10%), communicating through a defect of nasal bones. Extranasal gliomas that are usually paramedian are generally located at the glabella, but can be also present laterally or at the nasal tip (Uzunlar et al., 2001; Vuckovic et al., 2006). Intranasal lesions are usually located within the nasal passage medially to the middle turbinate bone. The intranasal type is more often associated with dural attachment (35%) than the extranasal type (9%) (Kennard & Rasmussen, 1990). Finally, combined intra/extranasal gliomas have a typical dumbbell shape with a connecting band (Vuckovic et al., 2006). Patients with NG may complain of nasal obstruction, epistaxis, and cerebrospinal fluid rhinorrhea. Moreover, the lesion can be associated with deformities of the adjacent bones and nasal cartilage such as widened nose and obstruction of the nasolacrimal duct. Hypertelorism, broadening of the nasal bridge, airway obstruction, and epiphora are secondary to growth of the mass (Bradley & Singh, 1982; Fitzpatrick & Miller, 1996). At endoscopic view, NGs appear as nonpulsatile, uncompressible, gray or reddish-blue to purple, soft or firm at touch, and polypoid-like lesion. The mass, which is present on the nasal dorsum and/or arises from the lateral nasal wall, may be associated with telangiectasias of the overlying skin (Hengerer & Newburg, 1990). Neuroimaging is mandatory to identify nasal lesions, to exclude its possible intracranial connection, and to plan the optimal surgical approach (Harley 1991; Hoeger et al., 2001). Because of its potential intracranial connection, excisional biopsy or fine needle aspiration cytology should not be performed due to the risk of meningitis or cerebrospinal fluid (CSF) leak (Claros 1998).
3.13 Juvenile angiofibroma
Juvenile angiofibroma (JA) is a highly vascular benign and locally invasive lesion that accounts around 0.05% of all head and neck neoplasms. The disorder typically occurs in adolescent males. Recently, some studies have reported that the lesion has an immunohistological and electron microscopic profile more consistent with a vascular malformation rather with a tumor (Beham et al., 1997, 2000). The site of origin of JA appears to be the sphenopalatine foramen or the bone of the vidian canal. From there, the lesion can expand to the nasopharynx, nasal fossa, paranasal sinuses, and pterygopalatine and infratemporal fossa. In some cases, involvement of the orbit and middle and anterior cranial fossa by bone erosion may be observed (Nicolai et al., 2003). Most patients present nasal obstruction associated with discharge and recurrent, spontaneous epistaxis. Due to enlargement of the tumor, facial swelling, proptosis, headache, cranial nerve palsies, and conductive hearing loss secondary to otitis media with effusion may also be observed. At
nasal endoscopic evaluation, JA appears as sessile, lobulated, rubbery and red-pink to gray mass covered by several vascular structures (Fig. 16).

Fig. 16. Juvenile angiofibroma (JA) covered by several fibrin due a recent bleeding in the left nasal fossa. Inferior turbinate (IT).

It occupies usually the nasopharynx and nasal cavity, and it bleeds easily when touched. It may sometimes have a polypoid or pedunculated aspect. Because multiplanar evaluation of the disease and detailed information on the relationship between the lesion and important adjacent structures are needed, MR is considered the gold standard diagnostic procedure. Moreover, before surgical treatment, preoperative diagnostic assessment of the vascular pattern of the lesion by angiography is required, which should be associated with angiographic embolisation to decrease intraoperative bleeding and, consequently, the risk of perioperative transfusion (Nicolai et al., 2003). A biopsy of the lesion is not indicated due to profuse bleeding (Antonelli et al., 1987).

4. Surgical technique and its influence on facial growth

Before describing the main surgical procedures, it is fundamental to highlight some general aspects of PESS. 1) The patient must undergo preoperative CT of sinuses to evaluate anatomy, likely type of disease, and extension to plan surgical management. 2) Preoperative
antibiotic and steroid therapy is also added to reduce inflammation and infection in the
sinuses. 3) PESS is always performed under general anesthesia. 4) Endoscopes of different
degrees (0°, 30°, 45°, and 70°) and size (4 and 2.7 mm), adult and pediatric instrumentation
sets for PESS, and microdebrider must all be available in the operating room. 5) Application
of cotton decongestant pledgets in nasal fossae for 10 minutes before surgical management
is helpful to increase the nasal space. 6) Surgical management must be conservative and
involves only the pathological sinuses. Herein, basal procedures about PESS are reported.
Since extensive and advanced endoscopic sinus procedures are beyond the scope of the
present chapter, these surgical treatments will not be presented.

4.1 Middle antrostomy
Submucosal injection of 1% mepivacaine chlorohydrate and 1:200,000 epinephrine is given
at the level of the root of the middle turbinate and uncinate process. The posterior edge of
uncinate process and, when evident, the main ostium of maxillary sinus are probed with a
small seeker. Next, partial uncinectomy with conservation of its upper third is performed
usually with back-biting forceps. When necessary, the natural ostium of the maxillary sinus
can be wided both posteriorly and inferiorly. The risk-areas are nasolacrimal duct,
sphenopalatine foramen, and lamina papyracea sited anteriorly, posteriorly, and superiorly,
respectively.

4.2 Anterior and posterior ethmoidectomy
After removal of the uncinate process, the anterior wall of the ethmoid bulla is evident and
may be opened. This surgical step may be performed by a microdebrider or Weil forceps,
and must be achieved medially and inferiorly avoiding damage to the orbit and roof of
sinus. At this point, basal lamella is exposed. When needed, the basal lamella is perforated
by Weil forceps or microdebrider at the infero-medial portion to prevent damage to the
lamina papyracea and fovea ethmoidalis which are situated laterally and superiorly,
respectively. Next, each bony lamella is opened and removed. During this surgical step, the
optic nerve, located posteriorly and superiorly, can be identified.

4.3 Sphenoidotomy
This is performed through transnasal, transetmoidal, or trans-septal approach, and the
opening of sphenoid sinus is achieved only if the pathology involves this sinus. In this
surgical procedure, instruments are utilized at an infero-medial angle to avoid injury of the
optic nerve and internal carotid artery, which lie at the lateral wall of the sinus.

4.4 Frontal sinusotomy
Frontal sinusotomy is only rarely performed in pediatric patients as sinusotomy of the
frontal sinus is highly challenging due to its small recess and anatomical position. A
standard anterior ethmoidectomy associated with opening of agger nasi is usually sufficient
to identify the frontal recess. If necessary, it can be enlarged using angle circular-biting
forceps. It is mandatory do not to strip mucosa to avoid a secondary frontal stenosis.

4.5 Potential effects of PESS on midfacial and sinus development
Even though the use of PESS is diffuse worldwide, its potential effects on sinus
development and midfacial growth are still object of discussion. In 1995, Wolf et al.
reviewed 124 children undergoing PESS for chronic recurrent rhinosinusitis. The mean age of patients was 12 years, with 3 children under 5 years. Based on a questionnaire about patient satisfaction and symptomatic relief, it was found that endoscopic surgical sinus surgery had no clinically relevant effects on facial bone development. In our opinion, these results might be influenced by the fact that only 25% of patients were under the age of 5 years, an age during which there is rapid growth of the sinuses. In 1996, Kosko et al. described 5 children who underwent PESS for recurrent rhinosinusitis at a median age of 30 months. After a mean follow-up of 42 months, these patients still complained of signs and symptoms of recurrent rhinosinusitis. For this reason, CT was performed in all children. Imaging revealed maxillary sinus hypoplasia in all patients without clinically apparent facial asymmetry. The authors concluded that this radiological finding might be related to endoscopic sinus surgery. In 2000, Senior et al. assessed the quantitative long-term impact of PESS on sinus development. In this study, 8 children who underwent PESS for periorbital or orbital sinusitis were reviewed after a mean follow-up of 6.9 years. Control groups included 9 adults without signs of rhinosinusitis on imaging and 10 adult patients with a clinical history of childhood sinus symptoms and CT-positive for rhinosinusitis. No significant differences in sinus volumes were observed among groups. In 2002, Bothwell et al. analyzed the long-term outcome of facial growth after PESS in a retrospective age-matched study. The study and control groups included 46 children who underwent PESS for chronic rhinosinusitis and 21 children who did not undergo intervention, respectively. Quantitative anthropomorphic and qualitative analyses were performed in all cases. No statistical differences in facial growth were identified between the two patient groups. In 2006, Van Peteghem & Clement evaluated the influence of PESS on facial growth in a prospective study. The patient cohort consisted of 23 children with cystic fibrosis of whom 13 underwent endonasal surgical treatment for massive nasal polyposis. After a follow-up of at least 10 years, cephalometric measurements were performed in the surgical patients and compared with those obtained in non-surgical group. No significant differences were found. Thus, even if the available evidence seems to indicate that PESS does not significantly affect growth and development of sinuses, analysis of potential surgical effects during rapid growth on facial skeleton has not been well assessed and warrants further investigation.

5. Conclusion

The introduction of rigid and flexible endoscopes has radically changed both diagnosis and therapeutic approaches to sinonasal diseases in pediatric patients. Endoscopy of the nasal cavities and nasopharynx permits observation of important anatomical areas that were previously not visible, evaluating macroscopic characteristics of the sinonasal lesions and their relationship with the endonasal structures. When associated with imaging of the sinuses, it may influence therapeutic planning. Consecutive endoscopic nasal procedures can also monitor sinonasal illnesses and their response to medical therapy. Subsequent development of PESS permitted the possibility to perform targeted and conservative treatments. In the post-operative period, rhinoscopy facilitates accurate debridement of nasal fossae and sinuses, promoting their healing. Finally, the performance of regular endoscopic nasal follow-up may identify early recurrences of sinonasal pathologies.
6. References


Advances in Endoscopic Surgery

Diagnostic and Therapeutic Sinonasal Endoscopy in Pediatric Patients


Surgeons from various domains have become fascinated by endoscopy with its very low complications rates, high diagnostic yields and the possibility to perform a large variety of therapeutic procedures. Therefore during the last 30 years, the number and diversity of surgical endoscopic procedures has advanced with many new methods for both diagnoses and treatment, and these achievements are presented in this book. Contributing to the development of endoscopic surgery from all over the world, this is a modern, educational, and engrossing publication precisely presenting the most recent development in the field. New technologies are described in detail and all aspects of both standard and advanced endoscopic maneuvers applied in gastroenterology, urogynecology, otorhinolaryngology, pediatrics and neurology are presented. The intended audience for this book includes surgeons from various specialities, radiologists, internists, and subspecialists.

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