

TITLE: Pediatric Rhinosinusitis

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Anatomy

The maxillary sinus is the first of the paranasal sinuses to begin development in the human fetus. They begin as outgrowths of the lateral nasal wall about day 65 of gestation. These sinuses slowly enlarge in utero but are not demonstrated on plain films until the infant is 4 to 5 months of age. Growth of these sinuses is biphasic with the first period of considerable enlargement during the first 3 years and the second phase between 7 and 12 years of age. During this second phase, the pneumatization extends laterally to the level of the lateral wall of the orbit and inferiorly into the alveolar process in conjunction with the eruption of permanent dentition. Slow expansion of the maxillary sinuses continues until age 18 to reach adult dimensions with an average capacity of 14.75 mL. The maxillary sinuses drain into the middle meatus.

The ethmoid cells begin development later in the third month of fetal development. The anterior ethmoids form as evaginations of the lateral nasal wall with the posterior ethmoids forming in the fourth month of gestation from outgrowths in the superior meatus. At birth these cells are fluid filled and are difficult to visualize on X-rays. By one year of age, the ethmoids can be detected on plain films and subsequently rapidly enlarge to reach adult dimensions by age 12. The cells number 4 to 17 cells on each side with an average total volume of 14-15 mL. The anterior ethmoid air cells drain into the middle meatus whereas the posterior cells drain into the superior meatus.

The frontal sinus begins development during the fourth month of gestation as an upward extension of the most anterosuperior ethmoidal cells. The frontal sinus is rarely visualized radiographically prior to age 5 or 6 after which it slowly grows to reach an adult size by late adolescence with a total volume of 6-7 mL. The pneumatization of the frontal sinus is variable with a developmental failure of one of the sides in 4-15% of the population. The frontal sinus drains into the frontal recess. The sphenoid sinuses originate during the fourth gestational month as paired evaginations of the mucosa in the superoposterior portion of the nasal cavity. They remain as small indentations in the sphenoid bone until age 3 when further pneumatization begins. Growth becomes rapid to reach the level of the sella turcica by age seven and reach an adult size by age 18 with a total volume of 7.5 mL. The sphenoid sinus drains into the superior

meatus along with the posterior ethmoid air cells.

The sinus mucosa consists of pseudostratified ciliated, columnar epithelial cells, goblet cells, and submucosal glands that produce a protective mucous blanket. The mucosal blanket traps bacteria and noxious materials, which are carried by ciliary motion to the ostium and into the nose for elimination. The orientation of the cilia within a given sinus is specific as secretions are propelled towards the natural sinus ostia and from there to the nasopharynx and oropharynx where they are subsequently cleared by swallowing. This mucosa is similar to that found in the nose and tracheobronchial tree.

Definitions

Pediatric rhinosinusitis is frequently broken down into categories depending on the course of illness: acute, recurrent acute, subacute and chronic. Symptoms of acute sinusitis include rhinorrhea, daytime cough, nasal congestion, infrequent low-grade fever, otitis media, irritability and headache. These symptoms can easily be confused with other illnesses, especially viral URI. The persistence of these symptoms beyond 7-10 days or the worsening of these symptoms at around 7 days with a predominance of cough and rhinorrhea is suggestive of acute sinusitis. Additionally, symptoms of a severe acute infection include purulent rhinorrhea, high fever (>40 degrees C) and periorbital edema. Recurrent acute sinusitis is usually defined as complete resolution between episodes of acute sinusitis and 3 or more episodes in 6 months or more than 4 episodes in one year. Subacute sinusitis is defined as signs and symptoms lasting three weeks to three months. Chronic sinusitis is defined as “low-grade” signs and symptoms lasting longer than three months.

Diagnosis

The most common complaints among parents seeking medical care for their children include: nasal discharge, cough, low-grade fever, fetid breath and painless morning periorbital swelling. The cough may be productive or nonproductive. Cough is usually present in the daytime and worse at night; cough present only at night is suggestive of resolving URI. Less frequently, children may present with URI symptoms that are more severe than usual. The fever may be higher, the nasal discharge more purulent; these may be associated with facial pain and swelling. School age children may report headache or dental pain, depending on their ability to localize discomfort.

The physical examination in pediatric patients with rhinosinusitis is often unrewarding. The physical examination is limited by the inaccessibility of the paranasal sinuses as well as the uncooperative nature of the pediatric patient. Younger patients may tolerate evaluation by an otoscope which may demonstrate nasal mucosal edema, erythema, or possibly purulent discharge in the nose. Examination in the oropharynx may reveal moderate injection of the oropharyngeal wall with postnasal drainage in the posterior pharynx. Occasionally there may be tenderness with palpation over the paranasal sinuses. Assessment of the face may reveal appreciable periorbital edema or dark discoloration of the lower eyelids. Flexible and rigid endoscopy may provide a more complete evaluation in an older, more cooperative child. The most specific findings for acute rhinosinusitis in a child include mucopurulence from the middle meatus (after topical vasoconstriction), periorbital swelling, and facial tenderness.

Workup

Computed tomography scanning provides an excellent tool for evaluation of sinus disease, particularly in cases of chronic rhinosinusitis. The CT can demonstrate disease that is not shown on routine X-rays. While CT scanning may demonstrate disease not shown on plain radiographs, the scan may not reveal the extent of disease actually present. CT scans are not necessary for the management of children with uncomplicated acute bacterial rhinosinusitis. Indications for CT scan include pre-operative workup and in cases where suppurative or impending suppurative complications are suspected.

Confirmation of the diagnosis of rhinosinusitis can be made by culturing an aspirate of the sinus secretions. While not completely free of morbidity as these children typically require a general anesthesia, a properly performed sinus aspiration allows for precise identification of the offending pathogen as well as the sensitivities of the organism to appropriate antibiotics. Indications for sinus aspiration in children include severe toxic illness, acute illness unresponsive to antibiotics within 72 hours, immunocompromised patients, suppurative complications, workup for fever of unknown origin. Unfortunately, nasal, oropharyngeal, and nasopharyngeal cultures correlate poorly with cultures of sinus aspirates. Therefore, it is not recommended to undertake these cultures as guides to the bacteriology and therapy of acute or chronic rhinosinusitis. Endoscopically guided culture of the middle meatus correlates well with maxillary or ethmoid sinus aspirates and is definitely less invasive than sinus puncture; this procedure requires a cooperative child. Random nasal swabs show little correlation with maxillary cultures.

Associated Conditions

In general, the pathophysiology of rhinosinusitis relates to impairment of mucociliary clearance, mucosal inflammation and any condition leading to decreased ventilation through a patent sinus ostium. Normal sinus drainage can be affected by:

1. **Viral upper respiratory infection:** This is the most significant predisposing factor for sinusitis. It is estimated that 5-10% of URIs are eventually complicated by an episode of sinusitis. Day care attendance is associated with a 3-fold increase in overall incidence of URIs. Breaking the cycle of chronic infection may require removing the child from day care for a time.
2. **Allergic rhinitis:** Allergy is a known contributing factor to both acute and chronic rhinosinusitis. Allergic rhinitis creates edema of the nasal passages, blocking proper drainage of the sinus cavities. This may lead to an episode of acute sinusitis or contribute to the chronic inflammation of those with chronic rhinosinusitis. Patients with refractory chronic sinusitis or history of atopy should be considered for allergy testing.
3. **Immune deficiency:** A history of frequent otitis media, pneumonia and sinusitis may suggest a primary or secondary immunodeficiency state. Serum IgG, IgA, IgM and IgE should be evaluated as well as ability to respond to polysaccharide capsular antigens of *S. pneumoniae* and *H. influenzae*. Patients identified with immune dysfunction may require IVIG therapy. Genetic counseling for the patient and family may be appropriate. Immunization against *S. pneumoniae* and *H. influenzae* are suggested.

4. Asthma: Sinusitis and asthma are frequently associated; controversy exists over whether they are manifestations in different parts of the respiratory tract of the same underlying disease process or whether a causal relationship exists wherein sinusitis worsens bronchial asthma. Zimmerman found a 31.2 % incidence of radiographic paranasal sinus abnormalities in asthmatic children compared to 0% in controls. Treatment of sinusitis, whether medical or surgical, has been shown in multiple studies to decrease use of broncodilators, normalize pulmonary symptoms and improve subjective asthma symptoms. Evidence for asthma effecting a worsening of asthma symptoms rests mainly on two theories, both controversial:
 - a. the sinonasal-bronchial reflex in which a trigeminal afferent-vagal efferent neural arc causes a reflex bronchoconstriction after irritant/allergic stimulus of the nose, or
 - b. aspiration of infected sinus secretions into the bronchial tree.
5. Gastroesophageal reflux disease: Recent studies suggest that patients with chronic rhinosinusitis have an increased prevalence of gastroesophageal reflux. Many patients, especially children, experience improvement in their chronic sinonasal symptoms after therapeutic trials of antireflux therapy. GER is theorized to have direct effects on nasal mucosa, initiating an inflammatory response associated with edema and impaired mucociliary clearance. Phipps in 2000 reported the results of a prospective trial in which pediatric patients referred for chronic rhinosinusitis were evaluated for gastroesophageal reflux. 19 of 30 patients (63%) were found to have esophageal reflux by pH probe. Six of the 19 patients (32%) demonstrated nasopharyngeal reflux. Fifteen of the nineteen patients had improvement of their sinonasal symptoms after treatment of GERD. Bothwell in 1999 reported that 89% of pediatric candidates for functional endoscopic sinus surgery avoided surgery with treatment for GERD.
6. Cystic Fibrosis: Cystic fibrosis is inherited as an autosomal recessive trait; the mutations associated with CF affect the CFTR protein which is expressed mainly in the epithelial cells of airways and gastrointestinal tract. Multiple different CFTR mutations have been characterized in CF patients. Patients with cystic fibrosis develop chronic pulmonary disease in childhood, sinusitis and nasal polyposis, pancreatic insufficiency and focal biliary cirrhosis. Cystic fibrosis patients presenting to the otolaryngologist usually have already been diagnosed with CF; however, some patients may be undiagnosed and present first to the otolaryngologist with sinonasal symptoms. Not all cystic fibrosis patients with chronic rhinosinusitis have nasal polyps but nasal polyps in the pediatric age group are rare. If cystic fibrosis is suspected, a sweat chloride test or referral for genetic evaluation should be made. Nasal cultures positive for pseudomonas or *S. aureus* are suggestive of CF. Recent studies suggest that heterozygous mutations in the CFTR gene are associated with chronic rhinosinusitis as well as isolated chronic pancreatitis, allergic bronchopulmonary aspergillosis and congenital bilateral absence of the vas deferens. Raman found that seven of fifty-eight pediatric patients (12.1%) with chronic rhinosinusitis harbored CFTR mutations; the expected rate is 3-4%. Wang found a 7% incidence of CFTR gene mutations in 123 chronic rhinosinusitis patients compared to 2% in a control group.

7. Primary ciliary dyskinesia: A history of chronic otitis media, chronic sinusitis and chronic bronchitis or bronchiectasis in an infant or child should provoke a workup for primary ciliary dyskinesia or immotile cilia syndrome. This may be associated with Kartagener's syndrome (sinusitis, situs inversus, bronchiectasis and male infertility). Diagnosis is confirmed by biopsy of inferior or middle turbinate mucosa or tracheal mucosa.

Responsible Organisms

Organisms commonly implicated in pediatric sinusitis include: *S. pneumoniae*, *H. influenzae*, *M. catarrhalis*, and group A streptococci. The microbiology of sinusitis in the intensive care unit and in patients with cystic fibrosis includes *S. aureus* and *P. aeruginosa*. Although there is increasing resistance to antibiotics commonly used to treat outpatient infections such as sinusitis, first-line therapies still have significant therapeutic advantage. However, patients referred to the otolaryngology clinic have frequently already been treated with multiple courses of antibiotics and required antibiotic coverage with increased spectrum. Other risk factors for antibiotic resistance include children two years of age or younger, day care attendance and antibiotic therapy within the last thirty days.

Treatment

Medical: This is the initial treatment modality in cases of acute or chronic rhinosinusitis. Most studies suggest that regardless of the data that indicate approximately 40% of cases of acute sinusitis will resolve without antibiotics, antibiotics allow for earlier resolution and may prevent recurrence. The American Academy of Pediatrics has published recent guidelines for the use of antibiotics which include:

- a. for young children with mild to moderate acute rhinosinusitis, amoxicillin is recommended at the normal or high dose
- b. patients with amoxicillin allergy should be treated with a cephalosporin such as cefdinir, cefuroxime, or cefpodoxime, whereas severely allergic patients should be treated with a macrolide such as clarithromycin or azithromycin
- c. children that do not respond to first-line therapy, children with more severe initial disease, and children who are considered high-risk for resistant *S. pneumoniae* (those who attend day care or have used antibiotics recently) should be treated with high-dose amoxicillin/clavulanate (90mg/kg of amoxicillin component)
- d. parenteral ceftriaxone should be used in those children who are vomiting and cannot take oral antibiotics

Duration of therapy is usually 10-21 days or until symptoms resolve plus seven days. Controversy exists over the benefit of adjuvant therapies such as nasal steroid spray, saline nasal spray, topical decongestants and antihistamines in the setting of acute rhinosinusitis.

For chronic rhinosinusitis, a four to six week course of a beta lactam stable antibiotic is appropriate, with three weeks of therapy being the minimum. Adjuvant therapy with topical nasal steroids is common. Mometasone furoate is the only nasal steroid approved for children who are two years of age or older. Fluticasone propionate is approved for children who are four years or older. Most of the other topical nasal steroids can be used in children 6 years of age and

older. The use of newer, nonsedating antihistamines may be beneficial in children with chronic rhinosinusitis and underlying allergy. Mucolytics such as guaifensin also may help thin secretions and improve symptoms.

Patients with acute rhinosinusitis rarely will ever need surgical intervention except in those cases that are complicated by orbital or nervous system complications. Subperiosteal abscess, orbital cellulitis, or intracranial abscess must receive aggressive surgical management. Subperiosteal abscesses can be managed by external ethmoidectomy or endoscopic techniques may be used to drain the abscess and treat the infected sinus. Intraconal abscesses/inflammation are much rarer and should be managed via external techniques in consultation with an ophthalmologist. Intracranial suppurative complications of sinusitis occur mostly in young adolescents and relate to sphenoid and frontal sinus disease. Appropriate antibiotic coverage, neurosurgical drainage as indicated and drainage of appropriate sinuses either externally or endoscopically is the usual treatment.

In children with chronic rhinosinusitis with moderate to severe nasal obstruction caused by adenoid hypertrophy, adenoidectomy has been shown to be beneficial. Adenoidectomy is almost always the first-line surgical intervention in pre-schoolers and often is the most appropriate first step in older children.

The current state of the surgical technique for treatment of rhinosinusitis is functional endoscopic sinus surgery (FESS). The technique frequently involves opening the osteomeatal complex and removal of sinus disease with minimal manipulation of the surrounding normal tissue. The treatment of pediatric rhinosinusitis by this method is a very controversial issue. Most authors suggest exhausting all other avenues of treatment prior to considering pediatric FESS. The literature indicates that in properly selected patients the results of FESS are good with minimal complications. Concern over sinus surgery affecting facial growth exists; recent data suggest that facial growth is not substantially affected.

While many may argue specific indications for or against surgery, the Consensus Panel for pediatric rhinosinusitis lists the following for absolute indications for FESS: 1) complete nasal obstruction in cystic fibrosis due to massive polyposis or closure of the nose by medialization of the lateral nasal wall, 2) antrochoanal polyp, 3) intracranial complications, 4) mucocoeles or mucopyocoeles, 5) orbital abscess, 6) traumatic injury in the optic canal, 7) dacryocystorhinitis due to sinusitis and resistant to appropriate medical treatment, 8) fungal sinusitis, 9) some meningoencephaloceles, and 10) some neoplasms. Possible indications are for children with chronic rhinosinusitis that persists despite optimal medical management (2-6 weeks of adequate antibiotics and treatment of any concomitant disease) and after exclusion of any systemic disease.

Complications:

1. Orbital: Bacterial infections of the orbit are caused by paranasal sinusitis in approximately 75% of patients. Spread of infection from the paranasal sinus to the orbit may occur directly through extension via osseous structures, indirectly via valveless venous plexuses surrounding the orbit and paranasal sinuses. Chandler classified orbital involvement in sinusitis into five categories: 1. inflammation with edema 2. orbital

cellulitis 3. subperiosteal abscess 4. orbital abscess and 5. cavernous sinus thrombosis. Approximately 3% of patients with paranasal sinusitis can develop orbital complications. Orbital complications occur more commonly in children than adults; children are more susceptible to upper respiratory tract infections, vascular foramina are larger, suture lines of the bone may be open and the osseous septa of the sinuses are thinner and more porous. In children, orbital inflammatory changes may be the only indication of underlying sinus infection. The most common extrasinus infectious complication in the pediatric age group is medial subperiosteal abscess. Presenting symptoms include periorbital cellulitis and orbital signs such as chemosis, proptosis or gaze restriction. Usually, recent history of URI can be elicited and physical exam will reveal signs of ipsilateral sinusitis such as middle turbinate erythema and edema and purulent middle meatus secretions.

2. Intracranial: Intracranial complications of sinusitis include meningitis, epidural abscess, subdural abscess, intracerebral abscess, Pott's puffy tumor and superior sagittal sinus thrombosis. The frontal sinus is most implicated source of intracranial spread followed by the ethmoid, sphenoid and maxillary sinuses. Venous drainage of the frontal sinus is via small diploic veins extending through the sinus wall; these, in turn, communicate with the venous plexi of the dura, periorbita and cranial periosteum. Septic thrombi can propagate or metastasize through this intimately connected venous network. Frontal sinusitis occurs most commonly in adolescent and young men; this corresponds to the time of peak vascularity in the diploic system and development of the frontal sinus. Frontal sinus osteomyelitis with erosion of the anterior sinus wall and subperiosteal abscess formation resulting in a "doughy" forehead swelling is Pott's puffy tumor.

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