Surgery for Chronic Rhinosinusitis in Children with Cystic Fibrosis

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Abstract

Rhinosinusitis is a very common disease in children with cystic fibrosis (mucoviscidosis), but may be under diagnosed. Several reasons may account for the disease being missed in children. The symptoms in the pediatric population are limited, and can be very similar to the common cold or allergic symptoms. Cough and nasal discharge may be the only symptoms present in these children. A high index of suspicion is necessary to make the diagnosis, especially if nasal polyps are seen. Most children are treated medically. Only a small number will require surgical intervention when medical treatment fails. Sinus surgery improves symptoms and quality of life, but likely does not improve pulmonary status. Recommended surgery includes endoscopic sinus surgery with wide opening of the sinuses. This population has unique surgical considerations. Complications of sinusitis, even though rare, can carry a high morbidity and mortality rate.
Introduction

Rhinosinusitis (RS) is a common disease in children with cystic fibrosis (CF) that is sometimes overlooked. Most children’s symptoms can be dismissed as allergic or secondary to recurrent upper respiratory tract infections [1]. The disease has great impact on the health care system and the quality of life of these children.

Patients with CF have a defect in chloride ion transport owing to an impaired CF transmembrane regulator, which plays a key role in hydrating the mucus [2]. Viscous mucus results in mechanical obstruction of the sinus ostia, predisposing the sinuses to local infection and inflammation resulting in chronic rhinosinusitis (CRS) and frequently in the development of nasal polyps. Additionally, there is evidence that the nasal mucosa itself is in a hyperinflammatory state, as suggested by increased expression of a Toll-like receptor 9, a component of innate immunity [3].

The clinical symptoms of RS in these children include nasal stuffiness, colored nasal discharge, and cough. Additionally, RS can contribute to frequent exacerbation of their lower airway disease. Sometimes, cough can be the only symptom recognized by the patient or family. Often, the cough is attributed to the lung condition. This is why it is estimated that only 10% of CF patients will complain of sinus related symptoms, whereas 80-100% of patients actually have RS [4]. Since children are numb to their sinus symptoms, it is extremely important that any child with suspected nasal polyps has work up for CF [5].

Case Study

Chief Complaint: Chronic rhinosinusitis

History of present illness: A 5 year old girl with a history of cystic fibrosis is referred by her pulmonologist for evaluation of chronic sinusitis. She was referred for chronic purulent rhinorrhea for the past three to four months despite three courses of antibiotics. She also has a persistent cough which keeps her parents awake, as well as low grade fever. Her parents note that even prior to these few months, she is “always sick.” She has also been diagnosed with otitis media by her pediatrician on multiple occasions.

Past medical history: Her past medical history is positive for cystic fibrosis diagnosed at birth at an outside facility. Genotyping is unavailable from that facility. Prior surgeries include the gastrostomy tube at age two for failure to thrive and a bronchoscopy one year ago. Family history is positive for cystic fibrosis in her father, which was diagnosed several weeks after the patient’s diagnosis.

Physical Examination: The patient is a healthy-appearing girl who is afebrile with normal vital signs. She is fairly cooperative for her age. Heart and lung sounds are normal. Ear exam shows healthy tympanic membranes without effusion or retraction. Anterior rhinoscopy shows much purulence bilaterally, midline septum, and no frank nasal polyposis. Oral cavity shows 2+ tonsils without erythema or purulence. She does exhibit posterior nasal drainage in the oropharynx. Neck exam shows no cervical lymphadenopathy. Auscultation shows equal breath sounds bilaterally and she has no cardiac murmurs.
Work up: Her sweat chloride and her genotyping at the outside facility are unavailable. Her most recent pulmonary function testing (PFT) shows a forced one-second expiratory volume over forced vital capacity (FEV1/FVC) of 81%. CT scan, obtained after 3 weeks of amoxicillin/clavulanate treatment, showed bilateral maxillary sinusitis, bilateral ethmoid disease, and mucosal thickening in the sphenoid sinuses. The frontal sinuses were not yet developed. (Figure 1)

Diagnosis of CRS in children

CRS in children is a clinical diagnosis. The diagnosis based on clinical presentation as well as duration of symptoms. In the first 7-10 days of nasal symptoms, the etiology is usually viral, unless symptoms worsen and a complication develops. If symptoms persist and are not improving by 10 days, then a diagnosis of acute rhinosinusitis needs to be entertained [6]. CRS is when symptoms persist beyond 12 weeks. Sometimes, acute exacerbations of these symptoms can occur. Allergic rhinitis can present with a similar clinical picture and must be distinguished based on timing of symptoms, as well as presence or absence of purulence.

The physical examination of a patient suspected to have CRS involves direct visualization of the nasal cavity and associated structures. Anterior rhinoscopy with an otoscope is relatively simple and should be a part of the initial evaluation. Attention is given to the nasal septum, to all visible turbinates, and to the presence or absence colored discharge. Nasal endoscopy in older or more cooperative children may be very helpful. Special attention should be made to whether there are polyps in the nose. The presence of polyposis in the nose should alert the physician to the possibility of CF.

Imaging studies are not necessary when the probability of sinusitis is either high or low. Imaging may be useful when the diagnosis is in doubt, based upon a thorough history and physical examination. Plain sinus radiographs may demonstrate mucosal thickening, air fluid levels, and sinus opacification. However, these are not as helpful as CT scans are. A CT scan is necessary in several instances, including when a complication is suspected, in children with polyps, or in those children who failed medical therapy and are considered for surgery [7].

Diagnosis of CF

CF is often diagnosed soon after birth due to newborn screening being implemented in all fifty states. Despite this, the diagnosis may be delayed until several years of age, especially with less common genotypes. Presence of nasal polyps in a child should prompt a work-up for CF [5]. The genetics and the various spectrums of the disease are well reviewed in the literature, and further reading is suggested [8]. Sweat chloride testing is often done as part of the workup for CF. Because values of sweat chloride change early in life in non-CF patients, and because it can change with time in CF patients, its interpretation can depend on the age of the patient, the presence of symptoms, the presence of siblings with the disease, and on repeat sweat chloride testing. The reader is encouraged to review the diagnostic guidelines of CF [9].

Treatment

The case presented meets the diagnostic criteria for CRS based on the complaints of chronic nasal obstruction, colored nasal discharge and cough, with symptoms for greater than 12 weeks, as well as a
finding of purulent discharge on anterior rhinoscopy. Her CT scan corroborated the diagnosis. Medical treatment continues to be the standard for initial care of CRS in CF patients. With appropriate medical treatment, many patients will not require sinus surgery. However, with failure of medical treatment, surgical management may be necessary. Medical treatment includes topical therapy and systemic therapy. Topical therapy includes nasal saline, steroid sprays, steroid rinses, topical antibiotics, and other medications such as dornase alfa. Systemic treatment can include antibiotics and steroids. Systemic antihistamines are in general not recommended, as they make thicker secretions, but of course treatment should be individualized.

Surgical Management

Surgical management for CRS in CF patients remains controversial. Presently the most common indication is symptomatic CRS, with the surgical goal to decrease symptoms. The literature does not support an improvement in pulmonary function with sinus surgery. Virgin et al in 2015 reviewed more than 5000 CF patients across 29 children’s hospitals [10]. They noted a significant variation in incidence of sinus surgery. The number of sinus surgeries per hospital ranged from 4 to 205 over a three-year period. The size of the CF center and the age of the patient were statistically significant predictors of sinus surgery [10]. A recent systematic review of surgical management of CRS in CF concluded that ESS yielded improvement as measured primarily by sinonasal symptoms and endoscopic findings, but it was unclear if it modified lower airway disease [11]. Hence, in symptomatic children, surgery may be indicated, but in the asymptomatic child, the decision is less clear. Indications for sinus surgery in CF patients are listed in Table 1, and include nasal obstruction not relieved by medical therapy, pulmonary exacerbations which are associated with nasal symptoms, symptoms of sinusitis not relieved by medical therapy, lung transplantation, paranasal sinus mucocele, and orbital or intracranial complications of CRS.

Pre-operative considerations specific to CF revolve around the pathophysiology of the disease, including coagulation disturbance, altered sinonasal anatomy, and diminished pulmonary status. Due to malabsorption of fats, CF can lead to deficiencies in vitamins A, D, E, and K. Because vitamin K is required for coagulation, it is recommended to acquire pre-operative coagulation studies. Poor pulmonary status may contraindicate surgery, as mucus plugging may make intubation dangerous. However, in appropriately selected patients, the surgery is thought to be safe [12]. Still, pre-operative clearance by both anesthesiology and pulmonology is prudent. Finally, it is known that due to the disease process, CF patients may have anatomic difficulties including underdeveloped ethmoid sinuses and sphenoid hypoplasia [13] and extensive polyposis. As with all sinus cases, the CT scan should be closely evaluated pre-operatively.

Also important are peri-operative considerations. When performed, the procedure of choice is functional endoscopic sinus surgery (FESS). FESS is a term for minimally invasive procedures designed to restore the natural drainage pathways of the paranasal sinuses. FESS is performed under general anesthesia. The nasal cavity is directly visualized and various specialized tools are used to relieve obstructive lesions of sinus outflow including polyps and diseased mucosa.

An important consideration in CF is the extent of sinus surgery. In the past, limited polypectomy was advocated. However, this was shown to increase the need for revision surgery, and to decrease the interval between surgeries [14]. More recently, wide pneumatisation of all sinuses, including a middle meatal antrostomy (MMA) wider than for routine FESS (so-called “mega-MMA”), has been advocated [8,
In CF patients, the clearance mechanism is compromised. Therefore, the purpose of the very wide antrostomies is to allow for the nasal irrigations done by the patient help the clearance of those sinuses. Studies have suggested that sinus irrigation is improved by larger sinus ostia [16]. Thus, it is advocated that in those patients a more extended approach is performed to allow for better irrigations [15].

Complications of sinus surgery in general include extensive blood loss, orbital complications, intracranial complications, recurrence of disease, and nasal complications such as septal perforation, synechia formation, and epistaxis. Duplechain et al reported that blood loss in ESS in CF patients was higher than non-CF patients (122 mL vs 98 mL respectively), as was operative time (113 minutes vs 89 minutes, respectively), but noted this did not affect their results [17]. A 20-year chart review of sinus surgery on cystic fibrosis patients found no increased blood loss requiring transfusions [18]. No increased incidence of pulmonary complication secondary to intubation was noted either [18]. Like the general pediatric population, CF patients may suffer synechiae after sinus surgery. Placement of steroid-eluting stents intra-operatively may help reduce this risk [19]. Empirically, we have found the careful placement of poly(ethylene terephthalate) gauze (“Telfa”), or even avoidance of use of this gauze, can decrease the chance of post-operative lateralization of the middle turbinate, and therefore reduce risk of synechiae between it and the lateral nasal wall.

Post-operative management is important, and can be difficult in the pediatric population. Parents are educated about the importance of post-operative care, and are encouraged to follow the regimen. The mainstays of post-operative care include early care including monitoring for mucosal edema, synechia formation, and brewing sinus infections, and late care including frequent nasal rinsing and nasal steroid sprays. Rinsing is most often done with saline, but may be medicated such as with steroids, antibiotics, or dornase alfa [20]. Rinsing may be more effective with increased size of sinus ostia, as suggested by a study of 17 non-CF patients using blue dye, in which the chance of sinus penetration by dye increased with increasing sinus osteal dimension [16].

Macdonald et al performed a review of the literature regarding FESS surgical outcomes in CF, though it was not focused on the pediatric population [21]. They report consistent improvement in symptoms of CRS such as headache, facial pain, obstruction, and olfaction, as well as improvement in endoscopy scopes in two of three reviewed studies. However, ESS in CF does not seem to improve forced one-second expiratory volume (FEV1), and it may or may not decrease the number of pulmonary admissions. The effect of ESS on sinus microbiology is encouraging. A study by Aanaes et al showed that ESS, coupled with regimented post-operative sinus washes with saline and antibiotic (colistimethate) as well as endoscopic debridement can improve sinus cultures [22].

Case outcome

The patient in the case study above would benefit from ESS under image guidance including bilateral “mega-MMA”, total ethmoidectomy, and sphenoidotomy. Adenoidectomy would be performed at the same time. Post-operative care including saline sinus rinses and topical antibiotics would be instituted.

Patient was recently seen for her 2 years follow up. She is doing great with widely open sinuses and no recurrence of her polyps.
Conclusion

CRS is a common disease in children which can significant impact on the quality of life of these children. Any child who is noted to have polyps in their nose should be worked up for cystic fibrosis. Surgery on the sinuses of those children varies by institution and by the expertise of the CF team and surgeon. Due to increased perioperative concerns, a multidisciplinary approach, including otolaryngology, pulmonology, and anesthesiology, is recommended. A more extended procedure for those children seems to have better outcome and also reduces the number of surgeries needed. Proper patient selection, counseling and follow-up are essential for a favorable surgical outcome.
Figure 1: CT scan of the sinuses with bone window showing axial orientation at the maxillary sinuses (A), axial orientation at the ethmoid / sphenoid sinuses (B), coronal (C), and sagittal (D) images. The patient had bilateral maxillary, bilateral ethmoid, and right sphenoid sinusitis with mucosal thickening and inspissated mucus. The frontal sinuses are not yet developed in this patient.
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<td>Paranasal sinus mucocele</td>
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References


