European Archives of Oto-Rhino-Laryngology (2024) 281:1131–1137
https://doi.org/10.1007/s00405-023-08307-z

REVIEW ARTICLE

Pediatric chronic rhinosinusitis

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Received: 25 September 2023 / Accepted: 18 October 2023 / Published online: 30 October 2023

Abstract

Purpose An up-to-date overview of diagnosis, differential diagnosis, comorbidities, and current medical and surgical management of pediatric chronic rhinosinusitis (PCRS).

Methods Review of current evidence-based literature on PCRS.

Results Diagnosis of PCRS seems to be improving based on recent evidence using nasal endoscopy as well as computed tomography scanning. Recent literature supports the fact that chronic adenoiditis can be an independent etiology of symptoms of chronic sinusitis, that are very similar to chronic adenoiditis. Allergic rhinitis and immune deficiency play important roles in the management of PCRS. Surgery for PCRS has evolved significantly in the last 15–20 years to include adenoidectomy as well as endoscopic sinus surgery.

Conclusions PCRS is very common in children causing poor QOL for these children. Medical management remains the main stay of treatment with attention to management of co-morbidities that may contribute to the disease severity. Making the correct diagnosis will help with the choice of surgical intervention if medical management fails.

Keywords Pediatric · Chronic rhinosinusitis · Chronic adenoiditis · Adenoidectomy · Endoscopic sinus surgery

Background

Rhinosinusitis is a common diagnosis encountered by providers of all disciplines. Prevalence rates for pediatric chronic rhinosinusitis (CRS) vary greatly depending on the country and criteria and can range between 5% and 15% [1]. Pediatric acute rhinosinusitis (ARS) is even more common, and children may experience multiple ARS episodes a year [2]. These diseases contribute to a large portion of healthcare expenditures [3]. One of the challenges with pediatric rhinosinusitis is making the correct diagnosis. Symptoms often overlap with that of other conditions, including the common cold, allergic rhinitis, and adenoiditis. Furthermore, symptoms may also differ by age as younger children may have different presenting symptoms as compared to older ones [4]. Cough and colored discharge are common presenting symptoms in younger children, whereas nasal stuffiness and facial pressure/headache are common symptoms in older children. Accurate diagnosis and prompt treatment of rhinosinusitis is important in children due to potential serious complications and impact on quality of life in these children [5, 6]. Medical management is the mainstay of treatment and is successful in most cases [5, 6]. When medical management fails, or a complication occurs, surgery may be required [5, 6].

Definitions

Rhinosinusitis is an inflammation of the nose and paranasal sinuses. It is classified based on the duration of symptoms. Acute rhinosinusitis (ARS) is when symptoms resolve in less than 4 weeks, whereas chronic rhinosinusitis (CRS) is when symptoms last for more than 12 weeks [5, 6]. Recurrent acute rhinosinusitis (RARS) is defined as 4 or more episodes of ARS per year with resolution of symptoms between episodes [5, 6].

Pediatric CRS requires the presence of two or more of the following cardinal symptoms lasting for 12 weeks or longer: nasal obstruction, nasal discharge (anterior or posterior), facial pain/pressure, and cough [5, 6]. Symptoms must be accompanied by objective evidence of inflammation,

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demonstrated on anterior rhinoscopy, nasal endoscopy, or radiography [5, 6].

Differential diagnosis

Differential diagnoses for rhinosinusitis can prove challenging. Clinical symptoms of nasal discharge and cough are very common in young children and can be a sign of rhinosinusitis; however, these symptoms can also be due to other factors, such as upper respiratory infections, allergic rhinitis, and/or adenoiditis [7]. Older children will present rhinosinusitis symptoms more similar to their adult counterparts, with nasal congestion and facial pressure/pain symptoms being common [4]. For older children, the adenoids would have regressed and will rarely be the cause of these symptoms.

Chronic rhinosinusitis vs. chronic adenoiditis

Children with chronic symptoms of nasal stuffiness, discharge, and cough may have chronic adenoiditis, chronic rhinosinusitis, or both. CRS often presents with chronic adenoiditis [8]. Making the correct diagnosis based on symptoms alone can be extremely challenging. Fortunately, medical treatment for both is essentially the same [7]. However, accurate diagnosis becomes critical to treatment planning when medical treatment fails, and surgical intervention is entertained [7]. Nasal endoscopic exam can be helpful with differential diagnoses; however, it may not be feasible in all children [8]. Conventional X-rays have poor sensitivity and specificity in diagnosing CRS and fail to correspond to CT scan findings in up to 75% of the patients [9]. Consequently, CT scan is the only gold standard in identifying the presence of CRS and the CT scan of the sinuses (Fig. 1) should be performed at the end of appropriate maximal medical management [9]. Scoring the scan using the validated Lund–McKay (LM) scoring system will allow for making the diagnosis of CRS [10]. This consists of scoring each of the 10 sinuses (five on each side) with a zero if it completely clear, one if it is partially clear and two if it completely opacified. Maxillary ostium blockage will also be scored in addition to the rest of the sinuses on each side as well. It will be scored zero if it completely open and two if it is partially/completely opacified. LM scores will range from 0 to 24 [10] and a CT LM score of five or higher having demonstrated an 86% sensitivity and 85% specificity in differentiating pediatric CRS patients from pediatric control patients [11]. Finally, magnetic resonance imaging (MRI) has a minimal role in diagnosis of pediatric CRS and is usually reserved for cases when complications or tumors are suspected [5, 6]

In addition to these objective tests, subjective measures of quality of life may also help to differentiate CRS from other disorders. The SinoNasal Quality of Life (SN-5) is a subjective self-report measure of quality of life for patients with sinonasal symptoms. The SN-5 has been found to correlate with objective CT scan scores among pediatric patients with CRS [12]. In a recent study in 2022, SN-5 scores successfully differentiate between pediatric patients with CRS and those with chronic adenoiditis [13].

Fig. 1 CT scan sinuses bone window coronal cuts of a 7-year-old male who failed appropriate medical therapy with an LM score = 6

Chronic rhinosinusitis vs. allergic rhinitis

Allergic rhinitis in children can also present with some of the same symptoms as CRS, such as nasal obstruction and rhinorrhea[14]. The symptom of colored discharge is not helpful for differential diagnosis as there is no evidence that a colored discharge is more indicative of rhinosinusitis as compared to allergic rhinitis. However, the duration of symptoms can be used for differential diagnosis, as allergic rhinitis symptoms are often intermittent, lasting for only a few days as compared to CRS symptoms which are more chronic. The fact that allergic rhinitis can be present in up to 27% of children with CRS indicates that the association between the two conditions is multifactorial and needs further investigation [14].

Epidemiology

Pediatric CRS is common with prevalence rates ranging between 5% and 15% depending on the country [1]. In the United States, it is diagnosed in up to 2.1% of pediatric patients in ambulatory clinic settings per year [3]. Rhinosinusitis symptoms may start initially as a viral upper respiratory infection, but the symptoms may progress to a chronic disease [2].

Risk factors

Several risk factors may predispose patients to the development of CRS. Assessing and addressing these risk factors may be important in the context of CRS treatment. A family history of CRS has been found to significantly increase the risk of CRS in children [15]. In addition to a familial genetic disposition, genetic diseases such as cystic fibrosis and primary ciliary dyskinesia may also predispose patients to the development of CRS. Non-genetic systemic risk factors may include immunodeficiency disorders, gastroesophageal reflux disease, allergic rhinitis, and asthma.

Cystic fibrosis

Cystic fibrosis is an autosomal recessive disease that affects the upper and lower airways. Patients with cystic fibrosis nearly always develop chronic mucosal inflammation and nasal polyposis which leads to mechanical obstruction of sinus ostia and increased risk of CRS [16]. These patients are often very refractory to treatment and require multidisciplinary care, including surgical treatment and topical therapies [17]. Children who present with nasal polyps should always be tested for cystic fibrosis via a sweat chloride test and/or genetic testing.

Primary ciliary dyskinesia

Primary ciliary dyskinesia, though uncommon, is another autosomal recessive disorder that increases the risk of developing CRS. It is caused by a defect in a specific element of mucociliary clearance and this buildup of mucus can lead to CRS [18]. A diagnosis of primary ciliary dyskinesia should be considered in cases of refractory CRS, especially when accompanied with chronic and recurrent otitis media [19]. Primary ciliary dyskinesia is often associated with situs inversus and bronchiectasis, known as Kartagener's syndrome [19].

Immunodeficiency disorders

Common primary immunodeficiency disorders include common variable immunodeficiency, Ig subclass deficiency, selective IgA deficiency, and specific antibody deficiency. The weakened immune system of patients with these disorders predisposes them toward persistent sinus infections and subsequent development of CRS [20]. However, the true incidence of primary immunodeficiency disorders in patients with CRS is not known.

Gastroesophageal reflux disease (GERD)

GERD may serve as another systemic risk factor contributing to CRS in children; however, the literature on this topic is limited and mixed [8, 21]. GERD may increase nasal mucus secretion and congestion, potentially leading to higher risk for CRS [21]. Recent expert opinion stated that routine treatment of GERD as part of CRS treatment is not needed and that if treatment is necessary, it will be for the GERD and not for the rhinosinusitis [5, 6, 8].

Allergic rhinitis

Allergic rhinitis is common in pediatric patients; however, the research on the relation between allergic rhinitis and CRS is mixed. Although some research suggests a common comorbidity between allergic rhinitis and CRS, there is currently no evidence that allergic rhinitis is causally related to CRS [22]. Studies have found that pediatric rhinosinusitis patients with allergic rhinitis had higher LM scores and higher recovery times after sinus surgery [22].

Asthma

Asthma is also prevalent among children and commonly comorbid with CRS. Research studies have indicated that asthma and CRS demonstrate a bidirectional relationship among children [23]. Symptoms of asthma such as nasal blockage and inflammation may increase the risk for developing CRS as well as exacerbate pre-existing CRS symptoms [23]. Furthermore, patients with CRS and comorbid asthma tend to have worse asthma and CRS outcomes [22, 23].

Etiology and pathogenesis

The pathogenesis of CRS is not fully elucidated and is the subject of ongoing research, but likely includes multiple contributing factors.

The paranasal sinuses are a group of paired, aerated cavities that drain into the nasal cavity via the sinus ostia. Though the true anatomic role of the paranasal sinuses is uncertain, their ability to clear normal mucous secretions depends on three major factors: ostial patency, ciliary function, and mucous consistency [24]. Any variety of inciting factors may irritate the sinus mucosa leading to inflammation, edema, bacterial proliferation, outflow obstruction, and mucociliary dysfunction. These in turn may contribute to the development of CRS. In patients with CRS, the paranasal sinuses were found to house a characteristic set of bacteria [25].

Similarly, the adenoids are also suspected to house pathogenic bacteria [26] which may cause obstruction and inflammation of the nasal passages [26]. Furthermore, the biofilms within the adenoids may not only support the proliferation of pathogenic bacteria but may also directly cause inflammation [26]. Consequently, a negative perpetuating cycle may be taking place within the adenoids consisting of bacteria causing nasal obstruction and inflammation which results in greater mucous retention and subsequent further proliferation of bacteria. The obstruction, inflammation, and bacterial proliferation may then lead to the development of CRS.

However, it is important to note that the role of microbes in pediatric CRS remains controversial. Bacterial and viral infections are not currently viewed as necessary components of the pathogenesis of CRS [27]. It has been hypothesized that bacterial infections, often times secondary to viral infections, cause mucosal edema and increased mucus production resulting in obstruction of the osteomata complex and decreased aeration and opacification of sinuses [28]. This in turn increases the risk for subsequent bacterial infections as well as CRS. Indeed, the literature is replete with studies showing favorable response to antibiotic treatment in CRS patients, suggesting a role for bacterial infection in CRS etiology [8]. As such, the use of antibacterial agents has remained a first-line treatment for many practitioners.

Viral infections are known to precede episodes of viral rhinosinusitis. These types of infections could facilitate the growth of pathogenic bacteria within the sinuses [29] and/or lead to permanent changes in the anatomy of the nasal passage [30] thereby increasing the risk for CRS. Unlike bacterial infections; however, viral infections are not usually targeted as a part of CRS treatment.

Finally, the role of inflammatory mediators in the pathogenesis of CRS in children is also uncertain. In adults, sinus diseases are posited to be related to inflammation in response to the presence of bacteria rather than to the action of these microbes. The finding of sinus mucosal infiltrates such as eosinophils, plasma cells, and lymphocytes suggests a process of "bacterial allergy" leading to inflammation [31]. However, it is likely that the pathogenesis of a wide spectrum of sinus diseases can be due to both inflammation resulting from infections as well as inflammation resulting from non-infectious etiologies.

Medical management

Antibiotics

The goal of medical management is to reduce sinonasal symptoms, reduce inflammatory burden, and restore normal function of the sinuses. The American Academy of Otolaryngology consensus statement recommended a 20-day course of an appropriate antibiotic for the treatment of pediatric CRS, which can be culture directed [8]. However, both the European Position Paper on Rhinosinusitis and Nasal Polyps [5] and the International Consensus on Allergy and Rhinology [6] did not support the use of oral or intravenous antibiotics for the treatment of PCRS due to lack of evidence for their effectiveness.

When oral antibiotics are used, high dose amoxicillin or amoxicillin–clavulanic acid are recommended as first-line treatments [6]. Cephalosporins (second or third generations) or macrolides can be used as a second-line treatments for patients with penicillin allergy [6]. There is no consensus on the duration of treatment, but most agree that it should be at least 3 weeks [8]. Antibiotics can be repeated depending on the response of the child. Antibiotic prophylaxis to prevent rhinosinusitis infection is controversial and there are concerns regarding increasing the prevalence of antibiotic resistant organisms. Despite this, the use of antibiotics as prophylaxis in pediatric patients is not uncommon and may be associated with a reduction in the number of rhinosinusitis episodes experienced [32]. Furthermore, antibiotic prophylaxis may be especially helpful for patients with cystic fibrosis, primary immunodeficiency disorders, and primary ciliary dyskinesia [33]. Although the use of antibiotic prophylaxis may help to reduce rhinosinusitis episodes, antibiotics have not been shown to prevent complications resulting from rhinosinusitis in pediatric patients [34].

Intravenous antibiotics for the treatment of pediatric patients with persistent or recurrent symptoms despite oral antibiotic management is not recommended when the rhinosinusitis is uncomplicated [8]. Parenteral antibiotics have not been found to contribute to a lasting resolution of symptoms in pediatric rhinosinusitis patients and thus should be reserved for complicated rhinosinusitis [35].

Adjunctive treatments

Adjunct treatments to antibiotic management typically consist of topical nasal saline sprays as well as intranasal corticosteroid sprays. Nasal saline sprays have been shown to be effective in several studies in improving mucous clearance, ciliary beat activity, and clearance of allergens and other mediators [36]. However, research on their adherence and compliance is limited and mixed [37]. Corticosteroid sprays such as fluticasone and mometasone are safe and effective as first-line or adjunct treatments for CRS with and without nasal polyps in children [6]. Short courses of systemic corticosteroids have been shown to be effective in treating inflammatory disorders of the sinuses; however, the potential for serious side effects should limit the use of systemic corticosteroids to only patients who are recalcitrant to conservative treatment [38].

Pediatric CRS patients who are treatment resistant or experience frequent exacerbations should also be tested for immune deficiencies and referred to an immunologist [39]. A recent review of pediatric CRS patients with primary immunodeficiency showed that intravenous immunoglobulin therapy was an effective treatment option for these patients [40].

Furthermore, a recent 2023 study of 39 children with cystic fibrosis who were treated with both lumacaftor and Ivacaftor showed improvements of paranasal sinus disease on MRI over a 7-month period [41].

Surgical management

Functional endoscopic sinus surgery (FESS)

The minimally invasive FESS procedure is designed to restore the natural drainage pathways of the paranasal sinuses. FESS is performed under general anesthesia, typically as a same-day procedure. The nasal cavity is directly visualized, and various specialized tools are used to relieve obstructive lesions of sinus outflow, including polyps and diseased mucosa. The affected sinus air ways are opened in a manner that augments natural mucociliary outflow. At present, FESS is considered one of the main surgical options for select children who meet strict criteria for CRS by the European Position Paper on Rhinosinusitis and Nasal Polyps, International Consensus on Allergy and Rhinology, and American Academy of Otolaryngology [5, 6, 8]. Many studies have demonstrated the efficacy of FESS to improve sinonasal symptoms, quality of life and endoscopic findings in children with recalcitrant CRS [42]. A 2013 systematic review found that FESS had success rates of over 82% with a major complication rate of 1.4% [43]. A similar systematic review and meta-analysis from 2013 on FESS also reported high rate of surgical success in pediatric patients with 71–100% experiencing improvement of CRS symptoms and quality of life and less than 1% experiencing any major complications [44].

Adenoidectomy

Over the last 20 years, adenoidectomy has proven to be successful in up to 80% of CRS patients and is recommended as the first-line surgical treatment [45]. Success rates may be lower in younger children less than 6 years of age [46]. Furthermore, success rates may also be lower in cases of concomitant disorders with rates as low as 28% for those with concomitant asthma [47]. Because of the average success rate with adenoidectomy alone for certain populations, a sinus wash at the time of adenoidectomy has been advocated [48]. The procedure consists of flushing the sinuses while at the same time obtaining a culture for antibiotic guidance. Success rates of up to 88% have been found for adenoidectomy with sinus wash [48].

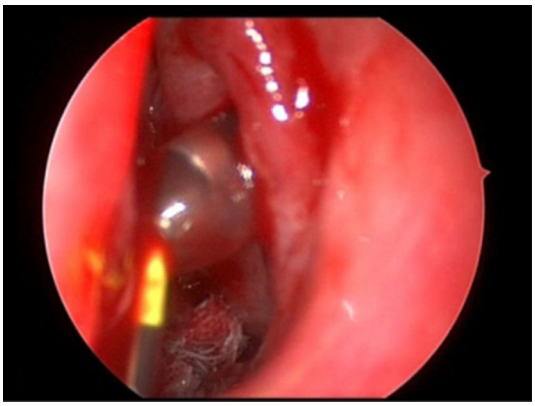

Fig. 2 Balloon catheter sinuplasty for the left maxillary sinus. Balloon is introduced behind the uncinate process (Asterix) into the natural ostium of the sinus

Balloon catheter sinuplasty (BCS)

Another surgical treatment that can be considered for pediatric CRS is BCS. It can be used at the time of adenoidectomy or endoscopic sinus surgery as a surgical adjunct to dilate the ostia as well as lavage the sinuses (Fig. 2). Prospective studies have shown that BCS is safe with minimal complications reported [49]. Several publications have demonstrated that when BCS of the maxillary sinuses was performed at the time of adenoidectomy, children had a better success rate (87%) than those who had adenoidectomy alone (54%) [49]. Larger prospective studies are still needed to determine the long-term efficacy of BCS; however, a recent study had found that success rate remained high 3–5 year post-dilation [49].

Summary

Pediatric rhinosinusitis is a common disease in children. Knowledge of the evidence available for medical as well as surgical treatment can help significantly in the management of these patients. There is significant consensus regarding medical management and growing evidence regarding surgical management. Making the diagnosis of CRS vs. chronic adenoiditis is challenging but of utmost importance for surgical management (Fig. 3). Adenoidectomy is a good first-line surgical treatment for CRS in children under 12 years of age.

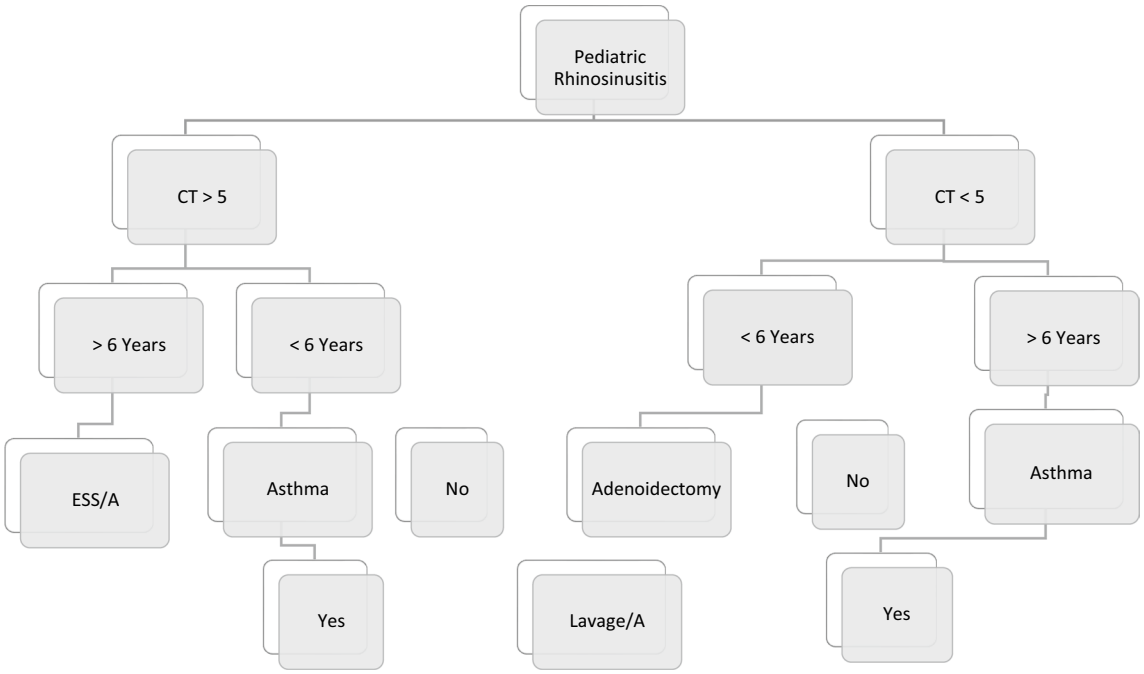

Fig. 3 Algorithm for surgical treatment of pediatric rhinosinusitis. *ESS* endoscopic sinus surgery, *A* adenoidectomy

Funding No funds, grants, or other support was received.

Data availability Not applicable.

Declarations

Conflict of interest The author has no relevant financial or non-financial interests to disclose.

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