

Chronic rhinosinusitis with polyps in children

Przewlekłe zapalenie zatok przynosowych z polipami u dzieci

FEDIR YUROCHKO ^{1/}, WOJCIECH DOMKA ^{2,3/}, LESZEK KŁĘBUKOWSKI ^{4/}

^{1/} Department of Pediatric Otorhinolaryngology, Lviv Regional Pediatric Clinical Hospital OHMATDYT, Ukraine

^{2/} University of Rzeszow, Faculty of Medicine

^{3/} Clinical Department of Otorhinolaryngology, F. Chopin Clinical Voivodeship Hospital No 1 in Rzeszow, Poland

^{4/} ENT Outpatient Clinic in Rzeszow, Poland

Przewlekłe zapalenie nosa i zatok przynosowych (chronic rhinosinusitis, CRS) z polipami występuje u dzieci rzadko. Najczęstszą chorobą układową związaną z polipami nosa w tej grupie wiekowej jest mukowiscydoza. Wśród różnych rodzajów polipów nosa prezentowanych u dzieci najczęściej występują polipy antrochoanalne. Podczas ustalania rozpoznania należy uwzględnić guzy i wrodzone wady rozwojowe. Leczeniem z wyboru w CRS z polipami u dzieci jest farmakoterapia, z glikokortykosteroidami jako najlepszą opcją postępowania. W przypadku nieskutecznej farmakoterapii należy rozważyć postępowanie chirurgiczne. W pracy omówiono epidemiologię, klasyfikację, objawy kliniczne, diagnostykę i postępowanie w CRS z polipami u dzieci.

Słowa kluczowe: polipy nosa, dzieci, przewlekłe zapalenie zatok, glikokortykosteroidy

Chronic rhinosinusitis (CRS) with polyps rarely occurs in children. The most common systemic disease associated with nasal polyps in this age group is cystic fibrosis. Among different types of nasal polyps presented in children, antrochoanal polyps are the most common. Tumors and congenital malformations should be considered when determining a diagnosis. The treatment of choice for CRS with polyps in children is pharmacotherapy with glucocorticosteroids as best option. In the case of non-effective pharmacotherapy, surgical procedure should be considered. The article presents epidemiology, classification, clinical symptoms, diagnostics and management of CRS with polyps in children.

Key words: nasal polyps, children, chronic sinusitis, glucocorticoids

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Adres do korespondencji / Address for correspondence

Wojciech Domka

Klinika Otorinolaryngologii, Kliniczny Szpital Wojewódzki Nr 1 w Rzeszowie, ul. Chopina 2, 35-055 Rzeszów

tel. +48178666362, +48605556026, fax. +48178666361

e-mail: w.domka@gazeta.pl

Introduction

Chronic rhinosinusitis (CRS) with nasal polyps in children is very rare before adolescence. The exact incidence of the disease in the pediatric population is difficult to establish because there is little research in this field [1, 2].

Nasal polyps are benign pathological lesions of the sinunasal mucosa. Once nasal polyps were considered a separate disease type, however polyps are now found to be more or less the same reaction of the nasal mucosa and sinuses to various factors and an end stage of various nasal diseases.

Epidemiology and classification

Epidemiology

Nasal polyps in children are sporadic with a frequency of 0.1%. In children with bronchial

asthma, they are more common, occurring at a rate of 1.8%. The incidence of polyps in children with cystic fibrosis (CF) is even higher, reaching 39.1%, with their frequency of occurrence increases with age. In the course of CF, polyps begin to develop from the age of 5. In children with aspirin triad, the incidence of polyps before the age of 18 is 100%. Nasal polyps are rare in children, but when they appear, their frequency is highest in patients over 10 years of age [2-11].

Nasal polyps in a pediatric population under the age of 2 suggest a probability of the defect in the neural tube (encephalocele). In the presence of nasal polyps in a child over the age of 2, the physician should first consider CF until another diagnosis is confirmed [8].

Nasal polyps in adults are more common in men than in women – in men are 2-4 times more

common [12-15]. Information on the difference in the frequency of the disease between the two sexes among children is not well described in the literature. According to information regarding children who have undergone surgery due to nasal polyps, the ratio is 1:1 [8].

Classification of polyps

There are five types of polyps according to the Stammberger classification (Table I) [16].

Table I. Clinical classification of nasal polyps according to Stammberger

Type	Classification
I	antrochoanal polyps
II	large, isolated (choanal) polyps
III	polyps with chronic rhinosinusitis, non-eosinophil-dominated
IV	polyps with chronic rhinosinusitis, eosinophil-dominated
V	polyps with specific disease (cystic fibrosis, malignancy)

Type I. Antrochoanal polyps are isolated unilateral polyps that arise from the maxillary sinus (usually from its posterior wall) and pass into the nasal cavity and through the posterior nostril to the nasopharynx. The combination of the two structures (sinus and nose/nasopharynx) has the appearance of a branch that comes out from the posterior fontanelle. In children, this type of polyps is common, accounting for 1/3 of all nasal polyps in children [3, 17].

Type II. Large, isolated (choanal) polyps are the only large polyps in the nasal cavity that come mainly from the contact of the two mucosal surfaces in the area of the ethmoidal sinuses or the sphenoidal recess. In children, this type of polyps is quite common.

Type III. Nasal polyps with chronic rhinosinusitis, non-eosinophil-dominated is a bilateral disease. They are rare in children.

Type IV. Nasal polyps with chronic rhinosinusitis, eosinophil-dominated. This group is most challenging to treat due to the high percentage of recurrences. It includes NARES (nonallergic rhinosinusitis with eosinophilia syndrome), PARES (pseudoallergic rhinosinusitis with eosinophilia syndrome), acetylsalicylic acid (aspirin) triad and allergic-fungal rhinosinusitis. Lot of such patients have bronchial hyper-responsiveness. This type is rare in children.

Type V. Nasal polyps with a specific disease (CF, malignancy). These nasal polyps are quite common in CF, primary cystic dyskinesia, and Kartagener syndrome. They are a particularly important type observed in children [4].

Clinical manifestation

Main symptoms include nasal occlusion, thick clear or purulent discharge, mechanical anosmia, watery eyes (epiphora), snoring, blocked nasal passage, and headache. With persistent polyp growth abnormal facial shape, including a broad nose and hypoplastic midface, may present over time. Blocking the sinus ostia leads to chronic or recurrent sinusitis. Nasal polyps may originate from one or more sinuses. In children, they more often arise from the maxillary sinus, and less so from the ethmoidal sinuses. Uncommon symptoms of polyps, particularly significant in size, include nasal bleeding, difficulty in swallowing, sleep apnea, and nausea [4].

In children with CF, the development of polyps is so slow that patients tend not to report ailments. It is more difficult to detect symptoms in younger patients, for example lack of smell.

Clinical manifestations of CRS with polyps in children are dependent on the severity of the disease [1, 2]. Small polyp do not cause symptoms and can be detected by chance during a routine examination when they are in the front or middle of the nasal cavity. Polyps in the posterior part of the nasal cavity are not visible in the routine anterior rhinoscopy. Therefore, they usually remain unrecognized until the onset of symptoms of CRS with polyps of the nose. Small polyps located in the middle nasal meatus may block ventilation and drainage of the sinuses, causing persistent symptoms of rhinosinusitis [18, 19].

Multiple polyps or the solitary large polyp may block the nasal cavity or nasopharynx causing sleep apnea and chronic respiration through the mouth [20, 21].

Quite rarely, patients with CF who have a lot of polyps may have damaged facial structures, causing exophthalmos, double vision and hypertelorism (Woakes' syndrome) [22].

The characteristic feature of CF is the presence of pseudomucocele of both maxillary sinuses. The medial displacement of the lateral wall of the nose caused by pseudomucocele narrows the nasal cavity and causes blockage of the nasal passage in children [4, 23].

Diagnostics

Recognition. In typical cases, there are single or multiple, smooth, shiny, white or grayish-yellow, circular elements, moving when palpated with a probe, and usually placed in the middle nasal or nostril. Polyps in children are more often unilateral. The duration of symptoms before diagnosis ranges from 2 months to 2 years [1].

Differential diagnosis. One should consider meningoencephalocele, juvenile angiofibroma, inverted papilloma, as well as nasal or pituitary tumors (Tab. II, III). In children up to the age of 2, meningoencephalocele or dermoid cyst may mimic a nasal polyp. Before biopsy or surgical removal doctor must perform CT and MRI.

Table II. Nasal diseases that need to be differentiated in a case of nasal polyp in a child [18]

Group	Disease
Congenital defects	Dermoid cyst
	Meningeal hernia
	Nasolacrimal cyst
	Nasal glioma
Benign neoplasms	Neurofibroma
	Craniopharyngioma
	Haemangioma
Malignant neoplasms	Rhabdomyosarcoma
Foreign body	

Table III. Criteria for differential diagnosis of nasal polyps in children

Disease	Clinical manifestations	Radiology
Juvenile angiofibroma	Heavy nasal bleeding in boys.	CT – bone destruction
Meningoencephalocele	Pathology is located at the frontal and ethmoid bones. Sometimes associated with hydrocephalus, eye abnormalities	CT – defect of the anterior skull base in the area of the cribriform plate
Inverted papilloma	Rarely in children, abnormal changes in the endoscopy of the nose. The biopsy is necessary	CT – an erosion of the lateral nasal wall
Glioma, dermoid cyst	The onset of symptoms from birth. Sometimes have an intracranial extension	CT – hypodense lesion, thin wall of lesion, extension under nasal bones in midline
Malignant neoplasms	Bleeding from the nose, nose and facial deformities, cranial nerve palsy	CT – spread on soft tissue, bone erosion. A MRI is necessary

The differential diagnosis of nasal polyps in children according to the age:

- Child under 2 – differentiation for congenital malformations (encephalocele, cystic dermoid);
- Child 2 years old or older – CF needs to be excluded as it is the most common cause of nasal polyps in a child.

Nasal examination

Examination of the nasal cavity in children is challenging and requires a lot of experience and a proper approach to the child. The anterior rhinoscopy start with the raising the tip of the nose (younger children have wider, round nostrils, which facilitates assessment of the inferior nasal concha). You can use an otoscope too. But anterior

rhinoscopy gives an insufficient information for diagnose CRS with nasal polyps, therefore endoscopic examination should be performed with a 2.7 mm diameter rigid endoscope in younger children. Nasal rigid endoscopy is more frequently diagnostic tool than a flexible endoscopy for different nasal and nasopharyngeal diseases [24].

Diagnostic tests

CT and MRI are radiological methods of choice for visualization of pathological changes in the nose and sinuses of children with CRS with nasal polyps. Before the surgical treatment, CT is preferred because it provides a best visualization of the bone structure. MRI is beneficial in cases of intracranial complications. CT is helpful in the diagnosis of CF, which clearly shows radiological findings such as demineralization and medial displacement of the lateral nasal wall, which is related to pseudomucocele [25-27].

CRS with nasal polyps in children is most often associated with CF [2, 11, 28]. Sweat tests to measure chloride levels or genetic testing for CF is necessary in all children with CRS with polyps of the nose. In children with allergic rhinosinusitis and CRS with nasal polyps, comprehensive allergy tests should be performed, including patient interviews, skin prick tests or specific IgE blood tests using a set of inhaled allergens. Cytological examination of nasal secretions helpful for the eosinophils counting. In case of high eosinophilia in nasal secretion, the treatment of choice is corticosteroids [29].

Treatment methods

Currently, there are recommendation for the following order of treatment for nasal polyps: pharmacological treatment as a first line therapy, followed by surgical treatment in case of failure of pharmacotherapy [6, 7, 25, 30-32]. This is particularly important in children.

Due to the high efficacy of topical corticosteroids, the discussion has ensued about the conservative and surgical treatment of nasal polyps [1, 5]. However, polyps are not homogeneous and therefore uniform therapy is not good for all cases of polyps.

The following treatment methods are recommended for the different types of polyps diagnosed in children (according to Stammberger).

Type I and II. Surgical treatment is an only option for these types. Surgery of choice is FESS. Corticosteroids have virtually no effect, so they are not prescribed before surgery or as monotherapy.

Type III. Initial treatment includes antibiotics, decongestant drugs, and topical corticosteroids. In

case of failure of pharmacotherapy, the patient may qualify for surgical intervention.

Type IV. This type is the most difficult to treat. This type responds well to corticosteroids, especially for long topical courses and short systemic treatment courses. Follow-up should be performed every 1-3 months. If such treatment is ineffective, the patient is eligible for the surgical procedure. After surgery, topical corticosteroids should be continued.

Type V. Pharmacotherapy of this type is similar to type III. However, a treatment of this type of polyps is mainly surgical. For CF, topical and systemic enzymatic therapy is prescribed.

Pharmacotherapy

Systemic and topical corticosteroids are designated as first line drugs in CRS with nasal polyps [5]. Antihistamines, decongestant drugs and sodium cromoglycate give a little benefit. Immunotherapy is used to treat allergic CRS associated with nasal polyps. Antibiotics are necessary in cases of bacterial infection [33].

The effects of corticosteroid therapy depend on the presence or absence of eosinophilia in the tissues, so patients with CRS with nasal polyps in combination with allergy or asthma have better benefits from such treatment. Oral corticosteroids are most effective in the treatment of eosinophil-dominated polyps. The maximum recommended dose for children is 1 mg/kg/day for 5-7 days with an assessment of the treatment's effectiveness after 1-3 weeks. There are studies on the efficacy of topical corticosteroid in the treatment of nasal polyps in pediatric patients with CF. The reported efficacy is 67.7% and most polyps are reported to regress [4]. Controlled studies have shown that intranasal mometasone and fluticasone (furoate and propionate) treatment did not affect the growth of the child and may therefore be considered as a drug of choice in children with CRS with nasal polyps [30, 34, 35]. There are no studies assessing the efficacy of topical corticosteroid therapy in pediatric CRS. Studies showing that topical corticosteroids are effective and safe in children with allergic rhinosinusitis may suggest that they can also be administered in the treatment of CRS. Nasal lavage and use of systemic and topical corticosteroids reduce bronchial hyperresponsiveness in asthmatic children. Nasal lavage is recommended in drops or aerosols (isotonic solutions at body temperature) as it helps to remove secretions and reduce swelling of the nasal mucosa. Complementary treatment consists of constant maintenance of nasal patency (mainly within the ostiomeatal complex), moisturizing of

the mucosa facilitating mucosal ciliary transport, stimulation of secretion, prevention of secretion retention in sinuses and immunomodulatory effects [24].

Surgical treatment

Surgical treatment is used when pharmacotherapy fails to cure inflammation of the sinuses. Findings contained in the international consensus for functional endoscopic sinus surgery (FESS) indications in children include these absolute indications:

- Complete nasal obstruction caused by massive polyposis or medial displacement of the lateral nasal wall
- Orbital abscess
- Intracranial complications
- Choanal polyp
- Mucus retention cyst
- Fungal sinusitis

Surgical treatment in children with frequent exacerbations of CRS is usually limited to a partial ethmoidectomy: removal of the uncinata process and opening of the ethmoid bulla. In CF with giant polyps, extensive ethmoidectomy may be necessary. FESS is a safe and effective method of treating CRS in children with disease refractory to pharmacotherapy. The most common complication is scarring within the ostiomeatal complex in the children. It is recommended that FESS should be used very carefully in children. This treatment has a little effectiveness in children under the age of 3. Perioperative intravenous administration of dexamethasone reduces edema and scar formation, especially in those children with asthma, minor changes in CT, those not exposed to tobacco smoke and those over the age of 6. There were no statistically significant differences in facial growth of treated children by FESS and children who not surgically treated [24]. Surgical procedure in the case of an antrochoanal polyp is based on the endoscopic removal of its nasal and intraosseous parts [3, 6, 7, 9].

Conclusion

Although CRS with polyps in children is rare, physicians should be able to diagnose the disease and select the correct treatment modality.

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