ORIGINAL ARTICLE

Incidence and evolution of nasal polyps in children and adolescents with cystic fibrosis

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Keywords: diagnosis, endoscopy, cystic fibrosis, polyposis, therapy.

Summary

Nasal polyps are a clinical sign of alert for investigating Cystic Fibrosis (CF). **Aims:** To study the incidence of nasal polyps in children and adolescents with cystic fibrosis, its possible association with age, gender, clinical manifestations, genotype and sweat chlorine level, and its evolution with topical steroid therapy. **Methods:** Clinical symptoms, sweat chlorine level and genotype were studied in 23 cystic fibrosis patients. Nasal polyps were diagnosed by nasal endoscopy and treated with topical steroids during 6 months, followed by a second nasal endoscopy. Fisher test was used for statistical analysis. **Results:** Nasal polyps were found in 39.1% of the patients (five bilateral, four unilateral), all older than six years, recurrent pneumonia in 82.6%, pancreatic insufficiency in 87% and malnutrition in 74%. No association was seen between nasal polyps and sweat chlorine level, genotype, clinical sings of severity and nasal symptoms. Seven patients improved in their nasal polyps with topical steroids, six showed complete resolution. **Conclusion:** The study showed a high incidence of nasal polyps in older children, who span the entire range of clinical severity, even in the absence of clinical nasal symptoms. Topical steroid therapy showed good results. An interaction among pediatricians and otolaryngologists is necessary for diagnosis and follow-up.

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Paper submitted to the ABORL-CCF SGP (Management Publications System) on October 4th, 2006 and accepted for publication on February 10th, 2007. cod. 3434.

INTRODUCTION

Cystic fibrosis (CF) is the most common lethal genetic disease in Caucasians, of autosomal, recessive transmission, at a rate of 1:2000 live births in this population[1,2], in Brazil, the incidence is of 1:9500 in Paraná[3], 1:8700 in Santa Catarina[4] and 1:10.0005 in Minas Gerais.

The variable severity associated with the clinical manifestations (distinctive phenotypes) depends partially on the genotype and results from the obstructive phenomena, thus characterizing the cystic fibrosis:

1. Chronic suppurative obstructive pulmonary disease;

2. Pancreatic insufficiency with digestion problems and malabsorption, resulting in secondary malnutrition;

3. Increased concentrations of chlorine and sodium, and age.

4. Adult male infertility.

Symptoms onset varies broadly, depending on mutation type, homozygote patients for the genetic mutation ▲F508 start having symptoms in the first 2-4 months of life. The classic clinical picture starts with dry cough, tachypnea, mild intercostal pulling, or, it may manifest itself as acute infection, like bronchiolitis. The clinical course evolves with recurrent pneumonia. Together with all of this, the patient has difficulty gaining weight, despite a voracious appetite, enlarged and more frequent foul-smelling defecation, diarrhea or steatorrhea (oily feces) [1,2].

Cystic fibrosis is diagnosed based on at least two of the four clinical-laboratorial aspects: family history of cystic fibrosis, pancreatic insufficiency, chronic suppurative obstructive pulmonary disease and high levels of chlorine and sodium (>60mEq/l) in their sweat secretion. Other clinical data that suggest the diagnoses are: meconium ileum and/or intestinal obstruction with atresia, deficient weight-height development, heat stress, chronic pansinusitis, nasal polyps, volvus and intusception, and azoospermia[6,7].

Clinical manifestations in the upper airways (UAW) happen to 100% of the patients, including recurrent sinusitis, rhinitis and/or nasal polyposis[8-11]. The incidence of nasal polyps has been reported in 6 to 48% of the cases[12,13], by the time cystic fibrosis is diagnosed, about 4% of the patients have some symptoms associated with nasal polyps. It is believed that about 14% of the patients with cystic fibrosis will require surgery to treat the polyps[8,10,11,14].

Based on these data from the literature, the departments of pediatric pneumology and otorhinolaryngology of the Botucatu Medical School - UNESP, decided to assess UAW involvement in patients with cystic fibrosis in the outpatient ward.

OBJECTIVE

The general goal of our paper was to assess nasal polyp incidence through endoscopy in children and adolescents with cystic fibrosis being followed in the outpatient ward. The specific goals were:

1- to assess age, gender, clinical symptoms and the genetic mutation of these patients, and the association between these data and nasal polyposis;

2- assess polyposis evolution with topical steroids.

PATIENTS AND METHODS

The present contemporary cross-sectional and prospective cohort was approved by the Ethics in Research Committee of the Botucatu Medical School - UNESP, under protocol # 1743/2005. The parents/guardians signed an Informed Consent Form.

In 2005 we assessed the 23 patients being followed at the Cystic Fibrosis Reference Center Outpatient Ward, with ages ranging between 1 year and 9 months and 22 years and 8 months.

From their charts, we obtained data related to gender, age, clinical manifestations of CF such as meconium ileum, malnutrition, pancreatic insufficiency and repetition pneumonia, and laboratorial exams to confirm CF, such as quantitative analysis of ion content in sweat[7] and genetic studies. All patients underwent otolaryngological evaluation and suffered nasal endoscopy. During the consultation we obtained information related to nasal obstruction, oral breathing, asthma and sinusitis.

Nasal endoscopy was carried out under topical anesthesia, using a flexible Storz pediatric bronchoscope of 2.4mm in diameter, or a rigid 30º, 2.4mm Storz scope.

In the nasal exam we described whether or not polyps were present, following the staging classification proposed by Johansson et al.15 (level 0 - absent, level I - polyp in the middle meatus, level II - polyp going through the middle turbinate with clear nasal floor, level III - polyp filling up the entire nasal cavity, whether or not there is secretion and its color, nasal mucosa aspect (color, edema, degeneration).

Those patients with nasal polyposis were prospectively followed up and submitted to clinical treatment with topical steroids in the habitual dose (mometasone 200 mcg per day) for 6 months. After this period, the nasal endoscopic exam was repeated.

For statistical analysis, the data obtained were described in their mean and standard deviation values. Age, gender, clinical symptoms and genetic mutations were associated with the presence of polyps. We used Fisher's Exact Test, at a significance level of $p<0.05$.

RESULTS

The median age of the 23 patients was of six years and four months, and 20 of them were males.