Braz J Otorhinolaryngol. 2016;xxx(xx):xxx–xxx

Brazilian Journal of
OTORHINOLARYNGOLOGY
www.bjorl.org

ORIGINAL ARTICLE

Nasal polyposis in cystic fibrosis: follow-up of children and adolescents for a 3-year period☆

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Received 21 February 2016; accepted 16 September 2016

KEYWORDS
Polyposis;
Cystic fibrosis;
Diagnosis;
Endoscopy;
Therapy

Abstract
Introduction: Nasal polyposis is often found in patients with cystic fibrosis.
Objective: To assess the incidence of nasal polyposis, the response to medical treatment, recurrence and the need for surgical intervention in children and adolescents with cystic fibrosis during a three-year follow-up.
Methods: Clinical symptoms (pulmonary, pancreatic insufficiency, malnutrition, nasal obstruction), two positive sweat chloride tests, and genotype findings in 23 patients with cystic fibrosis were analyzed. All patients underwent nasal endoscopy every 12 months from January 2005 to December 2007, to assess the presence and grade of Nasal Polyps. Nasal polyposis, when present, were treated with topical corticosteroids for 6–12 months, with progress being evaluated within the 3 years of follow-up.
Results: In the first evaluation, nasal polyposis was diagnosed in 30.43% of patients (3 bilateral and 4 unilateral), recurrent pneumonia in 82.6%, pancreatic insufficiency in 87%, and malnutrition in 74%. The presence of nasal polyposis was not associated with chloride values in the sweat, genotype, clinical signs of severity of cystic fibrosis, or nasal symptoms. In the three-year period of follow up, 13 patients (56.52%) had at least one event of polyposis, with the youngest being diagnosed at 32 months of age. Only one patient underwent surgery (polypectomy), and there was one diagnosis of nasopharyngeal carcinoma.

☆ Please cite this article as: Weber SA, Iyomasa RM, Corrêa CC, Florentino WNM, Ferrari GF. Nasal polyposis in cystic fibrosis: follow-up of children and adolescents for a 3-year period. Braz J Otorhinolaryngol. 2016. http://dx.doi.org/10.1016/j.bjorl.2016.09.005

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http://dx.doi.org/10.1016/j.bjorl.2016.09.005

Conclusion: The study showed a high incidence of nasal polyposis. Monitoring through routine endoscopy in patients with cystic fibrosis, even in the absence of nasal symptoms, is highly recommended. The therapy with topical corticosteroids achieved good results. Thus, an interaction between pediatricians and otolaryngologists is necessary.

PALAVRAS-CHAVE
Polipose;
Fibrose cística;
Diagnóstico;
Endoscopia;
Terapia

Polipose nasal em fibrose cística: seguimento em crianças e adolescentes durante um período de 3 anos

Resumo
Introdução: A polipose nasal é frequentemente encontrada em pacientes portadores de Fibrose Cística.
Objetivo: Avaliar a incidência de Polipose Nasal, a resposta ao tratamento clínico, a recorrência e a necessidade de intervenção cirúrgica em crianças e adolescentes com Fibrose Cística durante um seguimento de 3 anos.
Método: Os sintomas clínicos (pulmonar, insuficiência pancreática, desnutrição, obstrução nasal), duas pesquisas de cloro no suor positivas e genótipo de 23 pacientes com fibrose cística foram descritos. Todos os pacientes foram submetidos à endoscopia nasal a cada 12 meses durante o período de janeiro de 2005 a dezembro de 2007, para avaliação de presença e grau de Polipose Nasal. A Polipose Nasal, quando presente, foi tratada com corticosteroide tópico de 6 a 12 meses, e avaliada a evolução nos 3 anos de seguimento.
Resultados: Na primeira avaliação, a Polipose Nasal foi diagnosticada em 30,43% dos pacientes (3 bilaterais e 4 unilaterais), pneumonia recorrente em 82,6%, insuficiência pancreática em 87% e a desnutrição em 74%. A presença de Polipose Nasal não se associou aos valores de cloro no suor, genótipo, sinais clínicos de gravidade da Fibrose Cística ou sintomas nasais. Nos 3 anos de seguimento, 13 pacientes (56,52%) apresentaram pelo menos um evento de Polipose, sendo o mais jovem diagnosticado aos 32 meses de idade. Apenas um paciente foi submetido à cirurgia (polipectomia), e houve um diagnóstico de carcinoma da nasofaringe.
Conclusão: O estudo mostrou alta incidência de Polipose Nasal. O acompanhamento por meio de exames endoscópicos de rotina em pacientes fibrocisticos, mesmo na ausência de sintomas nasais, é altamente recomendado. A terapia com corticoide tópico mostrou bons resultados. Sendo assim, faz-se necessária a interação entre pediatras e otorrinolaringologistas.

Introduction

Cystic fibrosis (CF) is an autosomal recessive disease that affects the exocrine glands, involving multiple organs and progressing chronically and progressively. It is the most common lethal genetic disease in Caucasians, with an average frequency of 1:2000 live births.[1,2] In Brazil, studies have revealed an incidence of 1:9500 live births in the state of Parana,[3] 1:8700 in Santa Catarina[4] and. 1:10,000 in Minas Gerais.[5]

Respiratory infections leading to ultimate respiratory failure are the leading causes of death in CF patients. However, mortality has been reduced in recent years due to earlier diagnosis, greater attention to prophylaxis of recurrent airway infections, and better control of patients in specialized services.[1,2]

CF diagnosis is based on clinical and laboratorial criteria: family history of CF, pancreatic insufficient/pancreatic sufficient, chronic obstructive suppurative lung disease, and two high sweat chloride tests (>60 mEq/L) and/or detection of genetic mutations described in CF. Other clinical data that suggest the disease are: meconium ileus and/or intestinal atresia, hyponatremic dehydration, edema and hypoalbuminemia, chronic panrhinosinusitis, nasal polyposis (NP), volvulus, intussusception, bronchiectasis of unknown etiology, and azoospermia.[6,7]

Upper airway (UAW) impairment such as recurrent rinorhinosinusitis, rhinitis and/or NP occurs in over 90% of patients.[8–16] The incidence of NP, in particular, has been reported in 6–48% patients,[17,18] and is symptomatic in about 4% patients at diagnosis of CF.[8,10,11,19] The literature estimates that 14% of patients require surgical treatment of NP.[8,10,11,19]

To date, the pathophysiology of NP is still unknown.[20,21] Allergic processes have been reported as a possible cause of NP, but the prevalence of atopy in patients with CF is not higher than in the general population.[22]