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EAR, NOSE AND THROAT FINDINGS IN CYSTIC FIBROSIS PATIENTS*

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SUMMARY: Özçelik T, Özgirgin N, Özçelik U, Göçmen A, Gürcan B, Kiper N. (Department of Otolaryngology, Bayındır Medical Center, and the Chest Diseases Unit, Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey). Ear, nose and throat findings in cystic fibrosis patients. Turk J Pediatr 1997; 39: 51-54.

An ear-nose-throat survey was carried out on 30 children with cystic fibrosis (CF) between two and 17 years of age. There were three (10%) confirmed cases of secretory otitis media. One child had previous surgery for polyps. Ten children had chronic pansinusitis mildly responsive to medical therapy. Hypertrophy of the nasal conchae was detected in eight cases. The data showed that the risk of ear-nose-and-throat disease was increased in CF patients, but audiologic problems are not very common in CF patients. CF should be kept in mind in children who have nasal polyps, hypertrophied turbinates or sinusitis unresponsive to medical therapy. *Key words: cystic fibrosis, paranasal sinuses, nasal polyposis, ear problems.*

Cystic fibrosis (CF) is characterized by a defect in cyclic adenosine monophosphate (cAMP)-regulated chloride conductance, and is caused by mutations in the gene coding for the cystic fibrosis transmembrane conductance regulator (CFTR)[1]. The exocrine dysfunction of the acinotubular glands is manifested by abnormalities in the composition and physiochemical behavior of the secretions. The mucus produced is 30 to 60 times more viscid than normal.

Clinical symptoms commonly affect the gastrointestinal, upper and lower respiratory systems. The otolaryngological aspects of CF are dominated by involvement of the paranasal sinuses. This involvement is manifested as delayed development of the frontal sinus and opacification of the maxillary, sphenoid and ethmoid sinuses, as well as nasal polyposis. Middle ear problems can also be seen[2].

In this study we assessed the incidence of upper respiratory disease, in particular middle ear problems, in a group of CF children. In addition to clinical assessment, impedancemetry was used to estimate the incidence of secretory otitis media.

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Material and Methods

The study included 30 patients between ages two and 17. Patients diagnosed with CF had at least two sweat chloride values greater than 60 mEq/L and clinical and other radiological findings[3]. The patients were selected upon referral to the ear, nose and throat (ENT) department. The prerequisite for the patient to be included in the study was that he was free of acute illness and able to cooperate in an impedancemetric evaluation.

Each patient's parents completed a questionnaire on ENT symptoms, family history, recent respiratory symptoms and treatment. A full ENT examination, including bilateral otoscopy using operating microscope and pneumatic otoscope, rhinoscopy and endoscopic evaluation of the nasopharynx in cooperative patients, was carried out. Secretory otitis media was diagnosed clinically when fluid was suspected behind a dull or yellowish drum; drum retraction or reduced mobility on pneumatic otoscopy has also accepted as suggestive. The diagnosis of sinusitis was confirmed by x-ray.

Results

Three cases (10%) were clinically suspected of having secretory otitis media. Negative middle ear pressure and the absence of acoustic reflexes suggested the presence of effusion. Two cases resolved at the end of medical treatment. Ventilation tube insertion was planned for the third case.

One case (3%) had previous surgery for bilateral nasal polyps. Eight other cases (26%) were considered to have an abnormal nose with either purulent nasal discharge, congested mucosa or polypoid nasal conchae.

Ten (33%) cases had sinusitis symptoms such as facial pain, sinus headache or nasal obstruction sinusitis was confirmed by the detection of opacification of the paranasal sinuses on radiographs (Fig. 1).

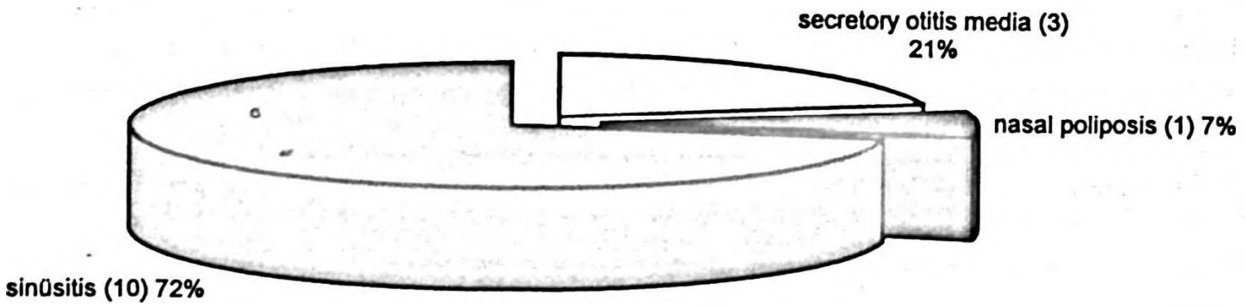

Fig. 1: Main ear nose and throat pathologies in CF patients.

Discussion

The literature is conflicting on whether there is an increased incidence of conductive deafness in CF, while an increased incidence of eustachian tube dysfunction and serious otitis media have been suggested[4]. The mucosal epithelium of the middle ear and the eustachian tube are in direct continuity with the upper respiratory tract. Thus it might be assumed that children with CF would have a higher than usual incidence of middle ear disease. Kulczycki et al.[5] reported a 26 percent incidence of conductive hearing loss in 70 children with CF. Jerger and Neely[6] observed a six percent incidence and Taylor et al.[7] found none. Kramer[2] indicated that the incidence of otologic disease in CF patients did not appear to be statistically greater than in the general population. Local experience does not suggest a significant increase in hearing problems in CF, although it could be argued that since we do not routinely perform audiograms, cases with hearing impairment could be missed. In our series, we found a 10 percent incidence of conductive hearing loss. The low incidence of serous otitis media in our own series may be attributable to the use of antibiotics for the slightest respiratory infection.

Nasal polyposis in cystic fibrosis children has been studied by several authors. six to 10 percent of cystic fibrosis patients have been found to have nasal polyps, most of which were bilateral[7]. Nasal polyposis was the first sign in one case (3.3%), and CF was diagnosed afterwards. Also, a hypertrophied, purplish turbinate accompanied by purulent rhinitis are features of CF that we found in eight (26%) children. The appearance is claimed to differ from that of the engorged turbinates of allergic rhinitis. Rulon et al.[8] reported that approximately 28 percent of the patients they studied had nasal polyps. However, to date there is no information as to why children with cystic fibrosis develop nasal polyps. It must be noted that nasal polyps in children without cystic fibrosis are rare, so in the presence of nasal polyps cystic fibrosis should always be considered in the differential diagnosis. Recurrence after surgery is the rule rather than the exception; spontaneous resolution may occur, many cases are asymptomatic, and the symptoms are tolerated with minimal discomfort in others. Nasal corticosteroids may be used in the treatment of nasal polyps and in order to prevent recurrences after the removal of the polyps[9].

Opacity of the paranasal sinuses on radiographs is an almost universal finding in cystic fibrosis; however, only rarely do symptoms arise from the sinuses. The symptoms described are sinus headache, facial pain and swelling. The possible causes of sinus involvement are excessive or unusually viscid mucus, infection and allergy[10]. Surgery is frequently unsuccessful and is best avoided[11]. In ten of our cases (33%), infection was detected clinically and radiologically in one or more sinuses and was mildly responsive to medical therapy.

In conclusion, the present data suggest that despite frequent involvement of the upper airways, audiological problems are not as common in CF patients as one would expect.

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