Nasal and paranasal disease in adult cystic fibrosis patients

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ABSTRACT: Children with cystic fibrosis frequently have nasal polyps and sinusitis. This study addresses (para-)nasal disease in 39 adult cystic fibrosis patients. Fifteen patients (39%) had recently had serious nasal symptoms and 26% sinusitis. Seventeen (44%) had nasal polyposis. Almost all sinus radiographs taken showed opacification, which was unrelated to symptoms. Polypectomies and antral irrigations were usually ineffective, whilst more extensive surgery generally gave better results.

It is concluded that a substantial number of adult cystic fibrosis patients frequently have upper airway symptoms. Sinus radiographs have little or no diagnostic value. Treatment of (para-)nasal disease in cystic fibrosis patients can be difficult; a guideline for treatment is suggested, calling for simple interventions coupled with intranasal steroids and nasal irrigation in early disease and for endoscopic to radical sinus surgery in recurrent advanced disease.

Keywords: Adult cystic fibrosis nasal paranasal polyposis sinusitis

Patients, materials and methods

Cystic fibrosis (CF) is the most common genetic disorder in Caucasians. Exocrine gland dysfunction leads to chronic obstructive pulmonary disease (COPD) and pancreatic insufficiency, eventually resulting in patient’s death. Whereas in 1970, the median age of survival was under 10 yrs, it now approaches 30 yrs in many CF centres, due to improved pulmonary treatment. Therefore, physicians will more often be confronted with adult CF patients.

Otolaryngologic complications in children with cystic fibrosis are well-documented. Nasal polyposis is reported in 7–26% of the younger patients [1–4]. Sinusitis occurs more frequently than in a normal population. Incidence, however, is unclear, largely because sinus radiographs are abnormal in almost all CF patients, and other criteria are hardly ever listed in reports addressing this matter.

Few reports deal with the otolaryngologic complications in adult CF patients. Nasal polyposis has been reported in 48% of adult patients [5], but the incidence may also be lower in adults than in children [2, 6].

There is no consensus in the literature on the treatment of otolaryngologic manifestations, while high recurrence rates are generally reported. The main purpose of this study was to evaluate the incidence of nasal and paranasal disease in adult CF patients and to establish whether these patients should be considered as a subpopulation that requires special attention for the otolaryngologist. Furthermore, results of earlier treatment and the contribution of sinus radiographs were analysed.

All 43 CF patients currently receiving treatment at the Department of Pulmonary Medicine of the University Hospital Rotterdam, Dijkzigt, were asked to take part in the study. Three patients did not wish to participate for various reasons and one patient was not considered to be representative. Thirty nine patients remained, aged 17–40 yrs (mean 26 yrs). Eighteen were males, 21 females. In all patients the diagnosis had been confirmed by sweat test. All patients visited the out-patient clinic of the otolaryngology department. History was taken using a standard questionnaire containing questions concerning medication, past or present nasal or sinus symptoms, visits to an otolaryngologist, treatment and results, relationship of symptoms with age or other factors, and other events. A routine Ear-Nose-Throat examination was performed and, if permission was given, sinus radiographs were made.

Special attention was paid to the present pulmonary status. Pulmonary status was scored as poor,
intermediate or relatively good, according to the following criteria. A poor pulmonary status was defined as having 4 out of 5 of the following: forced expiratory volume in one second (FEV$_1$) $<$60% predicted; forced vital capacity (FVC) $<$60% predicted; number of hospital admissions related to pulmonary exacerbations during the previous 2 yrs $>$3; continuous antibiotics; continuous steroids. A relatively good pulmonary status was defined as having 4 out of 5 of the following: FEV$_1$ $\geq$60% predicted; FVC $\geq$60% predicted; number of hospital admissions related to pulmonary exacerbations during the previous 2 yrs $\leq$1; discontinuous antibiotics; no steroids. Remaining patients were classified as intermediate.

The sinus radiographs were scored by J.D.F.K. and R.M.L.P. together. All sinuses were scored separately. A sinus was scored as "clear" if no mucosal swelling could be seen, "totally opacified" if no air contour was present, and "partially opacified" in the remaining cases. For analysis, "present" was defined as during the previous 6 months, while "past" was defined as longer than 6 months ago. Since it was impossible to retrospectively confirm the diagnosis of sinusitis, our criteria for having had sinusitis were: 1) being treated for sinusitis by an otolaryngologist; or 2) actively remembering having had symptoms suggestive of sinusitis i.e. severe periorbital headache worsening on bending over, combined with nasal obstruction and discharge.

Results

Visits to an otolaryngologist

Thirty out of the 39 patients (77%) had been treated in the past (>6 months ago) by an otolaryngologist for nasal or paranasal disease. Twenty three patients had been treated for severe nasal obstruction and 19 had had nasal polyposis. Fourteen patients had been treated for sinusitis. Nine patients had visited an otolaryngologist during the previous 6 months for nasal obstruction, headaches or nasal polyposis. Twenty three patients had nasal or paranasal disease. Twenty three patients had been treated for severe nasal obstruction and 19 had had nasal polyposis. Fourteen patients had been treated for sinusitis. Nine patients had visited an otolaryngologist during the previous 6 months for nasal obstruction, headaches or nasal polyposis. Of the 26 patients currently older than 20 yrs, 13 had been treated by an otolaryngologist after their twentieth birthday.

Symptoms

Fifteen patients (39%) complained of frequent or continuous nasal obstruction and 12 had frequent or continuous nasal discharge during the previous 6 months. Seven of 39 had symptoms suggestive of sinusitis during this period.

Physical examination

Seventeen patients (44%) presented with nasal polyposis. In two the polyps were classified as minor.

The remaining 15 had either intermediate-sized or large nasal polyps. Eight of these 17 patients (21% of the whole population) complained of nasal obstruction. Retracted eardrums were found in one patient.

Sinus radiographs

Thirty seven sinus radiographs were made. In one patient all sinuses were clear; this patient had never had any nasal or paranasal symptoms. In two patients there was partial opacification of only one maxillary sinus. In the remaining 34 patients, there was at least partial opacification of all sinuses. There was no correlation with symptoms.

Pulmonary status

Nine patients were classified as presently having a poor pulmonary status; eight were classified as relatively good, the remaining as intermediate. No correlation was found between pulmonary status and past or present nasal or paranasal disease. Also, although many patients could not give a clear answer and it was poorly recorded, no correlation seemed to exist between pulmonary exacerbations and nasal or paranasal disease.

Symptoms and age

Five patients reported that (para-) nasal symptoms increased with age, 12 experienced decrease of symptoms with age, usually after treatment and in the remaining patients the symptoms remained constant. An age-peak for symptoms could not be demonstrated in our patient group, nor could a correlation be found between age at CF diagnosis and nasal or sinus disease.

Treatment

Treatment and results were analysed. Due to small numbers, it was not possible to analyse treatment during adulthood separately. Over 50 polypectomies had been performed in 14 patients. This had resulted in long-term relief (longer than 1 year) in two instances. The remaining polypectomies had no or only short-term effect. More than 70 antral irrigations had been performed in at least 15 patients. In two patients this gave long-term relief but in most cases the effect was temporary. A Caldwell Luc intervention was performed in six patients, with good long-term result in two. In nine patients rhinoantrostomies were made, resulting in long-term relief in three cases. In four patients rhinoantrostomies were combined with an ethmoidectomy; three patients remained disease free. Radical sinus surgery,
consisting of a transantral ethmoidectomy, sphenoidectomy and removal of the lateral nasal wall, as first described by Denker [7], was performed in two patients, with good results so far.

Discussion

Cystic-fibrosis is a disorder affecting multiple organs. Otolaryngologic complications in children are well-documented and consist of nasal polyposis and frequent sinusitis. Although one could expect a high incidence of middle ear disease, due to mucosal pathology and frequent rhinitis or sinusitis, this had not proved to be the case [4, 8, 9].

Little is known about nasal and paranasal disease in adult CF patients. A study was undertaken to analyse the current and past nasal and paranasal status of the adult CF patients being treated at our institution.

The majority of the patients had been treated in the past by an otolaryngologist for nasal or sinus disease. Half of all patients were treated for nasal polyposis, while 36% had episodes of sinusitis. Approximately 25% of all our CF patients had visited an otolaryngologist during the previous 6 months, while more than 40% had serious symptoms during this period. Physical examination revealed nasal polyposis in approximately the same percentage. This is consistent with earlier findings [5] and is considerably higher than usually reported in children [1–4]. This might be attributed to the fact that nasal polyposis is frequently asymptomatic and will often only be discovered when actively looking for polyps. Approximately half of our CF-polyposis patients were symptom free.

Sinus radiographs in CF patients are known to be abnormal in >90% [1, 4, 10]. Opacification of sinuses on X-ray does not infrequently lead to the diagnosis of sinusitis. This is, in our view, incorrect. In 92% of our patients, at least partial opacification of both maxillary sinuses was found. This was not related to symptoms. Therefore, we feel that sinus radiography for diagnostic purposes is unnecessary.

A correlation between pulmonary status and nasal or sinus disease could not be demonstrated in our study. Drake-Lee and Pitcher-Wilmott [11] reported a significantly better preserved vital capacity in young CF patients with nasal polyposis, compared to nonpolyposis patients. Peak flow rate, forced expiratory flow, and residual volume to total lung capacity ratio were, however, similar in both groups. In our study, data were insufficient to demonstrate a possible correlation of pulmonary exacerbations and (para) nasal disease. UMETSU et al. [12] recently reported an improvement of pulmonary status after radical sinus surgery in a small number of patients.

STERN et al. [2] found a peak of nasal polyposis during mid-childhood. The high incidence of polyposis in our adult patient group does not concur with this finding. Furthermore, we could not demonstrate an age-peak for symptoms in our population, nor could a gradual increase or decrease with age or correlation with age at CF diagnosis be found.

To date, pathophysiology of nasal polyposis in CF patients remains unclear. It is possible that chronic mucopurulent discharge causes irritation of the mucosal lining, resulting in polyp formation. However, it was demonstrated that allergy does not play a major role [1, 2, 13], nor has an extensive infiltration of eosinophils been demonstrated [14]. Nevertheless, DONALDSON and GILLESPIE [15] found a positive effect on nasal symptoms with intranasal beclomethasone dipropionate in both polyposis and nonpolyposis CF patients. Others report little effect [2, 4].

Debate continues on surgical treatment. Some authors [1, 3, 13] advocate simple polypectomies, although recurrence rates of 60–80% are usual. Others [4, 9, 16] prefer more aggressive management, often polypectomy in combination with ethmoidectomy and Caldwell Luc intervention. In our experience, simple polypectomy for polyposis or antral irrigation in case of sinusitis has no long-term effect. Results from more extensive surgery were generally better, but numbers are small. A few endoscopic sinus procedures have been performed recently, but no results are yet available. DUPLECHAIN et al. [17], however, reported favourable results regarding endoscopic sinus surgery in CF children.

A rough guideline for management of nasal and paranasal disease in CF patients is suggested as follows. On first or second presentation of symptoms, a simple polypectomy or antral irrigation is performed, followed by administration of intranasal steroids and frequent nasal irrigations. In case of recurrent disease, meticulous endoscopic sinus surgery is performed. Post-operatively, intranasal steroids are administered and the patient is advised to irrigate the nose frequently with saline. If endoscopic sinus surgery proves unsuccessful, radical sinus surgery consisting of transantral ethmoidectomy, sphenoidectomy and removal of the lateral nasal wall may be considered. This procedure is, however, not recommended in children.

Conclusion

A high incidence of nasal polyposis and episodes of sinusitis in adult cystic fibrosis patients, as well as the difficulty of treatment of nasal and paranasal disease in these patients, require special attention from otorhinolaryngologists, who should preferably be working in a CF-team. Sinus radiographs appear to have little or no value in the diagnosis of sinusitis. Since many patients with nasal polyposis are asymptomatic, and no correlation with pulmonary status could be demonstrated, treatment should only be instituted if symptoms are present. Aggressive management of nasal and sinus disease is indicated in the case of frequent recurrent disease.
References