Upper respiratory tract in cystic fibrosis

Ear-nose-throat survey of 50 children

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Taylor, B., Evans, J. N. G., and Hope, G. A. (1974). *Archives of Disease in Childhood*, 49, 133. **Upper respiratory tract in cystic fibrosis: ear-nose-throat survey of 50 children.** An ear-nose-throat survey was carried out on 50 children aged 4 to 10 years with cystic fibrosis. 10% were shown to be transiently deaf, associated with eustachian tube dysfunction. There were no confirmed cases of secretory otitis media. 10 children (20%) were found to have nasal polyps or gave a history of polypectomy. Though a high incidence of middle ear problems in children with cystic fibrosis has been reported from the U.S.A., neither deafness nor secretory otitis media was found to be a problem in the group studied.

The upper respiratory tract, as well as the lower, is often abnormal in many children with cystic fibrosis (CF). They seem to have obstructive nasal symptoms more frequently than normal children, and nasal polyps, otherwise rare in childhood, are not infrequently found in CF.

The respiratory tract is abnormally liable to infection and though there is no direct evidence of abnormal mucus in CF, the mucosal secretions seem abnormally sticky. Children with CF might be expected to have a higher than usual incidence of middle ear problems.

Secretory otitis media (SOM) is not uncommon in normal child population surveys. Watson (1969) reported an incidence of 3.6% among 1700 5-year-olds. The diagnosis was based on clinical examination and pure tone audiometry. Using tympanometry, Brooks (1969) reported an even higher incidence among 1053 children; nearly 20% of 5-year-olds, 10% of 6-year-olds, 5% of 7-year-olds, and 2 to 3% of 9- to 11-year-olds.

Among CF patients, Forcucci and Stark (1972) found 30% of their children to have SOM, Jerger and Neely (1971) found only 4%, and Siegel and Taylor (1970) found none among 23 patients.

The diagnosis of deafness is usually based on audiometric criteria. Among CF patients, Kulczyki *et al.* (1970a) and Kulczyki, Herer, and Butler (1970b) in two studies found 25% and 27% of 41 and 70 children to be deaf with 15 decibel or greater

hearing loss on two or more test tones. Using the same criteria, Forcucci and Stark found 39% of their cases to be deaf.

Jerger and Neely (1971) considered these criteria to be unrealistic—if applied to their CF patients, 55% would have been deaf—and used the less stringent criteria of the American Academy of Ophthalmology and Otolaryngology (any two thresholds on either ear greater than 25 decibels hearing level). They found only 8% of their patients to be deaf by this standard. Nevertheless, there do seem to be regional differences for deafness and SOM in CF patients. Siegel and Taylor (1970) found none of their 23 patients to suffer from either condition. These results from various centres are summarized in Table IV.

It was decided to assess the incidence of upper respiratory disease, and in particular middle ear problems, in a group of English CF patients. In addition to clinical assessment and audiometry, tympanometry was used to estimate the incidence of secretory otitis media.

Subjects and methods

The study included 50 patients aged 4 to 11. Age distribution is shown in Table I. All the children between 5 and 10 years old regularly attending the CF clinic at this hospital were studied, and in addition 6 of the more co-operative 4-year-olds. No other selection was exercised. The respiratory tract status of the children therefore varied widely, some being completely symptom free and others being in respiratory failure when examined.

TABLE I
Age distribution of 50 patients

Years	4-5	6-7	8-9	10-11
No.	15	19	13	3

The parents answered a prepared questionnaire on ear, nose, and throat symptoms, family history, recent respiratory symptoms, and therapy.

Full ENT examination was carried out by one of the two ENT surgeons. Secretory otitis media was diagnosed clinically when fluid was seen behind a lack-lustre or yellowish drum; drum retraction or reduced mobility on pneumatic speculum testing were also taken as suggestive.

Audiometry was performed using a Kamplex pure tone audiometer, and tympanometry using a Madsen Impedance Bridge, type ZO 70. The criterion for deafness used was that adopted by the Schools Audiometric Screening Service. A child was deaf if there was more than 20 decibel hearing loss on two successive frequencies over the speech range. This is a level for diagnosing deafness midway between that used by other workers reporting results in CF.

Tympanometry distinguishes between fluid in the middle ear and the less serious eustachian tube dysfunction, where the tube is temporarily blocked, leading to pressure inequalities inside the middle ear compared with the outside. The Fig. shows examples of tympanometry (a) with a normal ear, (b) with eustachian tube dysfunction where the peak is displaced towards the side of negative pressure, and (c) with secretory otitis media where the curve is very flat. Any child in whom any abnormality was detected was completely reassessed at a later date.

Results

ENT symptoms. These were recorded at probably normal population incidence. 20 children had had ear ache and 11 had had ear infections at some time in their lives. Only one case had had a discharging ear. 17 had had at least one sore throat with fever in the preceding year, and some had had as many as 10 episodes. None had undergone tonsillectomy. 2 had undergone adenoidectomy for symptoms of nasal obstruction. 8 cases gave a history of sinusitis; 3 of these had polyps, 3 others had signs of nasal infection. There was no immediate family history of deafness.

During the month preceding the study, 38 of the children had received antibiotics (cloxacillin, trimethoprim/sulphamethoxazole, ampicillin, clindamycin, erythromycin, carbenicillin, or gentamicin aerosol). 21 gave a history of a recent cold.

Examination. Two cases were clinically

FIG.—Tympanometry. (a) Normal middle ear pressure with a peak at greater than -100 mm H_2O . (b) Eustachian tube dysfunction. Peak at more negative pressure. (c) Secretory otitis media. Flat curve due to fluid in the middle ear.

suspected of having secretory otitis media (Table II). However, the first case was not deaf and showed only bilateral eustachian tube dysfunction on tympanometry. The second case, aged 4, was unfortunately not able to co-operate with audiometry or tympanometry at the initial examination. At subsequent assessment both children were normal in all respects, giving no confirmed cases of SOM.

Ten cases of unilateral or bilateral nasal polyps were found. 4 of these had had previous polypectomies, 6 cases were largely symptomless. 11

TABLE II
Suspected secretory otitis media

	Clinical	Audiometry	Tympanometry
Case 1 Initial assessment	(R) Ear	Normal	Eustachian tube dysfunction
Case 2 Initial assessment	(L) Ear	Failed (no co-operation)	Failed (no co-operation)
Subsequent assessment in both cases	Normal	Normal	Normal

further cases were considered to have an abnormal nose with either purulent nasal discharge or congested mucosa.

Audiometry and tympanometry. 5 children failed the criteria taken for deafness (Table III). One of these was the case with clinical secretory otitis media who was failed initially because the test was unreliable. The other 4 were deaf in only one ear due to eustachian tube dysfunction. This was associated with upper respiratory infection in 3, and a resolving otitis media in the remaining one. All of these cases were completely normal at subsequent examination. 2 other cases had eustachian tube dysfunction, but neither was deaf, and subsequent examination was normal. There were no cases of sensori-neural deafness.

Discussion

Most of the difference in the incidence of deafness in CF reported by various authors seems to be due to

the criteria taken for deafness (Table IV). Using the criteria of Kulczycki *et al.* (1970a, b), 54% of our children would be deaf, including 4 of the younger subjects who were passed as normal with sweep audiometry at 20 decibels. Using the American Academy of Ophthalmology and Otolaryngology criteria (Jerger and Neely, 1971), none of our children would be deaf (excluding the case who was unable to co-operate with testing). Nevertheless, there does seem to be a regional difference. In Illinois, 16% of CF patients were deaf by the standards of the American Academy; in Texas, using the same criteria, 8%; in California there were no deaf children. Climate, seasonal variation, and social factors have been put forward to explain these differences. The skill of the audiometrician in obtaining the confidence of the children and in testing is a factor of major importance.

Similarly, the incidence of SOM varies from place to place. Probably 13% of the Washington, D.C. patients were affected and 30% of those in Illinois.

TABLE III
Deafness on audiometry and associated findings

Case no.	Impaired audiometry	Tympanometry	Clinical	Subsequent assessment
1	Left ear	Bilateral eustachian tube dysfunction	Upper respiratory tract infection	Normal
2	Right ear	Right eustachian tube dysfunction	Resolving otitis media	Normal
3	Left ear	Bilateral eustachian tube dysfunction	Upper respiratory tract infection	Normal
4	Right ear	Right eustachian tube dysfunction	Upper respiratory tract infection	Normal
5	Failed (no co-operation)	Failed (no co-operation)	Suspected secretory otitis media	Normal

TABLE IV
Reported incidence of deafness and secretory otitis media

Authors	Case no.	Decibel level for deafness (%)		Secretory otitis media (%)
		15	25	
Kulczycki <i>et al.</i> (1970a, b), Washington, D.C.	41 70	27 26	?	?13
Forcucci and Stark (1972), Illinois	31	39	16	30
Siegel and Taylor (1970), California	23	0	0	0
Jerger and Neely (1971), Texas	51	55	8	4
Present study, London	50	54	0	0

However, tympanometry was not performed in these series. In Houston the incidence was low-normal, and there were no confirmed cases in California or in the present study.

The finding of polyps in 20% of cases shows the high incidence of this known complication of CF. Shwachman *et al.* (1962) reported 50 cases (6.7%) from 742 CF patients. None of the 50 children studied had undergone tonsillectomy and only 2 had had adenoidectomy. This finding probably relates to the attitude to tonsillectomy taken by the paediatricians in charge of the clinics they have attended. It may be that their other health problems over-shadowed these symptoms.

Taking Brooks's figures for Manchester school-children as the 'normal' incidence of SOM, 6 of our children might have been expected to show evidence of this condition. Though there are no comparable epidemiological studies for deafness, nearly all patients with SOM would be deaf by our criteria. The reasons for the unexpectedly low incidence of middle ear complications found in this survey are difficult to explain. Patients in our clinic all receive postural drainage for their chests at least daily, which might keep the upper respiratory passages clear. Their exposure to antibiotics is much greater than that of the normal population. 76% of the children had antibiotics within 4 weeks of being

examined. Antibiotics given for chest infections might have an effect on the middle ear mucosa which results in this low incidence of SOM.

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