All patients with cystic fibrosis should have periodic audiologic testing in addition to their other regular physical and laboratory evaluations. Mild conductive hearing losses are more prevalent in children with cystic fibrosis; however, these patients respond promptly to corrective medical and surgical procedures.

Cystic Fibrosis and Hearing

Integrity of the Upper Respiratory Tract as Related to Hearing Sensitivity

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THIS study investigates whether the pathophysiologic disturbances in the upper respiratory tract of cystic fibrosis patients affect their hearing and evaluates the potential relationship between specific involvement of the upper respiratory tract structures and hearing sensitivity. There is a voluminous literature on cystic fibrosis, with more than 2,000 papers which deal with different aspects of the disease, yet no one seems to have explored this particular problem.

Background Considerations

Cystic fibrosis (C/F) is a congenital familial disease, transmitted through an autosomal recessive gene, affecting exocrine glands. Because the pancreas was the first organ attracting clinical and pathologic interest, this disease was described originally in 1936 to 1938 as fibrocystic disease of the pancreas, or cystic fibrosis of the pancreas.[1] Subsequent investigations revealed prominent gastrointestinal manifestations and pulmonary changes which indicated involvement of the mucus-producing glands; hence, the name of mucoviscidosis was coined by Farber and Shwachman, mucosis by Bodian, and mucositis by Michalowicz. The susceptibility of cystic fibrosis patients to heat prostration led to the discovery that the sweat glands excrete abnormal amounts of electrolytes. This indicated that practically all exocrine glands, both mucus- and nonmucus producing, are involved in cystic fibrosis patients.

The diagnosis of cystic fibrosis is established on the basis of specific exocrine gland manifestations, the patients' personal and family history, radiologic features of his respiratory tract, and several confirmatory laboratory findings, such as: deficits of pancreatic enzymes, elevated sweat Na^+ and Cl^- content, elevated Na^+ in nails, presence of Spock's factor or Mangos' factor.[2]

The incidence rate of cystic fibrosis is calculated as approximately one case per 1,000 Caucasian newborns, with a range from one per 600 to one per 2,000 newborns; [3] and at least ten times less in American Negroes. Thus about 4,000 children with this disease are born yearly in the U. S. A.

Despite improved diagnostic procedures, most of the cases are still not recognized, and several manifestations and complications may be treated under the wrong diagnosis. With upper respiratory tract involvement, the mis-

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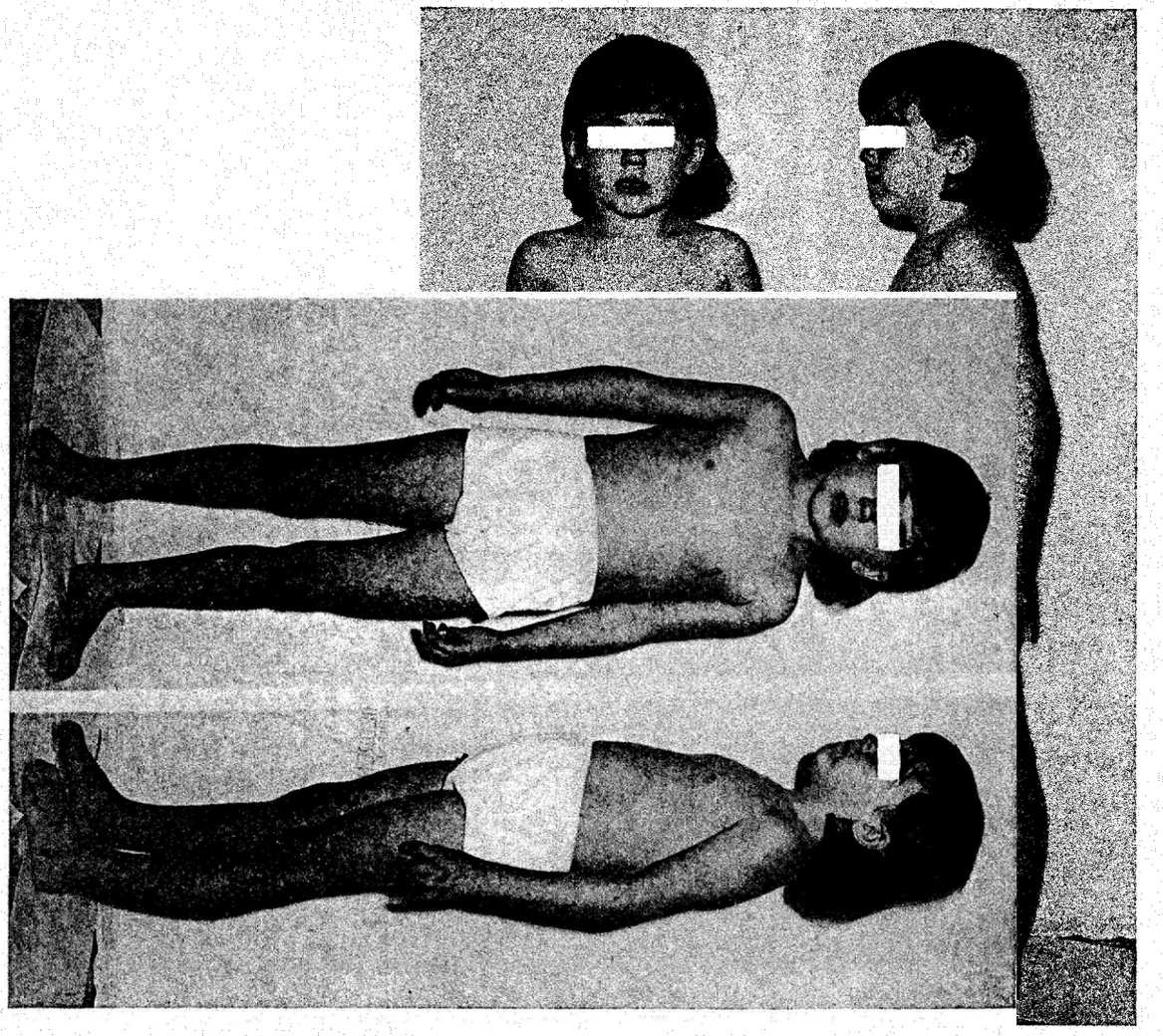


diagnoses may include: sinobronchitis, chronic sinusitis, nasal polyposis, chronic otitis media, URI of unknown etiology, etc.; with lower respiratory tract involvement: allergic bronchitis, emphysema, bronchiectasis, chronic bronchitis, or even tuberculosis.

Despite great clinical and research advances within the past 30 years, the basic defect in C/F is still unknown. Many manifestations can now be controlled, however, and many complications can be averted, provided the disturbance is recognized early enough (possibly within the first month of life), and preventive measures taken to arrest the progress of the disease.

The major preventive and therapeutic measures relate to: (1) adequate nutrition, including giving pancreatic enzymes; (2) adequate hydration, promoting thinning and natural flow of bronchial secretions; (3) active physical procedures to promote pulmonary drainage; (4) prevention of infections; intensive therapy should an infection develop.[5]

These steps, when faithfully and consistently adhered to, have resulted in increased longevity for thousands of cystic fibrosis patients. Several hundreds are now in their third decade of life—mature and productive members of the society. Some are college graduates and a few have even married; though all of

FIG. 2. Chest x-ray (PA and lateral views) of the girl in Figure 1 which represents diffuse changes, including pulmonary hyperaeration, PA diameter increased, mild dorsal scoliosis, prominent hilar areas, peribronchial infiltrates and pneumonia of right middle lobe (which cleared later).

these require access to specific medical and social facilities to keep alive.

More than 95 per cent of patients with cystic fibrosis suffer from respiratory tract involvement of some degree of severity, even though their physical appearance may be excellent (Fig. 1). Their tracheobronchial mucosa may be involved uniformly or focally. Their mucous glands produce tenacious secretions which cripple ciliary function and delay the flow of mucus. That is why maintaining good drainage of the respiratory tract is of fundamental importance for prevention of respiratory tract manifestations of all sorts (Fig. 2).

In cystic fibrosis patients, the nasal mucosa usually becomes hyperplastic, with prominent nasal turbinates occasionally concealing nasal polypi [6] (Figs. 3, 4). The resulting narrowing of the nasal passages and of the ostia to the paranasal sinuses prevents adequate sinus drainage. One finds paranasal sinus involvement in nearly 100 per cent of C/F patients and a 15 per cent incidence of nasal polyposis. With nasal polyposis the nasal bridge becomes broad and flat (Figs. 5, 6). Polypi also may form within sinuses, and erode neighboring bones.

A similar problem of drainage obstruction exists in the middle ear. The "edematouslike" mucosa of the eustachian tube prevents adequate flow of secretion from the middle ear, thereby predisposing it to chronic serous otitis and to recurrent otitis media. The tympanic membranes in approximately 50 per cent of cystic fibrosis patients have lost their luster and are retracted and dull; approximately 10 per cent of the patients show fluid levels or even bubbles behind the membrane, signs of serous otitis media.

The Present Investigation

The authors here report the results of studies to determine whether these pathophysiological changes in C/F patients affect their hearing sensitivity. This is a matter which has not been mentioned by previous investigators, even by those who have discussed sinusitis, nasal polyposis, and otolaryngologic evaluations in C/F patients.[8–10] The observations we have made are compared with results of stud-

ies of comparable school-age children who do not have C/F, which indicate a prevalence of hearing loss of 3 to 10 per cent.[11, 12]

We studied 70 patients, 67 Caucasians and three Negroes, affected with C/F. There were 35 girls and 35 boys, four to 19 years old. Table 1 lists their age and sex distributions, and the results of clinical evaluation of their progress and of their degree of conductive hearing loss.

Procedures Followed

1. The patients were studied as they reported to the medical clinic. The only restriction was that at the time of being tested they should be free of acute infection, and be able to cooperate in audiologic evaluations. A total of 96 audiologic and otolaryngologic examinations were performed on these patients over the past four years. In addition, 15 healthy siblings of C/F patients were examined similarly and used as a control group.

2. ENT evaluation consisted of a careful check on the status of nose, oral cavity, throat, upper pharynx, ears and neck.

3. The eustachian tube was politzerized and catheterized whenever fluid in the middle ear was suspected.

4. Tuning fork tests, using 256-Hz (Hertz), 512-Hz, and 1,024-Hz forks, were used to screen adequacy of bone and air conduction.

5. Hearing tests[14-16] were conducted by qualified audiologists under the direction of a co-author of this paper (G. R. H.). All testing was done in auditory test suites manufactured by the Industrial Acoustics Company. The over-all noise levels in these specially constructed rooms were less than 44 db (decibels) sound pressure level, ensuring that extraneous sounds did not interfere with the assessment of auditory thresholds.

Pure tone audiometers, calibrated to the International Standards Organization 1964 standards, were employed for all tests. Air conduction thresholds were determined at discrete test frequencies from 125 through 8,000 Hz. Bone conduction thresholds were assessed at octave intervals for test tones between 250 and 4,000 Hz. The calibration of the audiometers was checked regularly throughout the study.

When a child would cooperate for the procedure, contralateral masking was used during bone conduction testing. The masking source was a narrow band noise generator,

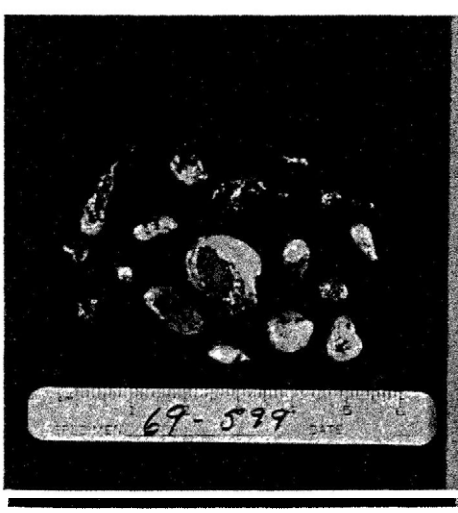


FIG. 3. Multiple nasal polyps removed from six-year-old girl with C/F (Patient 1).

FIG. 4. Histologic appearance of the polyp resected from Patient 1. Note increased vascularization, edema and leukocyte infiltration, distended mucosal glands.

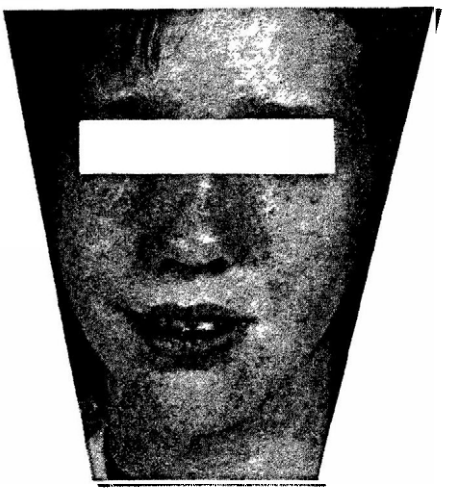


FIG. 5. Face of 13-year-old girl with C/F and nasal deformity.

with noise signal delivered to the subject's ear by an insert receiver.

For this study normal hearing was defined as air-conduction thresholds from 0 to 15 decibels (db) on the hearing-loss dial of the audiometer, *i.e.*, 0 to 15 db hearing level (H.L.). Some hearing was adjudged to be lost when air-conduction thresholds for two or more test tones, regardless of ear, were at the 20 db hearing level or poorer.

FIG. 6. X-ray of paranasal sinuses of C/F patient which indicates: opacification of the sinuses, swelling of lining mucosa, right nasal mass (polypi) narrowing nasal airway and deviating nasal septum.

An air-conduction loss between 20 and 30 db H.L. was deemed borderline, since in the speech frequencies (500 to 2,000 Hz) it only affects listening for very faint speech. A hearing loss between 30 and 45 db was considered mild since this degree of loss in the speech frequencies interferes with listening to soft conversational speech.

Audiologic Findings

Among the 70 C/F patients in this study, one had sensorineural hearing loss apparently unrelated to C/F, and 18 (26 per cent) had conductive hearing losses at the time of the first evaluation. Seven patients had hearing losses greater than 25 db H.L. All these patients had obvious pathologic changes in their upper respiratory tract, such as: serous otitis media, nasal polyposis, enlarged tonsils and adenoids. Five patients were treated surgically, after which their hearing returned to normal. The other two patients responded well to medical therapy.

Of the 11 patients with less pronounced hearing loss, all had unilateral or bilateral eardrum retraction without definite fluid level, hypertrophied nasal turbinates, occasional nasal polyp, and postnasal drip. They were treated medically with good audiologic improvement.

Among the 15 apparently healthy siblings of these C/F patients who were tested, only one had mild conductive hearing loss, associated with questionable changes in the left ear.

Table 2 summarizes the data for the first 12 patients, listing ages and dates when audiograms were conducted, pertinent clinical and laboratory results concerning the upper respiratory tract, and a general evaluation of their clinical progress.

Representative Case Histories

The following brief narratives describe eight patients with conductive hearing losses and one with sensorineural loss.

ANN (Patient 1) was hospitalized twice: (1) at the age of eight months because of acute pharyngitis and pyelonephritis; and (2) at the age of nine months because of malabsorption syndrome and anemia. During the second year of life, she developed a cough and recurrent bronchitis. The diagnosis of cystic fibrosis was made at the age of

TABLE 1. *Age, Sex and Status of Hearing Loss when Present in 70 Patients with Cystic Fibrosis*

	No. Patients	Age (Yr.)	Sex		Clinical Evaluation				Hearing Loss	
			F	M	Excellent	Good	Mild	Moderate	Borderline	Mild
	29	4–7	14	15	10	9	8	2	9	1
	25	8–11	14	11	7	12	6			6
	16	12–19	7	9	6	5	5		2	
TOTAL	70	4–19	35	35	23	26	19	2	11	7

two years two months. In spite of treatment, progression in peribronchial infiltrates and hyperaeration of the lungs took place, along with recurrent serous otitis media and prominent nasal turbinates. X-ray examination revealed opacification of paranasal sinuses. At the age of five years, nasal polyposis was discovered and an audiogram revealed a mild conductive hearing loss. An ear, nose and throat (ENT) examination confirmed bilateral nasal polyposis which required bilateral polypectomy. This was performed at the age of six years (Figs. 3, 4). The subsequent audiogram, done four months later, showed her hearing within normal limits.

TABLE 2. *Audiologic and Otolaryngologic Findings in First Twelve C/F Patients*

Patient	Age (Yr.) and Date of Audiogram	Hearing Threshold Hearing Loss: Borderline 20–30 db, Mild 30–45 db	U.R.T. Clinical Findings			Surgery	Clinical Evaluation
			Nasal Cavities	Ears	Throat		
1	4 4/12(2–67)	Normal	Clear bilat.	Neg.	Neg.		Mild
	5 11/12(9–68)	Borderline CHL	Bilat. polypi	TM dull	Neg.	P 10/68	Good
	6 4/12(2–69)	Improved bilaterally	Prom. turbinates	Neg.	Neg.		Good
2	4 6/12(3–67)	Borderline CHL	Polyposis	TM dull	Neg.	P 4/67	Mild
	5 5/12(2–68)	Normal range	Negative	Neg.	Neg.		Mild
	6 10/12(7–69)	Normal range	Negative	Neg.	Neg.		Mild
3	5 10/12(6–68)	Bilat. borderline CHL	Hyperemic mucosa	TM dull & ret.	Injec.		Excellent
	6 6/12(3–69)	Bilat. borderline CHL	Prom. turbinates	O.M.	Inject.		Excellent
	7 2/12(10–69)	Normal hearing	Prom. turbinates	Neg.			Excellent
4	11 5/12(6–68)	Very mild bilat. CHL	Polyps suspect.	Neg.	Post N. drip		Excellent
	12 2/12(3–69)	Very mild bilat. CHL	Clear bilat.	S.O.	Enl. tonsil		Excellent
	12 10/12(10–69)	Normal hearing	Clear bilat.				Excellent
5	6 5/12(2–67)	Rt. ear mild CHL	Negative	Neg.	Enl. aden.		Mild
	7 7/12(4–68)	Normal bilat. audiogram	Negative	Neg.	Neg.		Mild
6	5 6/12(5–67)	Bilat. CHL up to 40 db	Clear bilat.	Cerumen	Enl. aden.		Mild
	6 8/12(7–68)	Improved borderline CHL	Clear bilat.	TM ret.			Mild
	7 8/12(7–69)	Close to normal	Clear bilat.	TM sl. dull			Mild
7	13 4/12(7–66)	Borderline Bilat. CHL	Enl. turbinates	Neg.	Neg.		Good
	15 (3–68)	Bilat. normal range	Clear bilat.	Neg.	Neg.		Good
8	14 10/12(6–67)	Rt. borderline CHL	Deviated septum	Neg.	Hyp. aden.		Good
	15 7/12(3–68)	Normal hearing	Prom. turbinates	Neg.			Good
9	5 5/12(5–67)	Lt. borderline CHL	Polyposis	O.M.	T&A enl.	T&A, P 6/67	Good
	5 7/12(7–67)	Normal bilaterally	Negative	Neg.	Normal		Excellent
	7 8/12(8–69)	Normal bilaterally	Negative	Neg.	Neg.		Excellent
10	9 7/12(2–69)	Bilateral mild CHL	Prom. turbinates	TM dull	Inj. T&A	T&A, 5/65	Mild
	9 8/12(3–69)	Bilat. mild CHL (worse)		O.M.			Mild
11	7 2/12(10–65)	Bilat. CHL up to 1,000 H_z	Polyposis	O.M.	T&A enl.	T&A, P 11/65	Good
	8 7/12(3–67)	Lt. ear mild CHL	Negative	O.M.		M	Good
	9 10/12(6–68)	Bilat. CHL	Clear bilat.	O.M.		P.T.I.	Good
	10 5/12(1–69)	Normal audiogram	Negative	Neg.			Good
12	6 4/12(11–65)	Bilat. sensorineural Hearing loss	Clear	Neg.	Neg.		Good
	6 5/12(12–65)	wearing hearing aid					
	8 11/12(6–68)						Excellent

CHL = conductive hearing loss; P = polypectomy; T&A = tonsillectomy and adenectomy; M = myringotomy; O.M. = otitis media; S.O. = serous otitis; TM = tympanic membrane; P.T.I. = polyethylene tube inserted; Rt. = right; Lt. = left.

KEMP (Patient 2) was diagnosed as having cystic fibrosis at one month of age because of a family history of C/F, abnormally elevated sweat electrolytes and pancreatic enzyme deficit. In spite of early diagnosis and treatment, nutritional and respiratory tract manifestations developed which called for intensive care at home and frequent medical supervision.

At the age of three years nine months he was hospitalized for respiratory distress. Detailed studies for allergy conducted during his hospitalization were negative. Nasal and pharyngeal swabs were cultured and revealed the presence of *Staphylococcus aureus,* coagulase positive, and pseudomonas. He was bronchoscoped and received bronchial lavage with N-acetylcysteine. A detailed ENT evaluation indicated purulent rhinitis and slightly enlarged adenoids.

Six months later, ENT evaluation revealed a large polyp which completely occluded the right nasal passage and a small nasal polyp on the left. Subsequent auditory testing, performed in March 1967 at the age of four and one-half years, revealed a borderline bilateral conductive hearing loss (Audiogram 1a). Bilateral polypectomy was performed soon afterward.

The patient continued to receive intensive treatment for his pulmonary manifestations with relatively satisfactory progress. Repeat auditory testing, done at five years five months, revealed hearing within normal limits, bilaterally (Audiogram 1b). A third audiogram, done one and one-half years later, was normal and the patient's progress is satisfactory.

LORI (Patient 3) was diagnosed as having cystic fibrosis at the age of two years ten months because of a history of recurrent bronchitis, rectal prolapse, marked pulmonary hyperaeration and minimal prominence of the hilar areas. X-rays of the sinuses and lungs at six years of age showed haziness and mucosal thickening in both maxillary antra, but normal lungs. An ENT examination at four years seven months (March 9, 1967) showed slight retraction of both tympanic membranes, prominent nasal turbinates with copious mucosal secretions, clear throat, and slightly enlarged tonsils.

An audiogram at the age of five years ten months (June 10, 1968) revealed a borderline conductive hearing loss in both ears (Audiogram 2a). A follow-up ENT evaluation at the age of six and one-half years (March 7, 1969) indicated slight retraction of both tympanic membranes, fluid level in the left ear, clear throat with moderately enlarged tonsils, and questionable bilateral polyposis. Audiogram taken on the same day indicated again borderline conductive hearing loss in both ears (Audiogram 2b).

Nine months later, after intensive medical treatment, her audiogram showed normal hearing.

BRAD (Patient 4) was diagnosed as having cystic fibrosis at age three and one-half years. He has always had an excellent appetite and no gastrointestinal manifestations except for rectal prolapse at the age of six years. He had a history of rhinitis, spasmodic bronchitis and heat prostration at age five years.

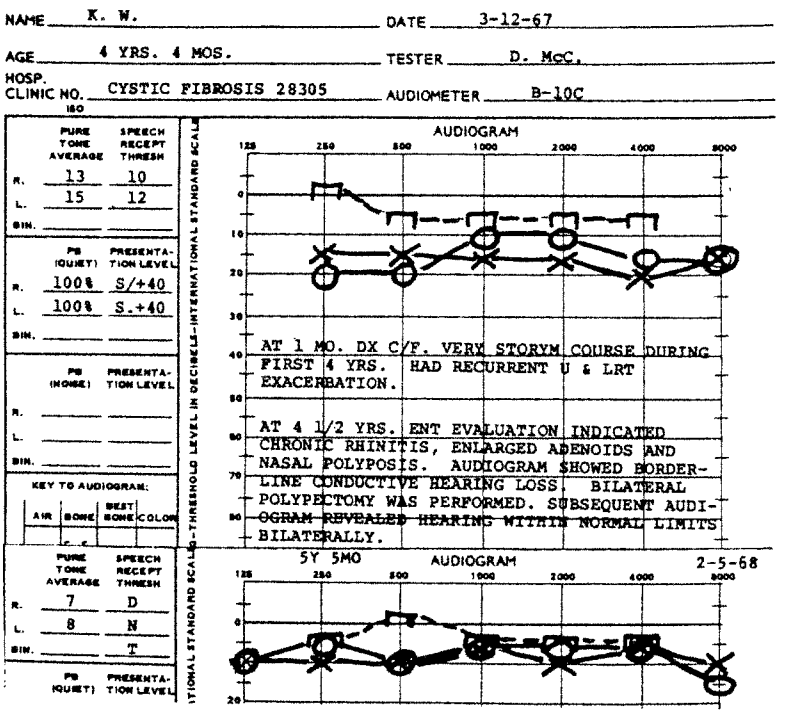
At ten years two months an ENT examination indicated enlarged tonsils, postnasal drip and questionable bilateral nasal polyposis. He was treated with antibiotics and nasal decongestants.

An audiogram at the age of ten years ten months indicated minimal conductive hearing loss (Audiogram 3a). His general condition remained good. An ENT examination done at 12 years two months showed tympanic membranes slightly retracted and dull, and tonsils moderately enlarged; audiogram revealed mild conductive hearing loss, more in the right ear than the left (Audiogram 3b). Another audiogram done nine months later indicated normal hearing and good clinical progress.

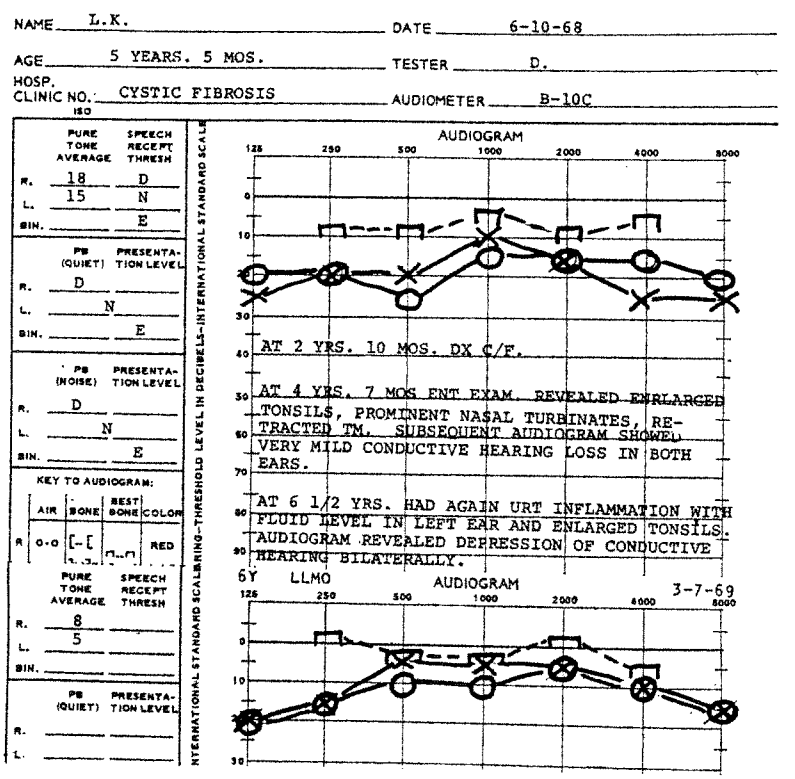
RONALD (Patient 5) was diagnosed at seven months of age. He had respiratory distress, nutritional deficit, abnormal sweat electrolytes and advanced pulmonary changes including bilateral bronchopneumonia. Repeated nasopharyngeal cultures revealed *Staphylococcus aureus,* aerobactor, proteus, and pseudomonas more recently. He has had intensive therapeutic treatment and attends school fulltime.

At age six years five months (February 1967) a hearing test indicated a mild bilateral conductive hearing loss (Audiogram 4a). An ENT evaluation revealed small tonsils and moderately enlarged adenoids. Another ENT examination at age seven years seven months revealed a mildly infected throat, patent nasal airways, no polyps, and no ear abnormalities; retests (Audiogram 4b) showed hearing within normal limits, bilaterally.

BECKY (Patient 6) not only had cystic fibrosis at the age of 20 months, but also well-compensated pulmonary artery stenosis. She was receiving an intensive home therapeutic program. ENT evaluation at the age of five and one-half years indicated impacted bilateral cerumen, which was removed, and bilaterally retracted tympanic membranes somewhat sluggish in movement. Tonsils were small, but adenoids large. Her first hearing test, done at the age of five and one-half years, revealed bilateral conductive hearing loss more in the left ear, on the order of 30 db for frequencies between 500 and 2,000 Hz (Audiogram 5a). Following treatment, a repeat hearing test, at the age of six years eight months, revealed definite improvement in both ears; however, there remained a mild residual conductive loss in both ears (Audiogram 5b). An ENT evaluation at that time showed good eardrum mobility. Her intensive medical treatment was continued. A repeat au-


NAME K. W. DATE 3-12-67
AGE 4 YRS. 4 MOS. TESTER D. McC.
HOSP. CLINIC NO. CYSTIC FIBROSIS 28305 AUDIOMETER B-10C
ISO
PURE TONE AVERAGE / SPEECH RECEPT THRESH
R. 13 10
L. 15 12
BIN.
PB (QUIET) / PRESENTATION LEVEL
R. 100% S/+40
L. 100% S.+40
BIN.
PB (NOISE) / PRESENTATION LEVEL
KEY TO AUDIOGRAM: AIR BONE BEST BONE COLOR
AUDIOGRAM
AT 1 MO. DX C/F. VERY STORYM COURSE DURING FIRST 4 YRS. HAD RECURRENT U & LRT EXACERBATION.
AT 4 1/2 YRS. ENT EVALUATION INDICATED CHRONIC RHINITIS, ENLARGED ADENOIDS AND NASAL POLYPOSIS. AUDIOGRAM SHOWED BORDER-LINE CONDUCTIVE HEARING LOSS. BILATERAL POLYPECTOMY WAS PERFORMED. SUBSEQUENT AUDI-OGRAM REVEALED HEARING WITHIN NORMAL LIMITS BILATERALLY.
5Y 5MO AUDIOGRAM 2-5-68
PURE TONE AVERAGE / SPEECH RECEPT THRESH
R. 7 D
L. 8 N
BIN. T
PB (QUIET) / PRESENTATION LEVEL
INTERNATIONAL STANDARD SCALE—THRESHOLD LEVEL IN DECIBELS—INTERNATIONAL STANDARD SCALE

Audiogram 1. a (top) b (bottom).


NAME L.K. DATE 6-10-68
AGE 5 YEARS. 5 MOS. TESTER D.
HOSP. CLINIC NO. CYSTIC FIBROSIS AUDIOMETER B-10C
ISO
PURE TONE AVERAGE / SPEECH RECEPT THRESH
R. 18 D
L. 15 N
BIN. E
PB (QUIET) / PRESENTATION LEVEL
R. D
L. N
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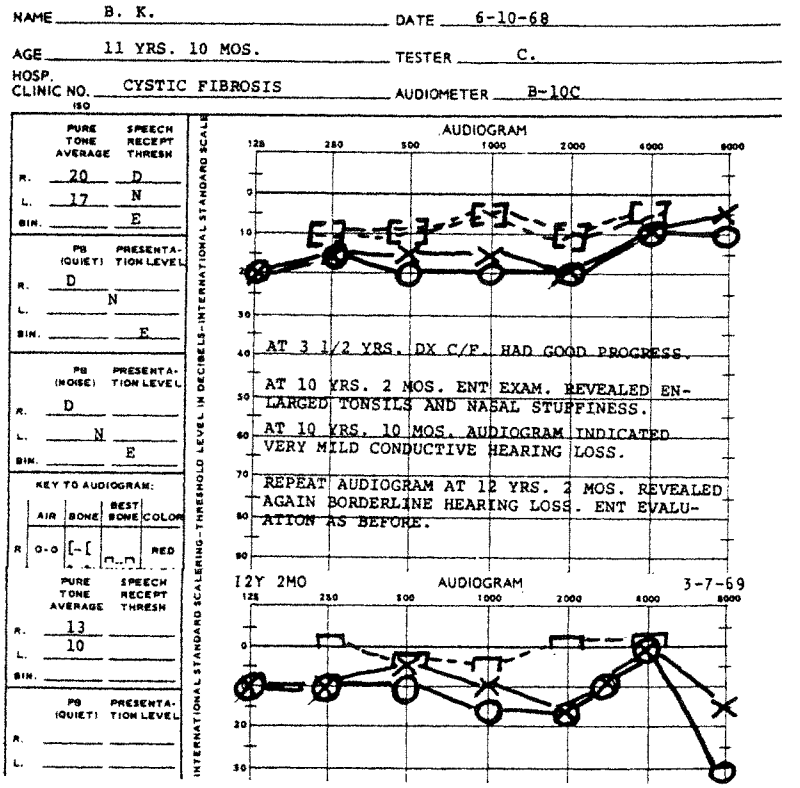
PB (NOISE) / PRESENTATION LEVEL
R. D
L. N
BIN. E
KEY TO AUDIOGRAM: AIR BONE BEST BONE COLOR
R o-o [-[RED
AUDIOGRAM
AT 2 YRS. 10 MOS. DX C/F.
AT 4 YRS. 7 MOS ENT EXAM. REVEALED ENLARGED TONSILS, PROMINENT NASAL TURBINATES, RE-TRACTED TM. SUBSEQUENT AUDIOGRAM SHOWED VERY MILD CONDUCTIVE HEARING LOSS IN BOTH EARS.
AT 6 1/2 YRS. HAD AGAIN URT INFLAMMATION WITH FLUID LEVEL IN LEFT EAR AND ENLARGED TONSILS. AUDIOGRAM REVEALED DEPRESSION OF CONDUCTIVE HEARING BILATERALLY.
6Y LLMO AUDIOGRAM 3-7-69
PURE TONE AVERAGE / SPEECH RECEPT THRESH
R. 8
L. 5
BIN.
PB (QUIET) / PRESENTATION LEVEL
R.
L.
INTERNATIONAL STANDARD SCALE—THRESHOLD LEVEL IN DECIBELS—INTERNATIONAL STANDARD SCALE

Audiogram 2. a (top) b (bottom).



Audiogram 3. a (top) b (bottom).

diogram four months later indicated further improvement and close to normal hearing.

LINDA (Patient 7) was diagnosed as having cystic fibrosis at age two years. She had an enormous appetite and abnormal stools, but fairly good weight gain. She had minimal pulmonary emphysema which was stationary. Respiratory-tract infections were infrequent and responded promptly to antibiotics.

At the age of 13 years four months, she had no complaints, an ENT examination showed only slightly prominent nasal turbinates. An audiogram showed minimal conductive hearing loss of no more than 20 db for test frequencies up to 500 Hz (Audiogram 6a). Her general progress was excellent. Chest x-rays revealed some hyperinflation and accentuated hilar markings.

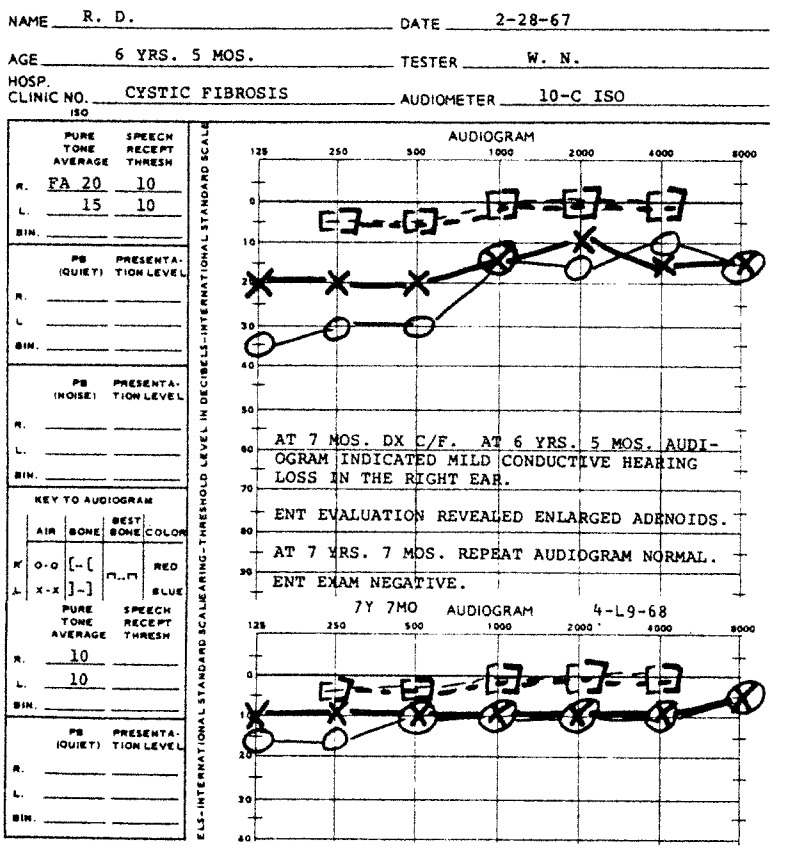
Another examination at age 15 years revealed slight dullness of the left tympanic membrane, but the audiogram indicated hearing within normal limits, bilaterally (Audiogram 6b).

PATRICK (Patient 8) was suspected of having cystic fibrosis from early infancy, but the diagnosis was not confirmed until age eight years nine months. His clinical condition was still very good, but radiologic examination indicated moderate to severe pulmonary involvement and complete clouding of all paranasal sinuses.

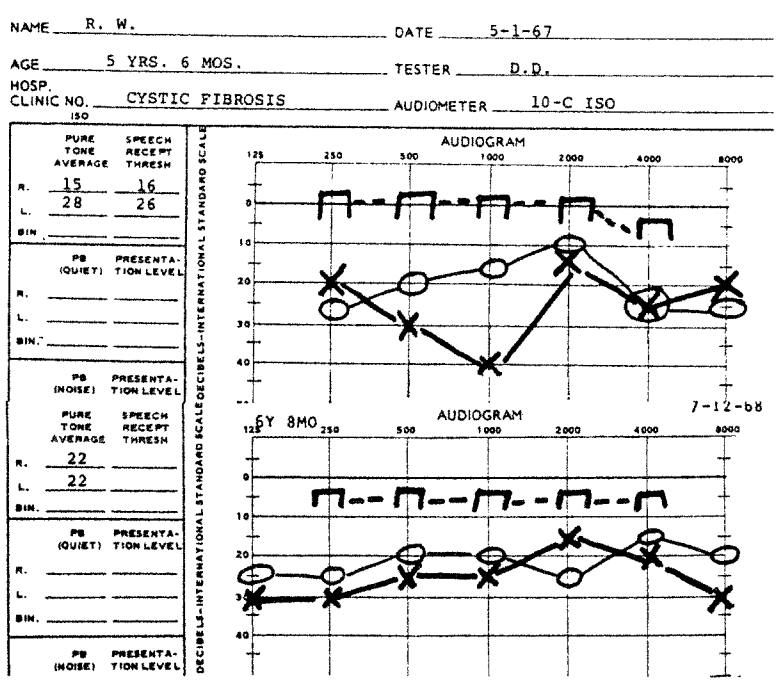
ENT evaluation at age 14 years ten months indicated retracted eardrums with possible fluid levels, hypertrophic adenoids, deviated nasal septum and hypertrophied nasal turbinates. His audiogram showed normal hearing in the left ear and borderline conductive hearing loss in the right ear.

He is following a long-term intensive respiratory home program, attends school regularly and physically compares favorably with his peers. Follow-up audiologic evaluation at the age of 15 years seven months showed hearing essentially within normal limits, bilaterally.

RICHARD (Patient 12) was recognized as having cystic fibrosis at age three months. At age six years, his chest appeared normal clinically and radiologically. He did not have hearing complaints, but his parents felt that he did not respond properly to sound. His initial audiogram revealed a moderate to severe sensorineural loss apparently unrelated to cystic fibrosis. There was nothing in his past history to suggest factors potentially related to the hearing impairment. It was concluded that the hearing loss was not related to his upper-respiratory-tract condition or to his cystic fibrosis. A hearing aid was provided, and his clinical progress has been excellent.



Audiogram 4. a (top) b (bottom).



Audiogram 5. a (top) b (bottom).

Confusion with Allergy

The multiple manifestations of cystic fibrosis may mimic various clinical problems. Patients with respiratory-tract involvement may be treated for acute or chronic rhinitis, sinusitis or otitis media, without the basic illness being recognized. Allergy is blamed frequently as a causative factor; the patient may then have prolonged allergic investigation, followed by usually unsuccessful antihistaminic and desensitization treatment.

The prevalence of allergy among C/F patients is 17 per cent, which closely corresponds with that among normal children.[17] Of 50 cystic fibrosis patients with nasal polyposis, all had paranasal sinus involvement (by roentgenograms).[6] In this series, 21 (41 per cent) had no evidence of atopy; they had negative skin tests, no nasal eosinophils in smears, and no family history of allergy.

In the above-cited series of 50 patients with nasal polyposis, three were less than three years old; 13 were between four and six years old; 18 were between seven and nine years old; 12 were between ten and 12 years old; and five patients were between 13 and 27 years old.

With superimposed respiratory-tract infection in cystic fibrosis the mucosa becomes more edematous, thick secretion accumulates, and lumenal obstruction favored. The patient with acute manifestations of rhinitis, sinus involvement, otitis media, may need help and relief urgently. We have heard of C/F patients with neurologic manifestations of meningismus and completely negative examination of the central nervous system and cerebral spinal fluid who became symptom-free after bilateral myringotomies were performed. However, none of the cases in the present study had neurologic or even otologic complaints. Sinus aspiration and drainage of both antra are occasionally required by C/F patients, but were not needed by the subjects in this study.

A striking aspect of the present series was that only one patient had otologic complaints, even when the local findings were positive. Subsequently, after otolaryngologic evaluation and therapy, their hearing returned to normal. The results of an audiologic examination may alert parents and physicians to incipient lesions of the upper respiratory tract in some C/F patients.

Radiologic studies of the upper respiratory tract can be of great diagnostic assistance. The frequent radiologic reports of chronic pansinusitis in the patients with C/F refer to anatomic and pathologic changes, not necessarily to an infection.

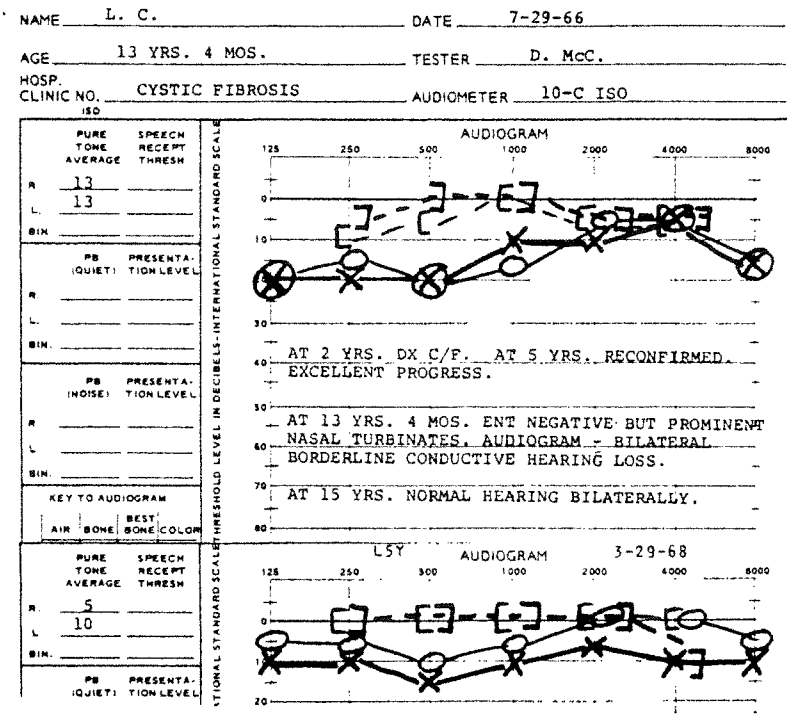
The sinuses seldom show a fluid level, and puncture and aspiration are usually negative. This chronic condition, specific to the cystic fibrosis entity, should have another name, such as "sinusosis," to distinguish it from the inflammatory processes which underlie what is currently termed acute or chronic sinusitis.

Serum Otitis Media

This currently is a popular otologic clinical and research topic. The pathophysiology of the upper respiratory tract presents a complex problem. There is multiple nomenclature and conflicting statements, particularly with reference to the factors contributing to eustachian tube blockage and serous otitis media.

Senturia *et al.*[18] made detailed studies which suggest that the basic cause of secretory otitis media is infection of the nasopharynx with retrograde inflammation of the eustachian tube. Suehs,[19] in addition to his own 372 cases of serous otitis media, analyzed 20 other reports with regard to its etiology, pathology and treatment. He concluded that upper-respiratory-tract infections are the chief predisposing causes, and that allergy accounted for only 10 per cent of the cases. Similarly, Robinson and Nicholas[20] underline the role of infection and state that "allergy has not been found to be an important factor in the etiology of otitis media with effusion." However, other investigators, such as Draper,[21] and Fernandez and McGovern,[22] indicate that serous otitis media occurs twice as often in allergic children than in control groups.

Apparently, secretory otitis media may have many causes, reflectd in dysfunction of the eustachian tube. The most frequently encountered factors refer to inflammation, allergy, mechanical obstruction by adenoid tissue, mucosal engorgement, inspissated secretion and nasal polyps.



Audiogram 6. a (top) b (bottom).

Recommendations

In patients with cystic fibrosis, all the above factors may be present. Hence detailed otolaryngologic and audiologic examinations should be performed during the patient's regular check-ups, and should include the following items.

Paranasal sinuses: If there is pus in the nose, x-rays of the paranasal sinuses should be obtained. Even though a fluid level in a sinus is rarely present, should fluid be suspected an antrectomy and repeat sinus wash-out should be done, bacteriologic culture obtained, and the patient treated accordingly.

Tonsils and adenoids: Adenoids should be removed only if they interfere with the eustachian tube performance and breathing. Tonsils should be removed when the patient has chronic cryptic tonsillitis, or when they are so large that they interfere with swallowing or breathing.

Other ENT findings: If there is chronic enlargement of the submaxillary glands, nothing needs to be done clinically, since this is usually a manifestation of cystic fibrosis or exocrinosis. The larynx should be examined because patients with cystic fibrosis often have an infantile larynx with an omega-shaped epiglottis.

The study indicates a definite relationship between hearing thresholds and the integrity of the auditory system and neighboring structures of the upper respiratory tract. There tends to be:

1. Chronic paranasal sinus involvement; an almost universal feature of the disease.
2. Occurrence of nasal polyposis in approximately 15 per cent.
3. Relatively common occurrence of serous otitis media which may be triggered by several mechanisms, notably exocrine glands pathology.
4. Chronic upper-respiratory-tract infections contributing to all the above.

Since even mild hearing loss may be depicted by the audiogram, this simple hearing test should be included in the periodic evaluations of C/F patients, as a sensitive detector of pathologic involvement of the upper respiratory tract.

References

1. Andersen, D. H. *et al.*: Problems in cystic fibrosis. Ann. New York Acad. Sci. 93: 485, 1962.
2. Di Sant'Agnese, P. A. and Talamo, R. C.: Pathogenesis and physiopathology of cystic fibrosis of the pancreas. New Eng. J. Med. 277: 1287, 1343, 1399, 1967.
3. Kulczycki, L. L., Macleod, K. I. E. and Shwachman, H.: A survey of school children for cystic fibrosis. Amer. J. Dis. Child. 100: 174, 1960.
4. ——, Guin, G. H. and Mann, N.: Cystic fibrosis in Negro children. Clin. Pediat. 3: 692, 1964.
5. Guide to Diagnosis and Management of Cystic Fibrosis. New York, National Cystic Fibrosis Research Foundation, 1965.
6. Shwachman, H., Kulczycki, L. L., Mueller, H. L. and Flake, C. G.: Nasal polyposis in patients with cystic fibrosis. Pediatrics 30: 389, 1962.
7. Kulczycki, L. L.: Some Distinctive Respiratory Features of Mucoviscidosis (Cystic Fibrosis). Basel, Proceedings of the 6th International Congress of Internal Medicine, 1960, pp. 255–261.
8. Pennington, C. L.: Paranasal sinus changes in fibrocystic disease of the pancreas. Arch. Otolaryng. 63: 576, 1956.
9. Lurie, M. H.: Cystic fibrosis of the pancreas and nasal mucosa. Trans. Amer. Laryng. Assoc. 80: 199, 1959.
10. Mendelson, R. S. and Cohen, B. M.: Otorhinolaryngologic aspects of cystic fibrosis. Arch. Otolaryng. 79: 312, 1964.
11. Eagles, E. L., Wishik, S. M., Doerfler, L. G., with Melnick, W. and Levine, H. S.: Hearing Sensitivity and Related Factors in Children. Monograph, Laryngoscope, 1963.
12. Robinson, G. C. *et al.*: A survey of hearing loss in Vaucouver school children. Canad. Med. Assn. J. 97: 1199, 1967.
13. Shwachman, H. and Kulczycki, L. L.: Long-term study of one hundred and five patients with cystic fibrosis; studies made over a five to fourteen year period. Amer. J. Dis. Child. 96: 6, 1958.
14. Newby, H. A.: Audiology. New York, Appleton, Century, Crofts, Inc., 1964.
15. Davis, H. and Kranz, F. W.: The international standard reference zero for pure-tone audiometers and its relation to the evaluation of impairment of hearing. J. Speech Hearing Res. 7: 7, 1964.
16. Herer, G. R.: Evaluation of hearing in infants and young children. Clin. Proc. Child. Hosp. D. C. 23: 18, 1967.
17. Kulczycki, L. L., Mueller, H. and Shwachman, H.: Respiratory allergy in patients with cystic fibrosis. JAMA 175: 358, 1961.
18. Senturia, B., Gessert, C., Carr, C. and Baumann, E.: Studies concerned with tubotympanitis. Ann. Otol. 67: 440, 1958.
19. Suehs, O. W.: Secretory otitis media. Laryngoscope 62: 998, 1952.
20. Robinson, J. M. and Nicholas, H. O.: Catarrhal otitis media with effusion. Southern Med. J. 44: 777, 1951.
21. Draper, W. L.: Secretory otitis media in children; a study of 540 children. Laryngoscope 77: 636, 1967.
22. Fernandez, A. A. and McGovern, J. P.: Secretory otitis media in allergic infants and children. Southern Med. J. 58: 581, 1965.

Deafness

Postnatal infections, particularly meningitis and otitis, are the most frequent causes of deafness in general. It is essential to test the hearing of every child, indeed of every baby, who has a history indicative of the possibility of partial or complete deafness.

Modern audiometric technics make it possible to detect deafness in one-year-old children and even in newborns. It is important to start the education of the completely or severely deaf child as early as possible to take advantage of the child's great receptiveness to auditory and visual stimuli during the first five or six years of life.— *From WHO Chronicle, January 1970.*