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Neuropsychological Correlates of Cystic Fibrosis in Patients 5 to 8 Years Old

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Intellectual, academic, and neuropsychological tests were administered to 20 children with cystic fibrosis (CF). Results were compared to test results from 20 controls matched for gender, age, and socioeconomic status. No differences between the groups were found. For children with CF, Verbal IQ, sensory-perceptual skills, and incidental learning correlated (*rs* = .39–.67) with Shwachman criteria of disease severity, with significant positive relations with the Growth and Nutrition measure, an index of the severity of the disease. Processing of tactile–perceptual information may be particularly vulnerable to disease severity. This study provides more information than previously available on the neuropsychological status of young children with CF, and it offers

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some hypotheses regarding the relation between disease severity and neuropsychological function.

There is evidence to suggest that children with cystic fibrosis (CF) show intellectual delays prior to the age of 5 years and that these delays disappear after that age (Kulczycki, Robinson, & Berg, 1969; Lloyd-Still, Hurwitz, Wolff, & Shwachman, 1974). However, the delays noted in younger children may persist as subtle brain-related deficits that could easily be missed with global intellectual measures. Certain characteristics of the disease, such as malnutrition in early life, the high vulnerability to respiratory infections, increased airway resistance, and the possibility of decreased cerebral oxygenation, may lead to neurobehavioral deficits. There is some suggestion that learning problems in school are common in this group of patients (Matthews & Drotar, 1984), and the possibility of mild neurobehavioral deficits in this group of children has been raised (Berg & Linton, 1989). A neuropsychological evaluation is more likely to detect brain-related deficits than intellectual testing alone (Lezak, 1983; Reitan, 1966), but there is currently no research that specifically evaluates neuropsychological function in children with CF.

For the purpose of control groups in studies of disabled and chronically ill children (Breslau, 1985; Breslau & Marshall, 1985; Stewart, Hiltebeitel, et al., 1991; Stewart, Silver, et al., 1991), CF has been considered an illness that does not involve the brain. However the lack of literature documenting the absence of central-nervous-system-related cognitive deficits in children with CF served as an impetus to directly examine neuropsychological function in this group of patients.

This study was designed to assess neuropsychological function in children over the age of 5 years with CF, as compared to a group of physically healthy children, matched for gender, age, and socioeconomic status (SES). A secondary purpose of this study was to obtain preliminary data on possible relations between neuropsychological function and severity of the disease, inasmuch as CF varies in severity.

METHOD

Participants

The CF group was composed of 10 boys and 10 girls, age 5 years 0 months to 8 years 8 months, recruited from the Children's Medical Center of Dallas CF clinic. The lower end of the age range was defined as 5 years because previous studies using global measures (Lloyd-Still et al., 1974) reported no deficits after this age and because more precise neuropsychological assessment instruments are not available prior to this age. The upper end of the age range was defined

as 8 years in order to remain within a single neuropsychological test battery. Exclusion criteria were (a) perinatal factors that could independently affect neuropsychological function—that is, premature birth (< 34 weeks gestational age), very low birthweight (< 2,000 g), or perinatal asphyxia—(b) coexisting diseases such as hepatic, renal, or endocrine disease; and (c) acute illness at time of study. Thirteen patients from this group were part of the control group in a previous study (Stewart, Silver, et al., 1991) and were included because they met inclusion and exclusion criteria for this study; the additional 7 patients were recruited to meet these criteria.

The Children's Medical Center of Dallas CF clinic serves the large majority of children identified as having the disease in the Dallas–Fort Worth region. The population served in this clinic represents the entire socioeconomic range and is of diverse ethnic membership. The method of recruitment was designed to maximize the likelihood that the children who participated in the study were representative of the larger group of CF patients in this area. Over the period of time that this study was conducted, there were 38 potential participants meeting inclusion and exclusion criteria for the study in our clinic. Potential participants were identified from their medical records, and their parents were contacted by telephone to request study participation, with formal informed consent obtained at the time of evaluation. Telephone calls were made sequentially from an alphabetical list, with recruitment efforts continuing until 10 boys and 10 girls were enrolled. In addition to the 20 patients recruited, four families were contacted who refused participation in the study. Three of these families lived significantly distant from Dallas. Although other families living at equal and farther distances did agree to participate, these three families found the extra trip to Dallas inconvenient. One local family refused on the basis that the child was involved in many activities and the time commitment was not worth their while.

All CF patients received a physician's evaluation within 30 days of study participation and were graded on the modified Shwachman Scale for CF (Doershuk, Matthews, & Tucker, 1964). This scale provides rating criteria ranging from 0 to 25—0–5 *(very poor)*, 6–10 *(poor)*, 11–15 *(fair)*, 16–20 *(good)*, and 21–25 *(very good)*—for four variables reflecting the severity of the disease. The variables are (a) general physical condition and activity; (b) pulmonary physical findings and degree of coughing; (c) growth and nutrition; and (d) chest x-ray for emphysema, bronchovascular markings, infiltration, or atelectasis. Scores in each of these areas are summed to obtain a total score. Scores for the group on the Shwachman criteria are presented in Table 1.

The control group consisted of 10 boys and 10 girls, age 5 years 1 month to 8 years 9 months. These patients were recruited by advertisement at Children's Medical Center and by word of mouth. The inclusion criterion was 5 through 8 years of age. Exclusion criteria were the perinatal factors described previously, acute illness at time of study, and past diagnosis of any chronic

TABLE 1
Demographic, Growth, and Disease Severity Measures for 20 CF Patients and 20 Medically Well Controls

	CF Patients		*Control Patients*		
	M ± *SD*	*Range*	*M* ± *SD*	*Range*	*p*[a]
Age (years)	7.3 ± 1.0	5.0–8.8	7.1 ± 1.3	5.1–8.8	< .75
Hollingshead SES	2.6 ± 1.19	1–4	2.7 ± 1.3	1–5	< .90
Weight	−0.85 ± 0.99	−3.00–1.57	0.74 ± 1.1	−0.95–2.44	< .0001
Height	−0.67 ± 0.75	−1.89–0.94	0.41 ± 0.91	−1.14–2.26	< .0003
Head circumference[b]	−0.57 ± 1.37	−2.83–3.60	.55 ± 1.12	−1.40–3.20	< .009
Arm muscle circumference[c]	97.5 ± 7.7	84–111	106.8 ± 8.5	90–120	< .001
Mean arm circumference[c]	93.1 ± 10.3	74–118	107.7 ± 13.5	90–129	< .009
Triceps skinfold thickness[c]	68.9 ± 35.7	32–175	125.83 ± 69.4	45–275	< .003
Physical condition/activity	21.0 ± 2.9	16–25			
Pulmonary findings	18.5 ± 3.6	12–24			
Growth and nutrition	18.4 ± 4.4	12–25			
X-ray results	18.2 ± 3.7	11–25			
Shwachman total	76.1 ± 12.5	52–90			

Note. There were 10 male and 10 female patients in each group. Of the 20 patients in each group, 17 were White and 3 were Hispanic or African American. CF = cystic fibrosis; SES = socioeconomic status.

[a]Based on an analysis of variance comparing the two groups. [b]Expressed as *z* score for age and gender based on normative data (Frisancho, 1981; Nellhaus, 1968). [c]Expressed as percentage of ideal for gender and age based on normative data (Hamill et al., 1979).

illness. An attempt was made to keep the two groups similar for gender distribution and SES (Hollingshead & Redlich, 1958) by approximate matching.

Although the CF patients received a combined score for the Growth and Nutrition component of the Shwachman criteria, for purposes of comparison to control children, height, weight, head circumference, and arm anthropometries were also obtained for the CF and control groups. These measures were converted for analysis based on available normative data (Frisancho, 1981; Hamill et al., 1979; Nellhaus, 1968). All measurements were performed by a dietitian specifically trained in the standardized methods as previously described (Stewart et al., 1988). Anthropometries are missing for 2 control children. *Z* scores were obtained for height or length, weight, and head circumference according to the following equation for conversion to standardization (Hays, 1973): *z* score = observed value − mean value for normal/standard deviation for normal (i.e., the standard deviation for the normal population of the same chronologic age and gender). Arm anthropometries

could not be converted because standard deviations are not available and were expressed as a percentage of ideal for age and gender based on available normative data. Comparisons of CF and control participants on demographic and growth variables are presented in Table 1. The two groups were equivalent for demographic variables but not for growth measures.

Psychological Measures

Testing was administered during a single day, with breaks provided as needed, and all children received a lunch break between intellectual and neuropsychological testing.

Intellectual testing. All patients were administered the appropriate Wechsler scales of intelligence—that is, the Wechsler Preschool and Primary Scale of Intelligence (WPPSI; Wechsler, 1967) or the Wechsler Intelligence Scale for Children–Revised (WISC–R; Wechsler, 1974), depending on the child's age. The WPPSI was used with children between 4 and 6 years of age, and the WISC–R was used with children over 6 years of age. The Wechsler scales are the most widely used comprehensive measures of children's intelligence. These scales are designed to measure cognitive abilities such as verbal comprehension, perceptual organization, and memory, and they yield three separate IQ scores: a Verbal IQ (VIQ), a Performance (or nonverbal) IQ (PIQ), and a Full-Scale IQ (FSIQ), which is a composite of the verbal and nonverbal scores. The tests were standardized on a cross-section of the U.S. population and allow comparison of a child's function to other children within 3 months of his or her chronological age. The standardized IQs have a mean of 100 and a standard deviation of 15.

Academic testing. The Wide Range Achievement Test–Revised (WRAT–R; Jastak & Wilkinson, 1984) was administered as a measure of academic achievement. This test assesses skill development in three basic academic areas: reading, spelling, and arithmetic. Performance in each area can be compared to that of other children in the U.S. population who are within 6 months of the child's age. Standard scores are yielded for each area and have a mean of 100 and a standard deviation of 15.

Neuropsychological testing. The Reitan–Indiana Neuropsychological Test Battery was chosen to assess neuropsychological function because it allows a comprehensive assessment of brain-related abilities by including multiple tests for each category of function and is sensitive to brain damage (Nici & Reitan, 1986; Reitan, 1987). Neuropsychological tests provide a more specific assessment of brain functions than do the global measures obtained from

the intelligence scales. The specific tests and the functions they assess are presented in Table 2.

For the neuropsychological tests, summary scores were calculated for each participant to represent his or her performance in each of the six categories of ability. Summary scores for each of the categories were computed as follows: Combined raw score distributions for the two participant groups for each variable were transformed into a normalized *T*-score distribution, with a mean of 50 and a standard deviation of 10. For those tests on which lower scores signify better performance (such as where the score represents the number of errors on a task), the scales were reversed when the *T*-scores were obtained. *T*-scores for all tests within each category of ability were summed and averaged to obtain the summary score for that category. The two participant groups were then reassembled for the statistical analyses.

Initial analyses of summary scores derived through *T* or *Z* distribution conversions have been used in other investigations of neuropsychological function (Nici & Reitan, 1986; Stewart, Hiltebeitel, et al., 1991; Stewart, Silver, et al., 1991) and are appropriate for the following reasons: Neuropsychological

TABLE 2
Abilities Assessed and Tests[a] Used to Measure Neuropsychological Function of Cystic Fibrosis Patients and Healthy Controls

Category of Ability	*Tests*
Motor	Finger Tapping—dominant hand
	Finger Tapping—nondominant hand
	Marching—dominant hand
	Marching—nondominant hand
	Marching—circles
	Tactual Performance Test—total time
Sensory-Perceptual	Imperceptions—auditory, visual, and tactile
	Tactile Finger Recognition—both hands
	Fingertip Symbol Writing—both hands
	Tactile Form Recognition—both hands
Incidental Learning	Tactual Performance Test—memory
	Tactual Performance Test—localization
Visual-Spatial Skills	Matching Figures
	Matching *V*s
	Star Drawing
	Concentric Squares Drawing
Abstraction, Reasoning, Logical Analysis, and Integration	Matching Pictures
	Category Test
	Color Form Test
	Progressive Figures Test
Language	Aphasia Screening Test verbal items

[a]Detailed descriptions of all tests may be obtained from the Reitan–Indiana Battery manual (Reitan, 1964).

skills are assessed using a variety of different tests for each category of ability, and individual raw scores are not directly comparable across tests. A summary score allows groups to be compared on performance in skill areas rather than on specific tests. Furthermore, initial analyses of summary scores significantly reduce the high likelihood of Type I error that would exist if the scores were compared separately for each of the tests.

The Neuropsychological Deficit Scale (NDS) total score was also obtained for each participant. This score, which provides an overall indication of brain-related functioning, is obtained by summing NDS scores in three areas: the level of performance on the separate tests, right–left comparisons for the Motor and Sensory-Perceptual tests, and the Aphasia Screening variables. Conversion to NDS scores was based on the normative data provided by Reitan (Reitan, 1987). Reitan's conversion results in a range of scores from 0 to 3 for each test and comparison, with the total possible score ranging from 0 to 150. The cutoff of 54/55 is designated by Reitan as separating brain damaged from normal subjects.

Data were obtained from all participants on all measures, with the following exceptions: Two patients in the CF group were below 6 years of age and had no school experience. They were not administered the WRAT–R. Two patients in the CF group and 4 patients in the normal control group could not tolerate the blindfold in the Tactual Performance Test (TPT) and therefore did not complete the test. Several of the younger children in both groups could not understand test demands for some individual tests. Their scores were not included, and where significant results are reported, any deviations from the full number of participants in the sample size are indicated.

Data Analyses

The two groups were compared for (a) performance level and (b) conversion to the NDS, an index of brain-related dysfunction (Reitan, 1987).

For performance level, the VIQ, PIQ, and FSIQ from the Wechsler scales and the reading, spelling, and arithmetic standard scores from the WRAT–R were compared using analyses of variance (ANOVAs).

The initial analyses for neuropsychological test data were conducted on the summary scores for each function using ANOVAs. Following the initial analyses, where differences reached significance (defined in our study as $p < .05$), the scores on the individual tests were also compared using ANOVAs. The difference between the two groups for the NDS total score was analyzed using ANOVAs. In addition, the number of participants in each group who fell above or below the cutoff of 54/55 was also compared using Fisher's Exact Test.

The relation between neuropsychological function and severity of the disease was assessed by obtaining Pearson correlation coefficients for the IQ,

academic, and neuropsychological summary scores to the total Shwachman score. In order to investigate which aspects of the disease related most closely to function, where the significance of the correlation was at a level less than .05, the correlations between the individual scores that make up the Shwachman total (i.e., physical condition and activity, pulmonary findings, growth and nutrition, and X-Ray results) and scores in the specific area of function were examined. In addition, as a sample of 20 patients was considered insufficient for parametric analyses, significant correlations were reanalyzed using the Spearman rho coefficient.

RESULTS

Means and standard deviations for CF and control patients on IQ scores, academic standard scores, and neuropsychological summary scores are presented in Table 3. None of the differences between the groups reached significance or trend ($p < .10$) levels. Means $\pm$ standard deviations for the CF versus the control group on the NDS total score were 36.5 $\pm$ 21.3 versus 31.0 $\pm$ 17.4. This difference was not significant. Five CF patients and 3 normal controls fell at or above the cutoff of 55. This difference also was not significant.

The relation between neuropsychological function and measures reflecting the severity of the disease are presented in Table 4. The relation between Sensory-Perceptual function and the Shwachman total score is presented in Figure 1. As significant correlations to the total Shwachman score on one-tailed tests of significance were found for VIQ, FSIQ, Sensory-Perceptual, and Incidental Learning test summary scores, the correlation between the individual components of the total Shwachman score and the measure of function was determined. The correlation between IQ and Shwachman measures was due primarily to a relation between VIQ and the Growth and Nutrition component of the Shwachman total score. For the Incidental Learning summary, only the Growth and Nutrition index from the Shwachman total accounted for the significant relation. For the Sensory-Perceptual measure, the strongest contribution to the overall correlation came from case history measures of Physical Condition and Activity; however, the other three measures contributing to the Shwachman total also were significantly correlated to the Sensory-Perceptual measure.

When additional nonparametric correlation analyses were used, all coefficients indicating the relation among IQ, academic, and summary scores with the Shwachman total score were lower. The correlation of VIQ, the Sensory-Perceptual summary, and the Incidental Learning summary with the Shwachman total were at trend ($p < .10$) levels (ρs = 0.35, 0.37, and 0.30, respectively), whereas the correlation between FSIQ and the Shwachman total score decreased to $p > .10$ ($\rho = 0.27$).

TABLE 3
Means ± Standard Deviations for 20 CF Patients and 20 Healthy Control Patients on IQ, Academic, and Neuropsychological Test Summary Scores

	CF Patients	*Control Patients*
Score	*M* ± *SD*	*M* ± *SD*
VIQ[a]	107.6 ± 18.8	113.1 ± 22.5
PIQ[a]	108.8 ± 16.9	110.6 ± 14.3
FSIQ[a]	108.9 ± 17.8	113.2 ± 18.5
Reading Standard Score[a]	101.8 ± 15.9	104.3 ± 17.1
Spelling Standard Score[a]	99.0 ± 13.6	99.8 ± 19.1
Arithmetic Standard Score[a]	94.6 ± 16.1	99.5 ± 15.8
Motor Summary[b]	49.1 ± 7.8	50.6 ± 6.9
Sensory-Perceptual Summary[b]	49.3 ± 7.3	50.6 ± 6.1
Incidental Learning Summary[b]	47.1 ± 10.7	53.2 ± 6.8
Visual-Spatial Summary[b]	50.1 ± 7.2	49.9 ± 7.0
Abstraction Summary[b]	48.5 ± 8.0	51.5 ± 5.8
Language Summary[b]	49.3 ± 9.0	50.7 ± 11.1

Note. CF = cystic fibrosis. The CF patients and control patients did not differ significantly ($p < .05$) on any of these measures. VIQ = Verbal IQ; PIQ = Performance IQ; FSIQ = Full-Scale IQ.

[a]$M = 100$, $SD = 15$ for IQ and academic standard scores in a normal population. [b]Obtained for each category of ability by combining and averaging normalized *T*-scores for all tests within that category. $M = 50$, $SD = 10$ for *T*-scores of the combined sample of CF children and healthy controls.

DISCUSSION

These findings support earlier reports (Lloyd-Still et al., 1974) indicating that, after the first 5 years of life, the intellectual function of children with CF is no different from that of healthy children. This study extends those findings to include academic scores and neuropsychological measures that are more sensitive to subtle impairment in brain function. Our findings are also consistent with the reports in the literature that, on many measures of psychological function, children with CF show as good performance as do children with other chronic illnesses (Drotar et al., 1981) and even children from the general population (Kashani, Barbero, Wilfley, Morris, & Shepperd, 1988).

The findings regarding the relation between disease-related measures and neuropsychological function suggest the following hypotheses: Children whose disease is least well controlled show more evidence of impact of CF on intellectual and neuropsychological function. VIQ, sensory-perceptual abilities, and incidental learning are most correlated with disease severity. Growth and Nutrition appears to be a particularly important correlate of intellectual and neuropsychological function in CF patients. It is important to note that delayed

TABLE 4
Pearson Correlation Coefficients Reflecting the Relation Between Measures of the Severity of the Disease and Measures of Intellectual, Academic, and Neuropsychological Functioning

	Total	*Physical Condition and Activity*	*Pulmonary Findings*	*Growth and Nutrition*	*X-Ray Results*
VIQ	.43*	.35	.33	.42*	.35
PIQ	.22				
FSIQ	.38*	.32	.32	.41*	.23
Reading Standard Score	.26				
Spelling Standard Score	.29				
Arithmetic Standard Score	.29				
Motor Summary	.36				
Sensory-Perceptual Summary[a]	.62***	.68***	.47*	.53**	.45*
Incidental Learning Summary[a,b]	.47*	.44	.45	.49*	.12
Visual-Spatial Summary[a]	.05				
Abstraction Summary[a]	.07				
Language Summary[a]	.08				

Note. VIQ = Verbal IQ; PIQ = Performance IQ; FSIQ = Full-Scale IQ.
[a]Obtained for each category of ability by combining and averaging normalized *T* scores for all tests within that category. [b]$n = 14$.
*$p < .05$, one-tailed. **$p < .01$, one-tailed. ***$p < .005$, one-tailed.

growth in itself does not result in intellectual and neuropsychological deficits: Our patients, who are significantly growth delayed as a group, do not have deficits as a group. This finding is consistent with findings from other growth-delayed populations, such as patients with juvenile rheumatoid arthritis (Cleveland, Reitman, & Brewer, 1965), who are not found to have deficits as a group.

Although growth has been shown to be an important correlate of intellectual function in infancy in numerous studies involving other clinical populations (Chang, Weisberg, & Fisch, 1984; Stewart et al., 1988; Stewart, Uauy, Waller, Kennard, & Andrews, 1987), in childhood, this relation has been found to be less apparent (Stewart et al., 1987; Stewart et al., 1989), with some indication that measures of growth during infancy rather than measures obtained during the childhood years may continue to predict intelligence over time (Nelson & Deutschberger, 1970). The vulnerability of the human brain is considered to be greatest during the first 2 years after birth (Dobbing & Smart, 1974), and nutritional deficits after that time are hypothesized to have less

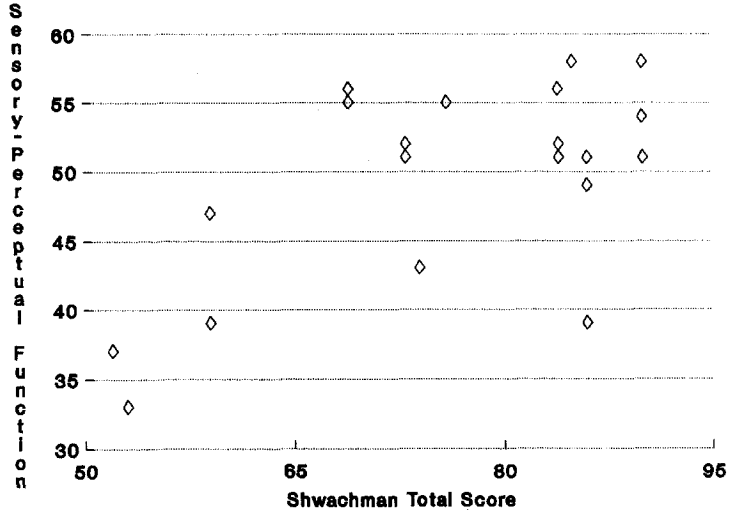

FIGURE 1 The relationship between Sensory-Perceptual summary score and Shwachman total score for 20 patients with CF.

profound effects on the brain. In addition, growth attained in later years may be more influenced by family-line effects, with smaller patients being simply the progeny of smaller parents and not just those most affected by the disorder. Possibly the reason that growth continues to be an important predictor of intellectual function in children with CF is that it is the most sensitive to disease severity of all the Shwachman variables and, therefore, is the best indicator of severity of illness, which is a predictor of intellectual function in other populations of children (Stewart et al., 1987).

Certain measures of intellectual and neuropsychological function are more closely related to disease severity than others in our study. The three measures that correlate with disease severity—VIQ, Sensory-Perceptual function, and Incidental Learning—do not have a common underlying basis to explain why they might be more vulnerable to disease severity than other test measures. VIQ was found in other studies to be sensitive to cerebral damage in children (Reitan & Davison, 1974). The tests that compose the VIQ assess the child's verbal comprehension and expressive skills and provide a relatively stable measure of knowledge acquired through accumulated experience (Sattler, 1988), unlike the other two groupings also affected by disease severity. Although VIQ would be affected by school absence, which could reflect disease severity, the academic tests would be expected to be even more sensitive to school attendance; however, the correlation between academic function and disease severity was not significant. Although sensory-perceptual skills and incidental learning would both be affected by decreased attention, which may

be the more general manifestation of poor health, other measures (e.g., PIQ) would be even more sensitive to alterations in attention, and these measures did not significantly correlate with Shwachman scores.

One common underlying basis for the specific tests that compose both the Sensory-Perceptual summary and the Incidental Learning summary is a reliance on tactile input, which is not important in most of the other tests administered in the battery. The one additional task in the battery that relies on problem-solving skills based on tactile input is the TPT total time score. This measure also has a significant motor demand and was included under the Motor summary, consistent with Reitan's groupings of measures (Reitan, 1964). In order to investigate the possibility that tactile information processing was impaired with disease severity, the relation between the Shwachman total score and the TPT total time score was obtained. Although the Motor summary score does not correlate significantly with the Shwachman total, there was an extremely high correlation ($r = .80$) obtained between the TPT total time score and the Shwachman total. Thus, one possibility is that disease severity diminishes the patient's ability to perform both simple perceptual and problem-solving tasks that rely on tactile perceptual skills.

Indeed, it would be interesting to assess, using item analysis, whether the delays found in children before age 5 in other studies might also be related to diminished tactile-perceptual skills. Tests for younger children usually contain many sensorimotor items, which would be more likely to be affected by tactile-perceptual deficits than would be scores on tests of global intellectual function. This difference in test composition might underlie the differences in findings before and after 5 years of age in children with CF.

It is important to emphasize the preliminary nature of our findings. Our participant groups were small. On several of the important measures, particularly the TPT, which yielded interesting results, the participant group was further reduced because of the difficulty some younger children had in performing the test. It is quite possible that these children had a higher level of impairment on these measures, and their elimination from the sample pool may have affected the findings. Furthermore, the relations between disease severity measures and neuropsychological function were assessed through univariate analyses, which provide only suggestions for further exploration. Much more meaningful information would be provided through multivariate analyses, which should include other potentially salient variables such as SES and gender. Such analyses were precluded in our study by the sample size. Finally, our study takes a cross-sectional perspective, making it difficult to fully elucidate the changes in the relation between disease progression and brain function from a developmental perspective. For example, disease severity and duration of severity may interact over time in ways that cannot be determined by the methodology of our study. Thus, the relations found in our study should be considered tentative and as hypotheses to be explored in future studies.

There are several additional limitations to our findings. The control group consisted of medically well children rather than clinical controls. Although comparison to medically well children does form a more stringent control, it is difficult to determine which of the findings related to disease severity might be the product of physical illness in general, as compared to the result of CF in particular.

In addition, the patients in our study had a well-managed disease. On all measures of severity of the disease, even the children with the least well-managed illness had Shwachman scores no lower than the *fair* category. With radical advances in medical care, the median life expectancy has shown significant increase over the last decade, and children in our participant age group would not be expected to be significantly impaired. One third of the population followed in our clinics are over the age of 20 years, with almost half of this group over the age of 30 years. Possibly if we studied older patients who, as a group, would be more likely to show disease progression, more neuropsychological deficits would be apparent.

CLINICAL IMPLICATIONS

The clinician working with these patients may be more alerted to those children who do have significant growth and nutrition disruption secondary to their disease. These children may merit a more careful evaluation of their cognitive abilities. If the disease progresses, some patients and families may benefit from the support that cognitive evaluation and school intervention could provide, and referral to a psychologist may be helpful.

In counseling the parents of children that have been newly diagnosed, the information that the children are cognitively quite intact, as long as the disease is well controlled, may be the source of some comfort. Our findings are heartening because they indicate that, despite the disruption of everyday life that children with even well-managed CF can face, the quality of their lives based on multiple measures of neuropsychological function appears to be good.

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