Children's Knowledge of Their Disease,  
Cystic Fibrosis

by:

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Introduction

Cystic Fibrosis is a hereditary disease, the effects of which are seen primarily in the lungs and digestive system. Mucus, which is normally thin and slippery, is more viscid and sticky in patients with CF. Consequently, this mucus tends to remain in the lungs, blocking bronchial tubes and providing a conducive environment for bacteria to multiply and cause infection. Mucus also tends to obstruct the ducts that carry enzymes from the pancreas to the small intestine where most digestion occurs. Without enzymes, food passes through the digestive tract only partially digested.

The disease is transmitted by recessive genes. One person in 20 is a carrier; there is no method at present to detect the carrier state. The chances of two carriers marrying is one in 400. Then, because of the recessive nature of the disease, there is a one in four chance that children of carriers will have the disease. The disease appears in approximately one in every 1600 live births. It is found primarily among Caucasians; it occurs infrequently in Blacks and rarely in Orientals.

Common symptoms of this disease include failure to thrive or gain weight; foul smelling, bulky stools; repeated lung infection; persistent cough; excessive mucus; difficult breathing; vomiting; and clubbing of the fingers.

The exact nature of the genetic defect and its metabolic processes are unknown. Consequently, there is no cure at present and treatment is symptomatic. Treatment may include daily postural drainage (a form of chest physical therapy in which lobes of the lung are vibrated or clapped to help clear the mucus), antibiotics to prevent lung infection, aerosols or mist therapy to thin the mucus, and in severe stages, oxygen may be used to facilitate breathing. Children are instructed to cough...
up the mucus. The digestive problems are treated with replacement enzymes which digest food. Children are taught to take these pills with meals and snacks. Additionally, a restricted diet is recommended, low in fat and high in both protein and calories. Such a diet aids in digestion and weight gain.

Recurrent infection causes scarring or fibrosis of the lung. Additionally, lobes may collapse and bronchial tubes may be destroyed as the disease progresses. Death usually results from severe pulmonary complications. Just twenty years ago, most of these patients died in early childhood. Today, with present treatment methods, life span has generally been lengthened and patients may live to young adulthood.

Review of the Literature

In our research we investigated the knowledge of their disease held by latency age children with cystic fibrosis. Our plan was to discover the extent of their knowledge about CF, to uncover their misconceptions, and to elicit their anxieties about the disease. Such findings would have important ramifications for patient education as a part of total pediatric pulmonary care.

Our literature review gave us basic information on which to build our research study, but we found no experimental study with cystic fibrosis such as the one we undertook. However, several articles underlined the importance of and the need for patient education in chronic and fatal illnesses. For example, Cotter and Schwartz note that "All children need to be given reasons for the treatment and the uncomfortable procedures they must undergo. Otherwise, treatment may be interpreted as punishment."\(^1\) In dealing with children who have fatal illnesses,
Morrissey writes, "The goals for helping children in this situation may well have to be limited -- to imparting a better understanding of their illness and medical procedures, to shoring up feelings of reassurance and support. Yet these limitations may be all the more reason for providing so sensitive and humane a service."² The importance of education was stressed, but research into educational efforts is sparse. As Spinetta et al. write, "few or no objectively based data have been gathered from the younger child himself on what he knows about his illness or what his psychological reactions are to it."³ Another researcher points out that "Patient needs for education are diverse, yet, they have not been differentiated. Even more important is the overall lack of quality of the service."⁴ It is further emphasized that "What we have, as the President's Council on Health Education found, is a great deal of health information but very little health education that helps the patient put the information to use."⁵ Others note that if the task of education is not undertaken "to the best of his {the patient's} ability to grasp it, he will seek this information elsewhere and will not obtain it as satisfactorily."⁶

Factual understanding is important since in uninformed children their concepts of the disease can give rise to anxiety - provoking fantasies.⁷ In fact the fantasies a child concocts to explain his disease may cause more anxiety than the truth itself. Moreover, as Singher writes, "Children can be worried by a conspiracy of secrecy and relieved by the opportunity to discuss with a sympathetic person his ideas and concepts."⁸ He adds, "I am convinced that a child worries most when he suspects the adult world is hiding something
from him."9 Cotter and Schwartz write, "For many children, especially	hose between 6 and 11 years of age, the procedures and side effects
of therapy will produce more anxiety than the illness itself."10 And
Singher notes that procedures can be explained and questions answered
to alleviate the child's anxieties. With an experimental group of
hospitalized children, researchers found that rational explanations of
projected procedures were valuable in limiting distorted perceptions.12
And a study of children with cystic fibrosis notes that "the fact that
the situation was discussed with them appeared to relieve some anxiety."13

Parents are often concerned about what the child should be told.14
One study of twenty-five families with cystic fibrosis patients found
that "60% of the parents indicated that they had never discussed the
diagnosis with the afflicted child."15 Nevertheless, as Waechter notes,
"the protectiveness of adults may not be entirely effective in preventing
children with fatal illness from experiencing anxiety..."16

Our investigation of the literature substantiated the need for
patient education and the existence of little research in this area. We
found no study that investigated how much cystic fibrosis children know
about their disease. Our hypothesis is that latency aged children with
cystic fibrosis do not have adequate knowledge of their disease and its
treatment requirements. If our hypothesis is true, this would support
the need for increased patient and family education to improve total
health care. We feel that this problem requires investigation because
our lack of research knowledge prevents health care teams from pro-
viding optimal care in treatment of patients with cystic fibrosis.
Knowledge of their disease may give patients and families a more
realistic view of their illness and may help alleviate anxiety. Our
research is relevant to the knowledge base of the social work profession because as a helping profession, we are committed to aiding people in fulfilling their life goals. When severe illness and anxiety interfere, we need to intervene as effectively as possible. Our choice of intervention must be based on knowledge. Therefore, we first need to know what information is lacking, what will be helpful, and what will be a source of comfort to our patients. This information may also be important in promoting effective family interaction.

Methodology

Our population consisted of CF patients between the ages of six and thirteen who regularly attend the Tulane Pediatric Pulmonary Clinic. We received permission to administer the questionnaire from both the parents and the children. Testing was carried out during their regular clinic visits, which occur approximately once every three months. We selected our sample from the latency age children that were available to us during clinic visits from August to October of 1979. Due to summer vacations, rescheduling, and hospitalization, we obtained 24 respondents out of an expected 30.

Our instrument was a questionnaire comprised of twenty-five items that measured the subjects' knowledge of CF, including the genetics, the physiology, diet and treatment; three of the questions elicited the children's feelings and perceptions; and a final question inquired where they had received their information about their disease. Of the twenty-five questions, fifteen required only a yes or no response. The other ten required only short answers.

We designed this questionnaire with the aid of the interdisciplinary
team members of the Tulane Pediatric Pulmonary Clinic. We interviewed the physicians, social workers, physical therapist, nutritionists, pulmonary technicians, and nurses involved in the care of these children. From these discussions, we devised a sample test which we then distributed to all team members for comment. After reviewing their recommendations, we revised the original and thereby constructed our present questionnaire. It was then approved by the committee on the Use of Human Subjects at Tulane Medical Center.

Because of the wide age range of our sample population, our test was constructed of age-appropriate questions, ranging from a simple one that requested the name of their illness to a more difficult one that asked why finger casts are made. With this structure of questions, we anticipated a wide range of scores. We purposely began with simple questions in an attempt to lower test anxiety.

Each author administered the test orally to approximately half of our sample. We conducted the questioning in one room which was free of distractions. We each were responsible for reading the questions and recording all answers. We were consistent in our instructions to the children, and prior to administering the test, we encouraged them to answer as well as they could but stressed that we did not expect them to know all the answers. We requested that they honestly say when they did not know an answer; we hoped to avoid guessing. We also told them that upon completion of the questionnaire, we would answer any questions they had. After each fifth question, the examiner provided positive reinforcement such as "Okay, you're doing just fine" or non-verbal encouragement to help further minimize test anxiety.
Results

Our population consisted of eight females, seven white and one black, and sixteen males, thirteen white and three black. Their ages ranged from six years to twelve years, nine months. All socio-economic categories were represented, with the majority of respondents coming from the middle class. The children's illness varied in severity. Eleven were classified as mild, eight as moderate, and five as severe. This determination was the physician's current evaluation of each child's health status. All parents of the children in this study gave permission for their children to participate. Only half of the parents examined the questionnaire and subsequently, one mother requested that question three (Can you outgrow cystic fibrosis?) and question thirteen (Is there a cure for cystic fibrosis?) be omitted. This mother felt that her son would not know the answers and would later seek them from her.

Scores ranged from a low of 20% (4 correct answers) to a high of 100% (25 correct). The mean score was 64.79% and the median was 64%. The mean score for the females was 65%, and the mean for the males was 63.43%. Thus, no significant difference was found between the sexes in this small sample. As expected, there was a positive correlation between the age group and level of knowledge about CF. Specifically, children between the ages of six and seven years, eleven months had an average score of 45%. 70.31% was the average score for children between the ages of eight years, one month and ten years, seven months. The oldest age group, from eleven years, two months to twelve years, nine months, averaged a score of 86%.

Our study also revealed a positive correlation between severity of illness and knowledge about CF. The average score in the children whose illness was considered mild was 55.27%; the average in those
seen as moderate was 66.8%; and the average in those in the severe stages of CF was 82.4%. Table II indicates the scores for each of these three categories.

We attempted to correlate knowledge of CF with the number of siblings who also had the disease. However, only three children had siblings with CF; the sample was too small for such analysis. Similarly only five children were diagnosed after the age of four years, and we could make no comparisons between level of knowledge and age of diagnosis.

Table III illustrates the number of incorrect responses to questions 1 through 18. The questions most frequently answered incorrectly were number 6 (Does CF affect your fingers?), number 9 (Does CF affect your pancreas?), and number 18 (Why are finger casts made?). With question 6, sixteen out of twenty-four children did not understand about clubbing, and this fact is consistent with our finding that nineteen out of twenty-four children did not realize that finger casts measure the degree of finger clubbing. Question number 9 was the one most frequently missed. Twenty out of twenty-four or 83.33% did not know that CF affects the pancreas. In fact, only four children were familiar with the meaning of the word "pancreas".

Only a small percentage of the children had difficulty with question 14 (Can someone else catch CF from you?) One child answered that CF could be contagious; another child was unsure. All children responded correctly to question 12 (Do your Mom or Dad have CF?)

**Question no. 4:** Can someone with CF eat anything they want? This question presented problems both to the examiners and to many of the children. The children had difficulty understanding that
the question was asking about dietary restrictions. However, we followed up the initial question with parts a and b (Name 3 foods you can eat, name 3 foods you cannot eat) and correlated all answers to determine if the child understood the association. Significantly, 43% of the answers supplied by the children about diet were incorrect or incomplete. Of the correct answers, children most frequently cited steak as a food that could be eaten (6 out of 24 or 25%). Their second most frequent response was baked potato (4 out of 24) or 16 2/3%. For foods that could not be eaten, ten children (41%) cited chocolate and eight children (33%) cited greasy foods. It was noticeable that only two children specified MCT oil and only three noted low fat products such as ice milk, or low fat milk and cheese as part of the CF diet.

**Question 15:** Why do you need postural drainage? Many children understood the purpose of postural drainage. Sixteen children or 66% knew it helped loosen the mucus so that it could be coughed out of the lungs. Two felt that it would keep them from getting sick; they could not explain why. Five children had no idea about the purpose of postural drainage, and one child mistakenly thought it would prevent coughing.

**Question 16:** Why do you take enzymes? Sixteen children or 66% knew the answer to this question; five did not know. Four were vague, stating that enzymes either helped or prevented them from becoming sick; they could not be more specific. One child would only respond, "Something bad will happen if I don't take them."

**Question 17:** Why do you come to this clinic? In answering this question thirteen children or 54% said they came to be checked, six said to be helped, and five did not know.
Question 18: Why are finger casts made? Nineteen children (79%) did not understand the purpose of finger casts. Only five gave an adequate response and only two of these specifically mentioned clubbing. Sixteen children stated they did not know, and the remaining three gave incorrect answers. One felt that a finger cast measured the length of the finger and another reported that it removed wrinkles from the finger. One child stated, "to see how much it affects the brain." Interestingly, question 18 was the only one that this child answered incorrectly.

Questions 19 through 22 were discussion questions which had no incorrect answers.

Question 19: What is the worst thing about having CF? The most common response (ten children) was related to treatment regimens. One child specified the mist treatment, another disliked pulmonary function tests, and a third said treatment in general. Five children disliked the postural drainage, and two of these mentioned that this treatment took so much time that their play and activities were restricted. Four children felt that symptoms were the worst aspects of CF. Three of these concentrated on stomach pains and coughing and one child, in the severe stage of CF, specified that the worst was a collapsed lung. Three children's answers indicated that CF had influenced their self image. One boy felt that he could not play sports, a second child that he could not do as much as other children, (eat and run around), and a third felt that he "can't be like other normal kids." The child reported that he looks "a little different" and he then pointed to his extremely clubbed fingers.
Two children reported that there were no bad aspects to CF. Only one child said that hospitalization was the worst part of illness and four reported they did not know.

**Question 20:** What is the best thing about having cystic fibrosis? Seven children responded that there was nothing good about CF. One child in particular expressed feelings of isolation due to her disease. Seven other children said they did not know. Three children felt that certain aspects of treatment were the best part about their illness and they specifically referred to hospitalization, medicine, and clinic. Getting out of school was the best aspect for two children. Another three children expressed positive consequences in terms of "getting out more"; one of these children was a CF poster child. One six year old boy felt he could get rid of CF, and another said "Let me put it this way: I'm not as bad off as some other people who have cancer or cannot walk." Two children would not express themselves any further than saying "It helps a little bit" and "It makes me part of a minority."

**Question 21:** What things do you think CF children can do better than other kids your age? Eight children replied that there was no difference between CF children and others; eight stated they did not know. Four referred to expertise related to medical issues: one child noted he could swallow pills better; another commented that she was better at treatment and drainage; a third felt they knew more about their insides than children without CF, and the fourth felt she was better at attending clinic.

Two children held misconceptions: One felt he could run better and faster (however, another child had emphatically noted that CF children could not run as well because they had more mucus). Another false
notion was that CF makes the child smarter. (This idea has had support in the past; however, recent studies indicate that while there is no difference in IQ, CF children tend to be more verbal than average and thus may appear more intelligent.\textsuperscript{17})

Our CF poster child believed that CF children are better at acrobatics. Finally, one child felt that children with CF are better because they "get more care."

\textbf{Question 22:} Where did you learn most of what you know about CF? Five children reported that their source was the clinic staff, six their mothers, and four both parents. Additionally, three children mentioned several sources which included parents, sister, and clinic staff. Two mentioned only the doctors and another stated she had learned information in the hospital. One child said he acquired his knowledge in school; and a six year old girl said "From nobody, I picked it up on my own." The child with the lowest score said he did not know.

\textbf{Shortcomings of the study}

The major shortcoming of our research was the small size of our sample, which was due to the small number of latency age children in the Tulane Pediatric Pulmonary Clinic. If this study were to be repeated and to include several cystic fibrosis centers, a large enough sample could be obtained to allow statistical analysis and the elucidation of significant differences.

From the results of the questionnaire, we feel that question number 4 was too ambiguous. Had the question been stated, "Because you have CF, can you eat ___?" we might have obtained responses that provided us with more information about the children's understanding of their diet.
Since no other test of this type exists, we had no measure of reliability for our own instrument. This was unavoidable.

Although we had an extremely wide range of test scores, there were slightly more high scores than we had anticipated, including one 100%. A redesign of the questionnaire might include more difficult questions. For example, children could be asked about diagnostic measures, current research, and medications.

Although we tried to minimize investigator effects, (for example, by standardizing the introductory speech to the children) two examiners administered the questionnaires and differences in style of questioning, personality etc. may have influenced responses. Use of a single investigator could have minimized this shortcoming.

Conclusions:

Our research confirmed our hypothesis that latency aged children with cystic fibrosis have an inadequate knowledge of their disease and its treatment requirements. It substantiated the need for increased patient education because although some children did well, the majority lacked considerable knowledge about CF (Tables I and II). The restricted diet appears particularly to call for further explanations. Almost all of the children had difficulty in this area; few could name even three foods they could and could not eat. Our study also indicated a need for education regarding all aspects of treatment. Our findings can now guide the Tulane Pediatric Pulmonary team in supplying this important information and thereby providing optimal care.
Unexpectedly, the questionnaire itself proved to be of therapeutic value to some children. The investigators provided a setting that offered the children an opportunity to ventilate feelings and gave the children permission to express negative thoughts about their illness.

While our results and conclusions are valuable in themselves, we feel that they also are an important foundation for further research. In particular, they can provide a baseline for a study investigating a correlation between knowledge of illness and level of anxiety.
ENDNOTES


5 Ibid.


9 Ibid.


BIBLIOGRAPHY


Questionnaire for Cystic Fibrosis Patients,  
(Latency Age: 6-13)

1. What is the name of your illness? ____________________________

2. Were you born with cystic fibrosis? _________________________

3. Can you outgrow cystic fibrosis? That is, will you ever reach an age when you won't have cystic fibrosis anymore? ____________________________

4. Can someone with cystic fibrosis eat anything they want? ______
   If response is yes:
   (a) What are 3 things you can eat? ____________________________
   (b) What are 3 things you should not eat? ____________________

5. Does cystic fibrosis affect your eyes? _______________________

6. Does cystic fibrosis affect your fingers? _____________________

7. Does cystic fibrosis affect your lungs? _______________________

8. Does cystic fibrosis affect your brain? _______________________ 

9. Does cystic fibrosis affect your pancreas? ____________________

10. Do cystic fibrosis children have lots of mucus phlegm? _______
    (If Yes,) Is it OK to cough up the mucus? ____________________

11. If you had a new baby brother or sister, could they have cystic fibrosis too? ________________________________

12. Do your Mom or Dad have cystic fibrosis? __________________

13. Is there a cure for cystic fibrosis? _________________________

14. Can someone else catch cystic fibrosis from you? ____________

15. Why do you need the postural drainage? ____________________
    (demonstrate clapping for their understanding)

16. Why do you take enzymes? _________________________________

17. Why do you come to this clinic? ____________________________
18. Why are finger casts made, (when you put your finger in the pink clay?)

19. What is the worst thing about having cystic fibrosis?

20. What is the best thing about having cystic fibrosis?

21. What things do you think cystic fibrosis children can do better than other kids your age?

22. Where did you learn most of what you know about cystic fibrosis?
Table I

Mean Scores
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<td>60%</td>
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</tr>
<tr>
<td>56%</td>
<td>56%</td>
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Table III

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<td>41%</td>
<td>29%</td>
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* One mother did not permit the use of questions 3 and 13. The figures for those two questions are therefore based on a population of 23 rather than 24.
INFORMED CONSENT

I understand that I am asked to give my permission for my child to participate in a study of children's knowledge about Cystic Fibrosis. I understand that my child will be asked questions about CF.

Before giving my consent by signing this form, I have been sufficiently informed of the purpose of the study and its administration.

I further understand that my child's participation in this research study is voluntary and that my child may withdraw whenever I or my child chooses. I understand that if I or my child have any questions regarding this study or this form, they will be answered so that I satisfactorily and completely understand.

I understand that my and my child's identity and all other information relating to me will be kept in strict confidence. I authorize release of information from this study to those agencies designated by the principal investigator and/or the granting agency.

I have read and understand this information stated above and I sign this consent form willingly.

In the case of a patient who is under the age of 18 years, this consent must be signed by a parent or legally authorized representative. This is true except for patients under the age of 18 years who are married and can sign their own consent.

Signature_________________________________ Date __________________

Witnessed by_________________________________ Date __________________

In the case of a minor who is twelve years of age or older, this consent form must be signed to indicate the minor's assent to participate in the study.

Signature_________________________________ Date __________________

Witnessed by_________________________________ Date __________________

I am unable to read but this consent form has been read and explained to me by ____________________. I understand the information stated above and I willingly sign.