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PSYCHOLOGICAL ASPECTS OF CYSTIC FIBROSIS IN CHILDREN

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Although altogether modern therapeutic procedures have considerably improved the survival and the quality of life of children with cystic fibrosis, the relevant psychological aspects have still been insufficiently considered. The aim of this study was the evaluation of the psychological characteristics and adjustment of CF children, together with how the family coped.

The psychometric instruments used were: Kohs Design Test, Eyzenck Personality Questionnaire, General Anxiety Scale, Emotional Profile Index, Human Value Test and two projective drawing test for 25 CF children (aged 10.13 ± 2.3 years) and Child Behavior Checklist for the mothers.

The unexpected good psychological results obtained from psychometrics instruments could be explained by the fact that CF children accept the real situation and express vivacity, but their deep feelings of fear impose on them a high level of self-control and resistance. Results from CBCL presented CF children as immature, with accentuated aggressiveness in interpersonal relationships. The most important problem is related to the delay of puberty changes in CF children leading to low self-esteem.

Generally, family members cope relatively well with disease in children, in spite of discrepancies in mother/child reports of child psychopathology, but divorce also occured in a few families, as well as the persistent influence of insecurity of the long-term prognosis for the functioning of the family. Psychological support for patients, family members especially mothers is necessary in the treatment of these children.

A need for a holistic approach in the assessment and therapy, including biofeedback techniques, was pointed out.

Descriptors: CYSTIC FIBROSIS-psychology

INTRODUCTION

Cystic fibrosis (CF) is considered to be the most common genetically based disease in white race populations, with an incidence of 1 in 1,500 - 2,500 live births, while in other races this ratio is 10 to 50 times lower. In Macedonia the incidence of CF is estimated to be 1 in 3,000 live births. Patients are usually diagnosed in early childhood as a result of meconium ileus, malnutrition, diarrhea, repeating respiratory difficulties and infections. The major diagnostic test is the sweet chloride determination (over 60 mmoll/l), but after 1989 DNA testing is also used. The most frequent CF gene mutation is ΔF508, present in about 70% of CF chromosomes worldwide, but altogether over 800 associated mutations have been described (28, 14). In our country genetic diagnosis as well as the antenatal carrier determination are available.

The main problems in patients with CF are related to chronic respiratory infection and the nutritional impact of intestinal malabsorption, but many other complications such as chronic liver disease, diabetes, distal intestinal obstruction, nasal polyps, rectal prolapse, pancreatitis, infertility etc. may also occur. Medical therapy in CF is very extensive in order to maintain optimal health and improve survival. All patients, as well as all family members must be educated and encouraged to participate in therapeutic procedures. Thus, patients' coping strategies and adaptation are of particular importance (16, 11, 18, 26).

Many researchers of CF have tended to focus on gross psychopathological problems in children and parents. It has been shown that CF children do not generally manifest significant psychopathology in comparison with the normal population of the same age. CF children are more maladjusted and experience significant psychological difficulty but do not exhibit major externalizing behavioral problems. Thomson et al. (30) note that CF children manifest fewer depressive and dysthymic diagnoses than psychiatrically referred children.

Important psychosocial problems in CF children's adjustment, comprise: a) acceptance of the disease, including understanding and compliance, b) freedom from severe psychopathology, c) normal or age-appropriate personality functioning and d) age-appropriate functioning in school, with family and peers. Maladjusted children manifest excessive anxiety, depression, school-related behavior problems or disciplinary problems at home.

The other issue is related to parental coping and adaptation to CF disease in children. Many parents cope relatively well and do not manifest major psychopathological problems. But there are critical times during which parental function-

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ing is strained, especially the role of the mother (21). Most of the family problems appear in the first year after the diagnosis of CF. On the following table the potential crises times for parents are presented (Table 1) (29).

Table 1. *Potential crises for parents of CF children*
Tablica 1. *Potencijalne krize u roditelja djece sa cističnom fibrozom*

• Diagnosis and explanation of the meaning of cystic fibrosis / Dijagnoza i objašnjenje značenja cistične fibroze
• Further pregnancies / Sljedeće trudnoće
• Learning to cope with new demands, which may occur each time a new treatment is instigated / Učenje kako se nositi sa novim zahtjevima koji se mogu pojaviti svaki puta kod uvođenja nove terapije
• First course of intravenous antibiotics and isolation of new bacteria from the sputum / Prvo davanje antibiotika intravenozno i izolacija nove bakterije iz sputuma
• Problems with school and employment / Problemi sa školom i zaposlenjem
• Increasing need for antibiotics / Povećana potreba za antibioticima
• Change from pediatric to adult care / Promjena s pedijatrijske na odraslu njegu
• Loss of responsibility such as allowing the child to perform their own treatment / Gubitak odgovornosti kao što je dozvola da dijete samo provodi liječenje
• Decision about transplantation / Odluka o transplantaciji
• Deteriorating health of a sibling with cystic fibrosis, friend, or one's own child / Pogoršanje zdravlja brata ili sestre s CF, prijatelja ili vlastitog djeteta
• Death of a sibling with cystic fibrosis, friend, or own child / Smrt brata ili sestre ili prijatelja s CF
• Bereavement / Žaljenje

Parents of CF children may express signs of depression, anxiety but also anger and hostility. Within a few years the family has more or less adjusted to the disease and equilibrium is restored, but still the process of stress and coping is a dynamic process and continues through the life span of the child and even after his death.

The aim of this study is a comparative evaluation of the personality profiles of school age CF patients with special attention to internalizing symptoms, and relating them to maladjustment and coping. Some controversial aspects of family functioning and child/mother discrepancies are also considered.

METHODS

The diagnostics and management of CF patients in our country began 20 years ago. The first children diagnosed died within a few months. Slowly, with the improvement of early-introduced therapeutic procedures and more exact diagnosis, the survival of CF children became significantly longer. Now, we have a group of 57 surviving CF children aged between 4 months and 22 years in a total population of two million people. The diagnostics is usually performed in the period of early infancy (1-5 months). We very rarely hospitalize children in preschool and school age where the diagnostics had not been established earlier. The more frequent mutation is ΔF508 and pulmonary symptoms are leading. Five year ago we founded the CF Association in Macedonia for the support of CF children and their parents.

We have studied 25 CF children, mean age 10.13 ± 2.29 years (23 boys and only 2 girls), selected from total of 57 children treated presently for CF. The children were examined in the period of improved health conditions (without superinfection, wheezing or gastrointestinal problems).

All results are compared with a control group of 25 healthy children of the same age selected by random from primary schools.

The psychological battery comprised Child Behavior Check List (CBCL), Kohs Design test, Eyzenck Personality Questionnaire (EPQ), General Anxiety Scale (GASC), Emotional Profile Index (EPI), Human Values Test (HVT) and two projective drawing test – the Drawing man test (Machover) as well as the Drawing family test (Corman).

CBCL (1) is designed to obtain the parent's descriptions of their children's behavior in a standardized format. There are 118 behavior problem items plus spaces for parents to write and score additional physical problems with no known medical cause. Two broadband groupings are focussed: internalized and externalized. They reflect a distinction between fearful, inhibited, overcontrolled behavior and aggressive, antisocial, undercontrolled behavior. The profile can contribute to a formal diagnosis by showing the degree of a child's deviance in behaviors that parents are more likely to observe than clinicians as well as help to structure effective training. CBCL is the test most used for the selection of behavior problems in chronically ill children but the agreement between the scales and a mental diagnosis was shown to be moderate (4, 14).

Kohs Design Test (17, 32) presents a simple test for global intellectual functioning. The results are correlated with the logical thinking (analytical-synthetic performance) of the person. In Macedonia standardization was made in 1978.

EPQ (13, 19) shows the four classical characteristics of the personality:

N - level of emotional stability/neurosis; E - dimension of extroversion/introversion; P - dimension of psychotic behavior and L - degree of dissimulation or social adaptability. Our previous experience with this method confirmed the validity, reliability and discriminitivity of the obtained results, especially in preadolescents (10-12 years).

EPI (2) gives the emotional structure of the patients in correlation with their personal characteristics. The basic theoretical concept of the test is the hypothesis that the personality traits are the results of primary emotions and emotional states. The obtained emotional profile indicatir the main conflicting area of the person, defined through eight dimensions (related to eight respective emotional states): incorporation (acceptance), non-control (impulsiveness), self-protection (fear), deprivation (sadness), opposition (refusal), exploration (self-control), aggression (destruction) and reproduction (vivacity). Bias represents the scale for assessment of socially favorable answers.

GASC (27) is a simple questionnaire chosen to show the actual anxiety of the patient. The level of anxiety correlates with fears from situations, persons and objects.

The drawings are used as projective tests in addition to other psychometric instruments (3, 10, 20). Two projective techniques are involved: Machover's analysis of the man drawing and Corman's analysis of the family dra-

wing. The Machover man drawing test is selective for actual problems and conflict within the child himself, and Corman family drawing shows the social and intimate relation of the family members through the development of the patients as well as in the actual situation. Our previous experience in the psychological assessment of children with organic disease through projective techniques is very positive (23, 24).

HVT (25) gives a quick overview of the motivational structure of the personality related to the super-ego component. The hierarchical values are correlated to the real personal needs. It is also a standard for the assessment of the interplay of social situations and the self.

This type of psychometric battery was chosen to obtain the global intellectual, emotional, behavioral and social functioning of CF children.

RESULTS

Firstly, we evaluated the global intellectual score of CF children with Kohs Block-Design test. The obtained score was IQ = 104 ± 32.78 which means that children had normal intellectual functioning.

CBCL obtained from mothers showed "normal" profile for the children' s age (Figure 1). However, three aspects of behavioral problems are more expressed, especially aggression, depression and compulsivity, but within "normal" T-scores (below 65 percentile).

The results obtained for EPQ in CF patients compared with the control group are presented in Table 2.

Generally, CF patients are similar in E, N and L dimensions of EPQ to the control group. Only the P dimension is statistically lower than in the healthy children, which means lower psychopathological traits.

The score obtained for GASC was M = 17.17,SD ± 11.69 (out of 35). The results show that CF children as a group manifest a moderate degree of actual anxiety, but the large standard deviation is related to large differences in anxiety between patients. For example, two of the examined children manifested very high actual anxiety (scores were 32 and 33 respectively), the reason was presumed to be the socio-economic deficiencies of the families. As is well known, the treatment and nurture of CF children are quite expensive, so that in a situation of unemployment the procedure may be compromised, influencing the child's malnutrition and emotional instability. However, generally, chronically ill children are overprotected, in attempt to satisfy most of their needs and wishes.

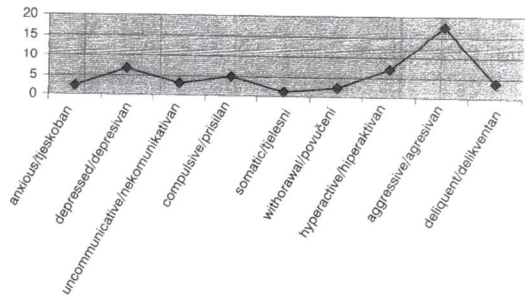

Figure 1. *CBCL from CF children*
Slika 1. *Lista ponašanja djece sa CF*

Table 2. *EPQ from CF and control group*
Tablica 2. *EPQ djece sa CF i kontrolne grupe*

Patients / Pacijenti	P score	E score	N score	L score
CF	6.67 ± 2.90	15.91 ± 2.54	11.67 ± 4.23	15.0 ± 3.81
Control / Kontrola	11.87 ± 6.23	13.16 ± 5.75	13.84 ± 5.31	12.64 ± 4.62
Student t-test	t=2.87 p<0.05*	t=1.66 p>0.05	t=1.18 p>0.05	t=1.45 p>0.05

* Statistically significant / Statistički značajno

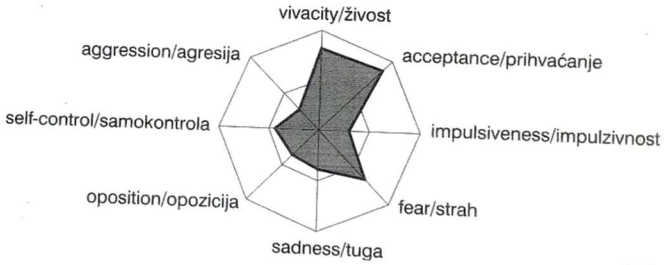

Figure 2. *EPI from CF children*
Slika 2. *EPI djece sa CF*

The results obtained from EPI (Figure 2) showed high bias scores related to dissimulation. Reproduction and incorporation are high, corresponding to the relatively good adaptation of CF children to everyday life. Scores obtained for fear (self-protection) and self-control were also relatively high.

The human values rank obtained from CF children was very interesting. As can be seen from Figure 3, the main value for children is good health, which is clearly understandable. The high value given to friendship and love express the need of support, and the following - freedom is stressed as necessary for living: these children need a feeling of freedom, the possibility to act and make decisions. Personal happiness is also ranked high in the human values related to self-development. The values quoted are generally highly ranked in adolescents and students, so that the obtained results could be interpreted as the earlier mental maturation of CF children. The low value given to beauty, belief in self, success and comfort in life could be related to low self-esteem and disappointment.

The drawings (Machover and Corman) obtained from CF children do not differ in general from those of the control group. Good family functioning, the interrelationship between family members as well as the identity of the pa-

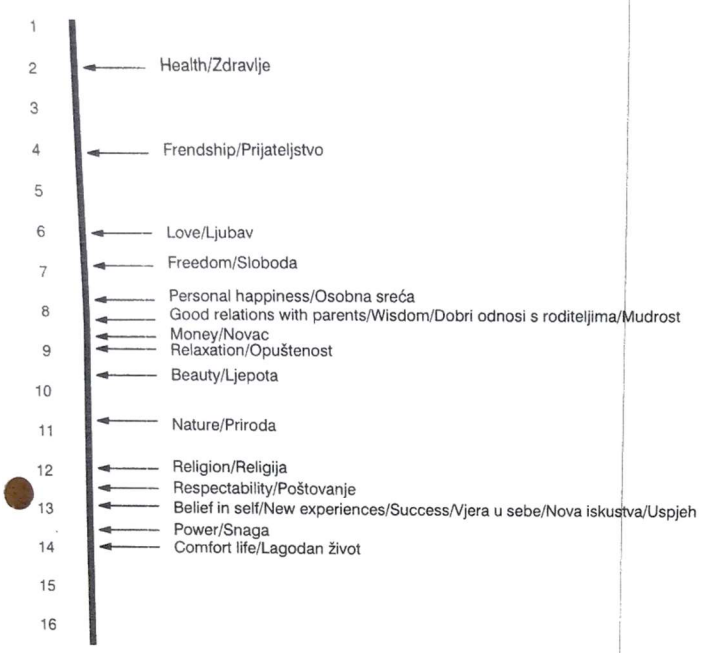

Figure 2. *EPI from CF children*
Slika 2. *EPI djece sa CF*

tients are interpreted as "normal". In three darwings obtained, figures with open arms could be related to an accentuated need for protection and love.

DISCUSSION AND CONCLUSION

1. The results obtained in our evaluation showed the normal intellectual functioning of CF children, which corresponds to the findings of other authors. In spite to chronic hypoxia, significant deficits in cognitive performance were not manifested. The scores obtained correlate to school achievement, so in spite of frequent absenteeism, CF children are good pupils. Moreover, some psychopathological problems happen to be even less frequent than in healthy children (results obtained from EPQ).

Thompson et al. (30) reported that CF children have higher internalized scores on a child behavior checklist and experience distress, anxiety and depression, albeit not more that non-chronically ill children. However, from our test results, interviews of parents and the observation of children, we could generally conclude that the behavior of CF children in our study appeared to be within the normal range. Only two children manifested psychological problems. One of them manifested a depressive reaction in his 17th year, after his mother's and grand mother's deaths. This boy also manifested staturoponderal deficit and sexual retardation. He is not adjusted to all the therapeutic procedures for CF. Another boy, 14 years old, also with staturoponderal deficit, manifested emotional immaturity and a sado-masochistic attitude toward his mother. This boy's scores for aggressiveness were very high. Generally, the results obtained showed that CF children have relatively high cognitive skills, they are moderate extroverts, do not manifest major psychopathological problems and they are not significantly anxious. This corresponds to Blair's (4) finding that most patients with cystic fibrosis are in robust psychological health.

The unexpected good psychological results could be explained by the fact that CF children accept the real situation and express vivacity, but the deep feelings of fear demand of them high self-control and resistance.

2. Most problems arise in the period of puberty and adolescence. The dystimic feelings and fear causing accentuated self-control and self-defense are frequently present. Disturbed self-image, deleted sexual characteristics, staturoponderal deficit, insecurity about the long-term prognosis are factors inducing behavioral and emotional problems in adolescent CF patients. None of our patients showed any sentimental relationship with the opposite sex, confirming the previous findings about the avoidance of close relationships with the opposite sex in adolescence (15).

3. All the families were well informed about the specificities of the disease. In general, they had no significant problems concerning normal family functioning. But, in 3 families divorce was noticed in the period of 1-2 years after the diagnosis, while in two the mothers died (from suicide and cancer, respectively). The poor economic situation is the biggest problem in some families.

As demonstrated in all studies like our own, the main psychological problems in the families are related to the mothers. The feelings of guilt, the everyday pressure related to nurture, medication and drainage procedures, the uncertainty of the future, were factors which influenced the mothers' mental stability. In addition, a difference between the aggressiveness expressed in patient self-evaluation and the opinion of mothers has been noted. Canning (5, 6) also concluded that there existed a discrepancy between parent and child reports concerning child psychiatric problems.

This was also noted by Canning in (7) where maternal distress was correlated with the number of disorders identified by mother, but not with those identified by the child. In our everyday experience fathers are rarely present during control/hospitalization. In some families we did not have any contact with father.

Parental coping with the disease appeared to be problematic, which corresponds with the findings of other authors (31, 22). Our results also agreed with Eddy's (12) concerning the agreement between parents associated with problems in compliance with treatment, which have an adverse impact on the disease and health status of the child with CF.

4. We have interwoven the psychological assessment and support of CF children and their families with the general therapeutic procedure. Psychological interventions varied depending on the type of problem presented and the environmental con-

text in which the child resides. Cognitive and behavioral treatment, combined with biofeedback relaxation procedures were used, and have shown to be quite efficient. Of course, parents and other family members were motivated to encourage the treatment. In our experience, the more frequent psychological problems during therapy were related to the child's adaptation and coping, especially in the period of puberty and adolescence, together with adherence to the medical regime and feeding, as well as parental coping linked to family dysfunction.

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Sažetak

PSIHOLOŠKI ASPEKTI DJECE SA CISTIČNOM FIBROZOM

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Premda su suvremeni terapijski postupci znatno poboljšali preživljavanje i kakvoću života djece sa cističnom fibrozom (CF), malo je pozornosti posvećeno psihološkim aspektima. Cilj ove studije bio je evaluacija psiholoških osobitosti i prilagođenost djece sa cističnom fibrozom, kao i obiteljsko prilagođavanje.

Upotrijebljeni su ovi psihometrijski instrumenti: Kohsov test inteligencije, Eysenckov upitnik ličnosti, skala za opću anksioznost, profil emocija ličnosti, skala ljudskih vrijednosti, kao i dva projektivna testa crteža za skupinu od 25 CF djece (uzrasta 10,13 ± 2.3 godina) kao i Lista djetetovog ponašanja (CBCL) za majke.

Dobiveni rezultati pokazali su normalno intelektualno funkcioniranje djece sa cističnom fibrozom, umjerenu ekstroverziju i aktualnu anksioznost, visoku samokontrolu i samozaštitu, koji odgovaraju osjećaju straha i prilagodbi bolesti. Dobro zdravlje, prijateljstvo i ljubav, kao i osobna sreća, najviše su stupnjevane vrijednosti. Rezultati dobiveni od upitnika CBCL prikazuju djecu sa CF sličnu zdravoj, ali s naglašenom agresivnošću u međuljudskim odnosima.

Članovi obitelji se srazmjerno dobro nose s djetetovom bolešću, premda postoje nesuglasnosti u izjavama majke i djeteta u odnosu na prisutnost djetetove psihopatologije.

Naglašena je potreba holističkom pristupu u procjeni i terapiji, uključujući biofidbek relaksacijske tehnike.

Deskriptori: CISTIČNA FIBROZA-psihologija

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