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Psychiatric Aspects of Primary Immunodeficiency Diseases: The Parental Study

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ABSTRACT

Primary immunodeficiency diseases (PID) consist of a group of long-term illnesses which had permanent psychiatric effects on the patients and their parents. This study was designed to find out the most important origins and aspects of stressor in parents of PID patients. To assess the impact of psychiatric aspects in parents of PID patients, a valid and reliable questionnaire was compiled based on patients' complaints and consulting professionals in PID and psychology.

Fathers of 26 PID patients (17 male and 9 female) were enrolled in this study. According to the result of this study, anxiety for long duration of disease of child (mean score= 4.42), anxiety for incurable diseases of child (mean score=4.23) and anxiety for side effects and complication of treatments on child (mean score=4.08) were the most important stressors of parents.

The comparison between specific PID groups showed that there were significant differences between total score of groups (XLA= 92.8±31.2, CVID=78.7±19.5 and other types of PID= 90.7±22.5, p-value =0.37).

Survey for finding fundamental stressors and continuation of psychological counseling are necessary to achieve successful management of PID patients and their parents.

Keywords: Chronic Disease; Community Psychiatry; Immunologic Deficiency Syndromes

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INTRODUCTION

Primary immunodeficiency diseases (PID) are characterized by chronic disorders with genetic

basis that affect components of the immune system and predispose patients to both infectious and non-infectious immune-mediated complications.^{1,2} This cluster of progressive and fatal diseases usually affect patients by prolix course from childhood to the end of life. Before the time of the death, the well treated patients showed relatively normal life span despite impaired physical or mental functioning, if they were under the monthly therapy with intravenous immunoglobulin (IVIG) and prophylactic antibiotics.^{3,4} The most significant clinical presentations in PID cases are recurrent infections, although the frequency of autoimmune diseases and malignancies are considerable.^{2,5-10} Complications of PID may lead to decrease in quality of life of both cases and their relatives.¹¹⁻¹⁷ PID, similar to the other chronic diseases, predisposes patient and his/her guardians to permanent emotional distresses which can, over the course of time, cause anxiety or despair.¹⁸ Therefore, PID patients and their family may suffer from significant and permanent interference psychiatric effects of the disease. PID children illustrate higher quality of life when they and their guardians have successful mastering on the physical, social, and emotional hardships associating with PID.¹⁹

Regarding the psychiatric problems that PID patients' guardians face, this study was designed to find out the most important origins and aspects which should be mentioned by pediatricians and psychiatrists.

MATERIALS AND METHODS

Study Population

The population of our study was selected from parents of PID patients who were diagnosed at the Children's Medical Center Hospital (Pediatrics Center of Excellence in Tehran, Iran) during 2008-2009. The diagnosis of all patients was previously confirmed based on the diagnostic criteria defined by European Society for Immunodeficiency (ESID) and Pan-American Group of Immunodeficiency (PAGID).²⁰ The Immunodeficiency Clinic at Children's Medical Center is a referral center for pediatric PID patients. Fathers of patients were enrolled in the study when their children were: (1) patients under regular IVIG replacement therapy with 400-500 mg/kg for every 3-4 weeks; (2) age range between 4 and 15 years; (3) No hospital admission during the previous month; (4) No major stress in previous 3 months; (5) No history of any

psychological diseases and problems.

Because of the reduction of bias in data collection, we restricted the gender of parents and the questionnaires were filled only by fathers of patients. Demographic data and overall score of psychiatric problems were evaluated for each participant. Prior to data collection, the study was approved in the ethics committee in Tehran University of Medical Sciences and informed consents were taken from all of the subjects.

Survey Approach

To assess a score of psychiatric aspects in parents of PID patients, a questionnaire was compiled based on patients' complaints and consulting professionals in PID and psychologist for content validity. An expert questionnaire compiler was recruited to design a pilot study to make the questionnaire reliable and valid (kappa coefficient=0.83, Cronbach's alpha=0.78).

The final version of questionnaire with 28 closed questions was prepared to evaluate different psychiatric aspects which parents of PID patients might face with their PID children.

The overall score of each participant was computed by summing the likert type scale value of each question from 1 to 5 score. Passing the exam was defined as answering more than 2/3 of the questions. The maximum possible measurement was 140 and the minimum score was 28.

Data Analysis

Correlation analyses were done using Pearson's product moment correlation coefficients. Statistical tests were two-tailed intra group. Kruskal-Wallis H was performed when comparison of scores was aimed between groups of diseases. Moreover, to handle as many observations as possible, missing data for repeated measurements were computed using an explicit regression model (i.e., repeated measure model with unstructured covariance matrix) that included previously observed scores of the participants as well as the important covariates.

RESULTS

Fathers of 26 PAD patients (19 males and 7 females), aged 4-15 (median=6) years, were included in this study. All participants were native Persian speakers and could easily understand the questionnaire. Before the

Table 1. Comparison of parents' psychiatric scores in different types of PID disease.

Groups	Number of Patients	Mean Score	Minimum Score	Maximum Score	Comparison with CVID*	Comparison with XLA*	Comparison with other PID*
CVID	16	78.7±19.5	37.00	106.00	-	0.645	0.788
XLA	5	92.8±31.2	43.00	127.00	0.645	-	0.548
Other PID	5	90.7±22.5	78.00	102.00	0.788	0.548	-
Total	26	84.3	37.00	127.00			

**p* values measured by Post Hoc analysis of Scheffe.

time of study, patients were followed for a total of 138patient-years with a mean follow-up of 5.0±3.7 years per patient.

After the pilot study and final modification of questionnaire, the mean score of participants was 84.30 ± 14.92. The lowest score was 43 (1.53 mean score for each question) and highest score was 127 (4.5 mean score for each question).

Fathers were equally concerned about their sons and their daughters (83.4±22.4 vs. 86.7±18.8,*p*=0.38).

The comparison of specific disease group showed that there were no significant differences between groups (XLA= 92.8±31.2, CVID=78.7±19.5 and other types of PID= 90.7±22.5; *F*=1.088, *p*-value of Kruskal-Wallis H analysis=0.24, Table1). Pair analysis of diseased group was abstracted in table1.

Table 2. The most important psychiatric stressors for parents of PID patients.

No	Stressor	Mean score	Percent of total
1	Anxiety for long duration of disease of child	4.42	5.24
2	Anxiety for incurable diseases of child	4.23	5.02
3	Anxiety for side effects and complication of treatments on child	4.08	4.84
4	Anxiety for future events of child	3.77	4.47
5	Anxiety for availability to PID necessary medical services	3.77	4.47
6	Anxiety for requiring to lifelong emergent services	3.77	4.47
7	Anxiety for abnormal growth and socioeconomically under development of child	3.73	4.42
8	Anxiety for precocious death of child	3.69	4.38
9	Anxiety for financial security and cost for expenditure of treatment	3.54	4.20
10	Anxiety for proscriptio from best available treatment for child	3.46	4.10
11	Anxiety for time consumption and missing days from work because of disease of child	3.23	3.83
12	Anxiety for lifelong need to the nursing services	3.23	3.83
13	Anxiety because of dangerous data giving by doctors	3.12	3.70
14	Anxiety for quality of medical services for child because of recurrent respects	3.04	3.61
15	Anxiety for supports of child when I am not exist	2.92	3.46
16	Anxiety for bearing another child with same genetically disease	2.81	3.33
17	Anxiety for shortage of data about the disease of child	2.81	3.33
18	Anxiety for destroying reliefs and opportunities because of child	2.77	3.29
19	Anxiety for lacking of relevant and families' supports	2.77	3.29
20	Anxiety for marriage and family making of child	2.65	3.14
21	Anxiety for tribulation of my development because of child	2.42	2.87
22	Anxiety for increasing problems of child in home	2.35	2.79
23	Anxiety for hiding disease of child from others	2.31	2.74
24	Anxiety for education and school attention of child	2.31	2.74
25	Anxiety for death due to disease because of existence of similar condition in our family	2.19	2.60
26	Anxiety for occurrence of disease in my other healthy child	1.81	2.15
27	Anxiety for presentation of my child in social environments	1.77	2.10
28	Anxiety for home remaining of child and I wish always she/he be admitted in hospital	1.35	1.60

There were no significant correlation between the scores of parents' psychiatric aspects and the age of the PID children ($r=0.123$; $R^2=0.015$).

The most important anxiety aspect of parents was anxiety for long duration of diseases because of childhood onset and long life suffering (mean score = 4.42; 5.2% of total stressor issues).

The second issue was treatment status of PID patients which was considered by parents (mean score = 4.23; 5% of total stressor issues). Therefore, concerns were high for the exact method of treatment and available choices for complete cure.

Side effects of current treatments such as immunoglobulin replacement therapy and long-term use of antibiotics were another serious stressor for parents of PID patients (mean score = 4.08; 4.8% of total stressor issues). Other stressors which were accepted in pilot study and their scores were listed in table 2.

DISCUSSION

Previous studies reported that 7-10% of all children had long-term diseases, if only serious chronic illnesses with primary physical origin are included.^{21,22} Albeit, it is estimated that 30-40% of children suffering from one or more long-term disorders, if visual and hearing impairments, mental retardation, and speech, learning, and behavior disorders are included in this category.²³⁻²⁵

Among such life-threatening illnesses, PID diseases showed influence on both physical and emotional functions of children and parents, in which usually are a non-limited course of disease and had a role in the child's overall development.^{26,27} PID patients had many sharing manifestations from five different classifications of long-term childhood disorders¹⁹. According to these classifications, PIDs are the result of abnormal hereditary traits (second classification) and cause neonatal/childhood recurrent infections and neoplasms (fifth classification).^{28,29}

Providing multidimensional care on PID patients' needs to promote realistic attitudes and decrease psychiatric stressors of children and their parents.³⁰ This aim should be in the plan of pediatricians and immunologists who care for developing children as socially competent and productive persons. Therefore physicians may be helped by awareness about the major causes for emotional stress in PID diseases.³¹

Parents of PID patients show a consequential pattern in adaptation with their difficult psychiatric situation as well as their children. Acceptance of child's disability and its impact on the family status is crucial in promoting a healthy personality development of the child.

This paper intends to assess the common forms of emotional stress, experienced by the parents of children with a long-term PID.

The results of our study showed that anxiety for long duration of disease of child, anxiety for incurable diseases of child and anxiety for side effects and complication of treatments on child are the most important stressor of parents. Moreover, we found that the psychiatric score did not differ between groups of diseases indicating similar impacts of PID disease on parents of cases.

Although the initial reaction of parents of chronically ill patients such as PIDs may usually include acute fear and anxiety related to the possible fatal outcome of the disease, and disbelief in the diagnosis,^{30,32} Scant information by the physician, uncooperative attitudes, mourning the loss of their desired normal child and self-blame are of their concerns.^{22,33}

Interestingly, the results of this study showed that parents of PID children had higher anxiety for unsuitable information of physicians rather than losing the data about the diseases.

Because of usually prominent prolonged parental over-concern in mothers, we decided to conduct this study on the fathers.

Inattentive position of physicians to common determinants of prolonged parental over-concern factors^{34,35} may lead to strong unresolved feelings of guilt, parental rejection or neglect of a PID child and to extreme parental denial of the severity of the illness.^{31,36}

Efforts for finding psychiatric aspects in parents of PID patients is helpful to support their psychological defense mechanisms coping with the constant strain to reduce blame crises and complications on the child, increase power of parents for guiding their child and reduce child sensing the parental rejection which encourages child to self-controlling, regular school attendance and promote their social activities.^{37,38} Despite introducing the coping mechanisms, association with other parents,³⁹ who have successfully adapted to the challenge of raising a PID patient is

helpful for many parents and should be programmed by responsible physicians.^{22,31,40}

Evaluation of psychiatric aspects related to PID patients' needs an individual study assessing psychological impact and final emotional, intellectual, and social outcomes in these patients. Some of stressors which should be verified is chronic time of pain and infections, recurrent hospital admissions, life-long nursing and treatment procedures, hereditary and progressive nature of PIDs, carrying multi-organ failure and poor prognosis, threats of exacerbations, lasting physical impairment, shortened life expectancy, mounting medical expenses and side effects of immunoglobulin replacement therapy and different types of prophylactic antibiotics, interference of illness with schooling/ leisure activities/ vocational training/ job opportunities and changes in coping behavior or adaptation of children.

PID is a long-term physical illness which affects psychiatric aspect of patients and their relatives. Survey for finding fundamental stressors and continuation of psychological counseling are necessary for achieving successful treatment of PID patients. The pediatricians and immunologists should pay attention to important role of parental psychiatrics' attitudes in encouraging PID patients during long process of treatment.

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