ORIGINAL ARTICLE
Iran J Allergy Asthma Immunol
March 2006; 5(1): 23-27

Health-Related Quality of Life in Primary Immune Deficient Patients

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Received: 27 November 2005; Received in revised form: 28 January 2006; Accepted: 15 February 2006

ABSTRACT

The primary immunodeficiency (PI) disorders are abnormalities in development and maturation of the immune system. Individuals with PI disease may experience frequent infections, which limit their abilities to exhibit physical and psychological well-being secondary to their illness. In this survey we compared health-related quality of life of primary immune deficient patients with healthy children.

The case-control study was designed for patients with PI disease who were referred to Children Medical Center in 2004-2005. Demographic information was taken and Pediatric Quality Of Life (PEDQOL) questionnaire were filled for 50 PI patients and 100 healthy children.

The mean age in PI patients was 12.62± 3.65 (range from 8 to 18) years and in the control group was 11.04± 3.3 years. In PI patients 68% were male and 32% female .Most patients with PI disease had a diagnosis of common variable immunodeficiency (54%) or X-linked agammaglobulinemia (24%). Patients with PI disease had great limitations in physical functioning and psychological well-being ($p<0.001$ and $p<0.001$ respectively) compared with children without a chronic health condition. Patients had lower PEDQOL scores in all age groups compared with normal sample ($p<0.001$). Long duration of disease significantly correlated with low psychological score. ($r = -3.23$. $P= 0.03$)

Children with PI disease experience poorer health related quality of life than healthy children, indicating more attention should be paid to early diagnosis and treatment of PI disease, as well as more attention to their social limitation. PI patients may need psychological consultation for better coping with their illness.

Key words: Immunologic Deficiency Syndromes; Pediatrics; Quality of life; Questionnaire

INTRODUCTION

Primary immunodeficiency disease (PID) consists of a group of disorders in which there is an intrinsic impairment in the body's immune system. The true prevalence of PID is not yet well established, although initial studies have estimated that 50,000 individuals in the United States have a kind of PID, with many more having an undiagnosed milder form of PID.[1] The incidence of these diseases vary, ranging from 1:400 to 1:500,000 live births in the United States.[2]

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Approximately 40% of those with a PI diagnosis are children and adolescents younger than 18 years.[2,3] As a result of their impaired immune system ,children with PID have an increased susceptibility to frequent and multiple infections and to other immune system disorders such as anemia, arthritis, and autoimmune diseases.[1-6] Clinical presentation is highly variable, ranging from various patterns of microbial susceptibility to episodes of allergy, lymphoproliferation or autoimmune manifestations.[7,8] Although survival rates and prognosis have greatly improved, children with PID are still at risk for physical, social, and psychological problems.[1,9] They are at an increased risk of school absenteeism, restricting participation in athletic and social activities and experiencing anxiety or depressive symptoms in response to living with their chronic unhealthy condition.[10]

It is now widely acknowledged that the personal burden of illness cannot be described fully by measures of disease status.[11] The quality, effectiveness, and efficiency of health care are often evaluated by their impact on a patient's "quality of life".[12] Health related quality of life (HRQOL) is a multidimensional concept that encompasses measurements of physical ,psychological and social well-being that assesses an individual's perception of the impact of illness on his or her life.[9,11,13] Health related quality of life is measured from the patient's perspective and is not determined by the clinician's evaluations.[9] Multiple populations with various chronic illnesses have been evaluated to determine their HRQOL, but patients with PID status have remained largely unstudied.[9] Therefore the purpose of this study was to provide a description of HRQOL in patients with PI disease and to compare it with healthy samples.

MATERIALS AND METHODS

Fifty patients with previously diagnosed primary immunodeficiency diseases, who were referred to immunologic clinic, were invited to participate in this study.

One hundred children without a chronic ill-health condition were recruited from several schools in Tehran as control group. This case-control study was carried out between the years 2004 and 2005. Patient's HRQOL was evaluated using the Pediatric Quality Of Life questionnaire (PEDQOL).[14] The patients were asked about their physical, emotional, social (friends, family) well being, learning issues, own physical perception and some questions about their condition such as pain, fear, general health and how they view their life in general. They were asked to comment on how they felt the week before, since this particular period has been demonstrated to be optimal for the self-evaluation of emotional reactions and conditions. The answer choices to question have been categorized in a four step scale for choosing between never, sometimes, often and always. A higher score indicated better HRQOL. The total items of the questionnaire were divided into two parts: the first 25 questions focused on their physical activity and the second 25 questioned on their psychological and social abilities.[14] The children were categorized into 3 groups: 8-11 years (prepubertal period), 12-14 years (pubertal period) and 15-18 years (post pubertal). Patients with PID were compared with healthy controls using student T test and χ^2. All data analyses were conducted using the SPSS statistical software version 10.

RESULTS

Demographic information is shown in table 1.

Table 1. Demographic information about the study population

Characteristic	Value
Age; year	
Mean ± SD (range)	12.62±3.65 (8-18)
Sex; N. (%)	
Male	34(68%)
Female	16(32%)
Medical history of allergy; N. (%)	
Yes	29(58%)
No	21(42%)
Vaccination; N. (%)	
Complete	34(68%)
Incomplete	16(32%)
Duration of disease; year	
Mean ± SD (range)	10.08±5.76 (2- 18)
Diagnosed age; year	
Mean ± SD (range)	7.61±5.65(1- 18)

Table 2. Illness characteristics of patients with primary immunodeficiency

Diagnosis	Number(%) of Patients
Common variable immunodeficiency	27(54%)
X-linked agammaglobulinemia	12(24%)
Chronic granolumatos disease (CGD)	4(8%)
Hyper immunoglobulin M	3(6%)
Ataxia-telangiectasia	1(2%)
Congenital neutropenia	1(2%)
IgG deficiency	1(2%)
Nijmegen breakage syndrome	1(2%)

There was no difference between sex for quality of life (P= 0.1). In this study, most patients with PI disease had a diagnosis of common variable immunodeficiency (54%) or X-linked agammaglobulinemia (24%). Illness characteristics of the patients are summarized in table 2.

In the control group, psychological and physical scores were almost similar; nevertheless the control group could not obtain the maximum score. Table 3 presents the mean and standard deviation (SD) for physical and psychological scores in different age groups of PI patients in comparison with healthy controls. Overall in PI patients, not only physical and psychological scores were significantly lower than the control group (p< 0.001, p< 0.001 respectively), but their total scores also were significantly lower too (p<0.001). Figure 1 indicates negative relation between total PEDQOL scores with age in PI patients. The patients who were older were associated with worse physical (P= 0.04) and psychological scores (p<0.001). Long duration significantly correlated with low psychological score (r = -3.23, P= 0.03). The medical history of allergy was significantly higher in PI patients in comparison with the healthy control group (P< 0.001). In control group, we had 100% complete vaccination while only 68% of PI patients had completed their vaccination (p<0.001).

DISCUSSION

HRQOL is a subjective perception of the effect of health status on physical, psychological and social functioning and wellbeing.[15] Although survival rate and prognosis have greatly improved in some of PI patients, children with PID are still at risk for physical, psychological and social problems due to their chronic ill health condition.[1]

Advanced age was associated with worse HRQOL. The inverse relationship with advanced age may be a reflection of showing the greatest decline in the ability to perform physical tasks and to participate in social activity secondary to their complication. Some studies reported worse HRQOL in female patients,[9] but in this survey there was not any difference between both genders for quality of life in the patients. PI patients had not completed their vaccination and because of this, they were prone to more infectious diseases, thus, this susceptibility could affect their quality of life.

Table 3. Mean and SD in total and different age groups in PI patients and control subjects.

Age range	Physical activities Score	P-value	Psychological Score	P value	Total HRQOL Score	P-value
8-11 years		0.002		0.07		<0.001
Cases	63.68±2.99		68.14±3.54		131.82±5.12	
Control	68.24±2.1		69.39±1.81		137.63±2.71	
12-14 years		<0.001		0.006		<0.001
Cases	62.64±4.61		66.55±1.92		129.18±5.67	
Control	68.45±4.16		69.19±3.86		137.656.17	
15-18 years		<0.001		0.055		<0.001
Cases	61.59±3.78		65.59±3.61		127.18±5.89	
Control	67.97±2.87		68.03±4.37		136±5.49	
8-18 years (in total)		<0.001		<0.001		<0.001
Cases	62.74±5.02		61.92±3.42		129.66±5.78	
Control	68.22±3.07		68.91±3.45		137.13±4.9	

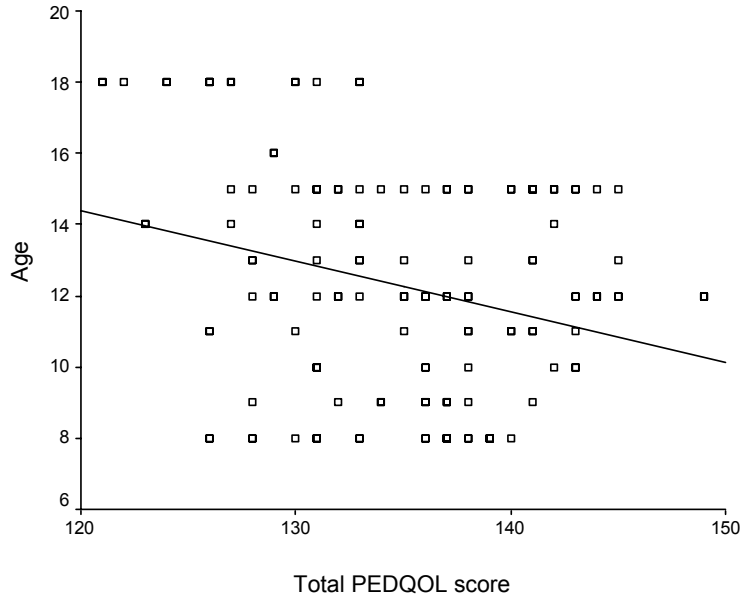

Figure 1. Negative relation between total PEDQOL score and age in PI patients.

Overall, patients with primary immunodeficiency in all different age groups had poorer perceptions of their health and greater limitation in physical and psychosocial activities than healthy control. Gradulf et al found that patients with hypogammaglobulinemia had a poorer HRQOL than the general population.[16]

Furthermore, due to their weakened immune defenses and the need to avoid unnecessary exposure to infectious agents, children with PID may limit their social interactions and their participation in physical and school–related activities,[9] and it could be one of the reasons of their lower psychological scores than physical scores. Due to the low prevalence of PI disease, these patients may feel that they have less social support or they are more misunderstood regarding their illness.

The relation between duration of disease and HRQOL may reflect psychotic effect of PID on PI patients, or may result the complication of the disease. But in some studies, they could not find any relation between duration of disease and HRQOL.[9] This finding indicates more attention to early diagnosis and treatment or prevention of PID and it implies that these patients may need psychiatric consultation for patients to better cope with their disease.

There were several limitations for the study. First limitation was the small sample size and this is due to the rarity of these diseases and short duration of the study.

Second, HRQOL was evaluated using the patient's report alone. Therefore, children's perceptions concerning the impact of a chronic health condition on their own psychological well-being may be discrepant with reality.

There are several potential directions for future study: Obtaining HRQOL information from multiple informants, including parents, teachers, and physicians may provide additional valuable knowledge. More studies are needed within larger samples of children with PI disease to examine specific health and treatment factors that affect children's HRQOL.

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