Child and caregiver psychosocial functioning in pediatric immunodeficiency disorders

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Background: Children with chronic illness have been found to be at an increased risk of behavioral and emotional difficulties. To date, children with pediatric immunodeficiency disorders (PIDDs) and their families have not been the focus of extensive published psychosocial research.

Objective: To determine if children with PIDDs and their caregivers have altered psychosocial function and whether the severity of the PIDD was associated with such difficulties.

Methods: Twenty children with PIDDs and 20 children with asthma were recruited for this study. Children and their caregivers completed various psychosocial questionnaire forms. Responses were compared with normative data for the appropriate measure and with other variables.

Results: Higher frequencies of children with PIDDs were found to have a number of elevated psychosocial concerns when contrasted with normative data, particularly from parent report. These concerns included depression, anxiety, somatization, social withdrawal, and social skills. The severity of the PIDDs was significantly associated with a number of behavioral adjustment issues, including receiving psychiatric diagnoses and special education services. Although children with PIDDs had significantly more psychiatric diagnoses than did asthmatic children, these groups did not differ significantly on questionnaire scores regarding child or caregiver psychosocial adjustment.

Conclusions: Children with PIDDs have significant behavioral problems. Children receiving intravenous immunoglobulin or immunomodulatory treatments were reported to have more problems than children not receiving them. This study highlights the need for further research in psychosocial functioning of children with PIDDs in an effort to develop interventions to promote their overall adjustment.

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INTRODUCTION

Children with chronic illness have been found to be at an increased risk of behavioral and emotional difficulties.[1–3] Much of this research has focused on common pediatric chronic medical conditions, such as asthma and cancer.[4–6] However, little research regarding behavioral difficulties for rare diseases, such as pediatric immunodeficiency disorders (PIDDs), has been performed.

Less than 1% of the population has a PIDD, and within this heterogeneous population more than 70 different disease entities exist, with varying severity and prognoses. Children with the most severe PIDDs may have numerous comorbid diagnoses, complex medical regimens, and uncertain life spans attributable to their immunodeficiency.[7] Specifically, all of these children are highly susceptible to infections[8–10] and have higher rates of lung disease and asthma,[11,12] cancer,[13,14] and gastrointestinal complications[10] than the general population. These additional diseases can contribute to the severity and complexity of illness that these children have. Furthermore, patients with severe immunodeficiency are often given monthly treatments of intravenous immunoglobulin (IVIG). These treatments can introduce more stress and complication to the daily functioning of these children and their families.

Most pediatric psychosocial studies include children with only one medical diagnosis, such as asthma or diabetes, and compare psychosocial functioning between illness groups and/or with a healthy comparison sample. For example, asthmatic children have been noted to have a higher incidence of anxiety disorders when contrasted with healthy control participants.[15] In addition, the diagnosis of clearly life-threatening illnesses, such as cancer[16] or diabetes,[17] has been noted to increase stress in children. Currently, it is unclear if children's adjustment difficulties in these chronically ill samples are resultant from the specific illness itself. Indeed, although Holden et al[6] found that asthmatic children were more adaptable and performed better in school than diabetic children, Hamlett et al[18] found that asthma was related to increases in child internalizing behavior problems. Consequently, the status of the literature is conflicted when it comes to determining whether psychosocial difficulties are disease specific or simply related to having a chronic illness in general. In fact, Siefert and colleagues[19] argued that having a child with any chronic illness, rather than an illness type, and the amount of perceived burden are the important factors related to adjustment. Consistent with this perspective, in adults with chronic

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illness, a strong linear association has been found to exist between the number of chronic diseases and depressive and anxiety symptoms.[20] This finding then suggests that perhaps children with multiple chronic conditions may have more adjustment issues as well.

In the case of children with PIDDs, their diagnosis goes hand in hand with having additional chronic health and potentially terminal conditions. It seems logical then that the compounded complicated medical regimens likely could add more perceived burden or stress to these families' daily lives than a single chronic disease. The purpose of this study was to determine if children with PIDDs and their families have significant psychosocial difficulties and whether the severity of the PIDD was associated with such difficulties. In addition, given the presence of multiple comorbid diseases with PIDD, we compared the prevalence of behavioral and emotional difficulties, as well as disease burden and morbidity factors, in our sample of children with PIDDs to that in a sample of children with asthma. It was hypothesized that children with PIDDs would have significantly greater psychosocial difficulties than would children with asthma and that disease severity would be associated with these difficulties within our sample of patients with PIDDs.

METHODS

Participants

All patients received medical care from a rural university-based pediatric clinic. Families who met the inclusion criteria were approached to participate during their child's routine medical visit. Written consent and assent were obtained from all caregivers and children, respectively, who agreed to participate. Caregivers completed the demographic form and parent-report questionnaires. Children 8 years and older completed a self-report questionnaire. Families completed the evaluation during the office visit. Families received a $10 gift certificate for caregiver completion of measures and an additional $5 gift certificate when the patient completed the child-report measure. Medical record reviews were conducted following the children's participation.

Inclusion and Exclusion Criteria

Participants were children ages 2 to 18 years (consistent with age norms for measures), with a diagnosis of a PIDD or asthma. All patients with PIDDs between the ages of 2 and 18 years were recruited for participation without regard to their specific immunodeficiency diagnosis and none declined. The PIDD severity was evaluated by whether the children received IVIG or immunomodulatory medications or not. Children with asthma were enrolled consecutively and matched for age and sex to children with PIDDs. Asthma disease severity was assigned according to the National Institutes of Health treatment guidelines with regard to medication use. Children younger than 2 years were excluded from study entry, because behavioral measures were age specific for children 2 years and older. The university institutional review board approved the study.

Measures

Demographic Information. A brief demographic form provided information about parental marital status, occupation, and education level, as well as family socioeconomic status and medical history and the ethnicity of the child.

Symptom Evaluation. The Symptom Assessment-45 Questionnaire[21] measures psychological distress in adults along 9 symptom dimensions or subscales (eg, depression, anxiety, obsessive-compulsive behavior). Caregivers completed this form based on their own behaviors. The Global Severity Index (GSI) combines information on the number of symptoms and intensity of distress across psychological constructs to provide a summary score of overall psychopathology. The GSI includes a statistical norm (much in the same way as a pediatric growth chart) that allows for sex-based standardized T-scores to be used in data analyses.

Parenting Stress Index–Short Form. The Parenting Stress Index–Short Form[22] is a 36-item short form of the Parenting Stress Index. The Parenting Stress Index–Short Form has 3 subscales (parental distress, parent-child dysfunctional interaction, difficult child), which are added to provide a total stress score. Higher scores reflect greater parental stress difficulties.

Behavioral Assessment System for Children. The Behavioral Assessment System for Children[23] (BASC) provides both parent-reported (BASC–Parent Rating Scales [PRS]) and self-reported (BASC–Self-Report of Personality [SRP]), objective evaluation of emotional and behavioral difficulties in children and adolescents ages 2 to 18 years. Age-appropriate BASC forms were used for child evaluation. The BASC yields scores on scales (eg, hyperactivity, conduct disorders, social skills, aggression, depression, withdrawal, social skills, leadership qualities, anxiety, self-esteem), which also combine to form composite scores (eg, externalizing, internalizing, adaptive behaviors for the BASC-PRS and school maladjustment, clinical maladjustment, personal adjustment for the BASC-SRP). Validity scales evaluate the respondent's pattern of responding to items. Scale and composite scores in the *at-risk* range indicate that behaviors may warrant clinical attention and evaluation, whereas *clinically significant* scores suggest that the reported behaviors represent a high level of maladjustment. For both child-report and caregiver-report measures, individual raw scores were compared with normative data for same-aged children, which resulted in standardized T-scores (mean ± SD, 50 ± 10). For the purposes of this study, *at risk* and *clinically significant* categories were combined to represent the rate of problematic behaviors or self-perceptions outside the normal range.

Statistical Analysis

Analyses between groups (children with PIDDs vs asthma; PIDDs vs nonsevere PIDDs) were performed with an independent samples t test for continuous data and a χ^2 analysis for categorical data. Because of the large number of analyses, point-biserial correlation and Pearson product-moment correlation coefficients were used to examine the strength of relations between BASC scale scores and other variables of interest.

RESULTS

Demographics

Twenty children with PIDDs (mean ± SD age, 10.6 ± 4.9 years) and a comparison group of 20 children with asthma (mean ± SD age, 9.2 ± 4.7 years), as well as their primary caregivers, participated in the study. Twenty-two children (55%) were male and 73 (93%) were white. Mothers' and fathers' median education level each was "some college," with a median annual family income in the \$25,000 to \$34,999 range. Asthma in children was diagnosed at a mean age of 4.0 years (SD, 3.6 years), whereas PIDDs in children were diagnosed with their immunodeficiency at a mean age of 5.6 years (SD, 4.1 years). Age when condition was first diagnosed was not significantly different between PIDD and asthma samples ($t_{37} = 1.3$, $P > .05$).

Disease Severity

The range of PIDD diagnoses was broad in this sample. Patients in this study had subclass deficiency with associated specific antibody deficiency (n = 5), IgA deficiency (n = 4), common variable immunodeficiency (n = 5), specific antibody deficiency (n = 3), chronic granulomatous disease (n = 1), pulmonary and gastrointestinal lymphangiectasia associated with hypogammaglobulinemia and lymphopenia (n = 1), and Kartagener syndrome (n = 1). Fifteen children with PIDDs also had asthma. Two children with PIDDs had severe asthma; 1 of these was oral steroid dependent. Nine children with PIDDs had moderate asthma, 3 had mild persistent asthma, and 1 had mild intermittent asthma. In the asthma control group, there were 3 patients with severe asthma (2 with previous prenatal intensive care unit admissions), 10 patients with moderate asthma, and 7 patients with mild persistent asthma. Children with asthma had comorbid diagnoses of gastrointestinal reflux disease, allergic rhinitis, atopic dermatitis, chronic sinusitis, migraines, and peanut allergy.

Within-Group Analysis of PIDD Sample

Disease Burden. Within the PIDD group, children were evaluated as having severe asthma (n = 11) if they were receiving IVIG infusions or other immunomodulatory medication. Children with more severe PIDD were taking significantly more routine medications (mean ± SD, 7.3 ± 2.7) than were children with less severe PIDD (mean ± SD, 4.1 ± 2.0) ($t_{18} = -2.7$, $P < .05$). The severe PIDD group also had significantly more physician visits in the previous year (mean ± SD, 14.3 ± 5.0) than did the less severe group (mean ± SD, 4.7 ± 3.7) ($t_{18} = -4.4$, $P < .001$). Lastly, the severe PIDD group had an average of 2.3 (SD, 2.8) hospitalizations during the previous year, whereas the less severe group had none, thereby yielding a significant group difference ($t_{18} = -2.1$, $P < .05$).

Psychosocial Outcomes. Within the PIDD group, greater disease severity was consistently associated significantly with psychosocial outcomes. First, all 6 children with a history of psychological or developmental delay diagnoses were classified in the severe PIDD group. Consequently, the group difference for this outcome variable was significant ($\chi^2_1 = 4.6$, $P < .05$). Furthermore, all children with PIDD who were receiving special education services (n = 5) were categorized as having severe disease as well, yielding a significance trend for severity group difference with respect to this variable ($\chi^2_1 = 3.6$, $P = .058$). However, the PIDD severity groups did not differ significantly in terms of having a history of grade repetition.

Psychosocial Questionnaire Data. The BASC-PRS measure revealed that caregivers of children with PIDDs noted several variables as being related significantly to their children's behavioral adjustment. Expressly, these caregivers noted that older children with PIDDs had lower social skills ($r = -0.63$, $P < .01$) and lower leadership skills ($r = 0.59$, $P < .05$) on the BASC-PRS. Caregivers for children with PIDDs who were receiving special education services also reported that their children had poorer social skills ($r = -0.57$, $P < .05$) and lower leadership skills ($r = -0.65$, $P < .01$). Caregivers of children with PIDDs who had repeated a grade during their schooling perceived their children to be more depressed ($r = 0.53$, $P < .05$) and to have greater attention problems ($r = 0.48$, $P < .05$). Finally, children with PIDDs missing an increased number of school days due to illness were noted to have increased somatization complaints ($r = 0.55$, $P < .05$), according to caregiver report.

Results with the BASC-SRP measure also demonstrated significant relations between morbidity variables and youth self-perceptions of their behavioral adjustment. According to self-report, older children with PIDDs reported greater general anxiety ($r = 0.64$, $P < .05$) compared with their younger counterparts. Children who had repeated at least one grade during their school history reported a greater sense of external locus of control ($r = 0.78$, $P < .01$), thus being more likely to attribute causes for their behavior to things outside their control. Children with PIDDs who had repeated a grade also reported an increased sense of inadequacy ($r = 0.73$, $P < .01$), dissatisfaction in their relationships with their parents ($r = -0.72$, $P < .01$), and little confidence in their own abilities (ie, self-reliance) ($r = -0.59$, $P < .05$). Children who were receiving special education services endorsed increased stress in their personal relationships ($r = 0.61$, $P < .05$), difficulty relating to others ($r = -0.61$, $P < .05$), and low self-esteem ($r = -0.59$, $P < .05$). More school absences due to illness was related to increased sensation seeking ($r = 0.71$, $P < .05$). Greater self-reported difficulties with atypical or highly individualized behavior ($r = 0.81$, $P < .01$), anxiety ($r = 0.60$, $P < .05$), depression ($r = 0.61$, $P < .05$), and low self-esteem ($r = -0.66$, $P < .01$) were all significantly associated with having an immediate family member with a primary immunodeficiency disorder.

PIDD vs Asthma Group Comparisons

Disease Burden. Several indices were evaluated to look at morbidity factors between children with PIDDs and asthma in this study. Children with PIDDs had significantly more comorbid diagnoses (mean ± SD, 3.7 ± 2.3) than did children with asthma (mean ± SD, 1.0 ± 0.9) ($t_{38} = 4.8$, $P < .001$) per medical record review. Examples of concurrent diagnoses in the PIDD population were hepatitis, thrush, atopic dermatitis, autoimmune neutropenia and thrombocytopenia, type 1 diabetes, recurrent herpes simplex, warts, hypothyroidism, growth hormone deficiency, *Nocardia* adenitis, sarcoma, epilepsy, gastroesophageal reflux disease, chronic sinusitis, chronic cutaneous *Staphylococcus* abscesses, and malabsorption syndrome. Other variables that reflect disease burden included number of medications taken, total number of physician visits during previous year, and number of inpatient hospitalizations during the past year, all of which were evaluated via medical record review. A significant difference was observed in the number of medications prescribed ($t_{38} = 2.5$, $P < .05$), with children with PIDD taking significantly more routine medications (mean ± SD, 6.2 ± 2.9) than children with asthma (mean ± SD, 4.4 ± 1.6). Children with PIDDs also had more physician visits in the last year, with children with PIDDs having an average of 11.0 visits (SD, 6.5 visits) in contrast to children with asthma having 3.4 visits (SD, 2.0 visits) ($t_{38} = 5.0$, $P < .001$). Finally, children with PIDDs had significantly more hospitalizations in the previous year (mean ± SD, 1.5 ± 2.5) compared with children with asthma (mean ± SD, 0.2 ± 0.5) ($t_{38} = 2.3$, $P < .05$).

Psychosocial Outcomes. In the PIDD group, 6 children (30%) also had a diagnosed psychosocial disorder, according to caregiver report and medical record review. Three children with PIDDs had received a diagnosis of depression or anxiety disorder and were taking psychotropic medication. Two children with PIDDs had a diagnosis of developmental delay. One child with PIDD had been diagnosed as having a conduct disorder. In contrast, only 1 child with asthma had been diagnosed as having an anxiety disorder, but no medication was prescribed. Thus, the PIDD and asthma groups differed significantly with respect to the number of children with psychosocial diagnoses ($\chi^2_1 = 4.3$, $P < .05$). Furthermore, 25% of children with PIDDs were receiving special education services, whereas 20% had repeated a grade in school, although these rates were not significantly different ($P > .05$) than those found in the children with asthma (6% in special education and 11% having repeated a grade).

Psychosocial Questionnaire Data

Caregiver Report of Child Adjustment. In a normalized distribution of standardized BASC T-scores, approximately 15.7% of children in the normative sample would yield elevated scores (ie, >1 SD above the mean; *at-risk* and *clinically significant* categories). It is noteworthy then to realize that the percentage of our PIDD sample surpassed this estimate in all but 1 (conduct problems) of the caregiver-report BASC scales. Indeed, the BASC-PRS estimates for elevated scale scores ranged from 15.9% (atypicality, aggression) to 21.2% (anxiety, withdrawal) to 26.4% (depression, social skills) to a high of 52.9% for the somatization scale in our PIDD sample. Results were roughly comparable for caregiver-report BASC scores with the asthma sample as well. For example, the percentage of caregivers rating their children as problematic exceeded normative estimates for all but 4 scales (aggression, anxiety, atypicality, and conduct problems). Rather, estimates ranged from 23.6% (hyperactivity, depression, withdrawal, and social skills) to approximately 34% (attention problems and adaptability) to 70.6% (somatization). Indeed, independent samples *t* tests yielded nonsignificant ($P > .05$) diagnostic group differences for the BASC-PRS externalizing behaviors, internalizing behaviors, and adaptive skills mean composite scores.

Child Self-report of Adjustment. In contrast to caregiver-report, BASC-SRP results yielded a smaller percentage of children reporting problematic scores. In fact, for the PIDD sample, only one scale (somatization) surpassed the expected rate of 15.7% at a frequency of 20.0%. In contrast, the sample of children with asthma exceeded the normal rate of scores for 6 of the scales. Specifically, the percentage of children reporting problematic concerns ranged from 16.6% (attitude toward school, atypicality, and self-esteem) to 24.9% (social stress) to a high of 37.5% (sensation seeking and somatization). Like caregiver-report, independent sample *t* test results yielded no significant ($P > .05$) between-group findings for the mean school maladjustment, clinical maladjustment, and personal adjustment composite scores.

Caregiver Self-report of Adjustment. An independent sample *t* test was conducted to determine differences between caregivers of children with PIDD and those of children with asthma with respect to the composite (GSI) score of caregiver adjustment on the Symptom Assessment-45 Questionnaire. Results were not statistically significant ($P > .05$; mean ± SD, 52.8 ± 11.0 for PIDDs; mean ± SD, 52.2 ± 9.4 for asthma). It is noteworthy that 20% and 10% of caregivers from the PIDD and asthma samples, respectively, received GSI scores that exceeded the clinical significance cutoff.

Similarly, an independent sample *t* test using the Parenting Stress Index–Short Form total stress score as the dependent variable revealed no significant diagnostic group difference for self-reported parental stress levels ($P > .05$; mean ± SD, 78.6 ± 23.5 for PIDDs; mean ± SD, 75.1 ± 25.8 for asthma).

DISCUSSION

We found that children with PIDDs in general had increased rates of psychological difficulties, with the exception of conduct problems, when compared with a normative sample. Problem areas with the highest frequency of participants being rated by caregivers as having elevated concerns were depression, anxiety, somatization, social withdrawal, and difficulties with social skills. In contrast, a higher prevalence of youth with PIDDs rated themselves as having elevated somatization levels when contrasted with average healthy children in the standardization sample for this questionnaire; no other self-report scales had greater frequency of elevations. Of interest, 20% of parents of youth with PIDDs also endorsed significant elevations of psychological distress in themselves. Perhaps such parental distress may account for some of the discrepancy between caregiver and child report on child behavior; specifically, distressed caregivers may view their child's behavioral functioning more negatively.

Regardless of discrepant reports, children with PIDDs seem to have significant psychosocial issues associated with their illness, particularly from the caregivers' perspective, when compared with normative data. These results are consistent with a meta-analysis of pediatric physical disorders that found that psychological adjustment risk was greatest in studies making comparisons to norms rather than study controls.[28]

We also chose to determine if the severity of the PIDD was relatable to behavioral adjustment issues. We compared children with PIDDs with more severe immunodeficiency diseases (receiving IVIG or other immunomodulatory drugs) to those children with less severe illness such as IgA deficiency. All youth with PIDDs who were receiving special education services fell within the severe category of this group, comprising nearly half of this severity subgroup. In addition, only the children with IVIG therapy received clinical diagnoses and therapy for psychological disorders, such as depression, anxiety, or conduct disorder, and developmental disabilities. These results consistently suggest that children with greater PIDD severity tend to have poorer psychosocial outcomes. Thus, clinicians should recognize that disease severity seems to be a good indicator for more closely monitoring psychosocial issues in patients with PIDDs. Furthermore, our findings support the suggestion that more research needs to attend to individual disorders and not simply interdisease comparisons.[28]

School variables (ie, repeating a grade, receiving special education) also were found to be related significantly to child adjustment in our sample of youth with PIDDs. For example, children with PIDDs who had repeated a grade reported greater problems with sense of inadequacy, had lower confidence in their own abilities, and were dissatisfied with the relationship with their caregivers. Similarly, children who were receiving special education services also reported higher levels of social stress and interpersonal difficulties, as well as lower levels of self-esteem. Taken together, it seems that these children may be experiencing symptoms of helplessness. Indeed, caregiver report resulted in significant relations between these school variables and ratings of lower social and leadership skills and higher depression symptoms and attention problems. Because all children who were receiving special education services were categorized as having severe PIDDs, caregivers may have taken a more authoritative approach to parenting as a protective mechanism. This style, in turn, may have resulted in their children feeling more dissatisfied with their relationship with their caregivers, as well as having a higher sense of inadequacy and difficulties with self-reliance.

Children with PIDDs and their families have not been the focus of extensive published psychosocial research, like other chronic illnesses. Rather, only one published study noted that children with PIDDs had significantly poorer health-related quality of life (eg, physical function, self-esteem, family activities) than healthy children but comparable to that of other chronically ill children.[24] Still, specific psychological functioning, such as diagnosed psychiatric disease, and broader evaluation of behavioral functioning were not specifically addressed in this study. Furthermore, this study used only parent reports and included only the more severe cases of PIDDs (children receiving IVIG infusions). We believe that the multiple illnesses that children with PIDDs (infection, autoimmune disorders) have can be significant stressors with which both children and families must cope. Children miss school frequently for medical appointments and may have limited participation in physical-, social-, and school-related activities to reduce exposure to infectious agents, given their weakened immune systems. These issues likely cause significant disruption in daily functioning. Other factors that interfere with the quality of life of children with PIDD include the large number of medications that they take for both their immunodeficiency and associated comorbid diseases.

In contrast to the limited psychosocial literature with PIDDs, extensive research has been conducted with various pediatric chronic conditions, such as asthma and cancer.[15–18,25–27] Conclusions from such research are mixed, such that some researchers argue that psychosocial difficulties in children stem from specific illness types, whereas other researchers purport that such difficulties result from having a chronic health condition, regardless of its type. Siefert and colleagues,[19] in particular, noted that they believe the pivotal factor in psychosocial adjustment is the family's level of perceived burden. Because of the more complicated medical regimen, as evidenced by significant group differences in terms of comorbid conditions, medications, physician visits, and hospitalizations, it was hypothesized that children with PIDDs and their families would report more adjustment difficulties than would families of children with asthma. We found that there was a significant difference in numbers of diagnosed psychiatric illnesses in the children with PIDDs compared with children with asthma. Contrary to hypotheses, however, we did not find significant group differences for general behavioral difficulties (ie, composite scores) in children, as well as parenting distress and psychological difficulties in caregivers, with questionnaire data. Yet, caregivers of youth with PIDDs, in general, tended to endorse more frequent rates of elevated behavioral concerns in their children across a broader range of scales or specific problem areas. In contrast, when examining the overall rate of elevated scale scores for self-report, youth with PIDDs seemed to report fewer behavioral difficulties than did youth with asthma. Despite the fact that it has been documented many times that youth and caregivers differ in their reports of child behavior,[29] these discrepant reports deserve further investigation with youth with PIDDs in future research.

Nonetheless, considering the small size of our sample, a sizable proportion of children were experiencing difficulties, which suggests a need for clinical intervention. Taking caregiver and child report together, it seems critical to recognize that significant behavioral and emotional concerns exist in this population and that such concerns need to be addressed (eg, referral for mental health services) to improve the daily functioning of these children. In particular, attention should be focused on those children receiving IVIG, those frequently missing school and having numerous medical appointments, and those having problems in school.

In summary, in our population of children with immunodeficiencies, we found significant numbers of diagnosed psychosocial disorders. In addition, these children had marked levels of depression, anxiety, and somatization. These children also had decreased social and leadership skills, which has implications for their long-term adult psychosocial functioning. Addressing these psychosocial issues in patients with PIDDs has the potential to greatly improve their quality of life.

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