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Quality of life in children with Prader Willi Syndrome: Parent and child reports

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ABSTRACT

Purpose: The purpose of this study was to evaluate the use of the Peds QL4.0 instrument to assess quality of life (QL) in children with Prader Willi Syndrome (PWS). This study also sought to compare differences in parent and child report as well as between children with PWS and without PWS.
Methods: Parents and children with PWS ($N = 44$) completed the PedsQL 4.0 instrument. A sub-sample of children completed the Peds QL 4.0 a second time to assess test-retest reliability. A comparison sample of children who were obese but without PWS ($N = 66$) also completed the PedsQL 4.0.
Results: PedsQL 4.0 showed acceptable internal consistency for the child report (αs >0.72) and was acceptable for 4 out of the 6 scales for the parent report (αs >0.66). Test-retest reliability coefficients showed support for the reliability of the instrument (ICCs > 0.64). Parents perceived lower QL than children with PWS. Children with PWS also showed lower QL than children without PWS.
Conclusions: This study provides support for the use of the PedsQL 4.0 instrument in children with PWS. As observed in other populations, parents perceive a lower QL for their children with PWS than the children themselves.

What this paper adds?

Prader Willi Syndrome (PWS) is a genetic disorder with both physical and psychological challenges. Previous research examining quality of life of individuals with PWS has focused on individuals over the age of 14 or assessed quality of life using parent report only. This paper extends previous research examining quality of life in individuals with PWS in several ways. First, it provides support for the use of the PEDS 4.0 instrument for both parent and child report of quality of life. Further, it makes comparisons between how parents of children with PWS and the children with PWS differ their perceptions of quality of life. Parents appeared to perceive the quality of life of their children lower than the child themselves. With one of the physical challenges of this disorder being obesity, a comparison of quality of life perceptions with children who are obese without PWS provides insight into the role of obesity in quality of life perceptions. Children with PWS and their parents report lower quality of life for the child than children who were obese without PWS and their parents.

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1. Introduction

Prader Willi Syndrome (PWS) is a complex genetic disorder that is characterized by physical and psychological challenges such as obesity, low muscle tone, coordination difficulties, behavioral issues and cognitive disabilities (Holm et al., 1993). This genetic disorder occurs with an alteration or lack of expression to the paternal chromosome 15 in the locus of 13–15q (Cassidy & Driscoll, 2009). Prader Willi Syndrome is a relatively rare disorder with prevalence estimates between 1 per 15,000 and 1 per 30,000 (Cassidy & Driscoll, 2009). A comprehensive approach to the treatment of PWS is often recommended including parent education, growth hormone replacement therapy, physical activity and diet (Cassidy & Driscoll, 2009).

Beyond the treatment of the disorder, there is a growing awareness of the need for improving the quality of life of individuals with PWS (Benarroch, Hirsch, Genstil, Landau, & Gross-Tsur, 2007). Previous research has demonstrated that physical and mental aspects of quality of life are lower in individuals with PWS than in a healthy population (Caliandro et al., 2007). Based on assessments from the short form-36 scale, individuals with PWS over the age of 14 showed significantly lower levels of physical functioning, role-physical, bodily pain, general health, social functioning, role emotional and mental health than the norms for a healthy population. The one exception was on the vitality subscale, of which individuals with PWS were about the same as the healthy population. In children under 14 years of age, quality of life was assessed by having parents complete the Child Health Questionnaire – Parent form – 50 (CHQ-PF50) (Caliandro et al., 2007). For those younger than 14 years of age, parents reported significantly lower levels on most domains than the healthy population including the domains of global health, physical functioning, mental health, psychosocial, and physical. These findings suggest that individuals with PWS report lower levels of quality of life than healthy norms.

Given that individuals with PWS are also typically obese, one might wonder how quality of life of individuals with PWS may compare to individuals who are obese but do not have PWS. In a study by Schwimmer, Burwinkle, and Varni (2003), children and adolescents who were obese reported significantly lower levels of health-related quality of life than healthy norms. In the same sample, parents of children and adolescents also reported significantly lower values for their children than the norms for a healthy population and these parent reports were also significantly lower than their children's reports (Schwimmer et al., 2003). These differences were across the physical health, psychosocial health, emotional functioning, social functioning and school functioning subscales. In fact, Schwimmer and colleagues reported that the rates of impaired levels of quality of life for those children who were obese were similar to those diagnosed with having cancer. This finding was replicated in another study that examined quality of life in children with a variety of chronic diseases (i.e., asthma, diabetes, cardiac conditions and obesity) compared to a sample of healthy children (Varni, Limbers, & Burwinkle, 2007a). Overall, quality of life as reported by children was lower in children who were obese when compared with healthy weight children and those with diabetes. Obese children also reported lower physical health, psychosocial health, and emotional health than healthy children but similar levels of school functioning (Varni et al., 2007a). Likewise, parents of children who were obese reported a lower quality of life than parents of children with a healthy weight with the exception of school functioning.

The difference between parent (a proxy report) and child report of quality of life is not surprising given that reviews have highlighted variability between parent and child reports (Eiser & Varni, 2013). For example, the Pediatric Quality of Life Inventory (PedsQL 4.0) has been validated for both child report (Varni, Seid, & Kurtin, 2001) and parent report (Varni, Limbers, & Burwinkle, 2007b). While knowing which report is more accurate may be important, it appears that the different perspectives on how the child feels and functions are also important (Eiser & Varni, 2013). Eiser and Varni (2013) suggested that healthy parents typically tend to rate their children's quality of life better than their child's rating; however the opposite is true for children with chronic health conditions where parents rate their child's quality of life as worse than the child reports (Eiser and Varni, 2013). As well, there seems to be more agreement between parent and child reports for the physical domains as opposed to more emotional domains (Eiser & Varni, 2013).

1.1. Purpose

This study has several purposes. The first purpose was to examine the reliability of the PedsQL 4.0 instrument as an assessment of quality of life in children with PWS. The second purpose was to examine differences between self-report and parent report of the PedsQL 4.0 instrument in children with PWS. The third purpose was to examine differences in the health-related quality of life of children with PWS and children who are obese but do not have PWS using both parent and child report.

2. Methods

2.1. Participants

Participants for this study were families enrolled in a physical activity intervention and the data obtained at baseline was used in the present study (Rubin, Wilson, Wiersma, Weiss, & Rose, 2014). Children ($N = 44$) with PWS along with one of their parents ($N = 44$) were included in this study. The most common type of genetic alteration in PWS was deletion ($n = 19$, 43.2%) followed by uniparental disomy ($n = 8$, 18.2%) and imprinting defect ($n = 4$, 9.1%). There were also several children that with unknown type but confirmed diagnosis by DNA methylation ($n = 13$, 29.5%). Children with PWS had an average age of 11.0