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Aberrant, autistic, and food-related behaviors in adults with Prader-Willi syndrome. The comparison between young adults and adults

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ABSTRACT

This study aims to explore the differences of age as well as genotype in regards to the severity of behavioral symptoms in Prader-Willi syndrome (PWS), with emphasis on the comparison between young adults and adults. The Food Related Problem Questionnaire (FRPQ), the Aberrant Behavior Checklist Japanese Version (ABC-J), and the Pervasive Developmental Disorders Autism Society Japan Rating Scale (PARS) were administered to 46 PWS patients, including 33 young adults (ages 18–28) and 13 adults (ages 30–45). To examine the differences between young adults and adults, Mann-Whitney *U* tests were conducted. Statistically significant differences were found in ABC-J ($p = .027$) and PARS ($p = .046$), with higher scores in young adults than adults. Such differences between the two age groups were still true for the subgroups having a paternal chromosome 15q deletion (DEL) for ABC-J ($p = .050$) and part of PARS (“Problematic behavior”; $p = .007$). By contrast, there was no significant differences between young adults and adults regarding FRPQ ($p = .65$). These results suggest that aberrant behaviors decline from around the ages of thirty, in PWS patients in general and in DEL subgroups in particular, while food-related behaviors give no indication of diminishing in spite of developmental growth.

1. Introduction

Prader-Willi syndrome (PWS) is a neurodevelopmental disorder, characterized by neonatal hypotonia, hypogonadism, hyperphagia, progressive obesity, and mild to moderate mental retardation (Prader, Labhart, & Willi, 1956; Crino et al., 2003; Cassidy & Driscoll, 2009). As a contiguous gene syndrome, PWS is caused by a loss of expression of the paternally derived genes in the q11-13

Abbreviations: ABC-J, Aberrant behavior checklist Japanese version; ASD, autism spectrum disorders; BMI, body mass index; FRPQ, food-related problem questionnaire; IQ, intelligence quotient; mUPD, maternal uniparental disomy 15; DEL, paternal deletion of 15q11-13; PDDs, pervasive developmental disorders; PARS, pervasive developmental disorders autism society japan rating scale; PWS, prader-Willi syndrome

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region of chromosome 15. A paternal deletion (DEL) of 15q11-13 is found in 70% of patients, and maternal uniparental disomy 15 (mUPD; when both copies of chromosome 15 are maternally inherited) is found in 25% (Buiting et al., 1995; Cassidy & Driscoll, 2009; Nicholls, Knoll, Butler, Karam, & Lalande, 1989; Ledbetter et al., 1981). Its prevalence is estimated at about 1 in 25,000 births (Diene et al., 2010; Vogels et al., 2004; Whittington et al., 2001). The syndrome is often associated with minor dysmorphic features: short stature, small hands and feet, hypopigmentation, and characteristic facial features, such as a narrow forehead, almond-shaped eyes and a triangular mouth.

Compared with other types of intellectual disabilities, individuals with PWS tend to show a wide variety of maladaptive behaviors. Those include hyperphagia (Holland et al., 1993), temper tantrums (Tunnicliffe, Woodcock, Bull, Oliver, & Penhallow, 2014), obsessive-compulsive behaviors (Descheemaeker et al., 2002; Dykens, Leckman, & Cassidy, 1996), repetitive and ritualistic behaviors (Greaves et al., 2006), self-injurious behaviors (Arron, Oliver, Moss, Berg, & Burbidge, 2011; Klabunde et al., 2015), autistic behaviors (Descheemaeker et al., 2006; Dykens, Lee, & Roof, 2011), and hyperactive/impulsive behaviors (Wigren & Hansen, 2005). Behavioral characteristics of this syndrome have been relatively well studied, most of which examined phenotypical differences between DEL and mUPD. It has been reported that the mUPD subtype has a higher risk for autistic-like symptomatology (Dimitropoulos & Schultz, 2007; Milner et al., 2005; Veltman et al., 2004).

These findings mainly based on Caucasian patients were consistent with a recent study about Japanese PWS patients, suggesting ethnicity-free phenotypical differences between the two main genotypes in PWS (Ogata et al., 2014). Aside from autistic behavior and psychosis (Vogels, Matthijs, Legius, Devriendt, & Fryns, 2003), the mUPD subtype may have a lower risk for other maladaptive behaviors than the DEL subtype, in terms of skin-picking (Dykens, Cassidy, & King, 1999; Symons, Butler, Sanders, Feurer, & Thompson, 1999), food-related problems (Dykens, Maxwell, Pantino, Kossler, & Roof, 2007) and obsessive-compulsive behaviors (Dykens & Roof, 2008).

To date, however, there is a paucity of data in regards to the developmental trajectory of problem behaviors. The majority of studies so far conducted have dealt mainly with childhood, due perhaps to the fact that between the ages of 2 and 3 years they start to present hyperphagia and associated food-related behaviors. Little is known how these behaviors progress over the course of development after chronological adolescence.

According to researchers (Dimitropoulos & Schultz, 2007; Dykens, 2004; Jauregi, Laurier, Copet, Tauber, & Thuilleaux, 2013; Sinnema, Boer et al., 2011; Sinnema, Einfeld et al., 2011), the relation between age and problem behaviors is a non-linear one, featured by gradual increase as children get older, young adults manifesting the highest degree of aggravation, and older adults with the decline of the behaviors. At the same time, a few behavioral problems are presumed to remain unchanged throughout patients' lives.

In addition to differences in behavioral profiles, it is likely that such a behavioral transition from childhood through adolescence to adulthood would have been influenced by phenotypical differences between the two main genotypes. Ogata et al. (2014) reported that there is a growing tendency of the autistic-like and impulsive behavioral problems, which are more severe in mUPD than in DEL that can manifest themselves later in adolescence. In adults with PWS much less is known about the effect of age and genotype on behavioral difficulties in adults with PWS.

This study aims to explore the effects of age as well as genotype have on the behavioral aspects of PWS, with special emphasis on the difference between young adults (ages 18–28) and adults (ages 30–45). The age of young adult is an important transitional epoch for two reasons. First, during this period young adults with PWS leave educational institutions and enroll in adult service systems which are more vocationally oriented. Second, for their medical care, pediatricians are, partly or entirely, replaced by physicians and psychiatrists. Such drastic changes of environmental factors are likely to have a negative impact on behavior and emotion of patients with PWS. This is because most of PWS patients are accompanied with intellectual disability, which can lead to low adaptability.

To highlight behavioral changes related to age, this study attempts to bring out the contrast between young adults and adults. Indeed, Dykens (2004) showed in “older adults” maladaptive and compulsive symptoms diminished significantly. However, researchers have not yet rigorously examined adults with PWS regarding a wide variety of behavioral symptoms.

2. Methods

2.1. Participants

46 Japanese participants with PWS were recruited from a single location. The Department of Pediatrics, Dokkyo Medical University Koshigaya Hospital was used for this purpose. All patients were diagnosed with PWS using fluorescence in situ hybridization or the methylation test. The participants consisted of 33 young adults (ages 18–28) and 13 adults (ages 30–45), including 23 young adults and 11 adults confirmed as having a DEL involving 15q11-13, and 10 young adults and 2 adults confirmed as having mUPD of chromosome 15 (Table 1).

2.2. The assessment of behavior

An extended battery of behavioral assessment was employed with regards to aberrant, autistic-like, and food-related behaviors, and intelligence. In all cases, the psychologist (H.O.) involved in collecting data was blind to the genetic status of each patient. For each participant, HO had 3–8 sessions in order to collect behavioral data. While behavioral instruments used in this study were originally constructed as self-administered or informant-based scale, some parts of questionnaire instructions are difficult to understand for informants, most of whom are nonspecialists with behavioral sciences. Considering this, all behavioral measures were

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