This study aims to compare maternal uniparental disomy 15 (mUPD) and a paternal deletion of 15q11-13 (DEL) of Prader–Willi syndrome (PWS) in regard to autism spectrum disorders (ASD). Forty-five Japanese individuals with PWS were recruited from a single recruitment center. The participants consisted of 22 children (aged from 6 to 12) and 23 adolescents (aged from 13 to 19). Six children and seven adolescents were confirmed as having mUPD. Sixteen children and 16 adolescents were confirmed as having DEL. Under blindness to the participants’ genotypes, a single psychologist carried out behavioral and psychological assessments, including the Wechsler Intelligence Scales, Pervasive Developmental Disorders Autism Society Japan Rating Scale (PARS), and ADHD-Rating Scale-IV (ADHD-RS-IV). Two comparisons were made: one between mUPD and DEL children and another between mUPD and DEL adolescents. In children, no significant differences were found between mUPD and DEL participants in terms of autistic (PARS childhood, $P = 0.657$) and impulsive behaviors (ADHD-RS-IV hyperactive/impulsive, $P = 0.447$).
**INTRODUCTION**

Prader–Willi syndrome (PWS) is a neurodevelopmental disorder, associated with neonatal hypotonia, hypogonadism, hyperphagia, progressive obesity, and mild to moderate intellectual disability. The physical manifestation of PWS includes short stature, small hands and feet, hypopigmentation, and craniofacial anomalies. This syndrome is a genetic disorder caused by a loss of expression of paternally derived genes on chromosome 15q11-13. The causes of this disruption include maternal uniparental disomy 15 (mUPD; when both copies of chromosome 15 are maternally inherited) and a paternal deletion (DEL) of 15q11-13. PWS is considered to occur, regardless of gender and race, with an estimated prevalence of one in 10,000 and one in 15,000 births [Cassidy, 1997].

The behavioral manifestations of this syndrome include hyperphagia, temper tantrums, obsessive–compulsive behaviors [Descheemaeker et al., 2002], repetitive and ritualistic behavior [Greaves et al., 2006], self-injurious behavior [Arron et al., 2011], and autistic behaviors [Descheemaeker et al., 2006; Dykens et al., 2011]. According to ample evidence, maternal duplications of 15q11-13 have an association with autistic spectrum disorders (ASD), but paternal duplications of 15q11-13 do not, suggesting the existence of maternally active gene(s) in chromosome 15q11-13 [Bolton et al., 2001; Veltman et al., 2005; Dimitropoulos and Schultz, 2007; Hogart et al., 2010]. So far, however, there has been insufficient research into the comparison between the two genotypes of PWS in relation to ASD.

This study aims to compare mUPD and DEL forms of PWS in regard to ASD. It has three advantages over previous researches. Firstly, all subjects with PWS were recruited from a single institution and were assessed neuropsychologically by a single clinical psychologist (H.O.). Hence, the psychometrical data of this study can avoid the risk of inter-rater variability caused by participants being assessed by multiple researchers. Secondly, this study takes into account the behavioral difference of children and adolescents. Prior studies have not considered behavioral changes across time in relation to physical development. Thirdly, this is the first neuropsychological study using Japanese PWS patients. To our knowledge, studies so far conducted have issued mainly from Western countries [Bolton et al., 2001; Milner et al., 2005; Descheemaeker et al., 2006].

**MATERIALS AND METHODS**

**Participants and Methods**

Participants included were 45 Japanese individuals with PWS 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 (FISH) or the methylation test. In terms of the patients who showed no deletion by FISH although consistent with PWS by methylation test, we performed a microsatellite analysis. These analyses revealed no biparental inheritance in all (only maternal inheritance), indicating that all patients were mUPD and with no imprinting defect. The participants consisted of 22 children (aged from 6 to 12) and 23 adolescents (aged from 13 to 20). Six children and seven adolescents were confirmed as having mUPD of chromosome 15. Sixteen children and 16 adolescents were confirmed as having a DEL involving 15q11-13 (Table I).

**The Assessment of Intelligence and Behavior**

An extended battery of neuropsychological assessment was employed. In all cases, the psychologist involved in collecting data was blind to the genetic status of each patient.

**MEASURES**

**Intellectual Ability**

To measure intellectual ability, a Japanese version of the Wechsler Intelligence Scale [Wechsler, 1991, 1997; Japanese WISC-III Publication Committee, 1998; Japanese WAIS-III Publication Committee, 2006] was administered. A full-scale IQ score, verbal and performance IQ scores, verbal comprehension, perceptual

**TABLE I. Patient Characteristics**

|                | Total Children |
|----------------|----------------|
| Number         | 22             |
| Male/female    | 14/8           |
| Mean age       | 8.95           |
| Age range      | 6–12           |

|                | Total Adolescents |
|----------------|-------------------|
| Number         | 23               |
| Male/female    | 15/8              |
| Mean age       | 15.83             |
| Age range      | 13–19             |

| DEL Children | Adolescents |
|--------------|-------------|
| Number       | 16          |
| Male/female  | 10/6        |
| Mean age     | 9.19        |
| Age range    | 6–12        |

| mUPD Children | Adolescents |
|---------------|-------------|
| Number        | 6           |
| Male/female   | 4/2         |
| Mean age      | 8.33        |
| Age range     | 7–11        |

|               |               |
|---------------|---------------|
|               |               |
organization, working memory, and processing speed were calculated for all individuals who completed the Scale.

The same clinical psychologist (H.O.) applied the tests in similar conditions of a calm environment and a comfortable atmosphere. When participants gave signs of fatigue or somnolence, the session was stopped for a break or adjourned until another day. The participants completed all the subtests in two or three sessions. Few participants showed a negative attitude towards the tests.

### Autistic Symptomatology

Autistic symptomatology was assessed using the Pervasive Developmental Disorders Autism Society Japan Rating Scale (PARS) [Adachi et al., 2006; Kamio et al., 2006]. This scale is a behavior checklist, developed as a screening questionnaire to determine Pervasive Developmental Disorders (PDDs). When assessing children using the PARS for these purposes, 33 items for children are applied for the evaluation of current autistic states. The PARS for adolescents is made up of five clinical subscores consisting of interpersonal skills (4 items), communication (7 items), obsessive-compulsive behaviors (3 items), problematic behaviors (9 items), and hypersensitivity (4 items). Likewise, when assessing adolescents and adults, 33 items for adolescents, partially shared by those for children, are applied for the evaluation of current autistic states. The PARS for adolescents is made up of five clinical subscores consisting of interpersonal skills (6 items), communication (7 items), obsession (6 items), problematic behaviors (11 items), and hypersensitivity (3 items). The PARS is applied widely to individuals with ASD and allows for the evaluation of the severity of a wide range of ASD symptoms [Yamada et al., 2007; Ito et al., 2012]. The reliability and the validity of the PARS have already been established [Adachi et al., 2006; Kamio et al., 2006].

### Inattention and Hyperactivity/Impulsivity

Japanese ADHD-RS-IV [Ichikawa and Tanaka, 2008] was administered to all participants. This scale is a Japanese version of the ADHD-RS-IV [DuPaul et al., 1998] that obtains parent ratings regarding the frequency of each ADHD symptom based on DSM-IV criteria. The scale consists of two subscales: inattention (nine items) and hyperactivity/impulsivity (nine items). Parents are asked to state the degree to which they best describe the child’s behavior over the previous 6 months. All items are scored on a 4-point Likert scale from 0 (“Rarely or Never”), to 3 (“Always or very often”), with higher scores reflecting higher degree of inattention and hyperactivity/impulsivity. The reliability and the validity of the Japanese ADHD-RS-IV have already been established [Ichikawa and Tanaka, 2008].

### Statistical Analysis

By means of a numerical coding system, all data were guarded under strict confidentiality and anonymity. The data were analyzed by SPSS 20.0J for Windows. Mann–Whitney U-tests were conducted to make two comparisons: one between mUPD and DEL children and another between mUPD and DEL adolescents. Wilcoxon signed-rank test for paired data were used to explore the differences between VIQ and PIQ for both groups of mUPD children and DEL children and for both groups of mUPD adolescents and DEL adolescents.

### Table II: Distribution of FIQ, VIQ, PIQ, VC, PO, WM, and PS Scores in the Groups and Comparison of the Two Genotype Groups

|                  | VIQ (Q1;Q3) | VIQ (Q1;Q3) | PIQ (Q1;Q3) | PIQ (Q1;Q3) | FIQ (Q1;Q3) | FIQ (Q1;Q3) | VC (Q1;Q3) | VC (Q1;Q3) | PO (Q1;Q3) | PO (Q1;Q3) | WM (Q1;Q3) | WM (Q1;Q3) | PS (Q1;Q3) | PS (Q1;Q3) | P-value |
|------------------|-------------|-------------|-------------|-------------|-------------|-------------|------------|------------|-------------|-------------|------------|-------------|------------|-------------|          |
| **Children**     | 47.00; 65.50| 52.25; 59.75| 45.00; 50.00| 43.25; 55.50| 54.25; 64.00| 54.25; 64.00| 50.00; 62.75| 50.00; 62.75| 50.00; 58.75| 50.00; 58.75| 50.00; 62.75| 50.00; 62.75| 50.00; 58.75| 50.00; 58.75|          |
| **Adolescents**  | 49.00; 55.25| 51.25; 63.00| 51.25; 63.00| 51.25; 63.00| 51.25; 63.00| 51.25; 63.00| 51.25; 63.00| 51.25; 63.00| 51.25; 63.00| 51.25; 63.00| 51.25; 63.00| 51.25; 63.00| 51.25; 63.00| 51.25; 63.00|          |
| **DEL**          | 46.50; 53.75| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00|          |
| **mUPD**         | 49.00; 58.00| 52.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00| 49.00; 55.00|          |

*P-values from the Mann–Whitney test.

*P < 0.05.*
RESULTS

Intellectual Ability

The median VIQ, PIQ, and FIQ scores and the median standard scores on the subtests in the groups and comparison of the two genotypes are presented in Table II. In children, statistically significant differences were found between mUPD and DEL in terms of PIQ (Wilcoxon, median = 39.5, 45.5; \( P = 0.012 \)), FIQ (median = 39, 46; \( P = 0.031 \)), and perceptual organization (median = 49, 50; \( P = 0.025 \)), with higher scores in the DEL group. In adolescents, significant differences were found between mUPD and DEL in terms of PIQ (median = 46, 50; \( P = 0.026 \)), perceptual organization (median = 49, 54; \( P = 0.017 \)) and working memory (median = 49, 53.5; \( P = 0.043 \)), with higher scores in the DEL group.

Table III presents the differences between VIQ and PIQ for both groups of mUPD children and DEL children and in both groups of mUPD adolescents and DEL adolescents. A significantly higher score for VIQ than for PIQ was observed in all of the four groups (Wilcoxon, mUPD children, median = 9; \( P = 0.027 \)), DEL children, median = 10.5; \( P = 0.001 \), mUPD adolescents, median = 9; \( P = 0.018 \), DEL adolescents, median = 7; \( P = 0.001 \)).

Autistic Symptomatology

In children, no statistically significant differences were found between mUPD and DEL participants in terms of autistic symptomatology indicated by the total score of the PARS (median = 13, 11.5; \( P = 0.657 \)). Also, no significant differences were found between the two groups of children in terms of clinical subscores in the PARS.

On the contrary, statistically significant differences were found between mUPD adolescents and DEL adolescents in terms of the total score of the PARS (median = 21, 11.5; \( P = 0.027 \)). In addition, significant differences were found between the two groups of adolescents in two among five subscores, such as interpersonal skills (median = 4, 2.5; \( P = 0.006 \)) and hypersensitivity (median = 2, 1; \( P = 0.003 \)) (Table IV).

The PARS total score in mUPD children was above the cut-off value cited in Adachi et al. [2006] based on normative data collected from 93 children. Also, compared with normative data collected from 95 adolescents [Kamio et al., 2006], the PARS score in mUPD children was above the cut-off value. On the other hand, the PARS total scores for both the DEL children group and the DEL adolescent group were below the cut-off value cited in each study (Fig. 1).

Inattention and Hyperactivity/Impulsivity

In children, no significant differences were found between mUPD and DEL participants in terms of both inattentive (median = 5.5, 2.5; \( P = 0.20 \)) and hyperactive/impulsive behaviors (median = 5.5, 2; \( P = 0.275 \)). By contrast, in adolescents, mUPD patients showed significantly more impulsive behavior (ADHD-RS-IV hyperactive/impulsive, median = 5, 5.5; \( P = 0.01 \)) than DEL patients. However, no significant differences were found between the two genotypes in terms of inattentive behaviors in either group (ADHD-RS-IV inattentive, median = 6, 2; \( P = 0.20 \)) (Table V).

Compared with normative data, the ADHD-RS-IV total score in mUPD children and that in mUPD adolescents were below the cut-off value cited in Ichikawa and Tanaka [2008]. Also, compared with normative data, the ADHD-RS-IV total scores in DEL individuals were below the cut-off value, regardless of age (Fig. 2).

DISCUSSION

This study is the first neuropsychological study so far conducted involving Japanese patients with PWS in a behavioral comparison of the mUPD and the DEL subtypes. Furthermore, the sole trained psychologist (H.O.), blind to participants’ genetic status, conducted detailed and systematic assessments, using well-established, reliable, and valid measures.

The FIQs in the both groups of children and adolescents in our sample are below 50. These scores are more than 50 points under the normative population score of 100, indicating a global impairment in intellectual abilities. The scores in our sample were lower than those reported by most other studies [Dykens et al., 1992; Roof et al., 2000; Whittington et al., 2004; Milner et al., 2005] from western countries. For example, conducting population-based study of children and adults, Whittington et al. [2004] reported a near-normal distribution of the FIQ around a mean of 60. Nevertheless, the scores in our study were very close to a finding from Taiwan [Shu et al., 2007]. Evaluating 20 patients (14 males/6 females), Shu et al. [2007] reported lower mean FIQ scores of about 50. Taking into account intellectual profiles in detail, we found the

| TABLE III. Difference Between VIQ and PIQ in the Total Group and the Two Genotype Groups |
|---------------------------------|-----------------|-----------------|-----------------|-----------------|
|                                | N               | Median (Q1;Q3)  | Min–Max         | P-value         |
|                                | Children        | Adolescents    | Children        | Adolescents    |
| Total group                    | 22              | 23             | 10.00 (5.75; 17.0) | 7.00 (6.00; 11.00) |
| DEL group                      | 16              | 16             | 10.50 (5.25; 17.0) | 7.00 (5.25; 9.75) |
| mUPD group                     | 6               | ?              | 9.00 (6.50; 13.75) | 9.00 (6.00; 14.00) |
|                                | Children        | Adolescents    | 0; 28           | –1; 24         |
|                                | Children        | Adolescents    | 0; 28           | –1; 24         |
|                                | Children        | Adolescents    | 0.001 **         | 0.001 **         |
|                                | 0.027 *         | 0.018 *        |

\( \sharp \)Median (Q1;Q3), PIQ, performance intellectual quotient, VIQ, verbal intellectual quotient.

\( \star \star \)P < 0.01.

\( \star \)P < 0.05.
VIQ > PIQ pattern in both groups of mUPD children and DEL children and in both groups of mUPD adolescents and DEL adolescents. This pattern characterized by a verbal intelligence higher than a performance intelligence resembles the intellectual profile of Asperger syndrome, one of the pervasive developmental disorders defined by autistic symptomatology without history of language delay [Klin et al., 1995].

We found a consistent pattern of increased autism-like behavioral impairments in the mUPD cases that were less observable in the DEL cases. These findings based on Japanese patients were in concord with those based on British samples, using either postal and telephone surveys [Veltman et al., 2004] or investigator-based assessment [Milner et al., 2005]. For example, Veltman et al. [2004] showed a consistent increase in reciprocal social interaction impairments among individuals with mUPD cases. Conducting a large study comparing mUPD and DEL forms of PWS, Milner et al. [2005] also demonstrated that mUPD exhibited significantly more autistic-like impairments on questionnaire, interview, and standardized observational measures. Our findings about Japanese PWS patients corresponding to previous researches issued from Western countries imply the possibility that mUPD cases are more prone to ASD than DEL cases, regardless of ethnoregional differences.

In our sample, the two groups of children did not show clinically relevant indices of ADHD, regardless of being mUPD or DEL. In adolescents, mUPD patients showed significantly more impulsive behavior (ADHD-RS-IV hyperactive/impulsive, \( P = 0.01 \)) than DEL patients. Even so, they did not reach the level of clinical ADHD, according to normative data cited in Ichikawa and Tanaka [2008].

So far, the relationship between ADHD and PWS has been rarely discussed. On the contrary, the close association of ASD with ADHD has often been considered and even shared heritability is postulated between the two conditions [Rommelse et al., 2010]. During physical growth, patients with PWS can develop inattention, distractability, and impulsivity. However, hypotonia and morbid obesity can prevent these behavioral characteristics from being recognized and standard assessment tools may fail to detect them. Applying Conners’ Parent Rating Scale (CPRS-48) to 58

---

**TABLE IV. Distribution of PARS Total Scores and Subscores in the Groups and Comparison of the Two Genotype Groups**

|                     | Total DEL (N = 23) | Adolescents DEL (N = 16) | Children mUPD (N = 6) | Adolescents mUPD (N = 7) | P-value |
|---------------------|-------------------|--------------------------|-----------------------|--------------------------|---------|
| Total score         | 11.50 (7.75; 18.00) | 12.50 (9.50; 20.50)     | 11.50 (8.25; 16.75)   | 12.00 (12.00; 32.00)     | 0.657   |
| Interpersonal skills| 2.00 (0.00; 3.00)  | 3.00 (2.00; 4.00)        | 2.50 (2.00; 3.00)     | 1.50 (3.00; 6.00)        | 0.701   |
| Communication       | 4.00 (3.00; 5.50)  | 5.00 (3.00; 6.00)        | 5.00 (2.75; 6.00)     | 4.00 (3.00; 6.00)        | 0.733   |
| Obsession           | 3.00 (1.50; 5.50)  | 3.00 (0.50; 4.50)        | 3.00 (2.00; 5.00)     | 3.00 (2.00; 5.00)        | 0.381   |
| Stereotyped behavior| 0.00 (0.00; 1.50)  | 0.00 (0.00; 1.00)        | 0.00 (0.00; 2.00)     | 0.00 (0.00; 2.00)        | 0.85    |
| Problematic behaviors| 0.00 (0.00; 2.50) | 0.00 (0.00; 4.50)        | 0.00 (0.00; 4.25)     | 0.00 (0.00; 2.25)        | 0.519   |
| Hypersensitivity    | 1.00 (0.00; 2.00)  | 1.00 (0.00; 2.00)        | 1.00 (0.00; 2.00)     | 1.00 (0.00; 2.00)        | 0.302   |
| Observed Score      | -0.69              | -6.5                     | 0.5                   | 1.86                     |         |

Q1, 1st quartile; Q3, 3rd quartile.  
P-values from the Mann–Whitney test.  
\(* P < 0.05.\)  
\(** P < 0.01.\)

---

**FIG. 1.** Difference between PARS total score and the cut-off value based on normative date [Adachi et al., 2006; Kamio et al., 2006].
patients with PWS (aged 5–18), Wigren and Hansen [2005] found that 26% of their sample reached clinically elevated indices of ADHD. In our sample, although failing to reach the cut-off point, mUPD adolescents showed a slight tendency of impulsivity that was not seen in DEL children.

This study considered the impact of biopsychological changes in chronological adolescence. The results suggested that the behavioral differences between mUPD and DEL cases in terms of autistic and impulsive symptoms tend to be unrecognizable in their childhood. In children, no significant differences were found between mUPD and DEL participants in terms of autistic symptomatology. On the contrary, in adolescence, statistically significant differences were found between the two groups in terms of autistic symptomatology. Among autistic characteristics, interpersonal skills and hypersensitivity were of prominence, where differences between mUPD and DEL were significant in adolescents, but not in children. Also, significant differences between mUPD and DEL participants in terms of both hyperactive/impulsive behaviors were not found in children, but were found in adolescents. Therefore, there is a growing tendency for the autistic and impulsive behavioral problems, which are more severe in mUPD than in DEL, to manifest themselves later in adolescence.

It is evident that a number of methodological limitations consist in the current study. Since this is a single-institution study to be aiming at a rare genetic disorder, the size of sample is relatively small. Besides, this study is cross-sectional rather than longitudinal. In fact, cross-sectional comparison between different age brackets cannot avoid inter-generational differences. For assessing behavior development across time, longitudinal studies by tracking the same cohort could make observing changes more accurate than cross-sectional ones. Furthermore, whilst this is the first study outside Western societies, applicability of the result to other non-Western populations remains unclear. Moreover, the assessment tools applied in this study were not developed for the examination exclusively of PWS individuals. Although these rating tools had solid external reliability, the indices of autistic and hyperactive behaviors require further validation in clinical assessment.

ACKNOWLEDGMENTS

This research was supported by a grant for Research Support Foundation from the Juntendo Institute of Psychiatry in the financial year 2013 (Heisei 25).

REFERENCES

Adachi J, Yukihiro R, Inoue M, Uchiyama T, Kamio Y, Kurita H. 2006. Reliability and Validity of the childhood part of the PARS (PDD-Autism Society Japan Rating Scale). Rinsyoseishinigaku 35:1591–1599 (in Japanese).

Arron K, Oliver C, Moss J, Berg K, Burbidge C. 2011. The prevalence and phenomenology of self-injurious and aggressive behavior in genetic syndromes. J Intellect Disabil Res 55:109–120.

Bolton P, Dennis N, Browne C, Thomas N, Veltman M, Thompson R, Jacobs P. 2001. The phenotypic manifestations of interstitial duplications of proximal 15q with special reference to the autistic spectrum disorders. Am J Med Genet 105:675–685.

TABLE V. Distribution of ADHD-RS Total Scores and Subscores in the Groups and Comparison of the Two Genotype Groups

|                  | Total   |       | DEL    |       | mUPD  |       | P-value |
|------------------|---------|-------|--------|-------|-------|-------|---------|
|                  | Children| Adolescents | Children| Adolescents | Children| Adolescents |       |
| Inattentive      | Median  | [Q1; Q3] | Median | [Q1; Q3] | Median | [Q1; Q3] | Median | [Q1; Q3] |       |
|                  | 2.00    | (0.00; 5.00) | 2.50   | (0.00; 5.00) | 2.00   | (0.50; 5.00) | 0.50   | (0.00; 3.00) |       |
| Hyperactivity/impulsivity | Median  | [Q1; Q3] | Median | [Q1; Q3] | Median | [Q1; Q3] | Median | [Q1; Q3] |       |
|                  | 2.00    | (0.00; 3.00) | 1.00   | (0.00; 3.00) | 2.00   | (0.25; 3.75) | 0.50   | (0.00; 1.75) |       |
| Total score      | Median  | [Q1; Q3] | Median | [Q1; Q3] | Median | [Q1; Q3] | Median | [Q1; Q3] |       |
|                  | 3.00    | (0.75; 7.25) | 4.00   | (1.25; 7.75) | 4.50   | (2.25; 9.25) | 3.00   | (1.25; 5.00) |       |

Q1, 1st quartile; Q3, 3rd quartile. *P-values from the Mann–Whitney test. **P < 0.05.

FIG. 2. Difference between ADHD-RS-total score and the cut-off value based on normative date [Ichikawa and Tanaka, 2008].
Cassidy SB. 1997. Prader-Willi syndrome. J Med Genet 34:917–923.

Descheemaeker MJ, Vogels A, Govers V, Borghgraef M, Willekens D, Swillen A, Verhoeven W, Fryns JP. 2002. Prader-Willi syndrome: New insights in the behavioural and psychiatric spectrum. J Intellect Disabil Res 46:41–50.

Descheemaeker M, Govers V, Vermeulen P, Fryns JP. 2006. Pervasive developmental disorders in Prader-Willi syndrome: The Leuven experience in 59 subjects and controls. Am J Med Genet Part A 140A:1136–1142.

Dimitropoulos A, Schultz RT. 2007. Autistic-like symptomatology in Prader-Willi syndrome: A review of recent findings. Curr Psychiatry Rep 9:159–164.

DuPaul J, Power J, Anastopoulos D, Reid R. 1998. ADHD-Rating Scale-IV: Checklists, norms, and clinical interpretation. New York: Guilford Press.

Dykens EM, Hodapp RM, Walsh K, Nash JJ. 1992. Profiles, correlates, and trajectories of intelligence in Prader-Willi syndrome. J Am Acad Child Adolesc Psychiatry 31:1125–1130.

Dykens EM, Lee E, Roof E. 2011. Prader-Willi syndrome and autism spectrum disorders: An evolving story. J Intellect Disabil Res 50:92–100.

Greaves N, Prince E, Evans DW, Charman T. 2006. Repetitive and ritualistic behavior in children with Prader-Willi syndrome and children with autism. J Intell Disabil Res 50:92–100.

Hogart A, Wu D, LaSalle JM, Schanen NC. 2010. The comorbidity of autism with the genomic disorders of chromosome 15q11.2-q13. Neurobiol Dis 38:181–191.

Ichikawa H, Tanaka Y. 2008. ADHD-Rating Scale-IV: Checklists, norms, and clinical interpretation. Tokyo: Akashisyoten (in Japanese).

Ito H, Tani I, Yukihiro R, Adachi J, Hara K, Ogasawara M, Inoue M, Kamio Y, Nakamura K, Uchiyama T, Ichikawa H, Sugiyama T, Hagiwara T, Tsujii M. 2012. Validation of an interview-based rating scale developed in Japan for pervasive developmental disorders. Res Autism Spectr Disord 6:1265–1272.

Japanese WISC-III Publication Committee. 1998. Nihonban WISC-III chinou kenshousou (Japanese Wechsler Intelligence Scale for Children, 3rd ed). Tokyo: Nihon Bunka Kagakusya.

Japanese WAIS-III Publication Committee. 2006. Nihonban WAIS-III chinou kenshousou (Japanese Wechsler Adult Intelligence Scale, 3rd ed). Tokyo: Nihon Bunka Kagakusya.

Kamio Y, Yukihiro R, Adachi J, Ichikawa H, Inoue M, Uchiyama T, Kurita H, Sugiyama T, Tsujii M. 2006. Reliability and Validity of the Pervasive Developmental Disorder (PDD)-Autism Society Japan Rating Scale (PARS): A behavior checklist for adolescents and adults with PDDs. Seishinigaku 48:495–505 (in Japanese).

Klin A, Volkmar FR, Sparrow SS, Cichetti DV, Rourke BP. 1995. Validity and neuropsychological characterization of Asperger syndrome: Convergence with nonverbal learning disabilities syndrome. J Child Psychol Psychiatry 36:1127–1140.

Milner K, Craig E, Thompson R, Veltman M, Thomas N, Roberts S, Bellamy M, Curran S, Spotikou C, Bolton P. 2005. Prader-Willi syndrome: Intellectual abilities and behavioral features by genetic subtype. J Child Psychol Psychiatry 46:1089–1096.

Rommelse NN, Franke B, Geurts HM, Hartman CA, Buitelaar JK. 2010. Shared heritability of attention-deficit/hyperactivity disorder and autism spectrum disorder. Eur Child Adolesc Psychiatry 19:281–295.

Roof E, Srone W, McLean W, Feuer I, Thompson T, Butler M. 2000. Intellectual characteristics of Prader-Willi syndrome: a comparison of genetic subtypes. J Intellect Disabil Res 44:25–30.

Shu SG, Chien S, Wu YC, Tsai PL, Yih JK. 2007. Anthropometric and intellectual evaluation of individuals with Prader-Willi syndrome. J Formos Med Assoc 106:509–512.

Veltman MW, Thompson RJ, Roberts SE, Thomas NS, Whittington J, Bolton PF. 2004. Prader-Willi syndrome—A study comparing deletion and uniparental disomy cases with reference to autism spectrum disorders. Eur Child Adolesc Psychiatry 13:42–50.

Veltman MW, Craig EE, Bolton PF. 2005. Autism spectrum disorders in Prader-Willi and Angelman syndromes: A systematic review. Psychiatr Genet 15:243–254.

Wechsler D. 1991. Wechsler Intelligence Scale for Children, 3rd edition. San Antonio, TX: The Psychological Corporation.

Wechsler D. 1997. Wechsler Adult Intelligence Scale, 3rd edition. San Antonio, TX: The Psychological Corporation.

Whittington J, Holland A, Webb T, Butler J, Clarke D, Boer H. 2004. Cognitive abilities and genotype in a population-based sample of people with Prader-Willi syndrome. J Intellect Disabil Res 48:172–187.

Wigren M, Hansen S. 2005. ADHD symptoms and insistence on sameness in Prader-Willi syndrome. J Intellect Disabil Res 49:449–456.

Yamada A, Suzuki M, Kato M, Suzuki M, Tanaka S, Shindo T, Taketani K, Akechi T, Furukawa TA. 2007. Emotional distress and its correlates among parents of children with pervasive developmental disorders. Psychiatry Clin Neurosci 61:651–657.