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This is the author's manuscript

Original Citation:

Availability:

This version is available <http://hdl.handle.net/2318/1858259> since 2022-05-08T17:46:01Z

Published version:

DOI:10.23736/S2724-5276.21.06447-8

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TITLE PAGE

Manuscript title:

Psychopathological profile in children with Prader-Willi syndrome as compared with autism spectrum disorder.

Running Title:

Psychopathology in children with PWS and with ASD.

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Full citation:

Bechis, D., Baietto, C., Caldarera, A. M., & Vitiello, B. (2021). Psychopathological profile in children with Prader-Willi syndrome as compared with autism spectrum disorder. *Minerva pediatrics*, 10.23736/S2724-5276.21.06447-8. Advance online publication. <https://doi.org/10.23736/S2724-5276.21.06447-8>

Psychopathological profile in children with Prader-Willi syndrome as compared with autism spectrum disorder.

Abstract

Background: Children with Prader-Willi syndrome (PWS) can present with social deficits and repetitive behaviours that are also encountered in autism spectrum disorder (ASD). This study aimed at ascertaining possible differences in psychopathology between PWS and ASD, with particular attention to obsessional thinking, repetitive behaviours, and impulsivity.

Methods: 71 children, aged 4-15 years: 24 with PWS, 23 with ASD, and 24 community controls, were assessed on two standardized parent-reported questionnaires: the Child Behaviour Check List (CBCL) and the Autism Spectrum Quotient (AQ). Group differences were tested with one-way ANOVA.

Results: ASD had higher CBCL internalizing symptom scores (67.50 ± 9.09) than PWS (56.62 ± 9.02 , Cohen's $d=1.20$). On specific CBCL items, PWS had more obsessional than ASD, which, in turn, showed more impulsivity than PWS. ASD had higher AQ scores than PWS, with small to medium effect sizes (d 's ranging from 0.22 to 0.53).

Conclusions: The PWS phenotype was characterized by intense obsessional, more marked than in ASD. ASD had greater psychopathology than PWS, especially of the internalizing type. Although limited by the small sample size, this study identifies obsessional as common feature in PSW. Such symptom, considering the negative impact on daily functioning, requires clinical attention for specific treatment approaches.

Keywords: Prader-Willi syndrome, Autism Spectrum Disorder, Psychopathology, Intellectual disability, Behavioural Profile

Introduction

Prader-Willi Syndrome (PWS) is a genetically determined neurodevelopmental disorder first described by Prader, Labhart and Willi in 1956 (1). It is caused by a failure of paternal expression of maternally imprinted genes at the 15q11–13 region, through one of two main mechanisms: either deletion from the paternal chromosome 15 (DEL), which is found in approximately 70% of cases, or inheritance of two copies of the maternal chromosome (maternal uniparental disomy (mUPD), which accounts for about 25% of the cases (2). Clinical manifestations include infantile hypotonia, increased pain threshold, neuroendocrine dysfunction, behavioural problems, self-abusive behaviour, compulsive eating disorder, anxiety and mood disorders (2, 3, 4).

Repetitive and ritualistic behaviours, often but not always linked to a preoccupation with food, are common, emerge early in life, and tend to increase with age (5, 6, 7). The classic compulsions typical of obsessive-compulsive disorder, such as hand washing, cleaning, and checking, are rare. Guinovart et al. (4) argued that in individuals with PWS, as it is for other patients with mental retardation (8), compulsions are not aimed at mitigating anxiety or stress, but have a self-stimulating. PWS is also characterized by cognitive rigidity, perseveration and difficulties in accepting changes of in routine (7, 9).

In recent years it has been reported that individuals with PWS are at increased risk for autism spectrum disorder (ASD), especially due to symptoms of social impairment and repetitive behaviours (6, 10). Salehi et al. (11) reported that individuals with PWS, compared with persons with other forms of intellectual disability, showed significant social deficits and maladaptive behaviour. There are also certain repetitive behaviours, such as tics and stereotypical behaviour, that are encountered in PWS, in addition to perseveration, reluctance to change, rituals and a restricted range of interests, all typical ASD manifestations (4). ASD traits may not be evident in young children with PWS, but appear later in development (7). Some reports suggest that children with PWS with the genetic mutation maternal UPD are at greater risk for autistic symptomatology than those with deletion (9, 12, 13). In particular, individuals presenting with maternal UPD may be at higher risk for social impairment than those with deletion (14).

ASD traits have been examined in studies comparing PWS with other neurodevelopmental syndromes (11, 13, 15, 16). Most of the comparative studies used a two-group design (5, 7, 16, 17), but one study included PWS and ASD together with a group of normally developing children as a reference (18).

Most psychiatric disorders in PWS arise in early childhood, except for depression and psychosis, which usually appear after puberty onset (3). According to some studies, the mUPD subtype may be more likely

to present with psychotic symptoms and have increased risk of psychotic recurrence, poorer response to treatment, and worse prognosis than the DEL subtype (19, 20). Hartley et al. (21) reported increased risk of depression, aggression, and dependent personality disorder in male patients with deletion PWS. Skokauskas et al. (22) found that PWS, compared to nonclinical controls, had higher scores of psychopathology on the Child Behaviour Checklist subscales *withdrawn-depressed*, *somatic complaints*, *social problems* and *thought problems*. Guinovart et al. (4) recalled that Symons et al. (23) found an 89% rate of self-harm, most commonly compulsive skin scratching (82%), often resulting in tissue injuries and infections. In a national survey undertaken on families of individuals with PWS (24), skin-picking was reported in 93% of adolescents with PWS. In another study, Dimitropoulos et al. (15) found that skin-picking started early in life and affected 40% of children with PWS. These repetitive, stereotypic behaviours, often with self-injurious consequences, while compulsive in nature, are distinct from the typical manifestations of obsessive-compulsive disorder (OCD). They may be part of the PWS phenotype and related to the level of intellectual disability and impulsivity (4, 25, 26, 27).

In sum, the available reports suggest that individuals with PWS present with a number of psychopathological features that are part of the ASD symptomatology and that significantly impair global functioning. Few studies however directly compared PWS and ASD with each other and with normally developing children.

The present study was meant to extend the current data on psychopathology in PWS by examining differences from ASD children and from community controls. The work was guided by the following three hypotheses:

H1. Both PWS and ASD children would present with higher levels of emotional and behavioural problems than normal community controls, but with greater psychopathology in ASD than in PWS.

H2. PWS would present with more ASD traits than community controls, but less marked ASD.

H3. Within the PWS group, there will be differences in psychopathology between the mUPD and the DEL genetic subgroups, with the former having greater severity.

Materials and Methods

Participants

A total of 71 children (age range 4-15 years) and their parents (age range 28-52 years) participated into the study. The sample included three subgroups: 24 children with PWS, 23 children with ASD, and 24

children from a community sample without known neurodevelopmental or psychiatric disorders. The 24 families of the first subgroup were recruited with the help of a Prader-Willi syndrome regional association and of the Prader-Willi national association. Children with ASD had been diagnosed according by an experienced child neuropsychiatrist according to DSM-5 criteria (28). Sample demographics are shown in Table 1. With regard to the genetic subtypes of PWS, 13 children (54%) had a mUPD mutation and 9 (38%) a DEL, while this information was not available for 2 children.

Ethical approval

Ethical approval for the study was granted by the institutional research ethics committee. Participants received a set of questionnaires, supplemented with a letter containing information about the study. After providing informed consent, the parents filled-in and returned the questionnaires.

Measures

The assessments included:

The **Child Behaviour Check List (CBCL)**, a standardized, parent-report questionnaire describing a range of symptoms in preschool ($\frac{1}{2}$ - 5 version, 29) and older children and adolescents (6 – 18 version, 30). The questionnaire is used in both clinical setting and research. The 1 $\frac{1}{2}$ - 5 years version includes 100 items and provides 7 empirical subscales (*emotionally reactive, anxious/depressed, somatic complaints, withdrawn, sleep problems, attention problems* and *aggressive behaviour*), two broad-band scales which are referred to as the dimensions of Internalizing (from the first four subscales) and Externalizing problems (from the is made of the last two subscales). The Internalizing Problems, Externalizing Problems and *Sleep Problems* scales are combined to generate the Total Problems scale. The 6 – 18 years scale has 113 items, and evaluates Internalizing (*Anxious/depressed, withdrawn/depressed, somatic complaints*) and Externalizing problems (*rule-breaking behaviour, aggressive behaviour*). In addition to the subscales composing the Internalizing and Externalizing scales, this version includes also *social problems, thought problems, and attention problems* subscales. Every item can be scored from 0 to 2 (0=not true; 1=sometimes true; 2=very/often true). Likewise the CBCL 1 $\frac{1}{2}$ - 5 version, also the 6-18 includes a Total Problems scale, calculated by combining the Internalizing Problems, Externalizing Problems and the *Other Problems* scales. In our analyses we were particularly interested in single items of the CBCL that assess behaviours typically associated to ASD or PWS symptomatology, specifically item 9 (*Can't get his/her mind off certain thoughts; obsessions*), item 66 (*Repeats certain acts over and over*), item 41 (*Impulsive or acts without thinking*), item 58 (*Picks nose,*

skin or other parts of body). Impulsiveness is a behavioural feature commonly observed in both PWS and ASD children (31, 32), and skin picking was reported to be a behaviour characterizing PWS compared to ASD (9). This approach of using single CBCL items in order to examine specific behaviours has already been used in child research (33, 34).

The **Autism Spectrum Quotient – Children version (AQ-Child)**, a 50-item parent-report questionnaire measuring autistic traits in children (35). The total score ranges from 0 to 150, with higher scores indicating more behaviours indicative of autistic features. AQ yields four subscales scores (*Mindreading, Attention to Detail, Social Skills and Imagination*), in addition to the *Total* score.

Statistical Analysis

Data analysis was performed by using SPSS 25. After running descriptive statistics, we tested differences between groups through univariate analyses of variance (one-way ANOVA), and looked both at statistical significance (p value) of the F-test and effect size, using the ω^2 , which, compared to other estimates, is less vulnerable to inflation from chance factors (36, 37). Post hoc pairwise comparisons were calculated with Bonferroni correction for multiple comparisons. We further examined specific differences between children with PWS and ASD through the Cohen's d . In order to examine differences, within the PWS group, according to genetic mutation, and in consideration of the very small size of the two subgroups, we used the non-parametric Mann-Whitney U test.

Results

CBCL

The scores of CBCL syndrome subscales, broad scales, and of the targeted single items are reported in Table 2, with statistical significance of between group differences and results of post-hoc pair-wise comparisons with Bonferroni correction and effect sizes.

General psychopathology (CBCL)

There were statistically significant differences between the three groups on all the CBCL broad and syndrome scales except for the *Somatic complaints* scale. As expected, PWS and ASD had higher scores (i.e., indicative of more psychopathology) than the community controls on most of the scales. One scale that was particularly discriminating between groups was PWS and ASD was the *Attention problems*

scale, with PWS reporting significantly higher scores than the community control sample, but significantly lower than ASD. There was a large effect size (Cohen's $d = .83$) on this scale between PWS and ASD. Large effect sizes between PWS and ASD were also found in the *Anxious/Depressed* syndrome scale ($d = .95$) and the *Internalizing* scale ($d = 1.20$), with the ASD group showing higher scores than PWS.

Targeted single CBCL items

Statistically significant differences between groups were found for items 9 (*Can't get his/her mind off certain thoughts; obsessions*), 41 (*Impulsive or acts without thinking*), and 66 (*Repeats certain acts over and over*), while for item 58 (*Picks nose, skin or other parts of body*) there was trend toward significance ($p=0.052$) (Table 2).

On item 9 (*Can't get his/her mind off certain thoughts; obsessions*), PWS had higher scores than ASD, and both were higher than CC. On 41 (*Impulsive or acts without thinking*), ASD was higher than PWS, and both were higher than CC. Medium to large effect size differences were found between PWS and ASD on item 9 ($d=.81$) and item 41 ($d=.75$).

On item 66 (*Repeats certain acts over and over*), while both PWS and ASD had significantly higher scores than CC, the difference between each other ($d=.47$) did not reach statistical significance.

Autism Spectrum Quotient (AQ)

The mean values of the AQ scores in the three groups are reported in Table 3, along with statistical significance of the between group differences, post-hoc multiple comparisons (Bonferroni-corrected), and effect sizes. Significant between group differences were found for all the examined variables, apart from the *Attention to detail* subscale. On the total AQ score, both PWS and ASD had significantly higher scores than CC. The effect size of the difference between PWS and ASD was $d=0.44$ (not statistically significant after Bonferroni correction). Likewise, on two of four AQ subscales (*Mindreading* and *Social Skills*), PWS and ASD scores were significantly higher than CC, with small effect sizes ($d=0.22-0.33$) between PWS and ASD (Tab. 3)

Characteristics of the two PWS subgroups according to genetic mutation: descriptive statistics

In the light of the studies which found that individuals with PWS present with specific behavioural features according to the genetic mutation (19), we examined differences, for the core variables (reported for the three groups in Table 3), between the two PWS subgroups, made of children with deletion and

children with maternal uniparental disomy. The Mann-Whitney U test showed no significant differences for all the considered variables. Descriptive statistics (mean and standard deviation) are shown in Table 4, attached to the present article as supplemental material.

Discussion

In this study we compared PWS to ASD and CC with respect to psychopathology and in particular autistic behavioural features. Overall, the results show that both PWS and ASD had greater psychopathology and autistic symptoms than CC. The PWS group shared some characteristics with the ASD group, but also differed from it for others, according to our hypotheses.

H1. Both children with PWS and children with ASD present with higher levels of emotional and behavioural problems when compared to general population, but it is possible to identify specific differences between children with PWS and with ASD.

Both the PWS and the ASD groups showed higher levels of behavioural difficulties compared to the nonclinical sample. However the ASD group showed higher scores at the *Anxious/Depressed* syndrome scale compared to PWS. This finding is in line with previous scientific literature, showing high rates of psychiatric disorders both in children with ASD and in children with PWS (3, 24, 38, 39).

Also the recent systematic review and meta-analysis by Glasson and colleagues (40) confirms the presence of psychopathology, as measured by the CBCL, in individuals with PWS.

The higher rates of psychopathology in the ASD group may be partially explained by the number of males in this group, compared to the other two. However, our results should be further investigated with a bigger sample and with more specific scales.

As regards the broad scales, the ASD group showed higher levels of psychopathology in all of the three scales, particularly for the Internalizing. The large effect size of the difference between PWS and ASD scores at the Internalizing scale, as it is for the *Anxious/depressed* scale, may indicate that children with ASD are much more prone to an internalization of symptom expression compared to children with PWS.

H2. Similarly to H1, children with PWS present with some traits consistent with autistic symptomatology when compared to general population, but, when compared with children diagnosed with ASD, it is possible to find specific differences between the two groups.

The multiple comparison (made by the post-hoc procedure following the ANOVA) between the three groups showed that both the PWS and the ASD group reported higher scores at the scales *AQ Total*, *AQ Mindreading* and *AQ Social Skills*, compared to the community sample. As regards the *Imagination* subscale, the post hoc procedure confirmed a significant difference only between the ASD and community sample, but not for the PWS group, which didn't show any significant difference, neither from the ASD group, nor from the community sample. This may indicate that the PWS and ASD group may both present an impairment, as compared to the non-clinical group, in relation to the social domain and to the mindreading abilities, but not for the other autistic domains. It is noteworthy that the differences for the mentioned domains were significant despite the ASD group including a higher number of male than female subjects, which, according to literature (41), should have the effect of increasing the scores related to ASD features.

Interestingly, our findings show that obsessive thoughts (CBCL item 9) are significantly higher in the PWS group compared both to the ASD and to the non-clinical group. Instead, scores related to impulsivity (CBCL item 41) in the ASD group are significantly higher than those in Prader-Willi and in the group from a community sample. This indicates that in our sample, obsessive thoughts seem to be a feature that differentiates children with Prader-Willi syndrome from children with ASD, whereas impulsivity stays in the core characteristics in the autism group. These findings are in line with what was described by Song et al. (7), that starting from middle school age, obsessive symptoms become particularly evident in PWS children.

H3. Within the PWS group, it is possible to identify specific features of children with deletion compared to children with mUPD

We didn't find significant differences between two major genetic subtypes (deletion and maternal UPD) at the AQ questionnaire and single CBCL items: however, this result may be due to the small sample size (6 children with deletion and 10 children with maternal disomy). Anyway, when looking at mean scores, although differences are not statistically significant, maternal disomy showed higher scores in many at the scales *AQ Total*, *AQ Mindreading*, *AQ attention to detail*, and lower scores at *AQ Imagination*. Also for the CBCL items, we found higher scores at item 9 (*obsessive thoughts*) for children with maternal disomy and higher scores for children with deletion on CBCL items 41, 58, and 66.

Study limitations and future directions

This study has several limitations. This was a sample of convenience, recruited from the local regional area and not epidemiologically representative. The sample size is small and, although consistent with the rare disease prevalence of PWS, limits the statistical power to detect significant differences. This limitation is especially evident when examining genetic subgroups. Another limitation is that, due to difficulties in recruitment, we couldn't completely balance for sex the ASD group, which had a greater proportion of males. Moreover, data on IQ or other cognitive measures were not available. For future studies, it would be important to be able to recruit a higher number of participants and use assessment tools more targeted to specific to PWS symptoms, such as compulsive skin scratching.

Conclusions

In conclusion, we found both similarities and differences between PWS and ASD with clear-cut differences from CC. ASD was characterized by greater psychopathology, in particular of the internalizing type, than PWS, while PWS showed more intense obsessional symptoms than ASD. Although limited by the small sample size, the study confirms that autism features are more common in PWS than among normal children. The data suggest that obsessionalism may represent a specific feature of PWS. If confirmed, this finding can have treatment implications and help develop more targeted interventions.

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Conflict of interest

In the last two years, BV has been consultant for Medice, Lundbeck, Goodwin & Procter, Haynes & Boone. The other authors have no conflict of interest to declare.

Source of funding

This research was supported in part by the Piedmont PWS Association and by the Children's Rare Disease Federation (Torino).

Ethics approval statement

The study was approved by the institutional research ethics committee on the 28th June 2018, with no. 0067250.

Authors' contributions.

Daniela Bechis and Chiara Baietto have given substantial contributions to the conception and the design of the manuscript, Angela M. Caldarera to acquisition, analysis and interpretation of the data. All authors have participated to drafting the manuscript, Benedetto Vitiello revised it critically. All authors read and approved the final version of the manuscript.

Acknowledgements

We would like to acknowledge the children and the families involved in this study: the Prader-Willi families and the Prader-Willi Italian Federation that supported this study.

We would like to acknowledge also Centro Ferrero in Alba for the help in recruiting ASD sample.

Table 1. Demographic Characteristics

	ALL	PWS ^a	ASD ^b	CC ^c
	(<i>n</i> = 71) (41 boys, 30 girls)	(<i>n</i> = 24) (10 boys, 14 girls)	(<i>n</i> = 23) (21 boys, 2 girls)	(<i>n</i> = 24) (10 boys, 14 girls)
Age (years)	Mean (SD): range 9.01 (2.52): 4-15	Mean (SD): range 9.76 (2.77): 5-14	Mean (SD): range 8.43 (2.79): 4-15	Mean (SD): range 9.13 (1.92): 5-11
Parents' age (years)	Mean (SD): range 42.56 (5.49): 28-52	Mean (SD): range 44.78 (3.50): 39-50	Mean (SD): range 39.57 (6.56): 28-52	Mean (SD): range 43.29 (4.81): 36-52
Parental education	N (%)	n (%)	n (%)	n (%)
<i>Junior High School</i>	10 (14.1)	2 (8.3)	7 (30.4)	1 (4.2)
<i>High School/ Vocational High School</i>	39 (55)	12 (49.9)	12 (52.1)	15 (62.5)
<i>University Degree (BA or MA/ PhD or Other)</i>	20 (28.1)	8 (33.4)	4 (17.4)	8 (33.3)
<i>Missing</i>	2 (2.8)	2 (8.3)	0 (0)	0 (0)

- a. PWS: Prader-Willy Syndrome
- b. ASD: Autism Spectrum Disorder
- c. CC: Community Control

Table 2. Child Behaviour Check-List (CBCL) in Prader-Willi Syndrome (PWS), Autism Spectrum Disorder (ASD) and Community Controls (CC)

	PWS N=24	ASD N=23	CC N=24	ANOVA F ^a	Effect Size ^b	Post-hoc Comparisons (Bonferroni) ^c	Effect Size ^d of the difference between PWS and ASD groups
	mean (SD)	mean (SD)	mean (SD)				
CBCL syndrome scales							
Anxious/Depressed	4.19 (3.70)	9.15 (6.41)	3.65 (2.60)	9.656***	$\omega^2 = .21$	ASD > PWS, Community	$d = .95$
Withdrawn/Depressed	6.67 (2.29)	5.35 (3.00)	1.78 (1.81)	12.025***	$\omega^2 = .26$	Community < ASD, PWS	$d = .57$
Somatic Complaints	2.48 (3.75)	3.90 (3.06)	1.74 (1.39)	3.100 <i>ns</i>	$\omega^2 = .06$	ASD > Community	$d = .42$
Social Problems	6.10 (2.88)	7.70 (4.71)	2.17 (2.06)	15.777***	$\omega^2 = .32$	Community < ASD, PWS	$d = .41$
Thought Problems	6.76 (4.30)	6.25 (3.57)	1.57 (1.47)	16.808***	$\omega^2 = .33$	Community < ASD, PWS	$d = .13$
Attention Problems	6.62 (3.76)	9.90 (4.12)	2.91 (2.71)	20.895***	$\omega^2 = .38$	ASD > PWS > Community	$d = .83$
Rule-breaking Behaviour	3.24 (2.61)	3.35 (1.66)	1.30 (1.40)	7.701**	$\omega^2 = .17$	Community < ASD, PWS	$d = .05$
Aggressive Behaviour	8.14 (6.30)	10.55 (5.88)	3.65 (2.46)	10.243***	$\omega^2 = .22$	Community < ASD, PWS	$d = .39$
Other Problems	6.71 (4.63)	5.75 (3.49)	2.43 (1.97)	9.134***	$\omega^2 = .20$	Community < ASD, PWS	$d = .24$
CBCL broad scales							
Internalizing T	56.62 (9.02)	67.50 (9.09)	54.27 (7.67)	14.725***	$\omega^2 = .30$	ASD > PWS, Community	$d = 1.20$
Externalizing T	54.95 (10.54)	60.05 (7.68)	49.41 (6.81)	8.707***	$\omega^2 = .19$	ASD > Community	$d = .55$
Total T	60.00 (9.64)	66.59 (9.40)	50.32 (7.42)	18.757***	$\omega^2 = .35$	Community < ASD, PWS	$d = .69$
Targeted single CBCL items							
Cannot get his/her mind off certain thoughts; obsessions (item 9)	1.43 (.60)	.90 (.71)	.17 (.49)	23.996***	$\omega^2 = .42$	PWS > ASD > Community	$d = .81$

Repeats certain acts over and over (item 66)	1.10 (.77)	.75 (.71)	.09 (.29)	15.148***	$\omega^2 = .31$	Community < ASD, PWS	$d = .47$
Impulsive or acts without thinking (item 41)	.62 (.67)	1.15 (.75)	.13 (.34)	15.343***	$\omega^2 = .31$	ASD > PWS > Community	$d = .75$
Picks nose, skin, or other parts of body (item 58)	1.05 (.83)	.85 (.88)	.48 (.59)	3.107 <i>ns</i> ($p=.052$)	$\omega^2 = .06$	<i>NS</i>	$d = .24$

a $*p < .05$; $**p < .01$; $***p < .001$; *ns* not significant

b. Omega squared (ω^2) values: $\omega^2 = .01$ small effect; $\omega^2 = .06$ medium effect; $\omega^2 = .14$ large effect (Field, 2013; Kirk, 1996)

c. The symbol “<” or “>” is used to indicate statistically significant differences, with $p < .05$ or $p < .01$

d. Cohen’s d values: $d = .2$ small effect; $d = .5$ medium effect; $d = .8$ large effect (Cohen, 1988)

Table 3. AQ scores in PWS, ASD and Community groups.

	PWS M (SD)	ASD	Commu nity M (SD)	ANOVA F ^a	Effect Size ^b	Post-hoc Comparisons (Bonferroni) ^c	Effect Size ^d of the difference between PWS and ASD groups
AQ-CHILD							
Total	67.44 (22.98)	76.31 (21.12)	48.83 (10.51)	12.671***	$\omega^2 = .28$	Community < ASD, PWS	$d = .40$
Mindreading	26.05 (10.11)	28.95 (7.43)	17.96 (4.51)	12.802***	$\omega^2 = .28$	Community < ASD, PWS	$d = .33$
Attention to detail	11.72 (6.96)	15.21 (6.23)	11.20 (4.07)	2.89 <i>ns</i>	$\omega^2 = .06$	<i>Ns</i>	$d = .53$
Social skills	19.83 (6.69)	21.53 (8.92)	14.17 (4.08)	7.298**	$\omega^2 = .17$	Community < ASD, PWS	$d = .22$
Imagination	8.50 (4.67)	10.05 (4.01)	6.71 (2.74)	4.176*	$\omega^2 = .09$	ASD > Community	$d = .36$

a $*p < .05$; $**p < .01$; $***p < .001$; *ns* not significant

b. Omega squared (ω^2) values: $\omega^2 = .01$ small effect; $\omega^2 = .06$ medium effect; $\omega^2 = .14$ large effect (Field, 2013; Kirk, 1996)

c. The symbol "<" or ">" is used to indicate statistically significant differences, with $p < .05$ or $p < .01$

d. Cohen's d values: $d = .2$ small effect; $d = .5$ medium effect; $d = .8$ large effect (Cohen, 1988)

SUPPLEMENTAL

Table 4. AQ and CBCL single items scores in PWS subgroups according to genetic mutation

	Deletion		Maternal Uniparental Disomy	
	M (SD)	N	M (SD)	N
AQ-CHILD				
Total	65.00 (25.35)	6	67.30 (23.69)	10
Mindreading	24.17 (11.55)	6	26.00 (10.40)	10
Attention to detail	10.00 (7.04)	6	11.80 (5.69)	10
Social skills	20.00 (6.75)	6	20.00 (7.48)	10
Imagination	10.00 (6.48)	6	8.20 (3.77)	10
CBCL				
Cannot get his/her mind off certain thoughts; obsessions (item 9)	1.29 (.49)	7	1.42 (.67)	12
Repeats certain acts over and over (item 66)	1.14 (.69)	7	1.08 (.79)	12
Impulsive or acts without thinking (item 41)	.86 (.69)	7	.33 (.49)	12
Picks nose, skin, or other parts of body (Item 58)	1.00 (.89)	7	.92 (.79)	12