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RESEARCH ARTICLE

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# Adolescent adaptive behavior profiles in Williams–Beuren syndrome, Down syndrome, and autism spectrum disorder

Carolina Grego Del Cole<sup>1,2\*</sup>, Sheila Cavalcante Caetano<sup>2</sup>, Wagner Ribeiro<sup>3</sup>, Arthur Melo E. e. Kümmer<sup>4</sup> and Andrea Parolin Jackowski<sup>1</sup>

## Abstract

**Background:** Adaptive behavior can be impaired in different neurodevelopmental disorders and may be influenced by confounding factors, such as intelligence quotient (IQ) and socioeconomic classification. Our main objective was to verify whether adaptive behavior profiles differ in three conditions—Williams Beuren syndrome (WBS), Down syndrome (DS), and autism spectrum disorder (ASD), as compared with healthy controls (HC) and with each other. Although the literature points towards each disorder having a characteristic profile, no study has compared profiles to establish the specificity of each one. A secondary objective was to explore potential interactions between the conditions and socioeconomic status, and whether this had any effect on adaptive behavior profiles.

**Methods:** One hundred and five adolescents were included in the study. All adolescents underwent the following evaluations: the Vineland Adaptive Behavior Scale (VABS), the Wechsler Intelligence Scale for Children (WISC), and the Brazilian Economic Classification Criteria.

**Results:** Our results demonstrated that the WBS group performed better than the DS group in the communication domain,  $\beta = -15.08$ ,  $t(3.45)$ ,  $p = .005$ , and better than the ASD group in the socialization domain,  $\beta = 8.92$ ,  $t(-2.08)$ ,  $p = .013$ . The DS group also performed better than the ASD group in socialization,  $\beta = 16.98$ ,  $t(-2.32)$ ,  $p = .024$ . IQ was an important confounding factor, and socioeconomic status had an important effect on the adaptive behavior of all groups.

**Conclusions:** There is a heterogeneity regarding adaptive behavior profiles in WBS, DS, and ASD. These data are important to better design specific strategies related to the health and social care of each particular group.

## Background

This study proposed to analyze differences in the performance of adaptive behavior between groups with genetic syndromes and neuropsychiatric disorders, such as is the case for Williams–Beuren syndrome (WBS), Down syndrome (DS), and autism spectrum disorder (ASD). These groups were also compared with a health control (HC) group. In addition, we have been concerned with

analyzing some of the variables that may interfere with the performance of adaptive behavior, such as IQ and socioeconomic level.

## Adaptive behavior

Adaptive behavior is the collection of conceptual, social, and practical skills that have been learned and are performed by people in order to function in their everyday lives [1]. A number of instruments are used to measure adaptive behavior, some of the more widely used ones include the following: the Vineland Adaptive Behavior Scales (VABS) [2], the Adaptive Behavior Assessment System [3], and the Scales of Independent Behavior—revised [4]. The VABS is composed of three

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main domains for adolescents: communication, daily living and socialization with each domain having three subdomains. The communication domain comprises receptive, expressive and written subdomains; the daily living skills domain comprises personal, domestic and community subdomains; and the socialization domain comprises interpersonal relationships, play, and leisure time and coping skills subdomains [2]. We chose to use VABS in this study because this instrument has been the most widely used of measures of adaptive functioning and semi-structured parent interview over checklists are less vulnerable to reporter bias [5].

Measuring adaptive behavior is of the utmost importance as it provides useful clinical information for the diagnosis of intellectual disabilities, seeing that, limitations in adaptive behavior, associated with deficits in intellectual functioning and age of onset prior to age 18, define intellectual disability [2, 6]. Furthermore, the adaptive behavior performance provides an information that allow establishing education and rehabilitation goals, once that, allow understanding of human functioning [2, 6]. For this reason, it is very important that there is a connection between policy and neurodevelopmental disorders research [7]. It is well known that individuals with certain genetic syndromes share not only physical features but also cognitive and behavioral characteristics. Indeed, different adaptive behavior profiles have been proposed for some genetic syndromes and neuropsychiatric disorders, such as is the case for WBS, DS, and ASD [8–11].

#### **Williams–Beuren syndrome**

Williams–Beuren syndrome is a multisystem genomic disorder, characterized by dysmorphic facial features, short stature, connective tissue abnormalities, and infantile hypercalcemia. People with WBS also have a specific cognitive and behavioral profile, which commonly includes mild intellectual disability (with a relative strength in language and verbal short term memory, and a weakness in visuospatial skills), hypersociality, attention deficit, and anxiety [12]. The condition is caused by the deletion of approximately 26–28 genes from chromosome 7 (7q11.23), and has a prevalence of 1 in 7500 [13]. This syndrome has complex medical, developmental, and behavioral features, requiring intensive multidisciplinary intervention [14].

Compared to healthy controls (HC), individuals with WBS have impairments in adaptive functioning [10]. Moreover, adaptive behavior has been observed to significantly decrease over time in adolescents and adults with WBS [15]. However, there is evidence of heterogeneity, with some individuals functioning at an extremely low level while others function at a chronological

age-appropriate level. This variability is likely to reflect inherent and environmental factors [16]. Regarding adaptive behavior profiles, adolescents and adults with WBS usually demonstrate better socialization [17–19] and communication skills but have a weaker performance in the daily living domain [19–21].

#### **Down syndrome**

Down syndrome is the most common autosomal abnormality in humans, with an incidence of 1 in every 800–1200 live births [22]. DS is not only characterized by classic phenotypic physical features, but also by its behavioral and cognitive profile including: intellectual impairment, other cognitive deficits (primarily in speech, language production, and auditory short-term memory) and difficulties in adaptive function [23]. Individuals with DS between 1.08 and 11.5 years age may present better adaptive behavior performance in social skills than in the daily living and communication domains [24]. In addition, significant effects of IQ level were observed on adaptive behavior in most functional skill areas such as communication, community use, functional academics, home living, health and safety, self-direction, social skills, and overall adaptive behavior score in young adults with DS. Participants with a higher IQ showed better outcomes in adaptive behavior and thus better competence in daily living [25].

#### **Autism spectrum disorder**

Autism spectrum disorder is an early-onset neurodevelopmental disorder whose prevalence is estimated to be 11.3 per 1000 [26]. Communication and socialization deficits are common features of individuals with ASD, who tend to respond inappropriately in conversations, to misread nonverbal interactions, and to exhibit impaired ability in building age-appropriate friendships, as well as usually being. Overly dependent on routine, highly sensitive to changes in their environment, or intensely focused on inappropriate items [27]. Intellectual disability may be one of the comorbidities in ASD [28]. Thus, measuring intellectual functioning allows differentiation between high- and low-functioning individuals. Furthermore, adaptive functioning positively correlates with intellectual profile, especially in the communication domain in ASD [29]. People with ASD between ages 4–23 years tend to demonstrate a better performance in communication and daily living, but a weaker performance in the socialization domain [30, 31]. A study comparing 40 high-functioning individuals with ASD with 30 healthy controls, both between 12 and 21 years of age, revealed global adaptive behavior deficits in ASD, with particularly prominent social skills impairments [32].

As the current emphasis of healthcare is on functional outcome, more information is needed regarding the various factors contributing to an individual's real life functioning and adaptive behavior [33]. Although the literature points towards a specific adaptive behavior profile for each disorder, there is no study comparing the adaptive behavior profiles among WBS, DS and ASD in order to verify the specificity of each profile. Several studies have shown that cognitive functions predict adaptive behavior performance [34–36]. The main objective of the present study was therefore to verify whether adaptive behavior profiles differ across diagnostic groups (WBS, DS, ASD), as compared with healthy controls and with each other.

A secondary objective was to explore the potential relationship between the conditions and the individuals' socioeconomic status, and the effect on adaptive behavior profiles. Previous studies have demonstrated a strong relationship between socioeconomic status and expressive communication during preschool years through third grade [37, 38]. Parental behavior can also be affected by socioeconomic status, consequently, there is effect on children. LeVine suggested that parental behavior is adapted to socioeconomic and demographic conditions [39]. However, in respect of WBS a study by Hahn et al. [20] found that income or maternal level of education did not influence performance in the communication domain or expressive communication subdomain in WBS or in a developmental disabilities group, with the exception of a statistically significant association between expressive communication and maternal level of education in the developmental disabilities group [20].

## Methods

### Participants

One hundred and five adolescents aged 11–16 years old and resident in the State of Sao Paulo, Brazil, were recruited. The sample comprised four groups: (1) 22 adolescents with WBS, (2) 22 adolescents with DS, (3) 37 adolescents with ASD, and (4) 24 healthy controls (HC).

Adolescents with WBS were recruited from the Brazilian Association of Williams–Beuren Syndrome and all individuals presented diagnostic confirmation by cytogenetic analysis by Fluorescence in situ Hybridization (FISH). Of the 28 individuals with WBS registered with the Brazilian Association of Williams–Beuren Syndrome (aged 11–16 years), 24 individuals agreed to participate in this study. One participant with WBS was excluded due to unfinished questionnaires, and socioeconomic class data were missing for another individual.

Individuals with DS were recruited from the Association of Parents and Friends of Exceptional Children, a non-profit organization that provides social services

to people with intellectual disability. The diagnosis of DS was confirmed by examining the karyotype in all individuals. One case was excluded due to comorbidity with ASD and another due to missing data.

Adolescents with ASD were recruited from the specialized clinic in ASD of the Child and Adolescent Psychiatric Unit at the Department of Psychiatry of the Federal University of Sao Paulo (UNIFESP) and from two psychosocial care centers for children and adolescents. Psychosocial care centers for children and adolescents are the main centers for the diagnosis and treatment of children and adolescents with ASD in Brazil. Specialized and experienced professionals in the field carried out the diagnosis of ASD in accordance with the DSM-IV diagnostic criteria, as these assessments took place between 2011 and 2013. One participant with ASD was excluded because of missing data.

The HC group comprised brothers, cousins and friends of the participants with WBS and this group were recruited from events organized by the Brazilian Association of Williams–Beuren Syndrome. The parents or legal guardians of all participants signed informed consent forms, as did the adolescents. The study was approved by UNIFESP's Research Ethics Committee.

### Instruments

We used the Vineland Adaptive Behavior Scales (VABS) to measure adaptive behavior. VABS evaluates the ability of individuals to cope with environmental changes, learn everyday skills, and demonstrate independence [2]. The scale is based on a structured interview, in which adaptive behavior information is obtained from a significant caregiver. Completion time was approximately 25–90 min. VABS is organized in a structure with three main domains: communication, daily living, and socialization. The raw scores obtained from the domains were weighted to adjust for chronological age, according to the VABS manual, and standardized to obtain a common metric [2]. VABS results can determine strengths and weaknesses in specific adaptive behavior areas, and the scale has extensive representative normative data as well as strong psychometric properties [2].

To estimate IQ we used the Wechsler Intelligence Scale for Children—Third Edition (WISC-III). This instrument evaluates children between 6 and 16 years of age [40]. We estimated IQ by using the weighted sum of the subtests Cubes and Vocabulary [41].

The Critério de Classificação Econômica Brasil (*Brazil Economic Classification Criteria*) developed by the Associação Brasileira de Empresas de Pesquisa [ABEP] (2011) [42] was used to measure socioeconomic status. The classification estimates the income of Brazilian families living in urban areas by evaluating their consumption

of durable goods and also assesses their educational level. The socioeconomic classes range from A (highest income) to E (lowest income) [42].

**Statistical analysis**

To describe the sample’s characteristics, the frequency of participants’ responses for each categorical variable was calculated. Continuous variables were described based on measures of central tendency (mean) and dispersion (standard deviation).

Mann–Whitney Wilcoxon post hoc test was used to adjust *p* values when bivariate comparisons of continuous variables were performed, as these variables had a non-normal distribution.

When a comparison between two diagnostic groups resulted in a statistically significant difference in any of the clinical scales, we ran a multivariate linear logistic regression model to control for the effect of demographic characteristics and IQ as potential confounders.

These comparisons resulted in three multivariate linear logistic regression models, in which the following pairs of diagnostic groups were compared: (1) WBS vs. DS, (2) WBS vs. ASD, and (3) DS vs. ADS. We repeated these three models for each of the following outcomes: (1) VABS total score, (2) VABS communication domain, (3) VABS socialization domain, and (4) VABS daily living domain.

Finally, we tested for interactions between diagnoses and socioeconomic classes through linear logistic regression models, controlling for IQ. The Vineland dimension (communication, socialization, and daily living) was defined as a dependent variable, whereas diagnosis, in interaction with economic classes, was entered as an independent variable.

In the linear logistic regression models, HC from the A/B classes were regarded as the reference category. For each diagnosis, when the difference between the A/B and C/D classes (e.g., WBS A/B classes vs. WBS C/B classes) was higher than 20%, we considered that this indicated an interaction between diagnosis and economic class, meaning that differences in performance between A/B and C/D classes were clinically significant. We arbitrarily established 20% as a parameter to define the interaction between diagnosis and economic classes, as this can be considered a clinically significant difference in adaptive behavior performance. Among many statisticians and epidemiologists, it is acceptable to set an “arbitrary” parameter such as this when the literature does not provide established parameters [43, 44].

The level of significance was set at *p* < .05. Effect sizes (Cohen’s *d*) were calculated as the difference between means divided by a pooled standard deviation (difference between HC and WBS, DS, and ASD groups). An effect

size of ≤.2 indicates a small change, between .2 and .5 a moderate change, and an effect size ≥.8 a large clinical change [45].

**Results**

**Sample characteristics**

There were no significant differences in age or socioeconomic class between diagnostic groups (WBS, DS, and ASD) and HC (*p* > .05). However, there was a significant difference in IQ between the HC and the diagnostic groups. For additional details see Table 1. The only significant gender difference was between the ASD and HC groups,  $\chi^2(1, N = 62) = 21.64, p < .001$ .

In a second analysis, we compared the WBS, DS, and ASD groups to each other. ASD had a significantly higher proportion of males compared to WBS,  $\chi^2(1, N = 62) = 11.12, p < .001$ . The proportion of males in the ASD group was also significantly higher than in the DS group,  $\chi^2(1, N = 62) = 18.92, p < .001$ . The WBS group had a significantly higher IQ than the DS group [ $z(43) = 3.10, p = .002$ ]. ASD group average IQ was also significantly higher than that of the DS,  $t(58) = -4.35, p < .001$ , and WBS group  $t(59) = -2.90, p = .004$ .

**Differences in adaptive behavior domains between groups**

All three diagnostic groups (WS, DS, and ASD) presented significantly lower scores in all VABS domains and total scores as compared to the HC group. Furthermore, the effect sizes were classified as moderate or large regarding adaptive behavior when we compared the HC group with each diagnostic group (WBS, DS, and ASD) (Table 2).

Differences between diagnostic groups (WBS, DS, and ASD) in relation to adaptive behavior performance are described in Table 3.

In multivariate linear logistic regression models, the WBS group still performed better than the DS group in the communication domain [ $\beta = -15.08, t(-3.45), p = .005$ ] and remained better than the ASD group in the

**Table 1 Sociodemographic characteristics of adolescents with WBS, DS, ASD compared to the HC group**

Groups	Gender	Economic class		Age	WISC (IQ)
	Male <i>n</i> (%)	A/B <i>n</i> (%)	C/D <i>n</i> (%)	Mean (SD)	Mean (SD)
HC	10 (42)	13 (54)	11 (45)	13.54 (1.84)	105.62 (16.77)
WBS	14 (61)	9 (41)	13 (59)	13.60 (1.90)	59.39 (9.07)**
DS	10 (45)	11 (50)	11 (50)	13.31 (1.49)	51.22 (3.62)**
ASD	36 (95)**	24 (63)	14 (36)	13.50 (1.62)	84.42 (28.98)*

HC healthy controls, WBS Williams–Beuren syndrome, DS Down syndrome, ASD autism spectrum disorder, WISC Wechsler Intelligence Scale for Children, IQ intelligence quotient

\* *p* < .05; \*\* *p* < .01

**Table 2 Effect size of adaptive behavior means for HC, WBS, DS, and ASD groups**

	1 HC	2 WBS	3 DS	4 ASD	HC vs. WBS Cohen's d/effect size r (p)	HC vs. DS Cohen's d/effect size r (p)	HC vs. ASD Cohen's d/effect size r (p)
	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)			
Socialization	112.45 (17.40)	60.30 (26.09)	55.86 (17.63)	56.52 (29.28)	2.35/.76 <sup>a</sup> (p < .001)	3.22/.85 <sup>a</sup> (p < .001)	2.32/.75 <sup>a</sup> (p < .001)
Communication	103.66 (13.57)	40.82 (18.95)	25.50 (7.46)	58.97 (28.51)	3.82/.88 <sup>b</sup> (p < .001)	7.17/.96 <sup>b</sup> (p < .001)	2.00/.70 <sup>a</sup> (p < .001)
Daily living	103.33 (18.89)	33.91 (15.57)	27.95 (11.79)	53.13 (30.74)	4.06/.89 <sup>b</sup> (p < .001)	4.81/.92 <sup>b</sup> (p < .001)	1.97/.70 <sup>a</sup> (p < .001)
Total VABS	108.62 (16.42)	41.39 (15.39)	32.54 (8.86)	53.63 (26.30)	3.60/.87 <sup>b</sup> (p < .001)	5.76/.94 <sup>b</sup> (p < .001)	2.50/.78 <sup>a</sup> (p < .001)

Socialization, communication, daily living skills of Vineland scale; total of Vineland scale (total VABS)

HC healthy controls, WBS Williams–Beuren syndrome, DS Down syndrome, ASD autism spectrum disorder

<sup>a</sup> Medium effect size

<sup>b</sup> Large effect size

**Table 3 Comparing adaptive behavior between diagnostic groups (WBS, DS, and ASD)**

	Total VABS Mean (SD)	Daily living Mean (SD)	Socialization Mean (SD)	Communication Mean (SD)
WBS	41.39 (15.39)*	33.91 (15.57)	60.3 (26.09)	40.82 (18.95)**
DS	32.54 (8.86)	27.95 (11.79)	56.52 (29.28)	25.5 (7.46)
WBS	41.39 (15.39)	33.91 (15.57)*	60.30 (26.09)	40.82 (18.95)*
ASD	53.63 (26.30)	53.13 (30.74)	56.52 (29.28)	58.97 (28.51)
DS	32.54 (8.86)*	27.95 (11.79)*	55.86 (17.63)	25.50 (7.46)**
ASD	53.63 (26.30)	53.13 (30.74)	56.5 (29.3)	58.97 (28.51)

Socialization, communication, daily living skills of Vineland scale; total of Vineland scale (total VABS)

WBS Williams–Beuren syndrome, DS Down syndrome, ASD autism spectrum disorder

\* p < .05; \*\* p < .01

socialization domain [ $\beta = 8.92, t(-2.08), p = .013$ ]. The DS group also performed better than the ASD group in the socialization domain [ $\beta = 16.98, t(-2.32), p = .024$ ]. Differences between groups lost statistical significance after IQ was controlled for, meaning that IQ acted as a confounding factor (Table 4).

**Association between adaptive behavior and conditions in relation to socioeconomic class while controlling for IQ**

Adolescents with delayed development (WBS, DS, and ASD) showed significant lower adaptive behavior scores when compared with HC from A/B economic classes in the bivariate analyses. The statistical differences were significant at  $p < .001$  for all comparisons between groups. However, when controlling for IQ, these differences remained only in a few dependent variables as shown in Table 5.

When differences between A/B and C/D classes were higher than 20%, we considered that there was an interaction between adaptive behavior and economic classes.

The following interactions occurred: (1) adolescents with WBS from the C/D classes had scores that were 83.8% lower than those from the A/B classes in communication; 88.5% in socialization; and 26.9% in daily living skills. (2) adolescents with DS from C/D classes had scores that were 34.4% lower than those from A/B classes in the socialization domain, and 21.8% lower in daily living skills. (3) adolescents with ASD from C/D classes had scores that were 44.3% lower than those from A/B classes in communication, 34.1% in socialization, and 24.2% in daily living skills.

**Discussion**

The findings of this study suggest that there are distinct adaptive behavior profiles in adolescents with WBS, DS, and ASD. Individuals with WBS had better adaptive behavior skills than individuals with DS, especially in communication, as well as better socialization skills than individuals with ASD. Furthermore, the DS group performed better than the ASD group in the socialization domain. These results remained significant after adjusting for IQ, since the groups were not matched for this variable when selected.

In spite of the limitations in the socialization and communication domains in participants with WBS when compared to the HC group, they still performed better in socialization compared to individuals with ASD, and in communication when compared to individuals with DS. This outcome is consistent with the literature with respect to adolescents and adults with WBS who showed satisfactory performance in social interaction [17, 18] and communication skills and poor performance in the remaining adaptive behavior domains [21]. One study showed that the performance of adolescents with WBS was better than that of adolescents with DS in all VABS domains [34], which is in agreement with the results presented in this study. It is reasonable to suggest that although

**Table 4 Multivariate logistic regression models comparing adaptive behavior between groups adjusting for demographics and IQ**

Groups	Coef. (95% CI)
WBS vs. DS	
Socialization	4.25 (−10.97 to 19.47)
Communication	−10.61 (−20.94 to −.28)*
Daily living	−.37 (−10.22 to 9.44)
Total VABS	−2.98 (−11.32 to 5.35)
Socialization	4.25 (−10.97 to 19.47)
WBS vs. ASD	
Socialization	−8.92 (−15.84 to −1.99)*
Communication	−1.14 (−6.72 to 4.44)
Daily living	1.80 (−4.39 to 8.01)
Total VABS	−1.83 (−6.72 to 3.04)
DS vs. ASD	
Socialization	−16.97 (−31.61 to −2.33)*
Communication	7.48 (−2.97 to 17.94)
Daily living	2.83 (−11.13 to 16.80)
Total VABS	−.19 (−10.70 to 10.30)

Socialization, communication, daily living skills of Vineland scale; total of Vineland scale (total VABS); demographics (gender, age, and economic class); intelligence quotient (IQ)

Coef. confidence interval, WBS Williams–Beuren syndrome, DS Down syndrome, ASD autism spectrum disorder

\*  $p < .05$

individuals with WBS within the age range of this study, 7.67–46.41 years of age have an interest in interpersonal relationships (socialization), they still have difficulty performing this skill to the same level as HC [46].

Thus, we suggest that the adaptive behavior profile for WBS, in terms of strongest to weakest performance in the three domains, would be in the order: socialization, communication, daily living skills. Our results agree with

the personality description of individuals with WBS as “hypersocial” [47].

Our results regarding the adaptive behavior profile of the DS group corroborate other studies that showed discrete differences between domains but a better performance in the socialization domain [34, 48]. Moreover, the DS group showed better performance in the socialization domain than the ASD group, but a worse performance in the communication domain when compared to individuals with WBS. Our results therefore agree with the pattern suggested by other studies that found relatively stronger social skills but weaker communication in individuals with DS between ages 1–11.5 years [9, 24]. DS acquire their adaptive skills ceiling scores at about the age of 12 years, furthermore, presented lower performance than HC children in 65% approximately [49]. A review study identified that the main difficulties in the DS cognitive profile are in communication, such as: auditory-verbal processing and short-term memory, expressive language, grammar, and pronunciation [7]. Therefore, the adaptive behavior profile for DS in this study would be, in terms of strongest to weakest domains: socialization, daily living skills, communication.

Previous adaptive behavior findings in children, adolescents and young adults with ASD showed significant deficits in the socialization domain, compared to a relative ability in the daily living skills and communication domains [30, 50, 51]. Thus, some results described in the literature corroborate with the clinical definition of ASD, which is characterized by impairment in social development. Accordingly, in our study the WBS and DS groups showed better performance in the socialization domain than the ASD group, after controlling for IQ. The adaptive behavior profile for ASD in this study would be, in terms of strongest to weakest domains: Daily Living Skills > Communication > Socialization.

**Table 5 Linear logistic regression testing for interaction between diagnoses and economic classes, controlling for IQ**

	Communication Coef. (95% CI)	Socialization Coef. (95% CI)	Daily living Coef. (95% CI)
HC + A/B	–	–	–
HC + C/D	17.78 (.38 to 35.17)	−1.05 (−19.71 to 17.60)	21.49 (3.26 to 39.73)
WBS + A/B	−17.34 (−38.70 to 4.01)	−7.84 (−30.75 to 15.06)	−16.00 (−38.40 to 6.38)
WBS + C/D	−31.80 (−52.04 to −11.57)*	−14.72 (−36.42 to 6.97)	−20.39 (−41.61 to .81)
DS + A/B	−38.39 (−59.58 to −17.19)*	−16.05 (−38.78 to 6.68)	−30.37 (−52.59 to −8.15)*
DS + C/D	−35.79 (−57.56 to −14.01)*	−21.59 (−44.95 to 1.76)	−23.78 (−46.61 to −.95)
ASD + A/B	−20.15 (−35.35 to −4.95)	−31.74 (−48.04 to −15.44)**	−19.89 (−35.83 to −3.96)
ASD + C/D	−29.04 (−46.89 to −11.20)*	−42.52 (−61.66 to −23.38)**	−24.61 (−43.32 to −5.90)

HC healthy controls, WBS Williams–Beuren syndrome, DS Down syndrome, ASD autism spectrum disorder, A/B A and B economic classes, C/D C and D economic classes

\*  $p < .05$ ; \*\*  $p < .01$

Subjects with WBS, DS, and ASD presented lower adaptive behavior performance when compared to the HC group. These difficulties in functional performance in all assessed diagnostic groups in this study are likely associated with limitations derived from the disorder itself. For some adaptive behavior difficulties, IQ might play an important role in adolescents with WBS, DS, and ASD, as previous studies have observed a positive correlation between adaptive behavior and IQ in ASD [31, 52–54], DS [25], and WBS [19]. Therefore, the main differences in terms of performance in adaptive behavior between the groups in this study can only be observed when taking into account IQ. For this reason, we believe it is essential describe the influence of intellectual disability in adaptive behavior performance.

Finally, the result of the analysis identified possible associations between adaptive behavior profiles and socioeconomic class, suggesting that being in the C/D classes might be related to increased adaptive behavior impairment. This result corroborates the theory that parental behavior is adapted by socioeconomic and demographic conditions with a subsequent effect on their children [39], with previous studies demonstrating a strong relationship between socioeconomic status and expressive communication during preschool years through third grade [37, 38]. A study evaluating the risk and protective factors of parental well-being compared parents of children with intellectual disability with control children and found that differences in economic hardship and self-rated health were the strongest predictors for well-being [55]. Thus, Olsson and Hwang [55] confirmed the importance of the relationship between socioeconomic status and the well-being of parents of children with intellectual disability.

As there are no parameters in the literature that define a significant difference in the Vineland scores, a difference of 20% was chosen as being clinically significant based on our clinical experience. The proposed 20% can be a good starting point to demonstrate the relationship between socioeconomic class and the condition, and the effect on adaptive behavior. Nonetheless, it would be worthwhile conducting more specific studies to test this parameter, which, if confirmed, possibly could indicate how interventions that improve families' socioeconomic conditions (i.e., the income and education level) most likely could help to alleviate the effects of genetic syndromes on adaptive behavior.

When interpreting the results of our study, some limitations should be considered. First, the sample size was defined based on the available number of adolescents with WBS, which is a rare syndrome. Despite this limitation, our statistical models were able to demonstrate differences between the groups that were consistent with

our hypotheses and with the literature. Second, only two subscales of the WISC-III were used to estimate IQ. Even though it has been shown that this approach provides scores that are similar to those obtained through the full WISC-III, it does not include other dimensions of intellectual function that may be related with adaptive behavior. The third limitation was not having collected information such as type of school (mainstream or specialist school), other co-morbid or medical conditions and maternal education. The final limitation was the gender differences between the ASD group and the other groups. However, our multivariate analyses showed that gender did not play a significant role as a potential predictor of adaptive behavior in our sample, although females with ASD tend to be somewhat more functional than males in relation to adaptive behavior [56]. Future studies should explore in more depth whether adaptive behavior profiles also differ between male and female individuals with ASD.

## Conclusion

Ours results can help devise intervention strategies that optimize developmental independence, family support, and community participation among adolescents with neurodevelopmental disabilities. Further research is necessary to measure the effectiveness of intervention strategies based on the adaptive behavior profiles for each specific condition.

Furthermore, our results also indicate that socioeconomic class has an important effect on adaptive behavior in adolescents with genetic syndromes and ASD. The possible effects of socioeconomic status; parental schooling; levels of family, health and educational support which affect adaptive behavior warrant further investigation.

## Authors' contributions

DCCG performed the data collection and wrote the manuscript; CSC contributed to the design of the research project, data collection and manuscript revision; RW performed the statistical analyzes in the study and revised the manuscript; KAM assisted in the analysis of the results and in their description; JAP oversaw the research project and reviewed the manuscript. All authors read and approved the final manuscript.

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#### Competing interests

The authors declare that they have no competing interests.

#### Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request. Furthermore, all data generated or analyzed during this study are included in this published article.

#### Consent for publication

Both legal guardians and adolescents signed informed consent in a form approved by the Research Ethics Committee of Federal University of São Paulo.

#### Ethics approval and consent to participate

This study was approved by the Research Ethics Committee of Federal University of São Paulo (297/10).

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