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HAND PREFERENCE IN THREE GENETIC SYNDROMES ASSOCIATED WITH INTELLECTUAL DISABILITY

George Kosmatos, Ioulia Nisiotou, Panayiota Stavroussi, Filippos Vlachos
University of Thessaly, Department of Special Education

ABSTRACT*: Research findings about handedness in sydromes associated with intellectual disability are ambiguous. We investigated the frequency of handedness in three syndromes associated with intellectual disability. A total of 80 age and gendre matched children and adolescents, (20 with Down syndrome, 20 with Sotos syndrome, 20 with Williams syndrome and 20 with typical development), participated in this study. Handedness was defined according to the Edinburgh Handedness Inventory and α classification we developed representing levels of handedness. Using the Laterality Quotients calculated for each group, we didn't find significant differences between these syndromes. However, our data indicate that atypical laterality between the three syndromes does not occur at the same rate, suggesting the existence of specific patterns of laterality in each syndrome, probably related to the underlying genetic basis*.

KEYWORDS: genetic syndromes, hand preference, down syndrome, sotos, syndrome, williams syndrome, intellectual disability.

INTRODUCTION

According to DSM-5, intellectual disability (ID) is defined as a neurodevelopmental disorder which involves significant limitations in both cognitive and adaptive functioning with an onset during the developmental period (American Psychiatric Association, 2013, Schalock, Luckasson & Tassé, 2019, 2021). The subgroup classification systems put an emphasis on adaptive functioning and intensity of support needs, rather than IQ scores (Papazoglou et al., 2014; Schalock et al., 2021). Schalock and his colleagues refer to a "multidimensional approach to subgroup classification" that considers adaptive behavior, support needs, and/or intellectual functioning (Schalock et al., 2021, p. 32-33).

Intellectual Disability is characterized by great heterogeneity and its etiology includes genetic and non-genetic factors. Within this context, there is an evolving research direction that focuses on etiology-specific conceptualizations on genetic syndromes associated with intellectual disability (Abbeduto et al., 2019; Hodapp, 2021). About 25% of people with ID have been diagnosed with a chromosomal abnormality. Chromosomal disorders occur in about 30% of known genetic causes, with Down syndrome (DS) being the most common with a prevalence of 22%. Other well-known genetic causes of ID are Fragile X Syndrome (FXS), Prader-Willi Syndrome, Williams Syndrome (WS) and Sotos Syndrome (SS) (Moser, 2004).

The most extensively researched indicator for laterality is handedness – a valid but coarse indicator (Ocklenburg et al., 2014). Several studies have investigated laterality

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in individuals with ID. Pickersgill and Pank (1970), in one of the first studies of hand preference in people with Down syndrome, showed higher incidence of left-handedness in non-DS people with ID (31%) than people with DS (18.7 %) and typically developing individuals (15.6%). A similar study showed a small difference between the two groups (DS: 27%, ID without DS: 29%) and a large difference between the above and the control group (11%) (Batheja & McManus, 1985). Vlachos and Karapetsas (1999) assessed hand preference in 7-9 years old and 13-15years old pupils with Down's syndrome (n=41) as compared with control schoolchildren (n=50) of the same ages. They found an increased incidence of left-handedness and mixed handedness in Down syndrome pupils compared to a typical developing population. Additionally, they report that younger Down syndrome pupils were less consistent in their hand preferences compared to the older ones and the normal controls. From the above it is clear that there are discrepancies between the findings of such studies, regarding the differences in preference between people with DS and typically developing people.

A recent meta-analysis indicated that individuals with intellectual disability were up to 166% more likely to be non-right-handed, and up to 98% more likely to be left-handed than typically developing individuals, suggesting that left-handedness is related to lower intellectual functioning (Papadatou-Pastou & Tomprou, 2015). However, as the studies investigating the relationship between handedness and intelligence rely on correlational evidence, causation cannot be determined. Thus, studies of hand preference in different genetic syndromes could be useful in identifying such causative agents (Niort & Hernández Vázquez, 2017).

Atypical handedness has been described in persons with developmental disorders or intellectual disability, with an increased frequence of non-right handedness in neurodevelopmental disorders (Bishop 1990 for a review; Leconte & Fagard 2006). Although several studies investigated hand preference in people with ID without a clear genetic etiology or in people with DS, limited research has been done in people with other genetic syndromes and only a few of them compare hand preference patterns between syndromes. In a relevant study, Van Strien et al. (2005) found an increased incidence of left-handedness in people with WS (26%). The differences were more pronounced in younger people with WS (5-15 years) than older people and in men than in women. The results were attributed to the slow brain maturation in individuals with WS, as previously suggested by Bishop (1990). Carlier and colleagues (2006) examined 45 children with DS and 34 with WS (mean age 13 years) and 85 typical developing children. They found that in the DS group there are more left-handed people compared to the other two groups, while the instability in the preference was higher in people with genetic syndromes than in the control group. The above researchers concluded that atypical laterality patern is different within the two syndromes, suggesting the existence of specific laterality paterns probably related to specific genotypes.

Comparative investigation of non-right handedness in people with different genetic syndromes could provide evidence for the impact of specific genetic basis on the development of handedness and its functional consequences. Children with genetic

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syndromes differ in clinical features and present difficulties in various areas during development.

In this study we suggest considering handedness, an index of brain laterality, as one of the developmental characteristics of each syndrome apart. The study aimed to assess hand preference of children and adolescents with three genetic syndromes (DS, WS and SS) associated with intellectual disability. Based on previous studies (Batheja & McManus, 1985, Carlier, et al., 2006, Van Strien et al., 2005, Vlachos & Karapetsas, 1999), we hypothesized that the incidence of non- right handedness within the three genetic syndromes will be higher compared to that found in typically developing individuals of the same age (1st hypothesis). In addition, the suggestion that atypical laterality is not the same between syndromes (Carlier, et al., 2006) led us to suppose that the incidence of non-right handedness will vary between the DS, WS, and SS groups (2nd hypothesis), reflecting different genetic defects.

METHODS

Participants

A total of 80 children and adolescents (44 boys and 36 girls, age range 5–16 years) participated in this study. We set up three groups of children with genetic syndromes (Down: N = 20; age range 5–16 years, M = 10,1 years, Sotos: N = 20; age range 5–16 years, M = 10,8 years, Williams: N = 20; age range 5–16 years, M = 10,5 years), which had a diagnosis of ID after assessment at state hospitals. The assessment was carried out by a psychologist and a special educator, and the criteria used included: (a) assessment of intelligence, (b) assessment of cognitive skills (i.e. visual discrimination, visual and auditory short-term memory, spatial orientation, laterality, etc.), (c) estimation of adaptive operation. Additionally, we set up a control group which comprised 20 typically developing participants (TD), which were matched according to age and gender with ID groups (1 DS /1 SS / 1 WS / 1TD). Greek was the first language of all participants. Ethical Approval for this research was provided by the University of Thessaly Ethics Committee.

Measures

Handedness was defined according to the Edinburgh Handedness Inventory (EHI; Oldfield, 1971), a reliable and well-validated instrument (Bryden, 1977). Numerous studies have confirmed the objectivity and reliability of EHI using other handedness questionnaires (e.g., Dorthe, Blumenthal, Jason & Lantz, 1995; McMeekan & Lishman, 1975; Ransil & Schachter, 1994; Williams, 1986). Test-Retest reliability of the EHI as measured by the Pearson r, Kendall τ, and Spearman r_s, ranged from 0.95 to 0.98 (Ransil & Schachter, 1994). The medium to high correlations of the EHI to other behavioral measures of handedness (for example the Purdue Pegboard test, a test of manual dexterity) underline the high concurrent validity of the test (Raczkowski, Kalat & Nebes, 1974; Triggs, Calvanio, Levine, Heaton & Heilman, 2000; Verdino &

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Dingman, 1998). The questionnaire comprises 10 items pertaining to hand preference in writing, drawing, throwing a ball, use of scissors, toothbrush, knife (without fork), spoon, broom (upper hand), striking a match and opening a box.

Procedure

All participants were examined separately on the 10 items of the EHI (Oldfield, 1971). To avoid misunderstanding due to possible reading deficiency, the examiner told the subject that some familiar activities were going to be named and the subject was to demonstrate how he or she ordinarily performed them. Each item was preceded by the phrase 'Show me how you …' and the examiner wrote down the answer on the questionnaire. Responses were scored 'right' or 'left' based on the hand used to demonstrate the activity.

Based on these responses we calculated a Laterality Quotient [LQ = (Right – Left)/(Right + Left)×100)] for each participant, which resulted in a score ranging from –100 to +100, where –100 indicated pure left-handedness and +100 indicated pure right-handedness. We classified respondents in three groups: children and adolescents who scored +50 to +100 were considered as right-handers (an EHI score +50 to +100), children and adolescents who scored -49 to +49 were considered as mixed-handers and those who scored -50 to -100 were considered as left-handers.

RESULTS

The mean EHI scores (LQ) were 60.00 (*SD* = 82.07) for the Williams group, 82.50 (*SD* = 43.99) for the Sotos group, for the Down group 72.00 (*SD* = 55.87), and for the control group 71.50 (*SD* = 60.19). The ANOVA test was used with EHI scores as a dependent variable and groups (three syndromes and control groups) as independent variables. The analysis revealed that the mean EHI score difference among the four groups was not statistically significant [F (3.76) = 0.44, and $p > 0.05$], indicating that the groups did not present significant differences in terms of lateral quotient scores.

Figure 1 shows the distribution of scores on the EHI for the three groups of genetic syndromes and the typical developing participants. As it is typical with handedness preference inventories, the distribution was J- shaped, with most children being right-handed in all groups.

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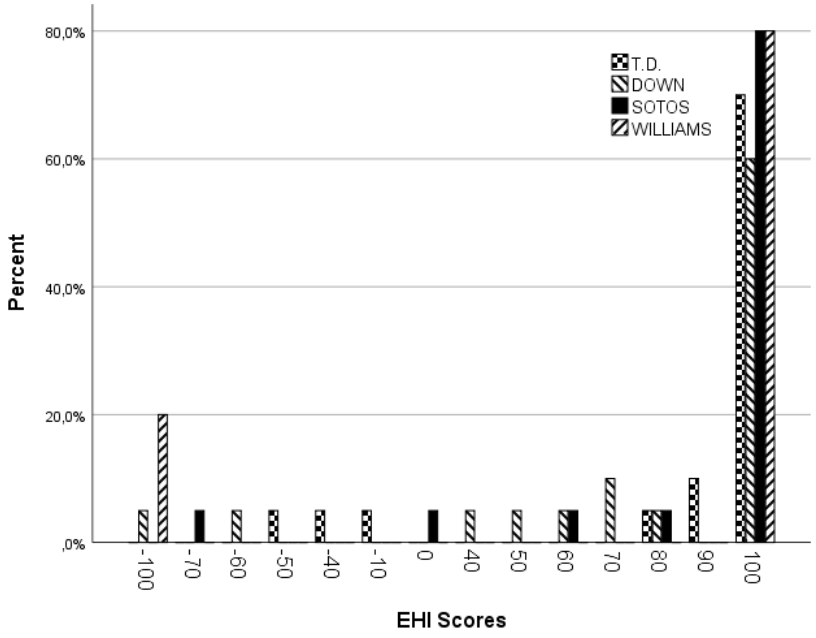

Figure 1. Percentages of EHI scores within the four groups of participants.

Table 1 shows the classification of our study population according to hand-preference, as it was mentioned above. As it can be seen in this Table, when participants were divided in three groups of right-handers (EHI +50 to +100), mixed-handers (EHI -49 to +49) and left-handers (EHI -100 to -50), there was a higher percentage of left-handedness in Williams syndrome group (20%) than in Down syndrome group (10%), TD children (10%) and Sotos syndrome group (5%). This difference was statistically significant ($\chi^2 = 8.36$, $df = 3$, $p = 0.04$), indicating small differences of hand preference between these three groups of genetic syndromes associated with intellectual disability.

Table 1. Percentages (%) of handedness classification within the four groups of participants

Groups	*Handedness classification*		
	Right-handers	*Mixed-handers*	*Left-handers*
Typical Development	85	5	10
Down Syndrome	80	10	10
Williams Syndrome	80	0	20
Sotos Syndrome	85	10	5

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DISCUSSION

In this study, we assessed hand preference in children and adolescents with one of three genetic syndromes (DS, WS and SS) associated with intellectual disability and a group of TD children. Our first hypothesis predicted that the incidence of non-right handedness in these three syndromes will be higher, compared to typically developing individuals. Based on the Laterality Quotients calculated for each group, we didn't find significant differences. Therefore, data of this study do not provide empirical support for this account. Such a result was not expected, given that literature and clinical experience report increased frequency of non-right handedness in people with ID compared to TD individuals (Papadatou-Pastou & Tomprou, 2015). The reason for this discrepancy may be linked to the fact that most studies have not used the Laterality Quotients comparisons, but they have compared the classification of handedness. In addition, various laterality measures have been used to assess hand preference in individuals with ID; variability in the degree of intellectual disability may also have influenced their results.

Our second hypothesis predicted that the incidence of non-right handedness would be different between the DS, WS and SS groups. We found a higher percentage of left-handedness in children with Williams syndrome (20%) versus children with Down syndrome (10%) and children with Sotos syndrome (5%). This finding is in line with the previous study of Van Strien et al. (2005) which found an increased incidence of left-handedness in people with WS (26%). Additionally, our data agree with the Carlier and colleagues (2006) assumption that the laterality pattern is not the same among different genetic syndromes. They found a higher frequency of left-handness in individuals with DS than in those with WS. Taken together, these data and observations suggest that different genes implicated in sydromes associated with intellectual disability may be involved in various handedness profiles usually observed in these populations.

Overall, our data based on handedness group classifications indicate that atypical laterality is not identical between these three genetic syndromes, probably due to specific patterns of laterality related to the molecular basis of each syndrome. Given the limited sample of our study, larger sample size and extension to other genetic syndromes may be worthwhile.

Our results provide support to recent studies which suggest genetic relationship between handedness and neurodevelopmental disorders and shows that detailed phenotyping will permit DNA genotyping to unravel the full complexity of handedness in neurodevelopmental disorders (Brandler & Paracchini, 2014). DNA studies are increasingly affordable nowadays and should be used in investigating chromosomal syndromes. They will contribute to the identification of genetic factors involved in brain lateralization and answer the question whether atypical asymmetry is a cause or a consequence of neurodevelopmental disorders. As Fidler & Nadel (2007) suggested, understanding the factors that may be associated with the specific features of known

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genetic syndromes through comparative research between them could contribute to the organization of more appropriate support programs and interventions.

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