Sensory Processing in Angelman Syndrome

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Aims

1) Explore sensory processing in Angelman syndrome (AS), including patterns of hypo and hyper-responsivity to sensory input and sensory seeking behaviours

2) Compare sensory processing across AS and Cornelia de Lange syndrome (CdLS) and fragile X syndrome (FXS), where unusual sensory processing has been reported.

Introduction

Angelman syndrome (AS)

• Rare genetic syndrome caused by missing information at the maternal chromosome 15q11-13 region.
• Clinical characteristics: severe intellectual disability, seizures, ataxic gate, hypopigmentation and abnormal EEG patterns (Boyd et al., 1988).
• Behavioural phenotype: short attention span (Walz & Benson, 2002), high levels of laughing and smiling (Horser & Oliver, 2006) and a fascination with water and crinkly objects (Didden et al., 2006).

Sensory processing in genetic syndromes

• Some genetic syndromes are associated with an unusual and distinct sensory profile, for example in Down syndrome (Wuang & Su, 2011)
• Understanding a syndrome's sensory profile can help with the design of syndrome sensitive interventions.

Sensory processing in AS

• Anecdotal evidence of unusual sensory interests including a preference for water-related items and shiny/reflective objects (Didden et al., 2006).
• Despite an initial suggestion of unusual sensory profile (Walz & Baranek, 2006) the literature on sensory processing in AS is limited.
• Aim of the current research was to explore sensory processing in AS in more details, and in comparison to individuals with fragile X (FXS) and Cornelia de Lange syndromes (CdLS) where unusual sensory processing has been reported.

Method

Participants

• 156 parents/carers of children aged 2-16 years with AS (N=86), fragile X syndrome (FXS; N=40) and Cornelia de Lange syndrome (CdLS; N=28).
• Participants were recruited from the database of families held at the Cerebra Centre for Neurodevelopmental Disorders and via online recruitment through syndrome support groups.

Measures

Sensory Experiences Questionnaire (SEQ; Baranek, 1999): The SEQ is a 42 item questionnaire which measures children's responses to everyday sensory events. Higher scores on the SEQ indicate greater degree in the frequency and nature of sensory processing difficulties. The SEQ is composed of several subscales including:

• Hypo-responsivity: under responsive to sensory input
• Hyper-responsivity: over responsive to sensory input
• Sensory seeking behaviours

Data analysis

1) Examine the scores from the AS group compared to SEQ norms: typically developing children (TD), children with a developmental disability (DD) and children with Autism using single sample t-tests.
2) Examine SEQ scores across the AS, CdLS, and FXS groups.

Results

Sensory processing in Angelman syndrome

Figure 1 shows the mean item score across three subscales of the SEQ for individuals with AS, CdLS, FXS, TD, DD and Autism.

1) Sensory processing in AS

Single sample t-tests between AS and TD, DD and Autism groups from the SEQ norms revealed:

• Significantly **higher levels of hypo-responsivity in AS** than TD and DD groups (TD: $t(137) = 7.45$, $p<0.01$; DD: $t(128) = 3.20$, $p<0.01$)
• Significantly **higher levels of hyper-responsivity** than TD group (TD: $t(137) = 5.95$, $p<0.01$)
• Significantly **higher levels of sensory seeking behaviours** across TD, DD and Autism groups. (TD: $t(137) = 8.03$, $p<0.01$; DD: $t(128) = 6.92$, $p<0.01$; Autism: $t(159) = 3.17$, $p<0.01$)

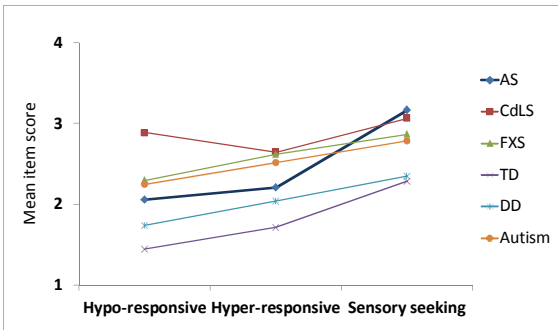

Figure 1: Mean SEQ item score across AS, CdLS, FXS, TD, DD and Autism groups.

2) Sensory processing in AS, CdLS and FXS

Kruskall Wallis tests revealed a significant effect of syndrome group on all subscale of the SEQ (Hypo: $X^2 = 25.18$, $p<0.01$; Hyper: $X^2= 14.37$, $p<0.01$; Seeking: $X^2=6.10$, $p<0.05$). Subsequent post hoc Mann Whitney U tests revealed:

• Significantly **lower levels of hypo-responsivity in AS** than FXS and CdLS (FXS: $Z = -1.99$, $p<0.05$; CdLS: $Z=-4.92$, $p<0.01$)
• Significantly **lower levels of hyper-responsivity in AS** than FXS and CdLS (FXS: $Z = -3.16$, $p<0.05$; CdLS: $Z=-2.89$, $p<0.01$)
• Significantly **higher levels of sensory seeking behaviours** in AS than FXS ($Z = -2.43$, $p<0.05$) and no significant difference in comparison to CdLS ($Z=-0.94$, $p =0.35$)

Conclusions

Unusual sensory processing in AS: Individuals with AS showed significantly higher score across all subscales in comparison to the TD and DD groups.

Differing patterns across syndrome groups: Although higher than typically developing children, the results suggests that levels of hypo and hyper-responsivity in AS were not as high as the FXS and CdLS groups.

Increased levels of sensory seeking behaviours: Individuals with AS showed significantly higher levels of sensory seeking behaviours in comparison to TD, DD, Autism, and FXS groups.

Impact on future interventions: The results suggest that individuals with AS may seek out sensory experiences, indicating a potential reinforcer for future behavioural intervention programmes.

References

Arron, K., Oliver, C., Berg, K., Moss, J., and Burbidge, C. (2011). Prevalence and phenomenology of self-injurious behaviour in genetic syndromes. *Journal of Intellectual Disability Research, 55,* 109-120.
Boyd, S. G., Harden, A., & Patton, M. A. (1988). The EEG in early diagnosis of the Angelman (happy puppet) syndrome. *European Journal of Paediatrics, 147,* 508-513.
Didden, R., Korzilius, H., Kamphuis, A. (2006). Preferences in individuals with Angelman syndrome assessed by a modified Choice Assessment Scale. *Journal of Intellectual Disability Research, 50,* 54–60.
Mount, R., Oliver, C., Berg, K., & Horsler, K. (2011). Effects of adult familiarity on social approach behaviours in Angelman syndrome. *Journal of Intellectual Disability Research, 55,* 339-350.
Oliver, C., Demetriades, L., & Hall, S. (2002). The effect of environmental events on smiling and laughing behavior in Angelman syndrome. *American Journal on Mental Retardation, 107,* 194-200.
Strachan, R., Shaw, R., Burrow, C., Horsler, K., Allen, D., & Oliver, C. (2009). Experimetnal functional analysis of aggression in children with Angelman syndrome. *Research in Developmental Disabilities, 30,* 1095-1106.
Tiger, J. H., Hanley, G. P. (2004). Developing stimulus control of preschooler mands: An analysis of schedule-correlated and contingency-specifying stimuli. *Journal of Applied Behavior Analysis, 37,* 517–521.
Walz, N. C. & Benson B. A. (2002). Behavioural phenotypes in children with Down syndrome, Prader-Willi syndrome, or Angelman syndrome. *Journal of Developmental and Physical Disabilities, 14,* 307-21.