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"Coming here, you will discover yourself, discovering others"
Romanian Prader-Willi Association

ASOCIACIÓN MADRILEÑA
PARA EL SÍNDROME DE
PRADER-WILLI



THE RELATIONSHIP BETWEEN TEMPER OUTBURSTS, REPETITIVE BEHAVIOUR AND A PREFERENCE FOR PREDICTABILITY IN CHILDREN WITH PRADER-WILLI SYNDROME

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INTRODUCTION: Children with Prader-Willi syndrome (PWS) often show 'temper outbursts' and certain types of repetitive behaviour. Some of these behaviours may be related to executive dysfunction. This study aims to describe the profile of executive functioning in children with PWS and explore potential relationships between this profile and aspects of the behavioural phenotype.

METHODS: 28 children with PWS (12 male; mean age 161m) and 28 typically developing children (11 male; mean age 103m) completed measures of executive attention including a computer task that can assess attentional switching. Carers of these children rated the frequency of repetitive behaviours on two informant report questionnaires. Carers of 46 children with PWS were interviewed about the environmental context of repetitive behaviour shown by their child

RESULTS: Group differences in developmental and chronological age were controlled for statistically. A mixed factor ANCOVA of reaction times in the computer task showed a significant interaction between group and switching ($F(1,52)=9.24, p=0.004$), with PWS children showing greater switching costs. Compared to typically developing children, children with PWS also showed significantly more preference for predictability, repetitive questioning, completing behaviour and persistent habits. Interviews showed that of the 80% of carers who reported temper outbursts, 91.9% reported that unpredictability occurred antecedent to these outbursts. Of 93.5% of carers who reported repetitive questions, 83.7% reported that these increased following unpredictability. A standard score of switch cost was significantly correlated to ratings of children's preference for predictability, but, interestingly, not to any other of the common repetitive behaviours.

DISCUSSION: Children with PWS show a deficit in switching attention that may underlie a preference for predictability that appears to be related to some challenging behaviours.