Title: Associations between Executive Functioning and Temper Outbursts: Evidence from Lowe Syndrome

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Introduction: Lowe syndrome (LS) is an X-linked disorder that occurs in 1 in 500,000 live births. It is a multi-system disorder that affects the kidneys, eyes and central nervous system. Challenging behaviour occurs more frequently in LS than in intellectual disability groups of heterogeneous aetiology (Kenworthy & Charnas, 1995; Arron et al., 2011). In particular, temper outbursts are highly prevalent in LS occurring in 80% of individuals (Arron et al., 2011). Deficits in executive functioning (EF) have been implicated in aggressive behaviour, temper-outbursts and more severe and persistent self-injurious behaviour, in genetic syndromes (Oliver & Richards, 2015; Woodcock et al., 2009). The aim of this study was to explore associations between EF and temper outbursts in Lowe syndrome (LS). Exploring associations between EF and temper outbursts in LS may help inform models of challenging behaviour, which may be applicable to broader intellectual disability populations.

Method: Participants were 26 males with LS, (mean VABS Language Composite = 5 years 10 months (range 2 years 6 months to 10 years 3 months); mean chronological age (CA)= 6 – 34 years). Participants completed a computerized battery of executive function tasks including two delay of gratification tasks, a go no-go task and a measure of task switching. Parents completed the Behaviour Rating Inventory of Executive Function—Preschooler Version (BRIEF-P; Gioia et al., 2003). The duration, frequency and severity of temper outbursts were measured using an adapted version of the Challenging Behaviour Questionnaire (CBQ; Hyman et al., 2002). The Vineland Adaptive Behavior Scales – II (VABS-II; Sparrow et al., 2005) were used as a proxy measure of ability.

Results: A high proportion of individuals with LS engaged in temper outbursts (75%), confirming previous reports of behaviour in this group. Poorer performance on a delay of gratification task was associated with a higher temper outburst composite score (R = -.52, p = .006). Greater executive dysfunction as measured by the BRIEF-P was strongly associated with a higher temper outburst score (R =.82, p <.001). Further analyses of the BRIEF-P subscales indicated that this association was underpinned by the inhibition, working memory and emotional regulation subscales. These associations remained significant once degree of adaptive behaviour was controlled for. Task switching was not associated with temper outbursts in LS.

Discussion: The results indicate that individuals with Lowe syndrome may have difficulties with inhibition, particularly in emotionally salient contexts where individuals are asked to wait for something they find gratifying. It may be that these difficulties increase the likelihood of temper outbursts occurring. Further research is needed to explore the phenomenology of temper outbursts in Lowe syndrome and to inform understanding of pathways from cognition to behaviour.

References/Citations: