THE PHENOMENOLOGY OF TEMPER OUTBURSTS IN INTELLECTUAL DISABILITIES

by

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Thesis Overview

Background: Despite a well-established literature on challenging behaviours there has been limited research on the nature and aetiology of temper outbursts in intellectual disabilities. This has implications for the development of effective behaviour management strategies and for quality of life for people with intellectual disabilities and their carers. This study explores the phenomenology of temper outbursts with specific reference to Lowe syndrome, a rare genetic syndrome affecting the eyes, brain and kidneys.

Systematic review: A review of the experimental functional analytic literature tested the hypothesis that temper outbursts frequently occur in response to thwarted goal-directed behaviour, and might therefore be strongly associated with a tangible reinforcement function. Operational definitions for all topographies of behaviour were extracted from 338 clearly differentiated functional analyses and the data were analysed for associations between behaviour and function. The review found evidence of a behavioural loading onto function for self-bite (tangible) and tearing objects (attention). No other associations were found. There was no support for the initial hypothesis.

Empirical research: Semi-structured interviews were conducted with caregivers of nine children (<18 years) and eight adults (≥ 18 years) with Lowe syndrome to provide a detailed descriptive account of the behavioural sequence, common antecedents and consequences of temper outbursts. Comparisons were made with similar work on Prader-Willi syndrome by Tunnicliffe, Woodcock, Bull, Oliver, & Penhallow, 2014. Outbursts in Lowe syndrome were found to be of high frequency and were associated with higher levels of physical aggression and property destruction than outbursts in Prader Willi syndrome.

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LIST OF ABBREVIATIONS

VOLUME ONE:

ASD	Autistic spectrum disorder
ADHD	Attention deficit hyperactivity disorder
fMRI	Functional magnetic resonance imaging
ID	Intellectual disability
LS	Lowe syndrome
MRI	Magnetic resonance imaging
NRES	National Research Ethics Service
OCD	Obsessive compulsive disorder
PWS	Prader Willi syndrome
QABF	Questions about behavioural function
VABS	Vineland adaptive behaviour scale

VOLUME TWO:

ASD	Autistic spectrum disorder
ADHD	Attention deficit hyperactivity disorder
BPS	British Psychological Society
CAMHS	Child and adolescent mental health service
CAMHS-LD	Child and adolescent mental health service – learning disabilities
CAPA	Choice and partnership approach
CBCP	Challenging behaviour care pathway
C-BIT	Cognitive-behavioural integrated treatment
CBT	Cognitive behavioural therapy
COPD	Chronic obstructive pulmonary disease
CORC	Child Outcomes Research Consortium
CPR	Clinical practice report
DSM-IV	Diagnostic and statistical manual, version four
HADS	Hospital anxiety and depression scale
MCMI	Millon clinical multi-axial inventory
NAT	Negative automatic thought
NICE	National Institute for Health and Care Excellence
NRES	National Research Ethics Service
OCD	Obsessive compulsive disorder
PPD	Paranoid personality disorder
RAID	Rapid assessment interface and discharge
RCP	Royal College of Psychiatry
SCAS	Spence children's anxiety scale
SLDOM	Sheffield learning disability outcome measure

SYSTEMATIC REVIEW:

WHAT DOES THE FUNCTIONAL ANALYTIC LITERATURE TELL US ABOUT THE AETIOLOGY AND MAINTENANCE OF TEMPER OUTBURSTS IN INTELLECTUAL DISABILITIES?

Abstract

Temper outbursts are a common form of challenging behaviour with significant deleterious effects on people with intellectual disabilities and their carers. There have been few studies, however, which examine this phenomenon in detail and there is a lack of consistency in operational definitions.

Aim: Evidence from studies in typical development indicate that thwarted goal-directed behaviour is a common antecedent to outbursts. If this were true for people with intellectual disabilities it might be hypothesised that behaviours commonly associated with temper outbursts, such as crying, screaming, hitting or property destruction, might load most frequently onto a tangible function of behaviour.

Method: A systematic review of the functional analytic literature tested this hypothesis by extracting operational definitions of behavioural topographies from 338 functional analyses in which a single social function of behaviour (tangible, attention or escape) was identified. A hypothetical temper outburst construct was developed and analysed for evidence of loading onto function group.

Results: The review found a strong association between self-biting and tangible function ($\chi^2 = 12.67$, $p = .002$), and between tearing objects and attention ($\chi^2 = 12.14$, $p = .002$). No other associations were found and there was no support for the initial hypothesis.

Implications: Future research on temper outbursts in intellectual disabilities may need to move beyond the behavioural approach to include changes in internal emotional and physiological arousal, which appear to be important components of this behaviour. Agreement is also needed on a consistent operational definition of outbursts to increase comparability between studies.

What does the functional analytic literature tell us about the aetiology and maintenance of temper outbursts in intellectual disabilities?

Temper outbursts are typically included under the rubric of challenging behaviour in published research on people with intellectual disability (ID) alongside behaviours such as self-injury and aggression. The reported prevalence in large sample studies is high, ranging from 24.9% to 34.9% (Smith, Branford, Collacott, Cooper, and McGrother, 1996) and the prevalence may be higher amongst those already exhibiting some form of challenging behaviour. In a sample of 1770 people with ID and challenging behaviour, 85% of adults and 74% of children were reported to evidence temper outbursts (Lowe et al., 2007). Additionally, high levels of hard-to-treat temper outbursts are reported in people with autism spectrum disorders (ASD; Adler et al., 2015; Konst, Matson, & Turygin, 2013) and pervasive developmental disorders (Aman et al., 2009). Higher prevalence figures than those identified for groups of people with ID of heterogeneous cause are also reported for specific genetic syndromes. Dykens, Hodapp, & Finucane, (2005) cite figures of 30-35% for people with Down syndrome, 67% for Cri-du-chat syndrome, 88% for Prader-Willi syndrome, and 94% for Smith-Magenis syndrome. High levels of temper outbursts have also been linked with Lowe syndrome (Kenworthy, Park, & Charnas, 1993). These prevalence data show temper outbursts to be relatively common and unequally distributed across groups defined by aetiology.

Temper outbursts are part of a typical developmental trajectory for children between the ages of 18 months and 4 years (Potegal & Davidson, 2003). They are a source of parental stress (Green, Whitney, & Potegal, 2011) and a common reason for referral for professional behavioural support (Eisbach et al., 2014). Wakschlag et al. (2007) differentiated between developmentally typical and clinically significant manifestations of temper loss and suggested

that a combination of unusual frequency, duration or severity (based on aggressive or destructive behaviours) indicated clinically significant emotional dysregulation requiring professional intervention in pre-school children. Similarly, Belden, Thomson, & Luby, (2008) defined five high risk patterns of temper outbursts leading to longer term behavioural difficulties. These high-risk patterns included frequency, duration, inclusion of aggression and destruction, self-injury, and inability to self-soothe. When severe behaviours extended into later childhood (8-10 years) Caspi, Elder, & Bem (1987) found a link between temper outbursts and negative life-course outcomes into adulthood. Angry, agitated outbursts or "rages", which mirror the behavioural sequence of temper outbursts in younger children (Potegal, Carlson, Margulies, Gutkovitch, & Wall, 2009), are a common cause of inpatient psychiatric admissions for children up to the age of 12 years (Carlson, Potegal, Margulies, Gutkovich, & Basile, 2009). These rages are linked to a range of psychiatric presentations, sometimes leading to seclusion or increased use of psychotropic medication (Carlson et al., 2009). The high prevalence of temper outbursts in intellectual disability populations in combination with these likely deleterious outcomes are cause for concern, and further research to determine the function and aetiological pathways for these phenomena is warranted.

The prevalence of temper outbursts in IDs is typically assessed using several standardised psychometric instruments but remains poorly defined (Tunnicliffe, 2012). The Disability Assessment Schedule (Holmes, Shah, & Wing, 1982) includes a single item - "Temper tantrums[1] – verbal abuse"- as does the Child Behavior Checklist (Achenbach, 1986) - "Temper tantrums or hot temper". The Aberrant Behavior Checklist (Aman & Singh, 1986)

[1] Temper outbursts are sometimes referred to as "tantrums" but the term will be avoided except for specific references in the literature due to potential negative connotations.

includes two items referring specifically to “temper tantrums” in the “anger, irritability, crying” subscale but all items in the subscale appear to be commonly associated with temper outbursts in the typical development literature. Some measures have more specific scales. The Multidimensional Assessment of Preschool Disruptive Behavior Questionnaire (Wakschlag et al., 2012) for example, uses a 14-item temper loss subscale combined with anger regulation items to explore content, triggers and contextual variables impacting on outbursts. However, the measure still omits a clear operational definition of the term “temper outburst” or “tantrum”.

In the typical development literature temper outbursts are described using lists of constituent behaviours. These include: crying, whining, yelling or shouting, screaming, hitting, kicking, stiffening body, pushing/pulling/grabbing, throwing objects, running away, (Potegal & Davidson, 2003), and head-banging, biting and breath-holding (Belden et al., 2008; Österman & Björkqvist, 2010). The number of items included varies between 10 items for the Temper Tantrum Grid employed by Giesbrecht, Miller, & Müller (2010) and 16 behaviours used by Eisbach et al. (2014). Although there is considerable overlap there is little consensus on defining criteria for an outburst. All descriptions include some form of physical aggression, usually hitting, but kicking and biting do not appear in every definition. Property destruction or throwing an object also appears in some form but in almost all cases an outburst can be recorded on the occurrence of “at least one” and any combination of the listed behaviours. Giesbrecht et al. (2010) provide a notable exception in that an outburst can only be recorded if a “strong facial expression” is present as well as at least one other behaviour. This identifies the importance of emotional state in temper outbursts. Potegal & Davidson, (2003) describe outbursts as negative emotional episodes but do not specify that vocal expression such as crying, shouting or screaming must be present and Potegal et al., (2009)

propose that all temper outburst associated behaviours can be considered as expressions of either anger or distress. Variation in included topographies clearly reflects the likely idiosyncratic pattern of behaviours but the absence of a consistent definition makes it difficult to identify a homogenous class of behaviours. This in turn compromises potential study of the aetiology of temper outbursts at an epidemiological level (Iwata, Pace, et al., 1994). Such an understanding could contribute to the development of more effective and generally applicable strategies for prevention and management.

The empirical literature on the determinants of challenging behaviour in people with IDs has been dominated for more than 30 years by a behavioural approach rooted in operant learning theory (Hanley, Iwata, & McCord, 2003; Beavers, Iwata, & Lerman, 2013). Frequent and severe temper outbursts are clearly recognised within this literature as a distinct form of challenging behaviour, as evidenced by their inclusion in psychometric assessments (Aman & Singh, 1986; Achenbach, 1986; Holmes et al., 1982; Wakschlag et al., 2012) and specific references to the form or cause of such behaviours (e.g. Beavers et al., 2013; Dykens et al., 2005; Tunnicliffe, 2012; Woodcock, Oliver, & Humphreys, 2009). Given the strong evidence of effective interventions based on functional analysis for challenging behaviours more generally (Hurl, Wightman, Haynes, & Virues-Ortega, 2016) it seems important to understand how temper outbursts could be understood using this operant framework.

Operational definitions are essential in functional analysis and the absence of an agreed definition in this literature may have hindered progress towards understanding the function(s) of temper outbursts. Systematic reviews by Hanley et al. (2003) and Beavers et al. (2013) found 22 papers over a thirty-year period which included temper outbursts as part of the functional analysis. Across both these reviews, differentiated results for temper outbursts were found in eight papers, of which three reported temper outbursts as escape maintained,

one attention maintained, two tangibly maintained and two maintained by multiple reinforcement. There is little consistency, however, in the definitions used. For example, Vollmer, Northup, Ringdahl, Leblanc, and Chauvin (1996) use the definition "screaming, crying, kicking or throwing objects", whilst Repp and Karsh, (1994) also include falling to the floor, tearing books and other task materials, non-compliance and elopement. It is often unclear how a distinction is made between temper outbursts and other categories such as aggression, which overlap with temper outbursts (hitting, kicking, throwing objects).

Beavers et al. (2013) recommended that all functional analyses should analyse each topography of behaviour separately. They argued that grouping behaviours together leads to an increase in undifferentiated or multiply reinforced outcomes which are harder to treat effectively from a functional perspective. The opposing argument for developing an agreed temper outburst construct is that informants often report a pattern of behaviours which cluster and are recognised as a single event (e.g. Bull, Oliver, Tunnicliffe, & Woodcock, 2015; Potegal & Davidson, 2003; Tunnicliffe, Woodcock, Bull, Oliver, & Penhallow, 2014). Such a cluster might reasonably be expected to load onto a single behavioural function for the individual. The evidence for a general loading of behavioural topographies onto specific reinforcers is sparse. However, there are some suggestions that this might be the case for some behaviours. Hanley et al., (2003) noted that aggression and disruption appeared to load more often onto escape than tangible or attention conditions, with stereotypies more strongly associated with automatic reinforcement. Although the direct evidence from functional analyses of temper outbursts suggests a variety of idiosyncratic functions (Vollmer, Northup, Ringdahl, Leblanc, & Chauvin, 1996), the number of papers is small and each paper describes only two or three individual cases. A common understanding of temper outbursts in young children is an expression of thwarted goal-directed behaviour (Österman & Björkqvist, 2010;

Vollmer et al., 1996). If this were true for people with ID we might expect temper outbursts and associated behaviours to load onto a tangible function more frequently than attention or escape functions in the functional analytic literature.

This brief overview of prevailing themes in the literature indicates that temper outbursts appear to be: 1) associated with aetiology of ID or co-occurring diagnoses such as autism spectrum disorder, 2) potentially different from other challenging behaviours due to recognition of an emotional component and 3) a response to thwarted goal-directed behaviour. Each of these observations is important as there is the potential for an exclusively operant learning perspective to be an incomplete explanation of the behaviours. A first step to address this possibility is to conduct a systematic review of the experimental functional analytic literature to appraise the evidence for functional accounts of temper outbursts. Given the theoretical perspective of a response to thwarted goal-directed behaviour it might be hypothesised that temper outbursts arise in response to situations where there is a "hot" motivational component, such as being denied access to a tangible object, and that behavioural indicators of temper outbursts may therefore load more frequently onto the tangible function. This can be tested by evaluating potential associations between defined topographies of behaviours and identified functions in the experimental functional analytic literature.

To evaluate these potential associations the review includes only papers which used experimental or quasi-experimental functional analysis incorporating at least two social functions of behaviour i.e. social positive (tangible or attention) and social negative (escape from demands) in addition to a control condition. The review does not include behaviours which were multiply reinforced or subject to automatic or sensory reinforcement. In the absence of a consistent definition of temper outbursts, operational definitions of all individual

behaviours included in the functional analysis were recorded. To test the hypothesis that temper outbursts, as opposed to clearly identifiable individual operant behaviours such as aggression, would be more strongly associated with the tangible function, a hypothetical temper outburst construct was created. This was based on a cluster of behaviours commonly associated with temper outbursts in the typical development literature, and on associations between behaviours identified during the review. Temper outbursts are generally understood to be expressions of negative emotions and so behavioural markers of change in an internal emotional state were included in the temper outburst construct (see Methods).

Methods

Procedure

A keyword search was made of the functional behavioural literature using PSYCHINFO, Web of Science and ASSIA. These databases provide good coverage of psychological and behavioural literature, as well as health, social sciences and education. Following Beavers et al.'s (2013) review, a test search on ERIC, a US based educational database, was also carried out but did not produce additional relevant articles.

A list of possible search terms was generated and circulated to eight academics with expertise and publications in behaviour analysis, intellectual disabilities and neurodevelopmental disorders, to ensure that the search terms were comprehensive. The final search terms used and the number of papers found are shown in Table 1.

Table 1: Search terms used and number of papers found

	Search Term	**Field**	**PSYCHINFO** **No. of papers**	**ASSIA** **No. of papers**	**WEB OF SCIENCE** **No. of papers**
	Date of search		*3/6/16*	*17/6/16*	*10/6/16*
	Time period covered		*1967 to May Week 4 2016*	*All years*	*1900 to May 2016*
1	"functional analysis.mp" or "exp. Functional Analysis" or "behavior* assessment.mp" or "behaviour* assessment.mp" or "exp. behaviour asst." or "exp. behavior analysis" or "behaviour* analysis.mp" or "behavior* support.mp"	Keyword	24,011		
2	"Functional analysis" or "behav* assessment" or "behav* analysis" or "behav* support"	Keyword		48,832	86,951
3	"reinforcement.mp" or "exp. reinforcement" or	Keyword	56,304		
4	"reinforcement"	Keyword		1,657	156,403
5	"intellectual disab*.mp" or "mental retardation" or "mentally retarded" or "learning disab*.mp" or "exp. learning disabilities" or "developmental disab*" or "exp. developmental disabilities" or "exp. intellectual development disorder" or "intellectual development disorder.mp"	Keyword	85,259		
6	"intellectual disab*" or "mental* retard*" or "learning disab*" or "developmental* disab*" or "intellectual development* dis*"	Keyword		16,992	219,454
7	1 and 3 and 5		291		
8	2 and 4 and 6			57	436
9	Limit "English" and "Peer reviewed articles only"		236	57	396
10	Initial exclusions after abstract review (see inclusion and exclusion criteria reported elsewhere)		190	29	309
11	Combined list after initial exclusions and removal of duplicates		413		

An abstract review was completed to identify papers for initial exclusion, resulting in a final selection of 413 papers for more thorough review and extraction of relevant data. A further 271 papers were excluded during this process using the inclusion and exclusion criteria. Behavioural topographies were recorded for each participant who met the individual inclusion criteria and exhibited a clearly identified single function for those behaviours.

After final exclusions during the process of data extraction the data set included 142 papers and behavioural topographies for 338 participants (see Figure 1).

Inclusion criteria

Empirical research papers describing participants with mild to profound intellectual disability were included if a systematic multielement experimental functional analysis of behaviour had been carried out based on procedures similar to those described in Carr & Durand (AB design; 1985), or Iwata, Dorsey, Slifer, Bauman, & Richman (ABC design; 1982/1994), or brief experimental functional analysis as in Northup et al. (1991). Papers based on descriptive analysis supported by standardised psychometric measures of behavioural function *and* a naturalistic quasi-experimental design were also included. Clearly operationalised topographies of behaviour had to be linked to identifiable individual participants. Participants were only included if a clear statement was present in the results section of a single function for the assessed behaviours.

Exclusion criteria

Papers were excluded if they met any of the following criteria:

- Review papers, meta-analyses or commentaries (n = 26).
- Functional analysis did not meet inclusion criteria e.g. descriptive or questionnaire-based functional analysis only without experimental testing of the hypotheses, preference assessment only, or only one experimental condition studied (n = 29).
- Inadequate behavioural descriptions (e.g. “aggression” without further operationalisation of the behaviours included; n = 2).
- Summaries of large scale studies where functions of behaviour could not be linked to an identified individual (n = 4).

- Experimental designs intended to assess the impact of parent/caregiver training only, or the impact of child characteristics on parental stress (n = 7).
- Intervention studies which assess the impact of behaviour modification without prior functional analysis (n = 51).
- Not available online (n = 21).
- No participants meeting the criteria outlined below (n = 131).

Individual participants were excluded for the following reasons:

- No evidence of intellectual or developmental disability.
- Participants for whom behaviour is primarily subject to sensory or automatic reinforcement.
- Participants for whom behaviour was multiply reinforced or undifferentiated during functional analysis.
- Participants for whom there is no clear statement of function in the results section.
- Identifiable duplicate participants described in another included paper by the same principal authors. The paper containing the most detailed behavioural descriptions would be used in this case.

Inter-rater reliability

During the initial coding of papers, if there was uncertainty about the inclusion of a paper or participant, or ambiguity about the function of behaviour, this was resolved through discussion with a post-doctoral researcher with expertise in behavioural methodology. After coding of all behavioural topographies, inter-rater reliability was sought for 20% of papers. Cohen's Kappa was calculated for two raters coding behaviours for 79 participants, resulting in κ = .83, (CI (95) 0.80 to 0.86; p < .001), which indicates strong agreement (κ >.80) between the two raters. The second rater was blind to the initial coding of behaviours.

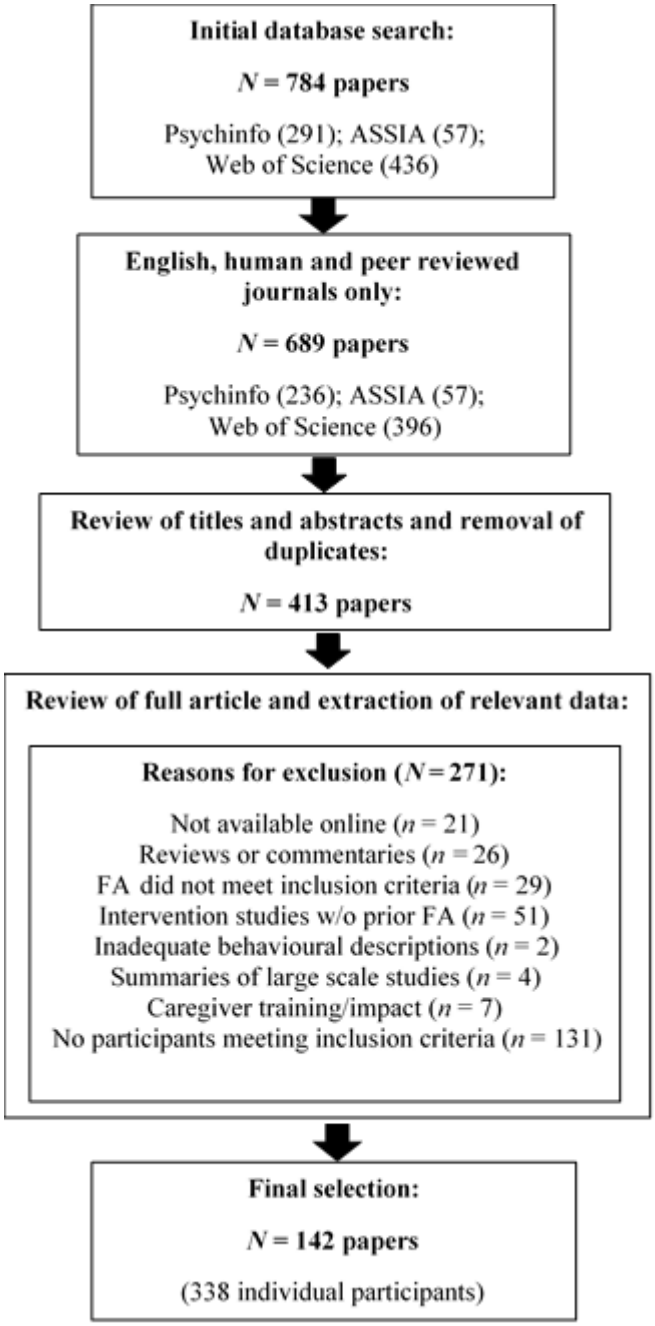

Figure 1: Flowchart of search strategy

Data extraction

Each selected paper was reviewed to check that the methodology described complied with the inclusion criteria for experimental functional analysis or for systematic quasi-experimental design supported by descriptive and questionnaire measures. The results of the functional analysis for each participant were then checked and those participants exhibiting a single social function of behaviour (tangible, attention or escape) were selected. Those showing undifferentiated, multiply reinforced or automatically reinforced behaviours were excluded. Each selected participant's demographic details were recorded on a spreadsheet with the function of behaviour. A tick was marked alongside every individual topography of behaviour included in the participant's functional analysis. Any behaviours which were exhibited by participants but not specifically included in the functional analysis were not recorded. For each new behaviour, a column was added to the spreadsheet to ensure that a comprehensive list of functionally assessed behaviours was produced.

After all papers had been reviewed, behaviours were grouped into categories for data reduction. Categories were generated based on similarity of behavioural topography. For example, face slapping was grouped with hitting other parts of the head with a fist or palm. All categories were then reviewed by two independent reviewers with knowledge of the functional behavioural literature to reduce the number of categories further. Idiosyncratic behaviours (e.g. intentional breath-holding) which could not be incorporated into another behavioural category, and were reported for less than ten participants, were excluded from the final analysis. Four aggregate variables were also constructed: physical aggression towards others, self-injury, property destruction and disruptive behaviour. The final list consisted of 31 categories of which 27 were included in the statistical analysis (see Results Table 2).

Analysis

Data from the spreadsheet (Appendix A) were transferred to a computerised statistical package for analysis. Data were first explored for associations between demographic characteristics, such as age, gender and genetic syndrome, and behavioural topographies or function group to check for the presence of confounding variables. Percentages were then calculated for the number of participants in each function group (attention, escape and tangible) displaying each category of behaviour. Pearson's chi-squared (χ^2) analysis was used to test for statistically significant differences between function groups for each behaviour, and for the four aggregate categories. Finally, the data were explored for evidence of associations between behaviours commonly linked to temper outbursts in both typically developing and ID populations.

Temper outburst construct

Although temper outbursts are poorly defined in the literature, the main aim of this review was to look for evidence to support a generalisable function of these behaviours. A temper outburst construct was therefore developed to reflect the fact that temper outbursts are not a single observable behaviour but can be made up of a variable range of individual topographies of behaviour. A defining characteristic of temper outbursts is a change in emotional state (Potegal & Davidson, 2003; Eisbach et al., 2014). Functional analysis is based on operant conditioning theory and therefore only includes overt observable behaviours. Internal states such as emotions are not considered to be accessible to objective scientific measurement and can only be inferred. Crying or loud vocalisations (which included screaming, yelling and shouting) were therefore used as proxy behavioural indicators of potential change in emotional state. Since either crying or loud vocalisations could indicate emotional arousal, a decision was taken to combine these two topographies to create a new

categorical variable (CLV; yes/no) which was scored as yes if either crying or loud vocalisations were present. After selecting cases based on this variable (CLV = yes) the data were explored for strong associations with other behavioural topographies. The resulting cluster of topographies (TO1), which consisted of crying or loud vocalisations plus at least one other behaviour from the associations found, was tested for differentiated function.

The initial temper outburst construct, as described above, did not include either physical aggression or self-injury. These behaviours are frequently mentioned in operational definitions of temper outbursts in the literature from both typically developing (Potegal & Davidson, 2003) and ID populations (e.g. Tunnicliffe et al., 2014). An alternative temper outburst construct (TO2) was therefore created to reflect the full range of behaviours described in the temper outburst literature. This categorical variable consisted of the CLV variable plus at least one other behaviour from physical aggression (any type), self-injury (any type), property destruction (any type), elopement, dropping, noncompliance or "tantrum". Although dropping and "tantrums" were initially excluded from statistical analysis of functions due to small numbers, they were included in the construct.

It is recognised that combining only one other behaviour with crying or loud vocalisations could be over inclusive, but reflects operational definitions used in the literature. Crying and self-injury, for example, could occur together in direct response to pain, but this combination sometimes appears in the literature labelled as a temper outburst (e.g. Marcus & Vollmer, 1996). A more conservative construct which required the inclusion of crying or loud vocalisations and at least two other behaviours from the above list, was also developed (TO3).

Statistical tests were used to examine associations between function group and each of the three hypothetical constructs. For all statistical analyses a Bonferroni correction was used to adjust for the number of tests carried out, resulting in the use of a *p*-value of .002 as the Alpha level for statistical significance.

Results

Data were extracted from 142 peer-reviewed articles. Operational definitions of behaviour from 338 individual functional analyses were recorded. The initial list of topographies included 92 different behaviours, which were later grouped into 27 categories plus four aggregate variables. The final categories and the number of participants displaying each type of behaviour are shown in Table 2.

The demographic characteristics of each participant for whom a single function of behaviour was identified is shown in Table 3. The final sample included 62.1% male and 33.7% female participants. The age range was between two years and 56 years, with a mean of 17 years (*SD* = 12.96). The majority (63.9%) were under the age of 18 years. The sample included 24.6% where ID had been confirmed but the level was unspecified, 13.0% mild or borderline, 16.9% moderate, 21.3% severe and 24.3% profound. A diagnosis of autism spectrum disorder (ASD) was reported for 29.3% of the sample, attention deficit hyperactivity disorder (ADHD) for 5.6%, pervasive developmental disorder for 4.1%, cerebral palsy for 3.6% and 7.1% had a seizure disorder. Genetic syndromes were identified for 30 participants (8.9%) and included Angelman syndrome (*n* = 4), Cornelia de Lange syndrome (*n* = 1), Dandy Walker syndrome (*n* = 1), Down syndrome (*n* = 9), fragile X syndrome (*n* = 6), Lennox-Gestaut syndrome (*n* = 1), Prader Willi syndrome (*n* = 1), Rubenstein Taybi syndrome (*n* = 1), Smith Magenis syndrome (*n* = 1), Sotos syndrome (*n* = 1), tuberous sclerosis (*n* = 1), XYY syndrome (*n* = 1), 3q29 deletion syndrome (*n* = 1) and 15q deletion

syndrome (n = 1). Thirty-four percent of the sample were non-verbal, and a further 22.2% had very limited verbal expression. A large proportion of the studies did not report on levels of mobility so the data could not be reliably analysed.

Table 2: Categories of behavioural topographies.

Category	Behaviours included	No. (%)[a] *N* = 338	
Hit	Hitting, punching or slapping another person forcefully using hand(s) or object.	153	(45.3%)
Push	Forceful pushing, grabbing, twisting or pulling of any body part, clothing or hair of another person.	110	(32.5%)
Kick	Kicking or stamping on any part of another person.	101	(29.9%)
Bite or pinch	Biting, scratching or pinching skin of another person between fingers.	117	(34.6%)
Headbutt	Forceful contact between head and any part of another person's body.	30	(8.9%)
Spit	Deliberate forceful projection of saliva, partly chewed food or vomit from mouth towards another person.	12	(3.6%)
Physical Aggression	At least one of the behaviours listed under hit, push, kick, bite or pinch, headbutt or spit.	173	(51.2%)
Head bang	Contact of head with a stationary object or hard surface.	82	(24.3%)
Face hit	Hitting or slapping face or head with own hand or object with sufficient force to cause reddening or bleeding.	96	(28.4%)
Self-hit	Hitting, slapping, punching or kicking own body parts (other than face or head) with closed fist, open palm, foot or object.	67	(19.8%)
Strike body	Striking body parts (other than head) against solid surface or object (including floor).	14	(4.1%)
Self-bite	Hand-mouthing or closure of teeth on skin of own fingers, hands, wrists or any other body part.	87	(25.7%)
Self-pinch	Pinching, scratching, digging, or picking at own skin with fingers or objects to cause reddening, bleeding or bruising, or poking or pulling at other body parts including eyes, ears or hair.	42	(12.4%)
Dangerous act[b]	Any behaviour likely to cause risk to self not listed under self-injury e.g. putting fingers in electrical sockets, deliberate overdose of medication without suicidal intent.	4	(1.2%)
Self-injury	At least one of the behaviours listed under head bang, face hit, self-hit, strike body, self-bite, self-pinch. (NB Does not include dangerous acts).	174	(51.5%)
Throw object	Throwing leisure materials or objects (not directly at another person).	112	(33.1%)
Tear object	Tearing, ripping, or crumpling of leisure or task materials, destruction of other objects including tearing paper from walls, or biting or tearing own clothing.	63	(18.6%)
Pull object	Pulling objects from shelves, swiping from table or knocking over objects (other than furniture).	33	(9.8%)
Hit object	Hitting, kicking, banging or stamping on walls, floor, furniture, windows or objects incl. door slamming.	59	(17.5%)
Damage furniture	Throwing, overturning or jumping on furniture.	33	(9.8%)
Destruction	At least one of the behaviours listed under throw object, tear object, pull object, hit object or damage furniture.	117	(34.6%)
Elopement	Leaving or attempting to leave the activity area.	12	(3.6%)
Crying	Crying, sobbing, or tearful.	22	(6.5%)
Loud vocalisations	Vocalisations above conversational level e.g. screaming, yelling, squealing, whining, growling, shouting, cursing or swearing.	50	(14.8%)
Perseverative speech	Perseverative, bizarre, inappropriate or delusional vocalisations irrelevant to task or to interest of others.	13	(3.8%)
Dropping[b]	Falling to the floor from a standing or seated position.	8	(2.4%)
Non-compliance	Non-compliance or refusal to take part in activity.	21	(6.2%)
Tantrum[b]	Tantrums or temper outbursts not otherwise specified.	4	(1.2%)
Disruption	At least one behaviour listed under elopement, crying, loud vocalisations, perseverative speech, dropping, non-compliance and tantrum	76	(22.5%)
Sexual[b]	Inappropriate sexual behaviour including remarks, or inappropriate touching of self or other.	5	(1.5%)
Stereotypy[b]	Stereotypical or repetitive movements, vocal tics or behaviours such as spinning objects, hand wringing or flapping, rocking or pacing, or repetitive ingestion of non-food items (pica).	6	(1.8%)

[a] Most participants demonstrated more than one behaviour so total percentages are greater than 100%.
[b]Separate categories not included in final analysis due to low numbers (N<10).

Table 3: Demographic characteristics of participants

Characteristic		**No. of participants *N* = 338**	**Percentage of total sample**
Gender:	Male	210	62.1%
	Female	114	33.7%
	Not recorded	14	4.1%
Age group:	Child (<18 years)	216	63.9%
	Adult (18 years or over)	117	34.6%
	Age not specified	5	1.5%
Level of ID:	Profound	82	24.3%
	Severe	72	21.3%
	Moderate	57	16.9%
	Mild/borderline	44	13.0%
	Level unspecified	83	24.6%
Diagnoses:	Autism spectrum disorder	99	29.3%
	Attention deficit hyperactivity disorder	19	5.6%
	Pervasive developmental disorder	14	4.1%
	Genetic syndrome	30	8.9%
	Cerebral palsy	12	3.6%
	Seizure disorder	24	7.1%
Communication:	Non-verbal	115	34%
	Single words or two-word phrases	43	12.7%
	Short sentences and >3-word phrases	32	9.5%
	Not enough information or unspecified	148	43.8%
Mobility:	Non- or partial ambulatory	16	4.7%
	Ambulatory	61	18.0%
	Not recorded	261	77.2%

Confounding variables

In view of the potential confounding influence of demographic characteristics, the data were first explored for associations between age, gender, level of ID, ASD and communication, and function group. The distribution of data for age was positively skewed due to the predominance of child participants and a non-parametric test was therefore used to compare the function groups. The independent samples Kruskal-Wallis test indicated that age in years did not differ across the function groups (p = .389). Pearson's chi-squared test indicated no significant differences between the function groups for level of ID, or degree of communication difficulties. The difference between function groups for ASD diagnosis approached significance with a p-value of .003, with a lower frequency of attention-reinforced behaviours in the ASD group. Statistical analysis was therefore run for each ASD group

(diagnosis present or absent) separately. Having a diagnosis of ASD resulted in no significant differences in the function of any of the behavioural categories measured. Gender made no significant difference to function of behaviour using a Bonferroni correction but a possible difference was found using a less conservative *p*-value of .05 ($\chi^2 = 9.663$, $p = 0.047$). In view of the high percentage of males (62.1%) in the overall sample, gender was therefore considered as a potential confounding variable.

Given the skewed age distribution in the sample, the data were explored for significant differences in the frequency of behaviours by age group (child < 18 years; adult ≥ 18 years). Pearson's chi-squared tests revealed significantly lower frequencies of physical aggression ($\chi^2 = 29.860$; $p < .001$), destruction ($\chi^2 = 26.522$; $p < .001$) and crying ($\chi^2 = 9.672$; $p < .001$) in the adult group. The likelihood of adults engaging in self-injury was higher than expected ($\chi^2 = 26.522$; $p < .001$) compared to children. The statistical significance of these results was confirmed across the whole age-range using a non-parametric Man-Whitney U-test of difference in the mean age of participants showing the behaviour and those who did not ($p < .001$). In view of these differences, the statistical analysis by function group was run separately for each of the age groups. Significant results and their potential impact on overall outcome is reported below.

Behavioural topographies

The main hypothesis to be tested in this study was whether behaviours associated with temper outbursts (including crying, emotional vocalisations, physical aggression, property destruction, self-injury, non-compliance and dropping) would be more frequently associated with tangible reinforcement, than attention or escape. The main variables of interest were topographies of behaviour, which were all categorical variables consisting of yes (behaviour present during functional analysis) or no (behaviour not reported as part of the functional

analysis). The three function groups were tangible (behaviour reinforced by access to a desired object or activity); attention (behaviour reinforced by access to attention); and escape (behaviour reinforced by escape from a task demand or other aversive stimulus). Low frequency behaviours exhibited by fewer than ten participants (dangerous acts, dropping, tantrums, sexual behaviours and stereotypies) were excluded from the statistical analysis. Results are provided in Table 4.

Table 4: Results of statistical analysis of behavioural function.

Category	**Number displaying behaviour**	**Percentage by function group**			**$\chi 2$** (df =2)[a]	**p-value**	**Post-hoc analysis[b]**
		Tangible %	Attention %	Escape %			
Hit	153	46.8	43.2	45.1	0.28	.867	NS
Push	110	27.3	31.5	35.5	1.84	.398	NS
Kick	101	35.1	28.8	27.6	1.30	.522	NS
Bite or pinch	117	37.7	30.6	35.5	1.22	.543	NS
Headbutt	30	7.8	11.7	7.2	1.66	.437	NS
Spit	12	6.5	0.9	3.9	4.31	.116	NS
Physical Aggression	173	53.2	47.7	51.3	0.79	.674	NS
Head bang	82	22.1	24.3	25.0	0.29	.863	NS
Face hit	96	32.5	24.3	28.9	1.60	.450	NS
Self-hit	67	23.4	20.7	17.1	1.25	.534	NS
Strike body	14	1.3	5.4	4.6	2.12	.347	NS
Self-bite	87	36.4	14.4	28.3	12.67	.002	A<T[c]
Self-pinch	42	6.5	17.1	11.8	4.76	.093	NS
Self-injury	174	57.1	45.9	53.3	2.32	.314	NS
Throw object	112	24.7	39.6	32.2	4.62	.099	NS
Tear object	63	10.4	28.8	15.1	12.14	.002	A>E[c]
Pull object	33	7.8	12.6	8.6	1.57	.457	NS
Hit object	59	18.2	20.7	14.5	1.66	.436	NS
Damage furniture	33	7.8	9.9	10.5	0.48	.786	NS
Destruction	117	27.3	40.5	33.6	3.53	.171	NS
Elopement	12	3.9	0.9	5.3	3.70	.158	NS
Crying	22	3.9	9.9	5.3	3.31	.191	NS
Loud vocalisations	50	15.6	14.4	14.5	0.05	.974	NS
Perseverative speech	13	1.3	8.1	2.0	8.19	.017	NS
Non-compliance	21	3.9	5.4	7.9	1.66	.437	NS
Disruptive	76	18.2	21.6	23.7	0.37	.830	NS

NS = Not statistically significant.

[a] Pearson's 2x3 chi-squared test calculated using SPSS.

[b] Post-hoc Bonferroni correction, $p \leq .002$.

[c] A = Attention; T = Tangible; E = Escape.

As can be seen from Table 4 only two significant differences were found across all the individual behaviours and the four aggregate variables. Self-bite was found to be less frequently associated with attention than with tangible reinforcement. Pairwise comparison

between attention and escape functions also approached significance at $p = .006$, suggesting that this behaviour was least likely to be reinforced by attention. Tear object was more frequently reinforced by access to attention than tangible. Pairwise comparisons for attention-escape and tangible-escape were not significant. No significant difference was found for any of the other variables.

Given the potential confounding influence of age, gender and ASD diagnosis on the interaction between behaviour and function, these two behaviour categories were further explored by running the analysis for each age group (child, adult), gender and ASD group. There was no significant difference in the function of self-bite for children, but for adults self-bite was less frequently reinforced by attention ($\chi^2 = 17.19$, $p < .001$). This was also true for those without an ASD diagnosis ($\chi^2 = 16.48$, $p < .001$), and approached significance for males only ($\chi^2 = 10.05$, $p = .007$). For tearing objects the overall result was for a higher frequency of attention reinforced behaviour compared to tangible and escape functions. The strongest evidence for this was found in the child only group ($\chi^2 = 20.53$, $p < .001$), and in the male only group ($\chi^2 = 16.80$, $p < .001$). The result for those without an ASD diagnosis approached significance ($\chi^2 = 10.26$, $p = .006$). For adults only and for females only there was no significant difference between functions for tear object.

Temper outburst construct

Exploratory analysis for clusters of behaviours associated with crying or loud vocalisations produced some significant results. Significant associations were found between the CLV variable and destruction ($\chi^2 = 11.76$, $p = .001$), non-compliance ($\chi^2 = 63.50$, $p < .001$) and "tantrum" (Fisher's exact, $p = .001$). Associations approached significance for self-injury ($\chi^2 = 7.25$, $p = .007$) and throw object ($\chi^2 = 7.88$, $p = .005$). There was no significant association between CLV and physical aggression of any sort. The first temper outburst

construct (TO1) therefore included CLV plus one other behaviour from destruction, non-compliance and "tantrums" but excluded near-significant and non-significant associations. Temper outburst constructs (TO2 and TO3) based on descriptions from the literature included CLV plus one (TO2) or two (TO3) other behavioural topographies from physical aggression (any form), self-injury (any form), property destruction (any form), elopement, dropping, non-compliance and "tantrums".

Pearson's chi-squared was used to test whether any of these hypothetical temper outburst constructs would load more frequently onto a tangible, attention or escape function. No significant difference was found for any of the temper outburst constructs.

Discussion

The primary purpose of this review was to examine the functional analytic literature for evidence of a relationship between behavioural function and temper outbursts. It sought to test the hypothesis that temper outbursts would be more frequently associated with tangible reinforcement than either attention or escape. This review found no evidence to support this hypothesis, either using individual behaviours or a hypothesised temper outburst construct. The review also found only limited evidence for a generalised loading of behavioural topography onto function (tangible, attention or escape) across the full range of challenging behaviours, with significant results for self-biting and tearing objects. Self-biting was most frequently reinforced by tangible rewards, and least likely to occur in response to attention. Conversely, tearing objects was most frequently reinforced by access to attention from a caregiver.

The significant findings for self-bite and tearing objects are noteworthy. The use of a Bonferroni correction to adjust for the large number of statistical tests on the data, means that

these results are unlikely to be due to chance. The number of participants in each category was also large enough to produce a reliable result (self-bite, $n = 87$; tear object, $n = 63$). Self-biting, particularly hand-biting has been shown to be highly prevalent in fragile X syndrome (Symons, Clark, Hatton, Skinner, & Bailey, 2003). It has also been noted as a common occurrence in laboratory studies of self-injury in animals. Stereotyped biting of forepaws has been associated with neurochemical dysregulation of the basal ganglia circuitry and in damage to the frontoparietal lobe in rats (Devine & Symons, 2013). Self-biting and crib-biting in horses has also been linked to stress and to environmental deprivation during juvenile development (Devine & Symons, 2013). This may suggest a relationship between some genetic disorders (e.g. fragile X) where self-biting is highly prevalent, and deficits in affect regulation in the presence of heightened emotional arousal. In the light of these findings, the increased likelihood of self-biting behaviour in the tangible condition in comparison to the attention condition is worthy of further investigation. It is harder to find a possible explanation for the loading of tearing objects onto the attention function. Further research to explore the function-behaviour link for this type of challenging behaviour may therefore be warranted.

Although there have been several reviews of the functional analytic literature over the last thirty years (e.g. Hanley, Iwata, & McCord, 2003; Beavers et al., 2013) there has not been such a detailed epidemiological analysis of the relationship between function and behavioural topography before. A total of 338 clearly differentiated functional analyses were examined, and detailed operational definitions of behavioural topographies were recorded. The outcome of this review appears to lend support to the argument that, in general, functions of behaviour are idiosyncratic and must be analysed on an individual basis. In spite of proven effectiveness (Hurl et al., 2016), experimental functional analysis remains a time-consuming process which

is not always possible outside a research setting. Alternative methods such as descriptive (Derby et al., 1992) or questionnaire-based functional analyses (e.g. Questions About Behavioral Function, QABF, Matson & Vollmer, 1995) have been shown to have good clinical utility but a more general understanding of the nature and possible aetiology of behaviours such as temper outbursts would still be of great benefit to caregivers not all of whom have access to professional assessment.

The lack of an association between temper outbursts and function warrants comment. Temper outbursts have previously been explored through functional analysis in typically developing children. For example, Carr & Newsom (1985) found evidence that temper outbursts in a school setting were associated with escape from demands. This finding was later questioned by Repp and Karsh (1994) who found that temper outbursts initially identified as escape-reinforced were later found to be more strongly related to positive reinforcement in the form of attention from teachers. Vollmer et al. (1996) argue that the function of temper outbursts is idiosyncratic and unique to each individual. They report functional analysis results for three children, one of whom exhibited tangibly-reinforced behaviour, another demonstrated behaviour which was reinforced by tangible and attention functions, and the other by multiple functions. There was no evidence from the current review of a general loading of temper outburst behaviours onto function, which could be taken as support for Vollmer et al.'s (1996) position. There are, however, several limitations to this study which could contribute to the absence of significant findings for temper outbursts.

The first limitation relates to the centrality of emotion to an understanding of temper outbursts in typical development (Potegal & Davidson, 2003; Green et al., 2011; Eisbach et al., 2014; Giesbrecht, Miller, & Müller, 2010), and the behavioural analytic constraints of the

functional analytic approach. In behavioural research, internal states such as emotions, pain, hunger or tiredness, are considered inaccessible to scientific enquiry and only overt observable behaviours are therefore monitored. Although some observable behaviours such as crying or loud vocalisations could be taken as indicators of emotional change, it was noticeable how rarely these appeared in the behavioural topographies in this study. Crying was present in only 6.5%, and loud vocalisations were mentioned in 14.8% of the studies. It seems unlikely that all the other participants were silent during outbursts of aggression or property destruction, but emotional vocalisations were not considered relevant to the analysis of behavioural function. In the absence of an agreed definition of temper outbursts in the literature, hypothetical constructs for temper outbursts were developed for this study based on significant associations between variables and on descriptions in the literature. Given the emphasis on emotional content of outbursts, as exemplified by Potegal & Davidson's (2003) anger-distress model, crying or loud vocalisations were considered to be a necessary defining characteristic to distinguish outbursts from other aggressive, destructive or disruptive behaviours. This resulted in the selection of a subset of the total sample who displayed crying or loud vocalisations (n = 52). Given the possibility that other participants also exhibited crying or loud vocalisations which were not recorded, the validity of the constructs based on this selection may be questionable. The constructs may include some combinations of behaviours which are not temper outbursts such as crying and self-injury which could be the direct result of pain. The construct is a combination of other behavioural variables and might therefore identify individuals with multiple challenging behaviours rather than temper outbursts as it is not possible to tell from the data whether these behaviours occurred simultaneously or separately. The constructs should therefore be considered as indicators of potential areas for further research rather than as a robust operationalisation of temper

outbursts. Further research is required to produce detailed descriptions of temper outburst behaviours in a range of intellectual disabilities to refine the operational definition.

The second limitation relates to the exclusive focus on the behaviours examined during the original functional analysis. Given the nature of this review, it would not have been possible to do otherwise, but conclusions cannot be drawn about the general nature of behaviours displayed by the individual participants from those chosen for functional analysis. It is possible that many, or even all, of the participants also displayed other challenging behaviours which were not recorded.

The sample is not representative of all people with intellectual disabilities or genetic syndromes. It has already been noted, that the age and gender distributions of the study sample were skewed, with a higher proportion of children (63.9%) and males (62.1%). The review only includes participants who appeared in published studies using some form of experimental functional analysis, and who displayed a clearly differentiated single function of behaviour. Topographies of behaviour had to be clearly linked to identifiable individuals, resulting in exclusion of several important papers which summarised the functional analysis results from large groups of participants (e.g. Iwata et al., 1994; Kahng & Iwata, 1998; Kurtz et al., 2003; Wallace & Iwata, 1999). Whilst these were necessary conditions of the research, the generalisability of findings is therefore limited.

Finally, the initial hypothesis was that a link might be found between temper outbursts and tangible reinforcement. All studies included were required to have a multielement design and explore a minimum of two different functions of behaviour. Iwata et al.'s, (1982/1994) functional analytic design, on which many of the papers were based, did not include a separate tangible condition. The original methodology compared social-positive (tangible or

attention) with social-negative (escape from demand or other aversive stimuli). It was only later that studies began to separate the social-positive function into tangible and attention conditions. Beavers et al. (2013) have argued that the inclusion of a tangible condition should only be considered where strong evidence already exists from carer report or descriptive analysis of potential tangible reinforcement. Their argument is that the inclusion of tangible conditions can skew the results of functional analyses and result in many more undifferentiated or multiply reinforced results. The tangible function group consisted of only 22.8% of the total sample, but it is impossible to know whether this is a true representation of the distribution of tangible reinforcement in the ID population, or reflects the exclusion of tangible conditions unless otherwise indicated.

Conclusions

This study examined the functional analytic literature for evidence of a link between behaviours commonly associated with temper outbursts and tangible function. The original hypothesis was based on findings from studies on temper outbursts in typically developing children as well as genetic syndromes which suggest that outbursts occur in response to thwarted desire for access to tangible rewards. The study found no evidence to support this hypothesis. However, the functional analytic literature is based on a behavioural methodology which does not consider internal states such as emotion or pain to be accessible to scientific enquiry. Given that temper outbursts are understood to be expressions of emotion this may have contributed to the outcome. Temper outbursts are a frequent cause of distress for parents and for people living with genetic syndromes. They can interfere with access to education and occupational opportunities, and are reported by parents to be difficult to manage using existing behavioural interventions. Whereas most topographies of behaviour can be clearly operationalised, outbursts consist of a number of different observable

behaviours which when grouped together are recognised as an outburst. The absence of a clear definition of temper outbursts provides a challenge for further research but one which needs to be overcome to understand the nature and aetiology of temper outbursts in intellectual disabilities as a basis for development of effective interventions.

Future research may need to move away from a purely behavioural approach, to include a way of capturing changes in internal emotional and physiological states. This provides a significant challenge when working with ID populations with a range of communication difficulties which may preclude self-report. In the first instance, where behaviours are of high frequency, video-recorded behavioural observations may offer an important addition to informant report interview or diary studies of temper outbursts. Naturalistic observations have been used to good effect to improve understanding of outbursts in Prader-Willi syndrome (PWS; Oliver, Woodcock, & Humphreys, 2009). Measurement of physiological arousal such as heart rate monitors or cortisol levels could be considered with appropriate ethical safeguards. Important work has also been carried out in the field of "behavioural neurogenetics" which looks at the potential behavioural impact of neurological differences in genetic syndromes (Reiss & Dant, 2003; Järvinen-Pasley et al., 2008; Meyer-Lindenberg, Mervis, & Faith Berman, 2006). Greater understanding of neural correlates of temper outbursts offers potential for innovative interventions. For example, studies of temper outbursts in PWS (Tunnicliffe et al., 2014; Woodcock et al., 2010) have identified a cognitive mechanism (task-switching) which is closely linked to outbursts. This has led to the development of promising interventions using signalling to alert the person with PWS to impending change which has been found to reduce the occurrence of outbursts significantly (Bull, Oliver, & Woodcock, 2017).

This review has provided an important overview of findings to date, from a functional analytic perspective, on the aetiology of temper outbursts in intellectual disabilities. The absence of a consistent operational definition of outbursts, however, limits the comparability of studies. The generation of detailed descriptive studies of outbursts in a range of intellectual disabilities and genetic syndromes could form the basis for agreement of a robust operational definition which would underpin further research on the biological and functional aetiology of these behaviours.

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EMPIRICAL RESEARCH PAPER:

A DESCRIPTIVE ANALYSIS OF TEMPER OUTBURSTS IN LOWE SYNDROME

Abstract

Background: Lowe syndrome is a rare chromosomal disorder causing multiple physical and intellectual impairments. Previous studies indicate high levels of challenging behaviour, with temper outbursts identified by caregivers as a particular difficulty.

Aim: This paper provides a detailed description of the behavioural sequence, antecedents and consequences of temper outbursts in Lowe syndrome from a caregiver perspective, with a view to improved understanding of the function of these behaviours.

Methods: Semi-structured interviews were conducted with primary caregivers of nine adults (18 years or over) and eight children (<18 years) diagnosed with Lowe syndrome. The study replicated work by Tunnicliffe, Woodcock, Bull, Oliver, and Penhallow (2014) on temper outbursts in Prader-Willi syndrome and comparisons are made with the results of that study throughout.

Results: Frequent temper outbursts in Lowe syndrome were associated with high levels of physical aggression and property destruction. Similarities were found with the pattern of behaviours in Prader-Willi syndrome and in typically developing younger children. Thwarted desire or being asked to do something they did not want to do was found to be the most common antecedent to outbursts in Lowe syndrome.

Implications: This study provides an important foundation for further research into the aetiology of temper outbursts in Lowe syndrome. Recent studies in Prader-Willi syndrome have found links between outbursts and cognitive task switching difficulties. It is possible that a similar or different executive function difficulty could be implicated in Lowe syndrome.

A descriptive analysis of temper outbursts in Lowe syndrome

Lowe syndrome (LS), also known as oculocerebrorenal syndrome, is a rare genetic disorder, affecting mostly males, with an estimated prevalence of 1 in 500,000 of the general population (Loi, 2006). The syndrome is caused by a mutation of the OCRL1 gene, which is located on the X-chromosome at Xq26.1 (Yuksel, Karaca, & Albayram, 2009). The mutation causes multiple impairments and impacts predominantly on the development of the eyes, brain, and kidneys (Lewis, Nussbaum, & Brewer, 2012). Affected boys are born with bilateral cataracts (dense clouding of the lens in both eyes) with approximately 50% developing glaucoma (a build-up of pressure behind the eye, causing damage to the optic nerve). Infantile hypotonia (poor muscle tone) can affect feeding, sitting, standing and walking. Kidney malfunctions lead to malabsorption of nutrients which can cause brittle bones and other complications such as spinal scoliosis (Loi, 2006). In older children and adults, kidney problems can result in renal failure with significantly reduced life expectancy (Lewis et al., 2012). All those affected have some level of intellectual disability (ID; 10-25% mild-borderline; 25% mild-moderate; 50-65% severe to profound; Lewis et al., 2012). Magnetic resonance imaging (MRI) studies have noted non-specific abnormalities in brain development, including delayed myelination and the presence of multiple small cystic lesions in the white matter but the implications of these findings are not yet understood (Allmendinger, Desai, Burke, Viswanadhan, & Prabhu, 2014; Yuksel et al., 2009). To date, there are limited published data available on the behavioural characteristics of LS but there are clear indications that challenging behaviour is a significant issue.

An association between challenging behaviour and poor quality of life for individuals with ID and their carers is well documented (Emerson & Einfeld, 2011; Moss et al., 2000; Hastings, 2002; Hayes, McGuire, O'Neill, Oliver, & Morrison, 2011). Understanding the

aetiology of such behaviours in LS is therefore important from a clinical perspective. Arron, Oliver, Moss, Berg, and Burbidge, (2011) reported a 60-70% prevalence of challenging behaviours, particularly self-injury and aggression, in people with LS (n = 56), compared to 10-15% prevalence in people with ID without a genetic syndrome (Emerson et al., 2001). These behaviours showed an association with impulsivity and repetitive behaviours. Moss, Oliver, Arron, Burbidge, & Berg (2009) noted that people with LS showed specific forms of repetitive behaviour, with hand stereotypies and lining-up behaviour being especially prevalent. In one of the few studies to focus entirely on behaviours in LS, Kenworthy, Park, & Charnas (1993) reported stubbornness, temper outbursts and repetitive behaviours in more than 80% of the sample. This finding has also been supported by parent surveys in which temper outbursts were reported as a daily occurrence by 50% of parents (Dolinsky, Jacobs, & Knight, 2008). Kutsch, Waite, Crawford, & Oliver, (under submission) report prevalence rates of 57.1% for self-injury, 60.7% for aggression and 74.1% for temper outbursts, amongst a sample of 28 boys with LS. Where elevated rates of challenging behaviour, over and above the rates found in other IDs, are reported for a genetic syndrome (e.g. Dimitropoulos, Feurer, Butler, & Thompson, 2001; Arron et al., 2011) it indicates a possible behavioural phenotype that warrants further investigation.

The majority of the literature on challenging behavior in IDs has adopted a behavioural perspective based on operant learning theory (Emerson, 1993). The theory suggests that behaviours are maintained by inadvertent positive and negative social, and automatic reinforcement. For example, an attempt to calm or soothe a child by providing attention or distraction with tangible items may reward behaviours. The child is then more likely to repeat these behaviours when in the same situation in the future (Carr & Durand, 1985). Kutsch et al. (under submission) used the Questions About Behavioral Function

standardized parent-report measure (QABF; Matson & Vollmer, 1995) to assess the functions of behaviour in LS and found a significant association between self-injury and escape from task demands, whilst temper outbursts and aggression were more frequently observed when access to tangible items was denied.

There is a strong evidence base for the functional analytic approach (e.g. Beavers, Iwata, & Lerman, 2013) . It is largely based on the effectiveness of interventions which disrupt prevailing environmental contingencies and provide an alternative response for the person with an ID (Hurl, Wightman, Haynes, & Virues-Ortega, 2016; Waite et al., 2014). This supports the argument that challenging behaviours are, at least in part, learned behaviours. An exclusively operant learning approach cannot however, adequately explain the evidence for identifiable behavioural phenotypes for different genetic syndromes.

The term "behavioural phenotype" is used to denote a characteristic pattern of behaviours which is demonstrated more frequently by people with a particular genetic syndrome than by those without the syndrome when developmental level is accounted for (Dykens, Hodapp, & Finucane, 2000; Waite et al., 2014). Temper outbursts have been shown to be more prevalent in several genetic syndromes, including Prader Willi, Cri-du-chat, and Smith-Magenis syndromes (Dykens et al., 2000) as well as in LS (Kenworthy et al., 1993). This partial specificity (see Dykens et al., 2000) is difficult to explain either from an exclusively biological or operant conditioning perspective. A theory which incorporates an interaction between the biological/developmental consequences of a genetic difference and environmental factors is needed (Tunnicliffe & Oliver, 2011).

In typical development temper outbursts are conceptualized as part of a normative developmental process (Potegal & Davidson, 2003). They are understood to reflect immaturity in language abilities (Österman & Björkqvist, 2010) and cognitive processes

involved in emotional and behavioural regulation (Giesbrecht, Miller, & Müller, 2010; Perlman et al., 2015). These cognitive mechanisms are commonly referred to as executive functions and include impulsivity, action planning and initiation, task switching, and control of emotional expression (Hunter & Sparrow, 2012). Peak prevalence of temper outbursts in typical development lies between the ages of 2 and 5 years (Potegal & Davidson, 2003; Bhatia et al., 1990). This age range coincides with important developments in executive function as well as communication and social skills. Descriptive studies of temper outbursts in young children (e.g. Potegal, Kosorok, & Davidson, 2003) have shown that typical behaviours include crying, screaming, shouting, hitting parents and siblings, aggression towards property such as hitting and kicking walls and furniture, breaking and throwing things, dropping to the floor, and deliberately hitting head against objects (Belden, Thomson, & Luby, 2008; Ö̈sterman & Björkqvist, 2010; Potegal & Davidson, 2003). These behavioural patterns have also been noted in older children referred for inpatient psychiatric treatment and referred to as "rages" or "angry-agitated outbursts" (Carlson, Potegal, Margulies, Gutkovich, & Basile, 2009). In these populations, outbursts are also understood as a failure to develop adequate self-regulatory executive function mechanisms (Carlson et al., 2009 ; Potegal, Carlson, Margulies, Gutkovitch, & Wall, 2009).

The rapid development of non-invasive brain imaging techniques has led to a growing interest in delineating the neurological and cognitive correlates of behaviour patterns associated with genetic disorders (e.g. Järvinen-Pasley et al., 2008; Meyer-Lindenberg, Mervis, & Faith Berman, 2006; Reiss & Dant, 2003). Evidence is emerging for the potential importance of cognitive deficits (brain-based processing difficulties) as an explanation for behavioural phenotypes (Tunnicliffe & Oliver, 2011). For example, recent work by Woodcock, Humphreys, Oliver, & Hansen (2010) using functional magnetic resonance

imaging (fMRI) techniques, has linked temper outbursts in Prader-Willi syndrome (PWS) to cognitive impairments related to task switching. If similar patterns of behaviour are found in LS it could be indicative of an underlying executive function or other cognitive deficit which interacts with environmental contingencies to explain the high prevalence of temper outbursts in LS.

It is important to remember the potential influence of other biological factors on behavior. It is possible, for example, that elevated behavioural difficulties in LS are related to visual impairment, other physical impairments or pain. Ek, Fernell, and Jacobson (2005) have observed higher rates of temper outbursts in children with bilateral optic nerve hypoplasia (a common cause of congenital blindness), in comparison to children with other types of visual impairment. These children also showed patterns of rigidity and dependence on routine, similar to those found in LS and other genetic syndromes. Kenworthy & Charnas (1995) anticipated the potential confounding effect of visual impairments and controlled for this by comparing rates of temper outbursts in boys with LS, with matched controls with similar IDs and visual impairments. The rates of challenging behaviours remained higher in the LS group. Kutsch et al. (under submission) found evidence of an association between physical impairments and temper outbursts in only three out of 19 participants (15.8%). It seems, therefore, that visual impairment and multiple health problems cannot account for outbursts in LS, and further research is needed to understand this phenomenon.

To date there are no detailed descriptions of the phenomenology of behaviour in LS but previous research has established that persistent challenging behaviours, including temper outbursts, are highly prevalent (Arron et al., 2011; Kenworthy et al., 1993; Kutsch et al., under submission). In view of this and the established detrimental effect of challenging behaviour on the lives of people with ID and their carers (Hastings, 2002; Moss et al., 2000) a

research project was developed to map the behavioural phenotype for LS and identify risk factors for the development of challenging behaviours. The current study forms part of this larger research project. Parents and carers of people with LS have identified temper outbursts as a particular problem which is difficult to manage using existing behavioural strategies (Dolinsky et al., 2008). This study therefore seeks to increase understanding of common antecedents to and the behavioural sequence during temper outbursts in LS, with a view to future development of more effective intervention strategies. It replicates the work of Tunnicliffe, Woodcock, Bull, Oliver, and Penhallow (2014), who developed a similar description of temper outbursts in PWS. This included antecedents, setting events, precursor behaviours, perceived emotional states and intervention strategies used by carers. Such a "bottom up" descriptive approach is essential in providing the foundations for further exploration of the aetiology of challenging behaviours. The replication of Tunnicliffe et al.'s work, allows for direct comparisons to be made across the two studies. Similar approaches have also been used in describing temper outbursts in typically developing children (Potegal & Davidson, 2003; Potegal, Kosorok & Davidson, 2003).

Methods

Participants

Eighteen primary carers of seventeen people with LS were interviewed about the behavioural sequence, antecedents and consequences of temper outbursts. To be included in this part of the LS study all respondents had previously confirmed that temper outbursts were a significant challenge. Fourteen mothers, one adoptive mother, and three fathers were interviewed, with one couple being interviewed together. All the people with LS were males and had been diagnosed either by a Paediatrician, Ophthalmologist or Geneticist. Ages ranged from eight to 37 years (M = 18.29 years; n = 9, under 18 years; n = 8, adults 18 years

or over). Adaptive functioning was measured using the Vineland Adaptive Behavior Scale – version II (VABS-II; Sparrow, Cicchetti, Balla, & Doll, 2005) and the results are reported in Table 5. Developmental age, as measured using the adaptive behaviour composite from the VABS, ranged from less than one month to 9 years 9 months (M = 4.18 years; n = 10, less than five years; n = 5, five years or more). All participants were living in the family home with the informant, except for one person with LS who had died six months prior to the interview, and had previously lived only part-time with the informant. The interview schedule was adapted slightly for this informant to reflect circumstances. For example, the informant was asked to describe temper outbursts in the last month of his son's life rather than in the last calendar month.

Table 5: Demographic information and adaptive behaviour scores for participants

Part. Ref.	Gender	Age (yrs)	Additional diagnosis	Adaptive behaviour: standard scores[a]					
				Comm	DLS	Social	Motor	ABC	AE[b]
1	M	25	OCD, Autism	48	52	43	72	47	9:4
2	M	20	-	75	71	80	81	83	9:3
3	M	8	-	90	76	85	67	82	4:8
4	M	8	-	72	71	83	67	74	4:2
5	M	8	-	65	66	66	67	65	3:3
6	M	9	-	72	71	82	61	74	4:4
7	M	28	ASD	21	29	52	44	31	5:7
8	M	9	-	62	62	62	64	62	3:3
9	M	37	-	na[c]	na	na	na	na	-
10	M	36	-	21	21	20	22	20	0:9
11	M	19	-	26	25	32	40	23	<0:1
12	M	15	-	45	30	48	56	39	2:1
13	M	21	Arthritis	69	63	89	70	72	9:9
14	M	9	Haemophilia	62	58	57	56	60	2:5
15	M	30	-	21	21	20	22	20	1:7
16	M	12	-	72	59	64	64	64	5:1
17	M	17	Autism	35	28	43	51	31	1:6

[a]Standard scores from VABS II (Sparrow et al., 2005). Standard scores represent level of functioning and correspond to the following categories: high: 130+; moderate high: 115-129; adequate: 86-114; moderate low: 71-85; low: 70 and below.
[b]ABC, adaptive behaviour composite; AE, age equivalent score for adaptive behaviour composite in years: months; ASD, Autism Spectrum Disorder; Comm, communication; DLS, daily living skills; OCD, Obsessive Compulsive Disorder; Social, socialisation.
[c] na, not available.

Participants in the PWS study (Tunnicliffe et al., 2014) were selected on the basis that one of their triggers for temper outbursts was a change in routine. This was not the case for the LS participants. While this may limit some of the conclusions that can be drawn from the comparisons, the PWS group provide a benchmark group where temper outbursts associated with routine, along with other antecedents, are well documented.

Recruitment

Participants were recruited as part of a larger LS study, via the LS Association in the United States of America (USA), and the Lowe Trust in the United Kingdom (UK). Seven participants were recruited from an existing database held by the Cerebra Centre for Neurodevelopmental Disorders at the University of Birmingham. Eleven participants resided in the USA, five resided in the UK and one family resided in Australia. Participants from the main study who identified temper outbursts as a significant behavioural problem were invited to participate in the temper outburst interview. A participant information sheet (Appendix B) and consent forms (Appendix C) were sent for the main study, with picture-based information (Appendix D) for those people with LS over 16 years of age who could give their own consent. Verbal confirmation of consent was also requested at the start of each interview.

Ethical considerations

Ethical approval was provided for the project by the NHS Research Ethics Committee (Wales-REC-4; Appendix E). All appropriate steps were taken to protect the identity and personal information of participants and data were handled in accordance with the UK Data Protection Act, 1988. Challenging behaviour has previously been shown to cause high levels of parental stress (Hastings, 2002). Interviews required caregivers to describe distressing behaviours in detail and care was taken to keep the research burden on families to a minimum. One participant had been recently bereaved and careful consideration was given to the

appropriateness of conducting this interview. Referral to local support services would have been made for any parent who became distressed during the interview but this provision was not required.

Procedure

All interviews were conducted by the same researcher, by telephone or video call, at a time convenient to the participant. Interview duration ranged from 54 to 86 minutes.

Materials

The semi-structured interview schedule (adapted from Tunnicliffe, 2012; Appendix F) included some open-ended questions designed to allow informants to provide their own description of idiosyncratic behaviours. Narrative descriptions were later verified using follow up questions. Informants were asked to list the behaviours observed during a typical outburst and estimate the frequency, severity and duration of each. Where necessary prompts were provided to ensure consistency in the level of detail given. Questions covered antecedents and consequences of behaviours, as well as a description of the sequence of events and behaviours in outbursts. Variation in setting events, possible triggers and caregiver responses were explored, together with caregiver perceptions of the emotions experienced by the person with LS during an outburst. The final question included a list of common triggers for temper outbursts in the general population and informants were asked to state whether the trigger had caused an outburst in their child during the preceding twelve months. This provided useful data to allow for comparisons with typically developing children and those with other genetic syndromes. The interview schedule consisted of 32 questions. Coding instructions for each question were taken from the original paper by Tunnicliffe et al. (2014, Appendix G) enabling quantitative analysis and direct comparisons with descriptions of outbursts in PWS.

Validity and reliability

The interview schedule was validated by Tunnicliffe et al. (2014) using convergent validity with behavioural diaries which were produced as part of a wider study on PWS. They found that 66-100% of the behaviours reported at interview were also reported in the diary records. Behavioural diaries were not used in the Lowe study to minimise research burden on families, many of whom were also taking part in other aspects of the LS research project. The bottom-up style of questioning included open-ended questions allowing for additional factors associated with LS to emerge. Some of the interview questions, were taken directly from the Challenging Behaviour Interview for which reliability is already established (Oliver et al., 2003; inter-rater reliability: 0.69, test-retest reliability: Pearson's $r = 0.90$). Five of the interviews were coded independently by two researchers to check inter-rater reliability. This was calculated as the percentage agreement on each question of the interview schedule. Agreement ranged between 60 and 100%, with an overall agreement of 85%. Fourteen out of 30 questions had 100% agreement.

Table 6: Categories of behavioural topographies

Categories	**Behaviours included**
Perseverative requests	Repetitive questions, or continuing requests for an item or object, or requests to avoid unwanted activity.
Non-compliance	Refusal to comply with request e.g. to use bathroom, put shoes on etc.
Facial expression	Angry facial expression, "screwing up his face", grimacing, scowling.
Physiological arousal	Red face, sweating, panting (as if out of breath).
Increased motor activity	Pacing, rocking, hand-flapping, twisting fingers, flailing arms and legs, stamping feet, biting or twisting tongue, gritting or clenching teeth.
Dropping	Throwing self to the floor from a seated or standing position, throwing body back in wheelchair.
Talking	Talking to self, talking to other.
Self-deprecating speech	"I'm so stupid", "I'm no good".
Verbal aggression	Verbal threats, insults, swearing at others, argumentative.
Emotional vocalisations	Shouting, yelling, screaming, squealing, growling, saying "I'm scared".
Crying	Sobbing, tearful.
Self-injury	Hitting self, hand-biting, pulling or twisting body parts, hitting self against furniture or hard surfaces.
Physical aggression (towards others)	Hitting, kicking, biting, scratching, pinching, digging nails into skin (drawing blood), headbutting, hairpulling.
Aggression towards property	Hitting or kicking walls, windows, floors, slamming doors, overturning furniture, throwing objects.
Antisocial acts	Spitting, deliberate defaecation, urination, rectal digging, smearing.
Destructive	Tearing, ripping objects, or spoiling an activity (e.g. overturning a game, taking toys from others.)
Avoidance behaviour	Walking away, ask to go to hallway, go to porch, go to bedroom.
Resumes activity	Sudden return to a calm state, goes back to what they were doing before the outburst "as if nothing has happened".
Relationship repair	Apologises, says sorry, asks for a cuddle, asks "mummy happy?", loving, kissing, hugging, makes tea for mother.
Exhausted	Tired, lies down, goes to sleep.
Other	Goes for a walk to self-soothe, has a shower to wash away bad feeling, lies down or falls asleep.

Coding and data analysis

To reduce descriptors of specific behaviours to a manageable number and allow comparison across participants, behaviours were grouped into categories (Table 6). A similar

procedure was used by Tunnicliffe et al. (2014) but a decision was taken not to automatically replicate those categories to allow for the emergence of different patterns of behaviour applicable to LS. Setting events, which increase the likelihood of a behaviour being triggered, were categorised into physiological, environmental and social factors according to McGill, (1999; Table 8).

Data were analysed using Pearson's chi-squared test for comparisons with data on PWS from the Tunnicliffe et al. (2014) paper. Fisher's exact tests were used to verify results where the assumption of five data points per cell was violated. Given the clinical importance of the study and the rarity of the syndrome leading to a relatively small number of participants, a Bonferroni correction ($p < .002$) was considered as too conservative hence an Alpha level of $p < .01$ was adopted.

Results

In this section, a descriptive analysis of the frequency, antecedents, behavioural sequence and consequences of temper outbursts in LS is presented. Where relevant, comparative data are also presented from Tunnicliffe et al.'s (2014) research on temper outbursts in PWS.

Frequency and duration

Data on the frequency and duration of outbursts are shown in Table 7. Temper outbursts were a frequent occurrence for all participants. Two informants expected to see the next outburst within 15 minutes, six in the next hour, six by this time tomorrow and two by this time next week. Only one out of the 17 informants would not expect to see another outburst until this time next month. Typical outbursts lasted less than a minute for two informants, less than five minutes for five informants, and another five informants reported duration of less than 15 minutes. Four informants stated that outbursts typically lasted

between 15 minutes and an hour, and only one stated that typical outbursts lasted more than an hour. Four informants reported duration of the longest outburst in the last month as greater than one hour, five less than an hour, four less than fifteen minutes and four less than five minutes. In total seven informants had ever experienced outbursts lasting more than an hour. The longest outbursts ranged from one and a half to four hours, with six informants reporting outbursts of between two and four hours.

Most informants could identify at least one factor likely to prolong an outburst, with only one parent unable to answer this question. Saying "no", "not getting his own way", or "being forced to do something he did not want to do" was cited by eight informants as the main reason for extended outbursts. "Frustration" was identified by three informants, anxiety by two, and ignoring or not paying sufficient attention by a further four. Obsessive behaviours and an inability to "let go" of an issue were mentioned by two informants.

Table 7: Frequency and duration of temper outbursts.

Response	**Frequency** ***N* = 17**
Timing of the next outburst:	
Within the next 15 minutes	2
Within the next hour	6
By this time tomorrow	6
By this time next week	2
By this time next month	1
Duration of longest outburst in the last month:	
Less than a minute	0
Less than 5 minutes	4
Less than 15 minutes	4
Less than an hour	5
More than an hour	4
Duration of typical outburst:	
Less than a minute	2
Less than 5 minutes	5
Less than 15 minutes	5
Less than an hour	4
More than an hour	1
Length of the longest outburst over one hour (minutes)	***N* = 7**
90 minutes	1
120 minutes	1
180 minutes	3
240 minutes	2

Setting events

Table 8 provides a list of the setting events identified. Twelve informants identified physiological or internal factors as setting events, including tiredness (*n* = 7), hunger (*n* = 4), anxiety/fear (*n* = 5) and physical pain or discomfort (*n* = 5). Low mood (*n* = 1) and thirst (*n* = 1) were also mentioned. Environmental factors included time pressure (*n* = 2), generalised change to routine such as being on holiday (*n* = 5), unfamiliar surroundings (*n* = 3) or arriving home from school (*n* = 1). Many informants noted that high ambient or unexpected noise levels (*n* = 9) or crowded situations (*n* = 4) increased the likelihood that an outburst would be triggered. Social factors also played a part with five informants reporting noticeable differences in the likelihood of outbursts depending on who the person with Lowe syndrome

was with. Embarrassment (n =1) and difficulties in relationships (with family or friends) were identified as setting events by five informants.

Table 8: Physiological, environmental and social setting events.

Setting event	***N*[a]**
Physiological (any of the below list)	**12**
Tiredness	7
Hunger	4
Thirst	1
Low mood	1
Anxiety/fear	5
Physical pain or discomfort	5
Environmental (any of the below list)	**17**
Time pressure	2
General change to routine (e.g. holidays)	5
Coming home	1
Unfamiliar setting	3
Crowds	4
Noise levels high or unexpected	9
Social (any of the below list)	**7**
When with certain person	5
Relationship difficulties	5
Embarrassment	1

[a]Some informants reported more than one setting event within each category.

Antecedents

Table 9 provides information about the principal antecedent for each individual. Nine out of 17 informants indicated that some form of thwarted desire was the most prevalent trigger for an outburst. This included frustrated goals (n = 1), delayed gratification (n = 1), "not getting what he wants" or "not getting his own way" (n = 6), "not being able to do something he wants to do" (n = 2). Two other informants stated that "being asked to do something he does not want to do" leads to most outbursts. Change to routine or uncertainty about expectations provoked regular outbursts for three people with LS. Two informants noted that unexpected change in auditory stimulation such as a car engine stopping, or the TV or radio switching to advertising, triggered outbursts. One informant identified boredom or frustration as the main trigger.

Table 9: Principal antecedents to each participant's temper outbursts.

Participant	Principal antecedents	Proportion of all temper outbursts preceded by principal antecedent	Does antecedent always lead to an outburst?	What is different on occasions when antecedent does not lead to an outburst?
1	Frustrated goals	8/10	No	How decision is presented, negotiation.
2	Change to routine	8/10	No	Environment - no outbursts in public.
3	Not getting what he wants	7/10	No	Environment – no outbursts at school. People - usually with mother or brothers, less often with father.
4	Not getting what he wants	9/10	Yes	Sometimes willing to negotiate.
5	Not getting what he wants	9/10	Yes	N/a
6	Wanting something and being tired	9/10	No	Environment – no outbursts in school or public.
7	Doing something he does not want to do	7/10	Yes	If he wants to go somewhere.
8	Not getting his own way	8/10	No	Environment – no outbursts in school or public. Parents more likely to negotiate in public.
9	Change in routine or expectation	8/10	No	Catch it quickly and acknowledge mistake.
10	Being asked to do something he doesn't want to do	9/10	No	Physical discomfort
11	Boredom or frustration	8/10	Yes	N/a
12	Something stopping (e.g. TV, radio, car engine)	5/10	Yes	Environment – no outbursts at school. People – more with mother than father. Gradual reduction in noise?
13	Not being able to do something he wants to do	9.5/10	Yes	N/a
14	Delayed gratification	10/10	Yes	People – having father around. Environment – no outbursts in school or respite.
15	Uncertainty	9/10	No	People – different carers, better with father.
16	Not getting what he wants	8/10	No	People
17	Noise (e.g. from kitchen), TV or radio going to commercial.	7/10	No	Not clear – possibly volume, or mood.

As well as identifying the most common individual trigger, informants were also asked whether a range of common triggers for outbursts in those without LS had triggered an outburst for the person they cared for in the last twelve months. All reported multiple potential triggers for temper outbursts, with the number of antecedents ranging from five to 18 out of 21 possible antecedents suggested. The results are presented in Figure 2 together with the results from Tunnicliffe et al. (2014) for parents/carers of people with PWS. The graph shows that all the LS informants (n = 17; 100%[2]) reported witnessing a temper outburst that was triggered by the person being asked to do something they did not want to do. This was significantly different (χ^2 = 7.24, p = .007) from reports of this antecedent for PWS informants (9/14; 64%). The next most commonly reported antecedents in LS were change in expectation (n = 16; 94%), change in own routine (n = 14; 82%), not getting something they want (n = 14; 82%) and interruption to preferred activities (n = 14; 82%). Interruption to preferred activities showed a significant difference from the PWS group (χ^2 = 7.04; p = .008). There was no significant difference in adherence to routines triggering temper outbursts, despite this being a selection criterion for the PWS study, suggesting that this may be an important antecedent for LS. All other differences were not statistically significant. Denial of food and disagreements were both reported in 76% (n = 13) of LS participants, and imperfections and concerns that belongings have been stolen were reported in 59% (n = 10) of participants. These results are consistent with the individual antecedents reported above. Other antecedents identified by more than 50% of informants in LS were change to another person's routine, reprimands about food, losing something, and believing that something is lost.

[2] Although small numbers (n < 20) would normally preclude use of percentages, they are shown here and throughout the paper where inclusion aids comparison with results from Tunnicliffe et al. (2014).

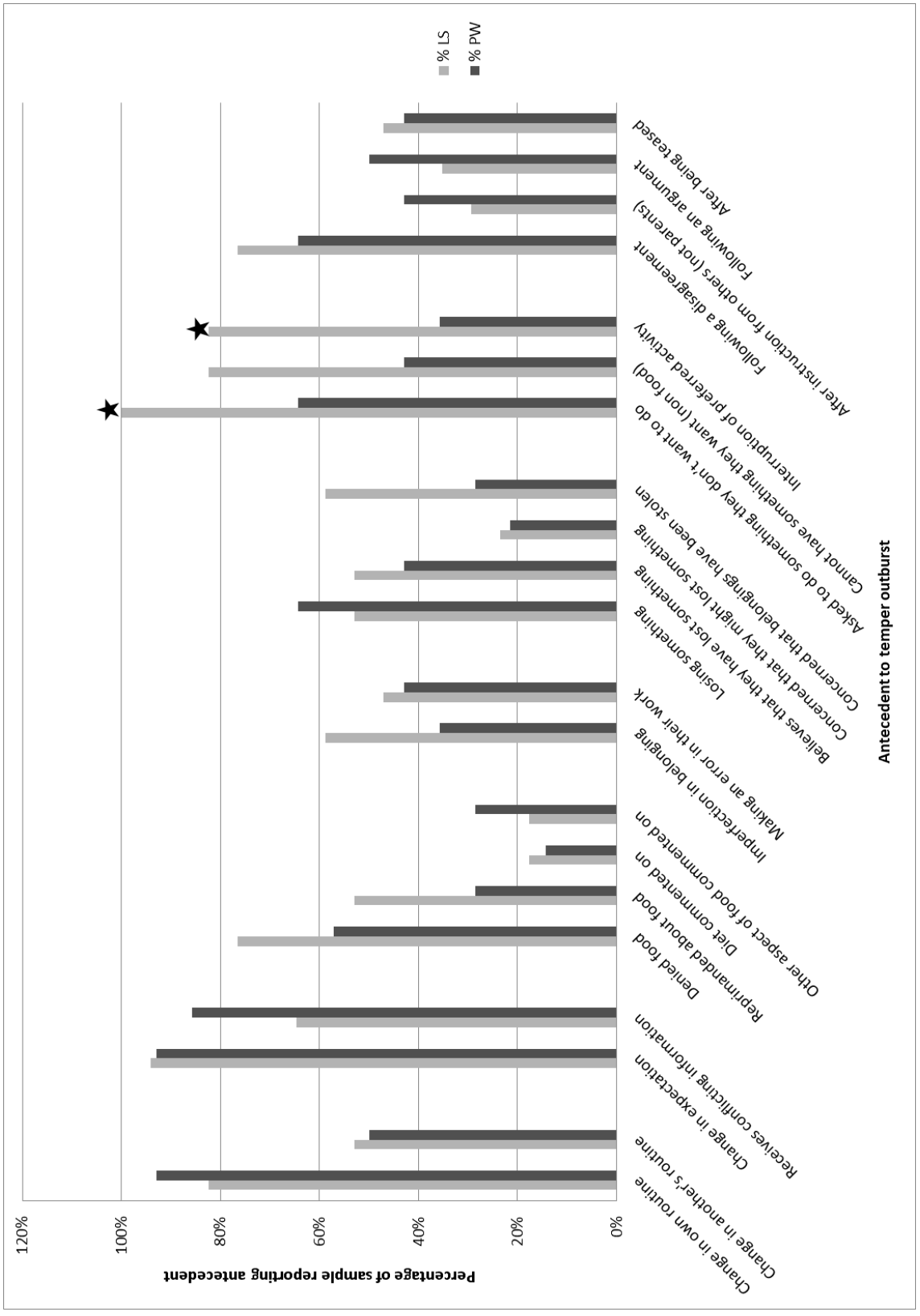

Figure 2: Percentage of informants reporting antecedent to temper outburst in preceding twelve months

★ Statistically significant difference between LS and PWS results.

Sequence of behaviours during an outburst

Individual behavioural sequences, using the coded behavioural topographies from Table 6, are shown in Figure 3. These are based on a description of the last bad temper outburst observed during the month preceding the interview. In LS 9/17 (53%) participants showed a predictable pattern of behaviours during temper outbursts. This compares with 100% (14/14) of the PWS participants described in Tunnicliffe et al. (2014). This difference is statistically significant (Fisher's exact, $p = .003$). Precursor behaviours which alert parents/carers to a potential outburst were identified by all LS informants, but only 7/14 of the PWS informants (Fisher's exact, $p = .001$). Seven of the LS informants reported perseverative requests or demands preceding outbursts, whilst four informants mentioned emotional vocalisations as a warning sign (e.g. shouting or yelling). Other precursors included: self-injury, verbal or physical aggression towards others, non-compliance with requests, increased motor activity or talking to self (verbalising thoughts of displeasure). Precursors were always present for 6/17 LS participants.

The most common behaviours during outbursts were emotional vocalisations ($n = 15$) and physical aggression ($n = 15$). Aggression to property such as kicking or hitting walls or throwing objects was reported by 12 informants, and verbal aggression (e.g. swearing or shouting directed at another person) was reported in six cases. Six participants exhibited self-injury. Of those showing externally directed aggression 14 showed multiple forms of aggression, with seven participants displaying aggression towards others and towards property and four displaying verbal and physical aggression towards other people and towards property. Destructive behaviour involving ripping, tearing or destroying objects was displayed by five participants. Crying, which is distinguished from other emotional vocalisations, was reported in the middle and towards the end of outbursts by eight

informants. This contrasts with reported crying behaviour in PWS (Tunnicliffe et al., 2014) which occurred at the start and end of outbursts, but never in the middle. Dropping also occurred both in the middle and at the end of outbursts. Specific behaviours coded as "antisocial" were not identified by Tunnicliffe et al. (2014) but occurred for five participants in the LS sample. These behaviours included spitting (n = 2) and deliberate urination, defaecation and smearing (n = 4), and were described as especially distressing by informants.

Behaviours during the end phase of an outburst showed two distinctive patterns. Eleven of the seventeen informants reported relationship repair behaviours including apologising, asking for a hug or seeking reassurance from caregivers. Seven informants reported that the person with LS would suddenly go back to their previous activity and emotional state as if nothing had happened.

The most frequently reported perceived emotions during an outburst were frustration (n = 12) and anger (n = 8). These sometimes occurred together. Anxiety or fear was reported by three informants and two informants felt that the person with LS was feeling out of control. Only one informant had a suspicion that the person with LS was excited and positively enjoying the outburst, and the same informant described the emotion at the end of the outburst as satisfaction.

Participant	1	2	3	4	5	6	7	8	9
Predictable chain?	Yes	No	Yes	No	Yes	Yes	Yes	No	No
Precursor behaviours	Pers. Reqs.	Pers. Reqs.	Non-comp.	Verbal Aggr.	Pers. Reqs.	Talking	Self-injury	Emot. Vocs.	Phys. Aggr.
Behavioural sequence during outburst	Incr. motor activity	Incr. motor activity	Verbal Aggr.	Destruct	Self-injury	Aggr. to property	Incr. motor activity	Phys. Aggr.	Non-comp.
	Self-injury	Facial express.	Phys. Aggr.	Aggr. to property	Facial express.	Drop	Pers. Reqs.	Talking	Phys. Aggr.
	Emot. Vocs.	Aggr. to property	Aggr. to property	Verbal aggr.	Emot. Vocs.	Emot. Vocs.	Emot. Vocs.	Verbal aggr.	Verbal aggr.
	Phys. arousal	Phys. Aggr.	Verbal aggr.	Talking	Drop	Anti-social	Self-injury	Aggr. to property	Emot. Vocs.
	Phys. Aggr.	Aggr. to property		Emot. Vocs.		Phys. Aggr.	Phys. Aggr.	Destruct	Anti-social
	Verbal aggr.	Emot. Vocs.		Phys. Aggr.		Crying	Crying		Self-injury
	Crying	Self-injury		Drop					Destruct
	Talking	Avoid							Drop
	Self-injury	Talking							
		Crying							
End phase	Talking	Resume activity or Exhausted	Exhausted	Rel. repair	Incr. motor activity	Rel. repair	Rel. repair	Talking	Talking
	Rel. repair							Rel. repair	Resume activity
Longest outburst	3 hrs	<5 min	<15 min	<15 min	<5 min	<15 min	<5 min	<15 min	<1 hr

Figure 3: Sequence of behaviours shown by each participant during temper outburst

Participant	10	11	12	13	14	15	16	17
Predictable chain?	Yes	No	No	Yes	Yes	No	Yes	No
Precursor behaviours	Emot. Vocs.	Emot. Vocs.	Pers. Reqs.	Inc. motor activity	Pers. Reqs.	Pers. Reqs.	Pers. Reqs.	Emot. Vocs.
Behavioural sequence during outburst	Incr. motor activity	Incr. motor activity	Aggr. to property	Talking	Destruct	Self-injury	Emot. Vocs.	Incr. motor activity
	Emot. Vocs.	Emot. Vocs.	Emot. Vocs.	Self-deprecating speech	Emot. Vocs.	Emot. Vocs.	Phys. arousal	Emot. Vocs.
	Self-injury	Aggr. to property	Phys. Aggr.	Verbal aggr.	Incr. motor activity	Aggr. to property	Drop	Phys. arousal
	Phys. Aggr.	Phys. Aggr.	Aggr. to property	Emot. Vocs.	Aggr. to property	Phys. Aggr.	Phys. Aggr.	Phys. Aggr.
	Aggr. to property	Incr. motor activity	Anti-social	Aggr. to property	Phys. Aggr.	Anti-social	Crying	Aggr. to property
	Drop	Phys. Aggr.		Destruct	Crying	Phys. arousal		Self-injury
				Crying	Anti-social			Crying
					Self-injury			
End phase	Phys. Arousal	Rel. repair	Avoid	Other	Emot. Vocs.	Rel. repair	Rel. repair	Rel. repair
		Resume activity	Resume activity	Rel. repair	Rel. repair	Resume activity	Resume activity	Resume activity
Longest outburst	<5 min	<5 min	3 hrs	4 hrs	<1 hr	<1 hr	<1 hr	3 hrs

Figure 3: Sequence of behaviours shown by each participant during temper outburst

Management strategies used by caregivers

A range of different intervention strategies were reported by informants. At the precursor stage the most successful strategy was distraction or redirection to an alternative activity for seven participants. Other strategies included calm reasoning (*n* = 2), removal of choice (*n* = 1), providing attention (*n* = 1), offering help (*n* = 1), reiterating clear routine (*n* = 1), removing other children from the room (*n* = 1) or giving in (*n* = 1). Informants estimated that success rates for avoidance of an outburst were between 40% and 90% at this stage. Only one informant felt that there was nothing that could be done even at the precursor stage. Critical points at which intervention was no longer possible were varied and included the start of physical aggression, aggression towards property, dropping to the floor, or a change in the pitch, tone or volume of vocalisations. Table 10 gives a list of principal strategies and the success rate for each.

Table 10: Principal strategies and success rates.

Preventative strategy at precursor stage	***N***	**Success rate**
Discussion/calm reasoning/negotiation	3	60-80%
Distraction/redirection (incl. use of humour)	7	50-90%
Consequences (e.g. removal of tangible or aversive consequence)	1	80%
Provide attention/offer help	2	40-80%
Give in to demands	4	70-100%
Withdraw person with Lowe syndrome from situation	2	90%
Nothing works	1	0%
Principal strategies during outburst		
Discussion/calm reasoning/negotiation	4	0-60%
Distraction/redirection (incl. use of humour)	3	0-50%
Consequences (e.g. removal of tangible or aversive consequence)	3	0-60%
Ignore/withdraw attention	3	0%
Withdraw person with Lowe syndrome from situation	3	0-60%
Restraint	2	Harm reduction. [a]
Other strategies described by individual informants	***N***	**Success rate**
Shouting	1	Not reported
Yelling "stop"	1	Not reported
Singing to him	1	Not reported
Provide choice	2	Not reported
Limit choice	1	Not reported

[a] 0% success in stopping outburst but used to prevent physical harm to self, carer, other person or property.

During an outburst, the chances of successful intervention reduced and the main aim of intervention at this stage appeared to be harm reduction, either to the person with LS, others at risk of aggression, or to avoid damage to property. Removal of the person with LS to a quiet location or withdrawal of the caregiver avoided further escalation but did not immediately stop an outburst. Seven informants found that the only way to stop an outburst was to accede to demands, although this was not always successful. Redirection, humour or distraction was reported to be successful in 60-90% of outbursts if the intervention was made early enough. Five informants felt that the only thing that brought about an end to the outburst was time and "waiting it out". Restraint was reported as a strategy employed for two people with LS who were at risk of serious self-harm if left alone.

The most common reason for variation in intervention strategies was whether the outburst took place at home or in a public place (n = 10). In the home, it would be easier to move the person with LS to a safe space or for the carer to withdraw. Concern for the judgement of others and risk to others' safety were given as reasons for variation in strategies when away from home. Informants also reported using a different response if the person with LS was hurting someone (including themselves). They would then be more likely to intervene directly rather than ignore behaviour. Some informants would need to withdraw for their own safety when the person with LS became aggressive. They might then need to defer the activity (e.g. changing bed sheets) until the person was in a calmer and more cooperative mood.

Comparison with Prader Willi Syndrome (Tunnicliffe et al., 2014)

The two samples were compared to check for significant differences in the mean age or adaptive abilities. No significant differences were found in adaptive abilities based on VABS adaptive behaviour composite (Mann-Whitney U, p = 0.377), but a difference was

found in the chronological age profiles of the two samples (t (29) = -1.44; p = .018), with a higher mean age in years reported for PWS. This age difference can be accounted for by the wider age range of adults in the PWS sample due to lower life expectancy in LS. When a comparison was made based on age group (< 18 years; ≥18 years) no significant difference was found between the two groups (p > .05).

The following differences were noted between the two samples. Crying (Fisher's exact, p = .008) and running away (Fisher's exact, p = .010) were more frequently reported in the PWS group. As previously reported physical aggression towards others was more frequently seen in LS (Fisher's exact, p = .010). Anti-social acts (spitting, deliberate defaecation or urination, or smearing) was not reported at all in descriptions of temper outbursts in PWS, but was reported by five informants in the LS study. This difference however was not statistically significant (Fisher's exact, p =.036). There was no significant difference in outbursts occurring in response to routine changes, but differences in the pattern of antecedents reported during the last twelve months were significant at p <.01 for "asked to do something they don't want to do" (χ^2 = 7.24; p = .007) and for "interruption of preferred activity" (χ^2 = 7.04; p = .008). Both these factors were reported more frequently in LS than in PWS. The sudden resumption of activities as if nothing had happened was not reported at all in PWS but was spontaneously mentioned by eight informants in the LS study (Fisher's exact, p = .003). No other significant differences were found.

Discussion

It has been established elsewhere that a higher prevalence of temper outbursts in people with LS than for people of typical development and people with intellectual disabilities is evident (Kenworthy & Charnas, 1995; Dolinsky et al., 2008; Kutsch et al., under submission). The primary aim of this investigation was to generate a description of temper

outbursts in LS based on informant accounts. Eighteen informants provided detailed accounts of the antecedents, behavioural and emotional sequence, and the consequences of temper outbursts in nine children and eight adults with LS. All participants were eight years or older, putting them above the expected chronological age of five years for reduction or cessation of temper outbursts in typically developing children (Potegal & Davidson, 2003). Developmental age, however, as measured using age equivalent scores from the VABS, showed that more than half the participants had a developmental age of below five years. The topographies of behaviour during outbursts in LS bear marked similarities to those described for temper outbursts in typically developing children aged 2-5 years (Österman & Björkqvist, 2010; Potegal & Davidson, 2003), and "angry-agitated outbursts" in paediatric inpatients (Carlson et al., 2009; Potegal et al., 2009). In pre-school children, Wakschlag et al., (2007) suggested that both quality of behaviours (severity) and pervasiveness (frequency and duration) should be considered when determining the degree of pathological emotional dysregulation. In the current study of people with LS most informants reported outbursts as a daily occurrence and nearly half reported them as hourly. Durations varied between less than five minutes and over an hour, compared with an average duration of three minutes in typically developing children (Potegal, Kosorok, & Davidson, 1996). The high prevalence of physical aggression towards others as well as verbal aggression and aggression towards property gives a clear indication of severity. Physical aggression took several forms, but included hitting, kicking, biting, scratching, pinching and hair pulling, of sufficient force to cause bleeding and bruising. Verbalisations included swearing, screaming and shouting at caregivers or other family members. Frequent reports of aggression towards property included door slamming, hitting and kicking windows and doors, or throwing or overturning furniture. Sometimes this caused permanent damage to property. Given this level of

aggression it is understandable why informants might describe such behaviours as a major challenge to their own wellbeing as well as that of the person with LS for whom they care.

The occurrence of behaviours such as smearing, deliberate defaecation or urination and spitting was a particularly distressing feature of outbursts for several informants. These behaviours were not noted in the PWS study (Tunnicliffe et al., 2014). The reasons for these behaviours are unclear. From a functional behavioural perspective, one possible explanation could be the increased likelihood of a reaction by carers to more extreme forms of behaviour. This also applies to extreme aggression towards carers, attacks on siblings or strangers, or dangerous behaviours such as kicking windows. This hypothesis was not tested as part of the current study but was proposed by some informants as an explanation for extreme behaviours.

When exploring the aetiology of temper outbursts in genetic syndromes it is important to consider the role of physical differences. LS is characterised by significant physical as well as intellectual disability (Lewis et al., 2012) with associated limitations to independent access to food and drink, and the possibility of physical pain and discomfort. Physiological setting events were commonly identified as increasing the likelihood of an outburst, including hunger, thirst, and pain. It is also interesting to note the environmental factors which impact on outbursts. Change in ambient noise or sudden changes in auditory stimuli were reported by more than half the respondents as increasing the likelihood of an outburst. Increased sensitivity to noise (hyperacusis) has been noted as a feature of other genetic disorders such as Cri-du-chat, and Williams syndromes but was not previously found to be associated with LS (Cornish & Pigram, 1996). Increased physiological arousal or anxiety caused by unusual sensitivity to sensory stimuli has been noted as a potential contributory factor in challenging behaviour in other disorders such as autistic spectrum disorders (ASD; Grapel, Cicchetti, & Volkmar, 2015) and Williams syndrome (John & Mervis, 2010). Another interesting aspect

of environmental setting is the reported absence of temper outbursts outside the home, and a difference in behaviours dependent on who the carer is (e.g. mother or father). Carlson et al. (2009) noted that 73.2% children admitted to psychiatric hospital for treatment of severe angry-agitated outbursts showed no or only one rage during their hospital stay. This suggests that some individuals may have a degree of control over the emotional and behavioural response to the same external triggers depending on the context in which they occur, or that the emotional salience of events differs between contexts. The "context-specificity" of outbursts may offer scope for environmental interventions to reduce the frequency or intensity of outbursts but further research would be needed to understand why self-regulation is possible in some circumstances but not in others.

This study has highlighted the potential importance of frustration intolerance as a factor in temper outbursts in LS. More than half of respondents spontaneously identified some form of thwarted desire as the principal antecedent (see Table 9). A statistically significant difference was also found between antecedents to temper outbursts in LS and PWS for "being asked to do something they did not want to do" and "interruption to preferred activity". The absence of difference in reports of routine change as an antecedent is interesting given that the PWS group were selected on this basis, and the LS group were not. It suggests that routine change may be an important challenge for people with LS. This similarity between the groups also adds strength to the argument that thwarted desire as a trigger may be particular to Lowe syndrome. The concordance between the open-ended questions about individual triggers and the responses to common antecedents lends further credence to this argument. Frustration was also the most frequently reported emotion during an outburst.

The relevance of frustrated desire was supported by the findings of Kutsch et al., (under submission) that temper outbursts in LS were most frequently triggered by denial of access to tangibles. In the current study temper outbursts were commonly triggered by denial of food. This has been reported as a characteristic response in PWS (Welham et al., 2015) in which failure to detect satiety is a known problem in appetite control, but is not a previously identified difficulty in LS. It is important to recognise that the same behaviours and antecedents (temper outbursts in response to denial of food) may result from different neurocognitive and genetic pathways and there may be a different explanation for the same phenomenon in LS. It may be that problems with satiety are not present in LS but denial of food is another form of thwarted desire which people with LS find difficult to tolerate.

The popular understanding of temper outbursts in young children is of the "spoilt child" who expresses extreme anger when their desires are not met with an immediate response by parents. Österman & Björkqvist (2010) described tantrums in typical development as a response to frustrated desire. They noted that the most rapid decline in outbursts occurs at the age of around four years when children start to develop more sophisticated language to express their emotions, including anger and frustration. It also coincides with the development of other social skills which enable them to get their needs met. In this study, there was no significant association between the communication or social abilities of participants and the frequency or duration of outbursts, but the small sample size may have led to a type II error and finding no association where one might conceivably exist. The current study is based on narrative descriptions from parents/caregivers, which may be influenced by the dominant discourse on the aetiology of temper outbursts. This would not, however, explain the apparent difference in the importance of thwarted desire between PWS

and LS as both groups of informants might be expected to be influenced in the same way by popular narratives about temper outbursts.

The inability to tolerate frustration in young typically developing children and in older paediatric psychiatric inpatients is thought to be due to immaturity of cognitive mechanisms which control and regulate emotions and behaviour, known as executive functions (Hunter & Sparrow, 2012). Similar deficits may also be implicated in temper outbursts in genetic syndromes. Executive functions cover a range of cognitive abilities including judgement, planning, impulsivity, behavioural inhibition and task switching. Change to routine has been noted as a potential trigger for temper outbursts in a number of genetic syndromes including PWS, LS, fragile X and Smith Magenis syndromes (Bull, Oliver, & Woodcock, 2017). A link has also been made between intolerance of change and repetitive behaviour as a precursor to outbursts (Moss et al., 2009). In the current study, perseverative requests were frequently reported as a precursor and change to routine or expectation was reported as antecedent to temper outbursts. In comparing antecedents and behavioural patterns in temper outbursts in PWS and LS it is important to note that the PWS participants were selected on the basis that change in routine had previously triggered outbursts (Tunnicliffe et al., 2014). Given that routines as a trigger did not differ between groups, this means that the apparent similarity between the antecedents to temper outbursts may underestimate the importance of preference for routine in LS.

In PWS a strong association has been found between task switching deficits, change to routine and temper outbursts (Woodcock et al., 2010). The cognitive challenge of moving from a well-rehearsed sequence of behaviour to adapt to a new task is thought to increase anxiety and overwhelm emotional coping skills. Recent findings from a study by Manning et al. (2016) have suggested possible involvement of the vagus nerve in emotion regulation in

PWS which offers new avenues for future research in this area. Preliminary results from the wider LS study suggest that executive function difficulties in the realms of inhibition, emotion regulation and working memory are also significantly associated with temper outbursts in LS ($r = 0.76$; $p = 0.001$; Waite, Crawford, Kutsch & Oliver, in preparation). Recent MRI studies show abnormalities in the brains of people with LS (Allmendinger et al., 2014) which may add weight to the argument for involvement of neurological difference in behavioural aetiology. The discovery of task switching difficulties in PWS has led to the development of promising interventions to support transitions between activities and reduce the incidence of outbursts (Bull et al., 2017). Although the use of vagus nerve stimulation to manage behavioural difficulties in PWS may be controversial, the study by Manning et al. (2016) may offer important insights into neural mechanisms associated with emotion regulation. A better understanding of executive functioning in people with LS is an important next step in developing effective interventions for management of temper outbursts in this group.

The semi-structured interview schedule included open-ended questions to allow for the emergence of a detailed descriptive account of temper outbursts in LS. It also provided sufficient structure for the collection of frequency data for comparison with behaviours in other populations. The interview schedule had the advantage of being previously published in a peer-reviewed journal and had been validated by Tunnicliffe et al. (2014) using convergent validity with behaviour diaries. Direct replication of a published study allowed for close comparisons to be made between two genetic syndromes associated with a high prevalence of temper outbursts. The schedule had been written however specifically for research on PWS and may therefore have overemphasised the importance of food related transactions which are a known problem in that population. The list of potential antecedents to temper outbursts used in the final question may have limited the potential responses unnecessarily or given a

false impression of the frequency of these triggers as the question asked if the situation had ever triggered an outburst. Concordance with responses to the earlier open-ended question about common antecedents suggested however that the responses could be relied upon to some degree. Reliance on informant report is also potentially problematic as caregivers will be influenced by their own personal perspectives, their attributions about the cause of behaviours that challenge, and their own levels of stress tolerance. Objective observation of temper outburst behaviours in either an experimental or naturalistic setting would provide additional scientific rigour to the descriptive accounts of temper outbursts. Naturalistic observations of infrequent behaviour by independent researchers would be very time consuming however, and the ethics of experimental provocation of highly emotional and potentially harmful outbursts is questionable.

Statistical comparisons have been made with PWS but should be treated with caution as the number of participants in each study is small. Percentages have been used to aid comparison between the two studies but could be misleading with such small numbers. It is also important to recognise that both samples only included informants who had identified temper outbursts as a significant problem. It might have been beneficial to interview caregivers of people with LS who did not show behaviours that challenge in order to understand how they were able to regulate their emotions more effectively. It would also have strengthened the research to have included interviews with the people with LS themselves about their own experience of temper loss, although this would have excluded those with very limited communication abilities. Alternative research methodologies were considered such as qualitative interviews using Interpretative Phenomenological Analysis, or Critical Incident Technique to assess situational factors which help or hinder emotion regulation. The benefits of being able to make direct comparisons to an existing published

study were thought to outweigh the disadvantages of the semi-structured informant interview approach. These alternative methods could however be useful in future research to explore the phenomenology of temper outbursts in LS and in other genetic syndromes.

Conclusions

This paper provides an initial descriptive analysis of temper outbursts in LS. The small sample size and nature of the study do not allow for definitive conclusions to be drawn about the nature or aetiology of outbursts. Plausible hypotheses have been generated based on parental attributions and comparisons with outbursts in other populations. Note has been made of the high prevalence of aggression in outbursts in LS, and the frequency of thwarted desire as a possible trigger. These observations may be worthy of further investigation in future research on LS. Experimental functional analysis of temper outbursts in LS could be a potential research option, with appropriate ethical safeguards, and could have direct benefits for individual behavioural interventions. Alternatively, detailed laboratory observations or video-recorded naturalistic observations similar to those employed by Oliver, Woodcock, & Humphreys (2009) to study outbursts in PWS, could offer an important addition to descriptive accounts. This would only be appropriate where frequency of outbursts is high and temper outbursts could be observed without need to artificially trigger an episode. Detailed diary studies would be a potential alternative and could be used alongside measurement of physiological arousal using heart rate monitors or measurement of cortisol levels. These methods have been utilised with some success in studies of PWS (Bull, Oliver, Tunnicliffe, & Woodcock, 2015).

One of the important aims of investigating challenging behaviours in genetic disorders is to develop effective preventative and management interventions to reduce distress for the individual and their carers. Further research is needed to determine whether under-developed

emotion regulation or other cognitive mechanisms may be contributing to the frequency and severity of outbursts in LS. Evidence is emerging from ongoing research to suggest that this is the case. Depending on the outcome of such research, successful interventions could be developed to strengthen emotional control or to reduce the cognitive challenge of particular situations or tasks. Promising research has been developed for managing task switching deficits in PWS (Bull et al., 2017) and also in the use of effective parenting techniques to teach emotional recognition and control to preverbal typically developing children (e.g. Douglas, 2007). With better understanding of the gene-environment-behaviour pathway (Tunnicliffe & Oliver, 2011) it is possible that these techniques could be adapted for children with LS and other syndromes in which temper outbursts are frequent.

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PUBLIC DISSEMINATION DOCUMENT:

THE PHENOMENOLOGY OF TEMPER OUTBURSTS IN INTELLECTUAL DISABILITIES

The phenomenology of temper outbursts in intellectual disabilities

Temper outbursts are a common form of challenging behaviour in people with intellectual disabilities which can restrict access to social, educational and occupational opportunities (Lowe et al., 2007). The prevalence of outbursts is especially high in some genetic syndromes such as Cri-du-chat, Smith-Magenis, Lowe and Prader Willi syndromes. Recent research in Prader Willi syndrome, has shown a link between temper outbursts and a cognitive difficulty in switching between tasks (Woodcock, Humphreys, Oliver, & Hansen, 2010). This has led to the development of promising clinical interventions (Bull, Oliver, & Woodcock, 2017). Further research on temper outbursts in other syndromes has the potential to make an important difference to the lives of people with intellectual disabilities and their carers.

This study explored the nature and function of temper outbursts in intellectual disabilities with specific reference to Lowe Syndrome, a rare genetic disorder affecting 1 in 500,000 people, mostly males (Kenworthy & Charnas, 1995). The study was in two parts: a systematic review of the functional behavioural literature; and a descriptive analysis of temper outbursts in Lowe syndrome from the perspective of caregivers. This research formed part of a larger study into the characteristic patterns of behaviour in Lowe syndrome by the Cerebra Centre for Neurodevelopmental Disorders at the University of Birmingham.

Systematic Review

Most literature on challenging behaviours in intellectual disabilities over the last thirty years has come from a functional behavioural perspective. In this model challenging behaviours are understood to perform a function for the individual, providing a way for them to get their needs met, although often at a cost to themselves and their carers (Emerson & Einfeld, 2011). Behaviours which initially occur by chance can become reinforced inadvertently by the way in which carers respond. For example, a parent may try to distract a

distressed child by offering a toy or food, leading to tangible reinforcement of the crying behaviour. Other common reinforcers are attention or escape from task demands. Experimental methods have been developed to test the function of an individual's challenging behaviour and intervene more effectively. These experiments are done on an individual basis but it was hoped that by examining many studies it might be possible to discern a pattern for the function of temper outbursts more generally. A systematic review of the functional behavioural literature was therefore carried out to test the hypothesis that temper outbursts are more likely to occur when access to tangible items, such as toys or food, is denied.

Method. Temper outbursts can include a wide range of behaviours including hitting, kicking, self-injury, property destruction, crying, shouting, non-compliance, dropping to the floor or running away. Operational definitions of all behaviours associated with a clearly differentiated single function of behaviour were therefore recorded. A total of 142 papers and 338 participants were found which met the inclusion criteria. Statistical analysis (Pearson's chi-squared test) was used to determine whether any of the behaviours were more likely to occur in response to one of three functions – tangible, attention or escape from demands. A hypothetical temper outburst construct was also tested made up of a cluster of behaviours commonly associated with outbursts.

Findings. Statistically significant differences were found between functions for self-biting (most frequently reinforced by access to tangible items) and tearing objects (most frequently reinforced by attention). No other significant associations were found. These behaviours are interesting in themselves but provided no support for the hypothesis that temper outbursts are more likely to be tangibly reinforced. There was no significant difference between functions for the temper outburst construct. Internal states such as emotions are not included as part of functional analysis as they are not directly observable and are therefore considered inaccessible to scientific enquiry. As temper outbursts are

closely linked to changes in emotion states this could account for the absence of clear findings from the review.

Empirical Research

The purpose of the empirical research was to describe the behavioural sequence, common triggers, consequences and strategies used by caregivers for temper outbursts in Lowe syndrome. Lowe syndrome, also known as oculocerebrorenal syndrome, is a rare chromosomal (genetic) disorder which is caused by a mutation on the X-chromosome. It affects mostly males who are born with cateracts (clouding of the lens) in both eyes, hypotonia (poor muscle tone), and kidney problems. All those affected have some degree of intellectual disability ranging from mild to profound.

Methods. A semi-structured interview was used to explore temper outbursts in Lowe syndrome. The parents/carers of 9 children (<18 years) and 8 adults (18 years or older) were interviewed and asked to describe the sequence of behaviours before, during and after an outburst. They were also asked what strategies they had tried to manage the temper outbursts. Findings from the interviews were compared to similar research on temper outbursts in Prader Willi syndrome (Tunnicliffe, Woodcock, Bull, Oliver, & Penhallow, 2014).

Findings. Temper outbursts amongst the Lowe syndrome participants were found to be of high frequency, occurring at least daily for 14/17 participants, with typical duration between one and 15 minutes. The most striking feature of the outbursts was the degree of physical aggression and property destruction reported by parents and carers who were frequently left with bruises or bleeding after providing personal care. Distraction appeared to be the most helpful management strategy but this was not always effective and often the only solution was to move the person to a quiet place and allow the outburst to take its course.

The pattern of behaviours was very like that described in Prader Willi syndrome (Tunnicliffe et al., 2014). The only significant difference was that the most common trigger for Lowe syndrome participants was being told "no", or being asked to do something they did not want to do. In Prader Willi syndrome most outbursts were triggered by a change in routine which was later found to be linked to a task switching difficulty (Woodcock, Humphreys, Oliver, & Hansen, 2010). Change in routine is also a common trigger in Lowe syndrome. This suggests that there may be a similar or slightly different mechanism underpinning outbursts in Lowe syndrome, involving a combination of functional reinforcement and cognitive factors, which would be worthy of further investigation.

Implications for clinical and research agenda

Temper outbursts present a significant management challenge for carers of people living with some genetic syndromes, including Lowe syndrome and Prader Willi syndrome. The degree of emotional arousal, physical aggression, property damage and self-injury also presents a risk to the wellbeing of the individuals themselves. To date the functional behavioural literature has offered little in the way of generalizable solutions to this problem, and the systematic review undertaken here was unable to advance understanding of this issue. Recent developments in the biological literature on genetic syndromes suggests that there are patterns of behaviour which are more likely to occur in one syndrome compared to another, which are called behavioural phenotypes. These provide a challenge to the functional behavioural approach which is unable to adequately account for these between-syndrome differences. Equally the biological model cannot account for within-syndrome difference leading to calls for an integrated model (Tunnicliffe & Oliver, 2011). Promising developments in understanding the pathway between genetic mutation and behaviour via specific cognitive deficits, such as that found in Prader Willi syndrome appear to offer an important way forward to a greater understanding of temper outbursts. Better understanding

of neurological factors impacting on temper loss in genetic syndromes could also contribute to management of aggression and disinhibited behaviour caused by neurological damage, such as traumatic brain injury or neurodegenerative disease. The detailed description of behaviours in Lowe syndrome provides an important starting point for developing effective interventions for this syndrome group and for expanding knowledge of how to manage other types of emotional dysregulation.

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Appendix A: Topographies of behaviour from functional analytic literature review

APPENDIX A: Topographies of behaviour from functional analyses

Reference	Function	Name	Gender	Age	Autism?	Hitting	Pushing	Kicking	Biting	Headbutting	Spitting	Any physical aggression	Headbanging	Face hitting	Self-hitting	Striking body against object	Self-biting	Scratching or pinching	Dangerous acts	Any self injurious behaviour	Throwing objects	Tearing objects	Pulling objects	Hitting objects	Damaging furniture	Any destructive behaviour	Elopement	Crying	Loud vocalisations	Perseverative speech	Dropping	Non-compliance	Tantrums	Any disruptive behaviour	Inappropriate sexual behaviour	Stereotypy
Allen, Baker, Nuernberger & Vargo (2013)	Attention	Nelly	F	48			✓					✓														✓			✓					✓		
Anderson, Freeman & Scotti (1999)	Attention	Ann	F	8		✓	✓	✓	✓			✓								✓	✓	✓		✓		✓										
Anderson, Freeman & Scotti (1999)	Attention	Tim	M	10		✓	✓	✓	✓			✓	✓	✓						✓	✓	✓				✓										
Asmus et al., (1999)	Escape	Luke	M	3		✓	✓		✓			✓	✓							✓	✓		✓			✓	✓		✓			✓	✓	✓		
Asmus et al., (1999)	Escape	Todd	M	4		✓	✓					✓	✓							✓	✓	✓			✓	✓	✓		✓			✓	✓	✓		
Asmus et al., (1999)	Escape	Trevor	M	5		✓	✓	✓	✓			✓	✓				✓			✓	✓	✓		✓		✓	✓		✓			✓				
Athens & Vollmer (2010)	Escape	Justin	M	7		✓	✓	✓	✓	✓		✓								✓	✓			✓		✓								✓		
Athens & Vollmer (2010)	Escape	Henry	M	8	Y	✓	✓	✓	✓			✓								✓	✓	✓				✓										
Athens & Vollmer (2010)	Tangible	Corey	M	9	Y	✓	✓	✓	✓		✓	✓								✓	✓	✓				✓										
Athens & Vollmer (2010)	Attention	Kenneth	M	6	Y	✓	✓		✓			✓								✓	✓					✓										
Athens & Vollmer (2010)	Tangible	Lana	F	4	Y	✓	✓	✓	✓			✓																								
Athens & Vollmer (2010)	Attention	George	M	10	Y	✓	✓	✓	✓			✓								✓	✓					✓										
Athens & Vollmer (2010)	Tangible	Clark	M	12	Y	✓	✓	✓	✓			✓																								
Bailey, McComas, Benavides & Lovascz (2002)	Attention	Jack	M	24		✓						✓								✓	✓			✓		✓										
Betz. Fisher, Roane, Mintz & Owen (2013)	Tangible	Casey	F	5														✓									✓						✓		
Betz. Fisher, Roane, Mintz & Owen (2013)	Tangible	Gordon	M	6		✓	✓	✓	✓			✓								✓	✓			✓		✓										
Betz. Fisher, Roane, Mintz & Owen (2013)	Tangible	Mikey	M	9		✓	✓	✓	✓			✓																								
Boesch, Taber-Doughty, Wendt & Smalts (2015)	Tangible	Mike	M	14	Y								✓							✓	✓			✓		✓										
Borrero & Vollmer (2002)	Tangible	Dan	M	9		✓	✓	✓	✓			✓								✓	✓			✓		✓										
Borrero, Francisco, Haberlin, Ross & Sran (2007)	Tangible	Todd	M	9		✓	✓	✓	✓			✓																								
Borrero, Vollmer & Borrero (2004)	Escape	Tobias	M	13		✓	✓	✓	✓			✓								✓	✓			✓		✓										
Borrero, Vollmer & Wright (2002)	Tangible	Dirk	M	15		✓	✓	✓	✓			✓																								
Borrero, Vollmer & Wright (2002)	Escape	Joey	M	4		✓	✓	✓	✓			✓																								
Borrero, Vollmer, Borrero & Bouret (2005)	Attention	Nellie	F	12		✓	✓	✓	✓			✓								✓	✓			✓		✓										
Borrero, Vollmer, Borrero & Bouret (2005)	Tangible	Antoine	M	8	Y	✓	✓	✓	✓			✓								✓	✓			✓		✓										
Borrero, et al., (2010)	Tangible	Greg	M	8	Y																								✓					✓		
Borrero, et al., (2010)	Escape	Greg	M	8	Y																			✓		✓										
Brown, et al., (2000)	Attention	Kelly	F	9									✓	✓			✓			✓																
Brown, et al., (2000)	Escape	Theresa	F	13									✓			✓				✓	✓															
Brown, et al., (2000)	Escape	Corey	M	5		✓	✓		✓			✓									✓	✓					✓							✓		
Camp, Iwata, Hammond & Bloom (2009)	Attention	Tara	F	28														✓		✓																
Camp, Iwata, Hammond & Bloom (2009)	Escape	Jerry	M	19													✓			✓																
Camp, Iwata, Hammond & Bloom (2009)	Escape	Kevin	M	54																	✓	✓			✓	✓										
Camp, Iwata, Hammond & Bloom (2009)	Escape	Travis	M	21																	✓	✓			✓	✓										
Carr & Britton (1999)	Attention	Christian	M	32																									✓					✓		
Carter (2010)	Escape	Anonymous	M	19		✓	✓			✓		✓									✓	✓				✓				✓						
Chen, McComas, Reichle & Bergmann (2015)	Tangible	Max	M	18	Y	✓		✓	✓	✓		✓	✓	✓	✓					✓	✓	✓			✓	✓								✓		
Cihak, Alberto & Fredrick (2007)	Escape	Haley	F	17																	✓															
Cihak, Alberto & Fredrick (2007)	Escape	Anne	F	16																									✓					✓		
Cihak, Alberto & Fredrick (2007)	Escape	Gail	F	17																																
Cihak, Alberto & Fredrick (2007)	Escape	Kyle	M	15																							✓									

APPENDIX A: Topographies of behaviour from functional analyses

Reference	Function	Name	Gender	Age	Autism?	Hitting	Pushing	Kicking	Biting	Headbutting	Spitting	Any physical aggression	Headbanging	Face hitting	Self-hitting	Striking body against object	Self-biting	Scratching or pinching	Dangerous acts	Any self injurious behaviour	Throwing objects	Tearing objects	Pulling objects	Hitting objects	Damaging furniture	Any destructive behaviour	Elopement	Crying	Loud vocalisations	Perseverative speech	Dropping	Non-compliance	Tantrums	Any disruptive behaviour	Inappropriate sexual behaviour	Stereotypy
Danov, Tervo, Meyers & Symons (2012)	Escape	Stan	M	6		✓	✓		✓			✓									✓	✓				✓										
Davis, Fredrick, Alberto & Gama (2012)	Escape	Todd	M	17	Y	✓						✓						✓		✓	✓	✓				✓		✓	✓					✓		
Davis, Fredrick, Alberto & Gama (2012)	Escape	Tony	M	12	Y																✓					✓	✓		✓			✓		✓		✓
Davis, Fredrick, Alberto & Gama (2012)	Escape	Mary	F	8		✓						✓																								
Davis, Fredrick, Alberto & Gama (2012)	Escape	Eli	M	18		✓						✓												✓		✓	✓							✓		
Day, Rea, Schussler, Larsen & Johnson (1988)	Tangible	David	M	9	Y												✓			✓				✓		✓										
Day, Rea, Schussler, Larsen & Johnson (1988)	Tangible	Mary	F	15									✓	✓						✓										✓						
DeLeon, Arnold, Rodriguez-Catter & Uy (2003)	Attention	Jeremy	M	21																													✓			
DeLeon, Kahng, Rodriguez-Catter, Sveinsdóttir & Sadler (2003)	Tangible	Grady	M	14		✓	✓		✓			✓																								
Derby, et al., (1994)	Escape	Jenny	F	28			✓					✓																								
Dicesare, McAdam, Toner & Varrell (2005)	Attention	David	M	18																				✓		✓										
Dixon, Benedict & Larson (2001)	Attention	Fernando	M	25																										✓			✓	✓		
Durand & Carr (1992)	Attention	Wendy	F	4		✓	✓					✓																✓	✓		✓		✓	✓		
Durand & Carr (1992)	Attention	Mark	M	4		✓	✓					✓									✓	✓				✓		✓	✓					✓		
Durand & Carr (1992)	Attention	Jonathan	M	3	Y																✓	✓				✓									
Durand & Carr (1992)	Attention	Dan	M	5																	✓	✓				✓		✓	✓					✓		
Durand & Carr (1992)	Attention	Brian	M	4																	✓	✓				✓		✓	✓			✓		✓		
Durand & Carr (1992)	Attention	Paul	M	4	Y																✓	✓				✓		✓	✓			✓		✓		
Durand & Carr (1992)	Attention	Jaynie	F	4	Y																							✓	✓			✓		✓		
Durand & Carr (1992)	Attention	Sam	M	5																	✓	✓				✓		✓	✓					✓		
Durand & Carr (1992)	Attention	Ted	M	3																	✓	✓				✓		✓	✓			✓		✓		
Durand & Carr (1992)	Attention	Ian	M	4																	✓	✓				✓		✓	✓			✓		✓		
Durand & Carr (1992)	Attention	Ray	M	3		✓	✓					✓									✓	✓				✓		✓	✓					✓		
Durand & Carr (1992)	Attention	Mike	M	5																								✓	✓					✓		
Durand (1999)	Escape	Matt	M	5													✓			✓								✓	✓					✓		
Durand (1999)	Tangible	Allison	F	15																								✓	✓					✓		
Durand (1999)	Tangible	Mike	M	3													✓			✓	✓					✓										
Durand (1999)	Attention	Ron	M	9	Y	✓						✓																								
Durand (1999)	Escape	David	M	11	Y								✓	✓						✓																
Ebanks & Fisher (2003)	Escape	Jim	M	19			✓					✓		✓	✓		✓	✓		✓	✓			✓		✓										
Edrisinha, O'Reilly, Sigafoos, Lancioni & Choi (2011)	Tangible	Alex	M	16	Y				✓			✓					✓			✓				✓		✓			✓			✓		✓		
Edrisinha, O'Reilly, Sigafoos, Lancioni & Choi (2011)	Attention	Josh	M	37	Y																								✓	✓				✓		
English & Anderson (2004)	Attention	Natalie	F	16	Y	✓	✓	✓	✓			✓	✓	✓	✓		✓			✓	✓					✓			✓					✓		
Fahmie, Iwata, Querim & Harper (2013)	Escape	Ellyse	F	16		✓	✓		✓			✓																								
Fahmie, Iwata, Querim & Harper (2013)	Attention	Brad	M	11																	✓	✓			✓	✓										
Fahmie, Iwata, Querim & Harper (2013)	Attention	Paul	M	22		✓	✓		✓			✓																								
Fahmie, Iwata, Querim & Harper (2013)	Attention	Elijah	M	7	Y														✓						✓	✓										
Fahmie, Iwata, Querim & Harper (2013)	Escape	Tim	M	4	Y												✓			✓																
Fahmie, Iwata, Querim & Harper (2013)	Escape	Kelly	F	11													✓			✓																
Fahmie, Iwata, Querim & Harper (2013)	Escape	Kim	F	12		✓	✓		✓			✓																								
Fahmie, Iwata, Querim & Harper (2013)	Escape	Cecille	F	12		✓	✓		✓			✓																								

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Reference	Function	Name	Gender	Age	Autism?	Hitting	Pushing	Kicking	Biting	Headbutting	Spitting	Any physical aggression	Headbanging	Face hitting	Self-hitting	Striking body against object	Self-biting	Scratching or pinching	Dangerous acts	Any self injurious behaviour	Throwing objects	Tearing objects	Pulling objects	Hitting objects	Damaging furniture	Any destructive behaviour	Elopement	Crying	Loud vocalisations	Perseverative speech	Dropping	Non-compliance	Tantrums	Any disruptive behaviour	Inappropriate sexual behaviour	Sterotypy
Fischer, Iwata & Mazaleski (1997)	Tangible	Mike	M	34										✓						✓																
Fisher, Greer, Querim & DeRosa (2014)	Escape	Evan	M	7	Y	✓	✓	✓	✓		✓	✓	✓			✓		✓		✓																
Fisher, Greer, Querim & DeRosa (2014)	Escape	Dan	M	7		✓	✓	✓	✓		✓	✓																							
Fisher, Greer, Querim & DeRosa (2014)	Escape	John	M	4	Y	✓	✓	✓	✓		✓	✓									✓			✓	✓	✓										
Fisher, Greer, Querim & DeRosa (2014)	Escape	Casey	M	4	Y	✓	✓	✓	✓		✓	✓									✓			✓	✓	✓										
Fisher, Kuhn & Thompson (1998)	Attention	Ned	M	9	Y	✓		✓	✓			✓										✓		✓		✓										
Frea & Hughes (1997)	Attention	Ned	M	18																										✓				✓		
Frea & Hughes (1997)	Escape	Donna	F	17																										✓				✓		
Fritz, DeLeon & Lazarchick (2004)	Escape	Curtis	M	19		✓	✓		✓			✓									✓	✓	✓		✓	✓										
Fritz, DeLeon & Lazarchick (2004)	Escape	Irene	F	12	Y	✓	✓	✓	✓			✓	✓	✓		✓		✓		✓	✓	✓	✓	✓		✓					✓			✓		
Fyffe, Kahng, Fittro & Russell (2004)	Attention	Matt	M	9																															✓	
Goh, Iwata & DeLeon (2000)	Tangible	Lisa	F	28											✓		✓			✓																
Goh, Iwata & DeLeon (2000)	Attention	Julia	F	43														✓		✓																
Goh, et al. (1995)	Attention	Bonnie	F	40													✓			✓																
Goh, et al. (1995)	Tangible	Laura	F	25													✓			✓																
Hagopian, Contrucci Kuhn, Long & Rush (2005)	Attention	Stephen	M	13		✓	✓	✓	✓			✓	✓					✓		✓	✓	✓		✓		✓										
Hagopian, Contrucci Kuhn, Long & Rush (2005)	Tangible	James	M	12	Y	✓		✓	✓			✓			✓		✓			✓	✓			✓	✓	✓										
Hagopian, Fisher & Legacy (1994)	Attention	Laurie	F	5		✓		✓	✓	✓		✓	✓	✓	✓			✓		✓	✓		✓	✓		✓										
Hagopian, Fisher & Legacy (1994)	Attention	Lynn	F	5		✓		✓		✓		✓	✓	✓	✓			✓		✓	✓		✓	✓		✓										
Hagopian, Fisher & Legacy (1994)	Attention	Wanda	F	5		✓		✓		✓		✓	✓	✓	✓			✓		✓	✓		✓	✓		✓										
Hagopian, Fisher & Legacy (1994)	Attention	Glenda	F	5		✓		✓		✓		✓	✓	✓	✓			✓		✓	✓		✓	✓		✓										
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Escape	1	NR	5		✓	✓	✓	✓	✓		✓	✓	✓	✓		✓	✓		✓	✓	✓	✓	✓	✓	✓										
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Escape	3	NR	11		✓	✓	✓	✓	✓		✓	✓	✓	✓		✓	✓		✓	✓	✓	✓	✓	✓	✓										
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Escape	4	NR	16		✓	✓	✓	✓	✓		✓	✓	✓	✓		✓	✓		✓	✓	✓	✓	✓	✓	✓										
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Escape	5	NR	10		✓	✓	✓	✓	✓		✓	✓	✓	✓		✓	✓		✓	✓	✓	✓	✓	✓	✓										
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Escape	6	NR	15		✓	✓	✓	✓	✓		✓									✓	✓	✓	✓	✓	✓										
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Escape	7	NR	9		✓	✓	✓	✓	✓		✓	✓	✓	✓		✓	✓		✓																
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Attention	8	NR	5		✓	✓	✓	✓	✓		✓									✓	✓	✓	✓	✓	✓										
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Attention	9	NR	2		✓	✓	✓	✓	✓		✓	✓	✓	✓		✓	✓		✓	✓	✓	✓	✓	✓	✓										
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Attention	11	NR	10		✓	✓	✓	✓	✓		✓	✓	✓	✓		✓	✓		✓	✓	✓	✓	✓	✓	✓										✓
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Attention	13	NR	12		✓	✓	✓	✓	✓		✓	✓	✓	✓		✓	✓		✓	✓	✓	✓	✓	✓	✓										
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Attention	14	NR	12		✓	✓	✓	✓	✓		✓									✓	✓	✓	✓	✓	✓										
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Attention	15	NR	14		✓	✓	✓	✓	✓		✓																								
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Attention	16	NR	10		✓	✓	✓	✓	✓		✓	✓	✓	✓		✓	✓		✓	✓	✓	✓	✓	✓	✓										
Hagopian, Fisher, Sullivan, Acquisto & LeBlanc (1998)	Tangible	17	NR	5		✓	✓	✓	✓	✓		✓	✓	✓	✓		✓	✓		✓	✓	✓	✓	✓	✓	✓										
Hammond, Iwata, Fritz & Dempsey (2011)	Tangible	Alex	M	14	Y	✓		✓	✓	✓		✓																								
Hammond, Iwata, Fritz & Dempsey (2011)	Tangible	Curtis	M	13	Y	✓		✓	✓	✓		✓																								
Hammond, Iwata, Fritz & Dempsey (2011)	Tangible	Abby	F	19	Y									✓						✓																
Hammond, Iwata, Rooker, Fritz & Bloom (2013)	Escape	Jacob	M	6	Y																✓		✓		✓	✓										
Hammond, Iwata, Rooker, Fritz & Bloom (2013)	Escape	Garrett	M	12	Y												✓			✓																
Hammond, Iwata, Rooker, Fritz & Bloom (2013)	Escape	Eddie	M	4		✓	✓	✓	✓			✓																								

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Hammond, Iwata, Rooker, Fritz & Bloom (2013)	Escape	Rosa	F	17		✓	✓					✓																								
Hanley, Iwata & Thompson (2001)	Tangible	Karen	F	29										✓	✓																					
Hanley, Iwata & Thompson (2001)	Tangible	Jake	M	34			✓		✓			✓					✓			✓																
Hanley, Iwata & Thompson (2001)	Attention	Julie	F	31		✓		✓	✓			✓				✓				✓																
Hanley, Piazza & Fisher (1997)	Attention	Hank	M	16			✓		✓			✓	✓	✓						✓																
Hanley, Piazza, Fisher & Maglieri (2005)	Attention	Jay	M	5	Y	✓	✓	✓	✓			✓		✓			✓			✓	✓	✓				✓										
Hanley, Piazza, Fisher, Contrucci & Maglieri (1997)	Attention	Tony	M	4		✓	✓	✓	✓			✓								✓	✓	✓		✓		✓										
Harper, Iwata & Camp (2013)	Escape	Ann	F	24	Y		✓					✓																								
Harper, Iwata & Camp (2013)	Escape	Penny	F	48	Y		✓					✓																								
Harper, Iwata & Camp (2013)	Escape	Will	M	52			✓	✓				✓																								
Harper, Iwata & Camp (2013)	Escape	Eve	F	13			✓	✓				✓																								
Healy, Brett & Leader (2013)	Escape	14	M	13	Y	✓	✓					✓																								
Healy, Brett & Leader (2013)	Escape	15	M	7	Y																✓	✓				✓										
Healy, Brett & Leader (2013)	Attention	16	M	12	Y	✓	✓					✓									✓	✓				✓										
Healy, Brett & Leader (2013)	Escape	19	M	16	Y								✓	✓						✓															
Healy, Brett & Leader (2013)	Escape	24	M	6	Y								✓				✓			✓															
Healy, Brett & Leader (2013)	Escape	31	M	19	Y	✓	✓					✓									✓	✓				✓									✓	
Healy, Brett & Leader (2013)	Tangible	32	F	8	Y	✓			✓			✓						✓			✓	✓				✓										
Horner & Day (1991)	Escape	Paul	M	12		✓		✓	✓			✓		✓				✓		✓						✓										
Horner & Day (1991)	Attention	Peter	M	14				✓				✓		✓						✓						✓										
Horner & Day (1991)	Escape	Mary	F	27	Y	✓		✓				✓		✓	✓					✓				✓		✓										
Horner, Day & Day (1997)	Escape	Clay	M	12	Y	✓	✓	✓	✓	✓		✓		✓			✓			✓	✓	✓				✓										
Horner, Day & Day (1997)	Escape	Pat	M	17		✓	✓	✓	✓	✓		✓						✓			✓	✓				✓										
Horner, Day & Day (1997)	Tangible	Karl	M	14	Y	✓	✓	✓	✓			✓	✓	✓			✓			✓	✓	✓				✓										
Iwata, Dorsey, Slifer, Bauman & Richman (1994)	Escape	1	F	4									✓		✓					✓						✓										
Iwata, Dorsey, Slifer, Bauman & Richman (1994)	Escape	3	M	13									✓				✓			✓						✓										
Iwata, Dorsey, Slifer, Bauman & Richman (1994)	Attention	5	M	13									✓	✓			✓			✓						✓										
Iwata, Pace, Cowdery & Miltenberger (1994)	Escape	Jack	M	12									✓	✓						✓						✓										
Iwata, Pace, Cowdery & Miltenberger (1994)	Attention	Millie	F	8									✓	✓						✓						✓										
Kahng, Abt & Schonbachler (2001)	Attention	Polly	F	15		✓	✓	✓				✓									✓															
Kahng, Iwata, DeLeon & Wallace (2000)	Attention	Julia	F	43									✓				✓			✓						✓										
Kahng, Iwata, DeLeon & Wallace (2000)	Attention	Nancy	F	50										✓	✓					✓						✓										
Kahng, Iwata, DeLeon & Wallace (2000)	Tangible	Lisa	F	27										✓	✓		✓			✓						✓										
Kahng, Iwata, DeLeon & Worsdell (1997)	Escape	Todd	M	50									✓							✓						✓										
Kahng, Iwata, DeLeon & Worsdell (1997)	Attention	Lynn	F	29											✓		✓			✓						✓										
Kahng, Iwata, DeLeon & Worsdell (1997)	Escape	Bob	M	45														✓		✓						✓										
Kahng, Iwata, Thompson & Hanley (2000)	Attention	Julia	F	43														✓		✓						✓										
Kahng, Iwata, Thompson & Hanley (2000)	Attention	Susan	F	31		✓		✓	✓			✓				✓				✓						✓										
Kahng, Iwata, Thompson & Hanley (2000)	Tangible	Matt	M	25											✓					✓						✓										
Kelley, Lerman & Van Camp (2002)	Tangible	Roger	M	10		✓	✓					✓								✓						✓										
Kelley, Lerman & Van Camp (2002)	Escape	Gary	M	9	Y	✓	✓	✓	✓			✓								✓						✓										

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Kelley, Lerman & Van Camp (2002)	Escape	Jennifer	F	10																✓	✓				✓										
Kurtz, Chin, Robinson, O'Connor & Hagopian (2015)	Escape	Noah	M	15	Y	✓	✓	✓	✓			✓		✓					✓	✓	✓	✓		✓		✓			✓					✓		
Kurtz, Chin, Robinson, O'Connor & Hagopian (2015)	Attention	Damien	M	7	Y	✓	✓	✓	✓			✓	✓	✓			✓		✓	✓	✓	✓		✓	✓	✓									
Kurtz, Chin, Robinson, O'Connor & Hagopian (2015)	Tangible	Jackie	F	7	Y	✓	✓	✓	✓		✓	✓							✓	✓	✓	✓	✓	✓	✓	✓			✓		✓			✓		
Kurtz, Chin, Robinson, O'Connor & Hagopian (2015)	Escape	Josh	M	10	Y	✓	✓	✓	✓			✓	✓	✓		✓	✓	✓		✓	✓	✓	✓	✓		✓									
Lalli, Casey & Kates (1997)	Tangible	Donny	M	3		✓	✓	✓	✓			✓									✓														
Lalli, Casey & Kates (1997)	Tangible	Tony	M	9									✓					✓		✓														
Lalli, Casey & Kates (1997)	Tangible	Harry	M	7									✓	✓			✓	✓		✓														
Lalli, Mace, Wohn & Livezey (1995)	Escape	Mary	F	15	Y	✓		✓				✓					✓			✓								✓					✓		
Lambert, Bloom & Irvin (2012)	Attention	Danny	M	3		✓	✓		✓			✓																						
Lambert, Bloom, Samaha, Dayton & Kunnavatana (2016)	Attention	Sam	M	31			✓					✓																						
Lancaster et al. (2004)	Attention	Participant 1	F	56																									✓				✓		
Langdon, Carr & Owen-DeSchryver (2008)	Escape	Timmy	M	6	Y								✓	✓	✓	✓	✓			✓														
Langdon, Carr & Owen-DeSchryver (2008)	Escape	Sally	F	7		✓			✓			✓					✓			✓														
Langdon, Carr & Owen-DeSchryver (2008)	Escape	Jake	M	18	Y												✓			✓														
LaRue et al. (2010)	Tangible	Ryan	M	9	Y	✓	✓	✓	✓			✓	✓	✓	✓					✓	✓		✓	✓		✓									
LaRue et al. (2010)	Tangible	Rupert	M	8	Y	✓	✓	✓	✓		✓	✓	✓	✓	✓					✓	✓		✓	✓		✓									
LaRue et al. (2010)	Tangible	Alex	M	20	Y	✓	✓	✓	✓			✓	✓	✓	✓					✓	✓		✓	✓		✓			✓				✓		
LaRue et al. (2011)	Escape	Mike	M	24	Y								✓	✓	✓					✓														
LaRue et al. (2011)	Attention	Sarah	F	7	Y																							✓					✓		
LaRue et al. (2011)	Attention	Paul	M	15	Y								✓	✓	✓					✓														
Le & Smith (2002)	Escape	Tom	M	37										✓	✓					✓														
Le & Smith (2002)	Escape	Fred	M	35									✓							✓														
LeBlanc, Hagopian, Marhefka & Wilke (2001)	Attention	Blair	F	11		✓	✓	✓	✓			✓									✓	✓		✓		✓									
Lerman, Iwata, Zarcone & Ringdahl (1994)	Escape	Stan	M	41						✓	✓	✓		✓	✓					✓	✓					✓			✓				✓		
Lerman, Iwata, Zarcone & Ringdahl (1994)	Escape	Peter	M	44													✓			✓									✓				✓		✓
Lindberg, Iwata, Kahng & DeLeon (1999)	Tangible	Dot	F	28										✓	✓		✓			✓														
Lindberg, Iwata, Kahng & DeLeon (1999)	Attention	Jodi	F	43														✓		✓														
Lindberg, Iwata, Kahng & DeLeon (1999)	Attention	Bridget	F	50									✓	✓	✓					✓														
Lomas Mevers, Fisher, Kelley & Fredrick (2014)	Escape	Sarah	F	14		✓	✓	✓	✓			✓		✓	✓		✓			✓	✓	✓	✓		✓	✓			✓				✓		
Mace & Lalli (1991)	Attention	Mitch	M	46																										✓			✓		
Marcus & Vollmer (1996)	Tangible	Sally	F	5		✓	✓	✓	✓			✓	✓	✓						✓															
Marcus & Vollmer (1996)	Tangible	Rob	M	4		✓	✓	✓	✓			✓																							
Marcus & Vollmer (1996)	Tangible	CJ	M	5	Y								✓	✓						✓				✓		✓		✓				✓	✓		
Marcus, Vollmer, Swanson, Roane and Ringdahl (2001)	Escape	Joel	M	5	Y	✓	✓	✓	✓			✓									✓														
Marcus, Vollmer, Swanson, Roane and Ringdahl (2001)	Escape	Matt	M	4		✓			✓			✓																							
Marcus, Vollmer, Swanson, Roane and Ringdahl (2001)	Escape	Joe	M	4		✓		✓				✓																✓	✓		✓		✓	✓		
Marcus, Vollmer, Swanson, Roane and Ringdahl (2001)	Tangible	Seth	M	6	Y	✓	✓	✓	✓	✓	✓	✓									✓														
Marcus, Vollmer, Swanson, Roane and Ringdahl (2001)	Tangible	Alvin	M	3		✓	✓		✓			✓																							
Mason & Iwata (1990)	Attention	Sally	F	6											✓					✓															
Mason & Iwata (1990)	Escape	Mort	M	18									✓	✓	✓					✓															

APPENDIX A: Topographies of behaviour from functional analyses

Reference	Function	Name	Gender	Age	Autism?	Hitting	Pushing	Kicking	Biting	Headbutting	Spitting	Any physical aggression	Headbanging	Face hitting	Self-hitting	Striking body against object	Self-biting	Scratching or pinching	Dangerous acts	Any self injurious behaviour	Throwing objects	Tearing objects	Pulling objects	Hitting objects	Damaging furniture	Any destructive behaviour	Elopement	Crying	Loud vocalisations	Perseverative speech	Dropping	Non-compliance	Tantrums	Any disruptive behaviour	Inappropriate sexual behaviour	Sterotypy
May & Howe (2013)	Escape	Jaylin	F	4																										✓		✓		✓		
Mazaleski, Iwata, Vollmer, Zarcone & Smith (1993)	Attention	Diane	F	32									✓	✓	✓					✓																
Mazaleski, Iwata, Vollmer, Zarcone & Smith (1993)	Attention	Brenda	F	42									✓	✓	✓	✓				✓																
Mazaleski, Iwata, Vollmer, Zarcone & Smith (1993)	Attention	Bonnie	F	40													✓			✓																
McClean & Grey (2012)	Escape	Aidan	M	21	Y	✓	✓	✓	✓			✓	✓							✓																
McClean & Grey (2012)	Escape	Brendan	M	15	Y	✓	✓	✓	✓		✓	✓					✓			✓																
McClean & Grey (2012)	Escape	Cormac	M	23	Y	✓	✓	✓				✓																								
McClean & Grey (2012)	Escape	Darragh	M	17	Y	✓	✓	✓	✓			✓		✓			✓			✓																
McComas, Hoch, Paone & El-Roy (2000)	Escape	Eli	M	8	Y																	✓				✓										
McComas, Hoch, Paone & El-Roy (2000)	Escape	Charlie	M	8	Y		✓					✓																								
McComas, Hoch, Paone & El-Roy (2000)	Escape	Ben	M	9	Y	✓			✓			✓																								
McComas, Thompson & Johnson (2003)	Attention	Dan	M	11		✓					✓	✓									✓					✓										
McComas, Thompson & Johnson (2003)	Escape	Abe	M	11	Y	✓		✓	✓			✓																								
McComas, Thompson & Johnson (2003)	Escape	Ari	M	12		✓		✓	✓			✓									✓															
McCord, Iwata, Galensky, Ellingson & Thomson (2001)	Escape	Debbie	F	43	Y	✓		✓	✓			✓			✓		✓			✓	✓			✓		✓										
McCord, Iwata, Galensky, Ellingson & Thomson (2001)	Escape	Sarah	F	41		✓	✓	✓	✓			✓		✓			✓			✓	✓			✓		✓		✓	✓					✓		
Mueller,Edwards & Trahant (2003)	Escape	Nelly	F	7		✓		✓	✓			✓												✓		✓		✓	✓			✓		✓		
Najdowski, Wallace, Ellsworth, MacAleese & Cleveland (2008)	Attention	Tom	M	45																															✓	
Najdowski, Wallace, Ellsworth, MacAleese & Cleveland (2008)	Tangible	Pete	M	5			✓					✓																✓	✓					✓		
Northup, et al., (1991)	Escape	Curtis	M	24		✓	✓		✓			✓																								
Northup et al.(1994)	Escape	Mike	M	5									✓	✓						✓																
Northup et al.(1994)	Escape	Heidi	F	11		✓	✓					✓																								
Northup, Fisher, Kahang, Harrell & Kurtz (1997)	Attention	Tom	M	Child		✓	✓	✓				✓									✓	✓	✓			✓										
Northup, Fisher, Kahang, Harrell & Kurtz (1997)	Attention	Alice	F	3		✓	✓		✓			✓									✓			✓		✓										
O'Reilly (1995)	Escape	Shawn	M	31		✓			✓			✓																								
O'Reilly et al. (2006)	Tangible	Sam	M	14	Y	✓		✓				✓	✓	✓			✓			✓																
O'Reilly et al. (2009)	Tangible	Terry	M	5	Y																✓					✓										
O'Reilly et al. (2012)	Tangible	Matthew	M	5	Y																						✓		✓		✓	✓		✓		
O'Reilly et al. (2012)	Tangible	Mark	M	9	Y							✓		✓			✓			✓																
O'Reilly et al. (2012)	Tangible	Luke	M	5	Y																						✓		✓		✓	✓		✓		
O'Reilly, Lancioni & Taylor (1999)	Attention	Rick	M	10		✓						✓																								
O'Reilly, Lancioni, King, Lally & Dhomhnaill (2000)	Attention	Ole	M	22			✓		✓			✓																								
O'Reilly, Lancioni, King, Lally & Dhomhnaill (2000)	Attention	Lena	F	9													✓			✓	✓				✓	✓										
Pace, Iwata, Cowdery, Andree & McIntyre (1993)	Escape	Mary	F	3									✓					✓		✓																
Pace, Iwata, Cowdery, Andree & McIntyre (1993)	Escape	Wally	M	17											✓					✓																
Pace, Iwata, Cowdery, Andree & McIntyre (1993)	Escape	Lynn	F	2													✓			✓																
Pence, Roscoe, Bourret & Ahearn (2009)	Escape	Gina	F	10	Y								✓	✓						✓																
Pence, Roscoe, Bourret & Ahearn (2009)	Escape	Casey	F	10	Y	✓	✓	✓	✓			✓																								
Pence, Roscoe, Bourret & Ahearn (2009)	Attention	Jake	M	8	Y	✓	✓	✓	✓			✓																								
Pence, Roscoe, Bourret & Ahearn (2009)	Attention	Larry	M	19	Y																	✓	✓	✓		✓										
Peyton, Lindauer & Richman (2005)	Escape	Suzie	F	10	Y																										✓			✓		

APPENDIX A: Topographies of behaviour from functional analyses

Reference	Function	Name	Gender	Age	Autism?	Hitting	Pushing	Kicking	Biting	Headbutting	Spitting	Any physical aggression	Headbanging	Face hitting	Self-hitting	Striking body against object	Self-biting	Scratching or pinching	Dangerous acts	Any self injurious behaviour	Throwing objects	Tearing objects	Pulling objects	Hitting objects	Damaging furniture	Any destructive behaviour	Elopement	Crying	Loud vocalisations	Perseverative speech	Dropping	Non-compliance	Tantrums	Any disruptive behaviour	Inappropriate sexual behaviour	Sterotypy
Piazza et al. (1999)	Attention	Ike	M	11		✓	✓	✓	✓	✓		✓					✓	✓			✓	✓	✓	✓		✓										
Piazza et al. (1999)	Attention	Paul	M	11	Y	✓	✓	✓				✓									✓															
Potoczak, Carr & Michael (2007)	Escape	Ursula	F	9																											✓		✓		
Potoczak, Carr & Michael (2007)	Escape	Howie	M	7	Y																✓					✓	✓			✓		✓		✓		
Potoczak, Carr & Michael (2007)	Escape	Darryl	M	17																	✓					✓	✓							✓		
Potoczak, Carr & Michael (2007)	Escape	Larry	M	8																							✓					✓		✓		
Reed, Pace & Luiselli (2009)	Tangible	Larry	M	16													✓	✓		✓	✓	✓			✓	✓								✓		
Rehfeldt & Chambers (2003)	Attention	Vince	M	23	Y																									✓				✓		
Richman & Hagopian (1999)	Attention	Don	M	6		✓	✓	✓				✓			✓		✓			✓	✓	✓				✓										
Richman & Lindauer (2002)	Attention	Tim	M	14																																✓
Richman, Wacker, Asmus & Casey (1998)	Escape	Amy	F	27	Y																✓					✓			✓		✓		✓			
Richman, Wacker, Asmus, Casey & Andelman (1999)	Escape	Kim	F	6																									✓		✓		✓			
Roane & Kelley (2008)	Attention	Gail	F	16									✓	✓	✓					✓																
Robertson, Wehby & King (2013)	Tangible	Nicholas	M	2	Y	✓	✓	✓				✓									✓		✓			✓			✓				✓			
Samaha et al. (2009)	Tangible	Greg	M	8	Y																								✓				✓			
Schieltz et al. (2010)	Escape	Andy	M			✓	✓	✓	✓			✓	✓					✓		✓	✓	✓		✓	✓	✓				✓			✓			
Schieltz et al. (2010)	Escape	Kurt	M			✓						✓									✓			✓		✓				✓			✓			
Schmidt, Shanholtzer, Mezhoudi, Scherback & Kahng (2014)	Escape	Michael	M	14		✓	✓	✓	✓	✓		✓									✓		✓	✓		✓			✓				✓			
Shabani et al.(2013)	Escape	Adam	M	19			✓		✓			✓		✓						✓	✓			✓		✓										
Shabani et al.(2013)	Escape	Lester	M	18					✓			✓					✓			✓						✓			✓				✓			
Shabani et al.(2013)	Tangible	Tanya	F	15	Y	✓		✓	✓			✓	✓	✓						✓	✓			✓		✓				✓			✓			
Shirley, Iwata, Kahng, Mazaleski & Lerman (1997)	Tangible	Rick	M	29									✓	✓	✓					✓															
Shirley, Iwata, Kahng, Mazaleski & Lerman (1997)	Tangible	Sue	F	24										✓	✓		✓			✓															
Shirley, Iwata, Kahng, Mazaleski & Lerman (1997)	Escape	John	M	39									✓				✓			✓															
Sigafoos & Tucker (2000)	Attention	Brad	M	19	Y	✓						✓																								
Slifer, Ivancic, Parrish, Page & Burgio (1986)	Escape	Jimmy	M	13		✓	✓	✓	✓			✓																								
Slifer, Ivancic, Parrish, Page & Burgio (1986)	Attention	Jimmy	M																										✓				✓		✓
Smith, Iwata, Goh & Shore (1995)	Escape	Walter	M	31													✓			✓															
Smith, Iwata, Goh & Shore (1995)	Escape	Landon	M	44									✓	✓	✓		✓			✓															
Smith, Iwata, Goh & Shore (1995)	Escape	Olivia	F	27										✓						✓															
Smith, Iwata, Goh & Shore (1995)	Escape	Larry	M	46										✓			✓			✓															
Smith, Iwata, Goh & Shore (1995)	Escape	Carl	M	38									✓	✓			✓			✓															
Smith, Iwata, Goh & Shore (1995)	Escape	Stan	M	47									✓	✓	✓					✓															
Smith, Iwata, Goh & Shore (1995)	Escape	Helen	F	28										✓	✓					✓															
Smith, Iwata, Goh & Shore (1995)	Escape	Everlyn	F	33									✓	✓	✓		✓			✓															
Smith, Iwata, Goh & Shore (1995)	Escape	Milt	M	43									✓		✓					✓															
Smith, Iwata, Vollmer & Pace (1992)	Escape	Jerry	M														✓			✓															
Smith, Iwata, Vollmer & Pace (1992)	Escape	Bill	M											✓						✓															
Sprague, Flannery & Szidon (1998)	Tangible	Anonymous	F	13							✓	✓																	✓				✓			
Steege et al. (1990)	Escape	Ann	F	5													✓			✓															
Steege et al. (1990)	Escape	Dennis	M	6													✓			✓															

APPENDIX A: Topographies of behaviour from functional analyses

Reference	Function	Name	Gender	Age	Autism?	Hitting	Pushing	Kicking	Biting	Headbutting	Spitting	Any physical aggression	Headbanging	Face hitting	Self-hitting	Striking body against object	Self-biting	Scratching or pinching	Dangerous acts	Any self injurious behaviour	Throwing objects	Tearing objects	Pulling objects	Hitting objects	Damaging furniture	Any destructive behaviour	Elopement	Crying	Loud vocalisations	Perseverative speech	Dropping	Non-compliance	Tantrums	Any disruptive behaviour	Inappropriate sexual behaviour	Sterotypy
Thompson, Fisher, Piazza & Kuhn (1998)	Attention	Ernie	M	7		✓		✓	✓			✓																								
Tiger, Fisher, Toussaint & Kodak (2009)	Escape	Carl	M	10		✓	✓	✓				✓									✓	✓	✓	✓												
Tincani, Castrogiavanni & Axelrod (1999)	Tangible	Darren	M	26	Y	✓	✓		✓			✓																								
Tincani, Castrogiavanni & Axelrod (1999)	Attention	Lorie	F	30	Y								✓		✓		✓			✓																
Tincani, Castrogiavanni & Axelrod (1999)	Attention	Bobby	M	30	Y	✓	✓		✓			✓																								
Travis & Sturmey (2010)	Attention	Mr. Jones	M	26																									✓				✓		
Van Camp et al. (2000)	Tangible	Timon	M	8													✓			✓															
Van Camp, Lerman, Kelley, Contrucci & Vorndran (2000)	Tangible	Roger	M	13		✓	✓	✓	✓			✓																								
Volkert, Lerman & Vorndran (2005)	Attention	Meadow	F	7		✓	✓		✓			✓																								
Volkert, Lerman & Vorndran (2005)	Escape	Nick	M	6					✓			✓		✓						✓	✓					✓										
Volkert, Lerman & Vorndran (2005)	Tangible	Tyler	M	3		✓	✓		✓			✓																								
Volkert, Lerman & Vorndran (2005)	Escape	Max	M	7		✓	✓	✓	✓			✓	✓	✓				✓		✓																
Volkert, Lerman & Vorndran (2005)	Escape	Adel	F	9													✓			✓									✓		✓		✓		
Vollmer et al. (1998)	Escape	Dana	F	22		✓			✓			✓																								
Vollmer et al. (1998)	Attention	Alan	M	6											✓			✓		✓	✓	✓		✓		✓										
Vollmer et al. (1998)	Escape	Matthew	M	16		✓		✓	✓			✓									✓	✓				✓										
Vollmer, Borrero, Lalli & Daniel (1999)	Tangible	Dale	M	9	Y	✓	✓					✓																								
Vollmer, Borrero, Lalli & Daniel (1999)	Tangible	Todd	M	9		✓	✓	✓				✓																								
Vollmer, Iwata, Zarcone, Smith & Mazaleski (1993a)	Escape	Randy	M											✓			✓			✓																
Vollmer, Iwata, Zarcone, Smith & Mazaleski (1993a)	Escape	Karen	F										✓	✓	✓					✓																
Vollmer, Iwata, Zarcone, Smith & Mazaleski (1993a)	Attention	Sharon	F										✓	✓	✓					✓																
Vollmer, Iwata, Zarcone, Smith & Mazaleski (1993b)	Attention	Diane	F	32									✓	✓	✓					✓																
Vollmer, Iwata, Zarcone, Smith & Mazaleski (1993b)	Attention	Bonnie	F	40													✓			✓																
Vollmer, Iwata, Zarcone, Smith & Mazaleski (1993b)	Attention	Brenda	F	42									✓	✓	✓					✓																
Vollmer & Vorndran (1998)	Tangible	Denise	F	29														✓		✓																
Wacker et al. (1990)	Tangible	Bobby	M	7	Y												✓			✓																
Wacker et al. (2011)	Escape	Jose	M	4		✓			✓			✓					✓			✓	✓					✓										
Wallace, Iwata, Hanley, Thompson & Roscoe (2012)	Tangible	Shelby	F	29										✓	✓		✓			✓																
Wallace, Iwata, Hanley, Thompson & Roscoe (2012)	Attention	Susan	F	32		✓						✓				✓				✓																
Wallace, Iwata, Hanley, Thompson & Roscoe (2012)	Tangible	Matt	M	35													✓			✓																
Watson, Ray, Turner & Logan (1999)	Escape	Matt	M	10									✓	✓	✓	✓		✓		✓								✓				✓	✓			
Winborn-Kemmerer, Ringdahl, Wacker & Kitsukawa (2009)	Attention	Jack	M	7			✓		✓			✓																								
Winborn-Kemmerer, Ringdahl, Wacker & Kitsukawa (2009)	Tangible	Sally	F	20		✓			✓			✓	✓		✓		✓			✓																
Worsdell, Iwata, Conners, Kahng & Thompson (2000)	Attention	Brenda	F	52									✓	✓	✓					✓																
Worsdell, Iwata, Conners, Kahng & Thompson (2000)	Attention	Dolly	F	38									✓							✓																
Worsdell, Iwata, Conners, Kahng & Thompson (2000)	Attention	Jenna	F	44														✓		✓																
Worsdell, Iwata, Conners, Kahng & Thompson (2000)	Attention	Beth	F	46													✓			✓																
Worsdell, Iwata, Conners, Kahng & Thompson (2000)	Tangible	Rich	M	33										✓	✓		✓			✓																
Worsdell, Iwata, Conners, Kahng & Thompson (2000)	Tangible	Sheila	F	29										✓	✓		✓			✓																
Worsdell, Iwata, Hanley, Thompson & Kahng (2000)	Tangible	Jed	M	33													✓			✓																
Worsdell, Iwata, Hanley, Thompson & Kahng (2000)	Tangible	Max	M	37									✓							✓																

APPENDIX A: Topographies of behaviour from functional analyses

Reference	Function	Name	Gender	Age	Autism?	Hitting	Pushing	Kicking	Biting	Headbutting	Spitting	Any physical aggression	Headbanging	Face hitting	Self-hitting	Striking body against object	Self-biting	Scratching or pinching	Dangerous acts	Any self injurious behaviour	Throwing objects	Tearing objects	Pulling objects	Hitting objects	Damaging furniture	Any destructive behaviour	Elopement	Crying	Loud vocalisations	Perseverative speech	Dropping	Non-compliance	Tantrums	Any disruptive behaviour	Inappropriate sexual behaviour	Sterotypy
Worsdell, Iwata, Hanley, Thompson & Kahng (2000)	Attention	Janet	F	44														✓		✓																
Worsdell, Iwata, Hanley, Thompson & Kahng (2000)	Attention	Annette	F	31		✓		✓	✓			✓				✓				✓	✓															
Worsdell, Iwata, Hanley, Thompson & Kahng (2000)	Tangible	Shonnie	F	29										✓	✓		✓			✓																
Yarbrough and Carr (2000)	Escape	Ray	M	18	Y								✓		✓	✓	✓			✓																
Yarbrough and Carr (2000)	Tangible	Kim	F	11	Y								✓		✓	✓	✓			✓									✓				✓			
Zarcone et al. (1993)	Escape	Lisa	F	39										✓						✓																
Zarcone et al. (1993)	Escape	Karen	F	26										✓	✓		✓			✓	✓					✓										
Zarcone, Iwata, Smith, Mazaleski & Lerman (1994)	Escape	Kate	F	27										✓						✓																
Zarcone, Iwata, Smith, Mazaleski & Lerman (1994)	Escape	Chris	M	38									✓				✓			✓																
Zarcone, Iwata, Smith, Mazaleski & Lerman (1994)	Escape	Jesse	M	40									✓	✓	✓	✓				✓																

Appendix B: Participant information sheet

The Cerebra Centre for Neurodevelopmental Disorders

UNIVERSITY OF BIRMINGHAM

Behaviour in Lowe Syndrome
Information Sheet

(17/3/2015 – Version 1)

Please read this information carefully before deciding whether you wish to take part in the study. If you have any further questions or have a medical issue that makes this information difficult to read please contact Dr. Jane Waite on [redacted]

You can watch a short film about this research project on our website at: http://www.findresources.co.uk/lowe-syndrome-project-launched

Please complete the enclosed consent forms and return them to us in the prepaid envelope provided if you and your child/person you care for would like to take part. You can also complete consent forms online at [insert web address] logging in with the password: **cere1bra**.

Background

We are conducting a research study at the Centre for Neurodevelopmental Disorders, University of Birmingham. Your child/person you care for does not need to show behavioural difficulties to take part; we are interested in learning about the differences between people who do and do not show these difficulties. We are conducting this study because:

- Around 70-80% of individuals with Lowe syndrome show behavioural difficulties.
- The presence of behavioural difficulties (self-injury, aggression, destruction of property and temper outbursts) in people with intellectual disability can lower quality of life.
- We know of a number of factors that can impact on the development and maintenance of behavioural difficulties.
- Despite robust evidence of these risk markers for development of behaviour in other groups, they have not been studied in individuals with Lowe syndrome.
- We hope that exploring the causes of behavioural difficulties will help to better target more effective interventions for behaviour and improve quality of life for people with Lowe Syndrome.

Aims of the study

We aim to further understanding of the causes of behavioural difficulties in individuals with Lowe Syndrome. Eventually we hope that our results will help to improve the quality of life of individuals and their families, and more effectively target interventions for behavioural difficulties.

What will happen if you/ the person you care for decide(s) to participate?

Where will the research take place?

The research will take place at either our Centre in the University of Birmingham, at your home, a syndrome support day or another location that is convenient to you.

Who will be involved in collecting the data?
Members of the research team at the Cerebra Centre for Neurodevelopmental disorders including Dr Jane Waite, Prof Chris Oliver, Helen Cressey and Alicia Kutsch will collect data.

How long will participation in the study take?
The time spent taking part will depend on which stage of the study it's convenient for you to take part in. We can discuss the stages of study over the telephone with you. If you would like to take part in the entire study you should set aside **approximately one day**. This will involve a member of the research team meeting with your child/person you care at a location of your choice, and asking you some questions about the person's behaviour.

We will be collecting information from participants between June 2015 and Jan 2017. After this we will spend some time understanding the information we have collected and writing reports. This means that the study will be finished in Jun 2017.

What will your child/person you care for be required to do during the study?
We will meet with your child/person you care for and will complete structured assessments to measure how your child processes information and how they interact with the world around them. We will measure these abilities using engaging table-top activities, computer based tasks and observations. The tasks are suitable for children over the age of four years and adults irrespective of their degree of intellectual disability or visual impairment.

What will you be required to do in the study?
You will be asked to take part in an interview about behaviour and adaptive abilities that will be conducted by researchers either over the phone or in person. We would like to discuss the behaviour of your child/person you care for. We will ask parents and caregivers to complete a questionnaire online (paper copies available on request).

Will assessments/interviews be recorded?
During the assessments, your child's behaviour and the behaviour of people in your child's immediate surroundings will be recorded using a video camera. These observations will be video recorded in order to check the accuracy of the observations with another researcher.

During the interviews that we will conduct with you, the interviewer's questions and your responses may be audio recorded to ensure with accurate data collection.

The University of Birmingham will hold the copyright for the audio/video recordings so that the confidentiality of these recordings will be protected. But, the University of Birmingham will not be able to edit or use the recordings for teaching purposes unless you give us your written permission to do this.

We may contact you again in the future to ask your permission to use some of the recordings for teaching purposes. At that time you will be able to decide whether or not you are happy for the recordings to be used for these purposes. Agreeing to participate in this study **does not** mean that you will have to give your permission for the use of these recordings in the future.

Confirmation of genetic status
If you decide to take part we would like to ask your permission to contact your G.P or consultant to request written confirmation of your child's/person you care for's genetic diagnosis. We would like to obtain this information as it helps ensure research findings are published in the highest quality scientific journals and also helps us learn more about how genetic markers might be linked to behaviour. It is entirely up to you and/or the person you care for whether you give us permission to do this and it will not impact on your participation in the study.

Are there any risks that individuals taking part in the study might face?

We will not expose your child/person you care for to any assessment that we have not previously discussed with you. We will not ask your child/person you care for to participate in any assessment that you feel may potentially cause distress.

Participating in the research study means that your child would be put in a situation where they would have to meet new people. We will provide you with detailed information in advance about which researchers will be working with your child/person you care for and will follow your advice about how best to introduce ourselves in a way that would be easiest for your child/person you care for.

What are the potential benefits for participants from taking part?
You will receive a personalised feedback regarding your child/ the person you care for. This study will help us to find out more about the lives of people with [insert syndrome] and the difficulties that these people face. The results might help us to improve things for people [insert syndrome] in the future.

Where will data be stored?
The data collected will be kept in locked or password protected storage at the University of Birmingham. All information will be stored in locked cabinets.

Information gathered about you and your child will be stored separately from any information that would allow someone to identify who you or your child are (e.g. your full names, your address, your contact details). We will only be able to trace the information we have collected about you and your child back to you using a reference number.

The video and/or audio recordings are considered to necessarily contain personal identifying information. We will, therefore, store the recordings of you and your child separately to the other information we have collected. These recordings will not be labelled with your names or any other personal identifying information but will be labelled with your reference number.

The data collected via online questionnaires will be collected using a tool called 'Limesurvey'. Information collected this way is stored temporarily on high security servers at the University of Birmingham. The University of Birmingham adheres to stringent security practices; however, as is always the case when using the internet, there is a possibility that agents (e.g. 'hackers') might attempt to access the information. Please only participate if you are comfortable with this risk. In the unlikely event of abuse being identified, this information will be disclosed by the research workers.

Only members of the research team at the University of Birmingham and our will have access to information that we collect about you. Personal identifying information will be treated as strictly confidential and handled in accordance with the provisions of the Data Protection Act 1998.

<u>If you/ the person you care for decide(s) to participate, what will happen after that participation?</u>
You and your child/person you care for will receive an individual feedback report describing the results of all of the assessments that were carried out during the study. If requested, we can circulate this report to other interested individuals who you tell us about. Descriptions of research findings will be published in newsletters of the support groups and educational institutions involved. Any request for advice concerning the participant will be referred to Professor Chris Oliver, Clinical Psychologist. The researchers will publish the findings from the study in scientific journals and will present the results at relevant conferences.

What will happen to the data afterwards?
The information that you provide will be locked in a filing cabinet at the University of Birmingham or held on a password protected database. All personal details will be kept separately from the information collected. You will be able to decide whether or not you want to make your research data available to any professionals or clinicians working with you and the person you care for should they wish to see it. Any recording we have made of your and your child will be destroyed 5 years after the end of the study unless you have given us your written consent for the recordings to be used for teaching and/or dissemination.

If participant is not known to us already: At the end of the study, your personal details will be ***destroyed unless you tell us otherwise.*** This means that we would no longer be able to trace the results of your assessments back to you. Before you finish taking part in the study we will give you some information about our regular participant database and you can decide whether you want your details to be retained.

If participant is known to us already and has previously agreed for us to keep their details and contact them for future research: Since you have previously been involved in our research projects at the University of Birmingham and have agreed to be contacted by the research team with information about future research work, we have a copy of your personal details on the 'Regular Participant Database'. This database is password protected and only approved members of our research team have access to your details. We do not share your details with anyone outside the research team.

What happens if I decide that I no longer want my details on the Regular Participant Database?
All you would need to do is contact Chris Oliver on [redacted] Your details would be removed from the database immediately.

Confidentiality
The confidentiality of participants will be ensured. If published, information on the participant will be presented without reference to their name or any other identifying information.

Withdrawal
You can withdraw participation from the study at any point without given a reason. This will not restrict your access to services or your right to treatment. You can withdraw research data from the study up to one month after taking part.

Review
The study has been approved by the National Research Ethics Service (NRES) Committee. The study forms part of an educational project and has been reviewed by the Research Support Group, University of Birmingham.

Complaints
If you would like to make a complaint about this research please contact Brendan Laverty Head of Research and Governance & Ethics, Research Support Group, University of Birmingham.
Email: [redacted]

Appendix C: Lowe Syndrome Study Consent Forms

IMPORTANT:

You need to decide whether your child/the person you care for is able to understand enough about the study to make an 'informed' decision independently about whether or not they would like to participate and to communicate this decision to you. If you are unsure whether or not your child/person you care for is able to understand enough to make a decision independently then we can provide you with some guidelines to help you to assess this. A picture information sheet can also be made available to you if this would be of help. Please contact Dr Jane Waite [redacted] or [redacted] to request a copy of this.

Please choose from one of the following options:

1. **My child/ the person I care for is <u>able to understand</u> what is involved in the study and what will be required from them if they participate and has communicated their decision to me:**

If you think that the person **<u>is able</u>** to understand enough about the study in order to make an 'informed' decision and they decide that they would like to participate then please ensure that they complete **Section 1 of <u>Consent Form A</u>** enclosed, or that you complete it with them, on their behalf. A parent/carer will need to complete **Section 2 of <u>Consent Form A</u> in** order to indicate that they also agree to participate in the study. Please contact the research team if you would like a copy of a picture information sheet or if you need us to adapt this information further, in order to suit your child's needs. Please return the consent form along with the questionnaire pack to us in the prepaid envelope provided. This consent forms can also be completed at [insert web address] logging on with the password: **cere1bra.**

2. **My child/ the person I care is <u>under</u> the age of 16 and is <u>unable to understand</u> what is involved in the study and what will be required from them if they participate (either because they are too young to understand or because they are unable to understand) and cannot communicate their decision to me:**

If you are reading this information on behalf of someone you care for who is **<u>under the age of 16 years</u>** and you decide that the person ***<u>is not</u>*** able to make an 'informed' and independent decision about whether or not they would like to participate, then we would like to ask you to decide whether or not you think that it is in your child's best interests for them to participate in the study and whether you would like to provide your consent to participation on their behalf. If you would like your child/person you care for to participate in this study, please complete **<u>Consent Form B</u>** attached. Please return the consent form along with the questionnaire pack to us in the prepaid envelope provided. This consent forms can also be completed at [insert web address] logging on with the password: **cere1bra.**

3. **My child/ the person I care for is <u>over</u> the age of 16 and <u>unable to understand</u> what is involved in the study and what will be required from them if they participate and cannot communicate their decision to me:**

If you are reading this information on behalf of someone you care for who is **<u>over the age of 16 years</u>** and you decide that the person ***<u>is not</u>*** able to make an 'informed' and independent decision about whether or not they would like to participate, you need to decide whether you wish to act as a personal consultee on their behalf. Please read the attached information on acting as a personal consultee and if you decide to participate in the study complete **<u>Consent Form C</u>** attached. This consent forms can also be completed at [insert web address] logging on with the password: **cere1bra.**

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The Cerebra Centre for Neurodevelopmental Disorders

<u>Consent Form A:</u> For individuals who are able to provide consent to participate in the study

<u>Lowe Syndrome Study</u>

Study Director: Professor Chris Oliver

<u>SECTION 1:</u> Please complete this section if you are a person with Lowe Syndrome:

1. Has somebody else explained the project to you? YES/NO
2. Do you understand what the project is about? YES/NO
3. Have you asked all of the questions you want? YES/NO
4. Have you had your questions answered in a way you understand? YES/NO
5. Do you understand it is OK to stop taking part at any time? YES/NO
6. Do you understand that we may want to make a video of your assessment so that the researchers can check them afterwards? YES/NO
7. Do you understand that your parent/guardian/carer may complete some questionnaires about you? YES/NO
8. Are you happy for your parent/guardian/carer to complete questionnaires online? YES/NO
9. Are you happy to take part? YES/NO

If you don't want to take part, don't sign your name!

If you <u>do</u> want to take part, you can write your name below

You can also choose if you want to say 'yes' to these questions:

10. If your Dr asks to see your results from this project is that OK? YES/NO
11. Can we ask your Dr for information about what caused your genetic syndrome? YES/NO
12. Are you happy for us to contact you again in the future? YES/NO

Your name: _____

Date:_____

The person who explained this project to you needs to sign too. If you are under the age of 16, this should be your parent/guardian.

Name:_____ Sign [paper version only]:_____
Date:_____

<u>SECTION 2:</u> Please complete this section if you are a parent/carer/guardian of a person with Lowe Syndrome who has provided their consent to participate in the study.

1. I confirm that I have read and understood the information sheet dated 7/3/2015 Version 1 for the above study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily. ☐

2. I understand that my participation and that of my child/person I care for is voluntary and that I am free to withdraw at any time without giving any reason, without my or that of my child's/person I care for's medical care or legal rights being affected. ☐

3. I understand that all information collected during the study will be confidential. Only members of the research team at the Cerebra Centre for Neurodevelopmental disorders will know who has participated in the study. All information collected during the study will be stored in locked cabinets that only members of the research team will have access to. No names will be published in any reports. Information will be treated as strictly confidential and handled in accordance with the provisions of the Data Protection Act 1998. ☐

4. I understand that as part of the above study, video/voice recordings of participants and members of participants' families who are involved in the research may be made and stored for further review. ☐

5. I understand that the University of Birmingham will hold the copyright of any video/voice recordings collected during the study but that this does not entitle the University of Birmingham to edit, copy or use the videos for teaching purposes without my written permission. ☐

6. I am happy to be contacted in the future by the University of Birmingham regarding the use of video/ audio recordings for teaching purposes. ☐

7. I understand that I have the option to complete an online questionnaire as part of this study. I understand the risks of using the internet and will only participate in this aspect of the study if I'm comfortable with these risks. (Please note: you can request paper copies of the questionnaire if you prefer; tick here to indicate this ☐) ☐

8. I agree to take part in the above study. ☐

Optional clause: The statement below is <u>optional</u>:

1. I agree to the University of Birmingham research team sharing my research data with any professionals or clinicians working with me and the person I care for should they request to see them. ☐
2. I agree to my child's/person I care for's GP being informed of my participation and that of my child/person I care for's in the study, where access to my child's/person I care for's medical records is required. ☐

Name: _____ Telephone number: _____
Address: _____Email: _____
Relationship to participant:_____ Signature [paper versions only]: _____
Date: _____

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The Cerebra Centre for Neurodevelopmental Disorders

<u>Consent Form B:</u> For Children under the age of 16 who are <u>not</u> able to provide consent.

<u>Lowe Syndrome Study</u>

Study Director: Professor Chris Oliver

SECTION 1: Please complete this section if you are a parent/ guardian of a child (under 16 years) with Lowe Syndrome who is not able to provide consent.

1. I confirm that I have read and understood the information sheet 17/3/2015 Version 1 for the above study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily. ☐

2. I understand that my participation and that of my child/person I care for is voluntary and that I am free to withdraw at any time without giving any reason, without my or that of my child's/person I care for's medical care or legal rights being affected. ☐

3. I understand that all information collected during the study will be confidential. Only members of the research team at the Cerebra Centre for Neurodevelopmental disorders will know who has participated in the study. All information collected during the study will be stored in locked cabinets that only members of the research team will have access to. No names will be published in any reports. Information will be treated as strictly confidential and handled in accordance with the provisions of the Data Protection Act 1998. ☐

4. I understand that as part of the above study, video/voice recordings of participants and members of participants' families who are involved in the research may be made and stored for further review. ☐

5. I understand that the University of Birmingham will hold the copyright of any video/voice recordings collected during the study but that this does not entitle the University of Birmingham to edit, copy or use the videos for teaching purposes without my written permission. ☐

6. I understand that I have the option to complete an online questionnaire as part of this study. I understand the risks of using the internet and will only participate in this aspect of the study if I'm comfortable with these risks. (Please note: you can request paper copies of the questionnaire if you prefer; tick here to indicate this ☐)

7. I am happy to be contacted in the future by the University of Birmingham regarding the use of video/ audio recordings for teaching purposes. ☐

8. I agree to take part in the above study. ☐

9. I agree to my child/person I care for taking part in the above study ☐

Optional clause: The statement below is optional:

10. I agree to the University of Birmingham research team sharing my research data with any professionals

or clinicians working with me and the person I care for should they request to see them.

11. I agree to my child's/person I care for's GP being informed of my participation and that of my child/person I care for's in the study, where access to my child's/person I care for's medical records is required. ☐

Name: _____Name of person you care for: _____

Address:_____Email: _____

Telephone number:_____Relationship to participant: _____

Signature [paper version only]: _____Date: _____

<u>Consent Form C:</u> For individuals <u>over</u> the age of 16 who are <u>not</u> able to provide consent.

Before deciding whether to participate, please ensure you read the information on acting as a <u>personal consultee (attached/link)</u> for the person you care for.

[Paper: By initialing the boxes you are acting as a personal consultee and consenting on behalf of the person you care for to participate in this research].

[Online: By ticking the boxes and clicking on 'Next' at the bottom of the page you are acting as a personal consultee and consenting on behalf of the person you care for to participate in this research].

Please read the following statements and indicate each one you agree with.

Please initial [online: tick] box…

1. I have been consulted about the person I care for's participation in the research project titled 'Understanding Behavioural Difficulties in Lowe Syndrome'. I have read the information sheets (version: ...) and had the opportunity to ask questions about the study and understand what is involved. ☐
2. In my opinion he/she would have no objection to taking part in the above study. ☐
3. I understand that I can request he/she is withdrawn from the study at any time without giving any reason and without his/her care or legal rights being affected. ☐
4. I understand that all information collected during the study will be confidential. All information collected during the study will be held on secure servers by the hosting website and then transferred to locked cabinets that only members of the research team will have access to. No names will be published in any reports. Information will be treated as strictly confidential and handled in accordance with the provisions of the Data Protection Act 1998. ☐
5. I understand that as part of the above study, video/voice recordings of participants and members of participants' families who are involved in the research may be made and stored for further review. ☐
6. I understand that the University of Birmingham will hold the copyright of any video/voice recordings collected during the study but that this does not entitle the University of Birmingham to edit, copy or use the videos for teaching purposes without my written permission. ☐
7. I understand that I have the option to complete an online questionnaire as part of this study. I understand the risks of using the internet and will only participate in this aspect of the study if I'm comfortable with these risks. ☐

(Please note: you can request paper copies of the questionnaire if you prefer; tick here to indicate this ☐)

8. I agree to take part in the study titled 'Understanding Behavioural Difficulties in Lowe Syndrome'. ☐

Optional clause: The statement below is <u>optional</u>:

11. I agree to the University of Birmingham research team sharing research data with any professionals or clinicians working with my child/the person I care for should they request to see them. ☐

12. I agree to my child's/person I care for's GP being informed of my participation and that of my child/person I care for's in the study, where access to my child's/person I care for's medical records is required. ☐

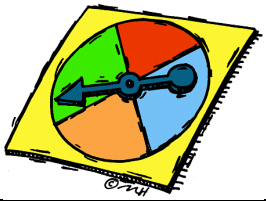
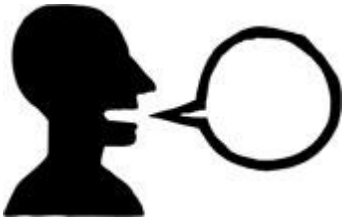
Name: _____Name of person you care for: _____

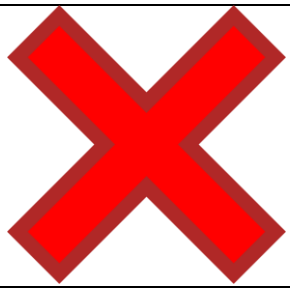
Address:_____Email: _____

Telephone number:_____Relationship to participant: _____

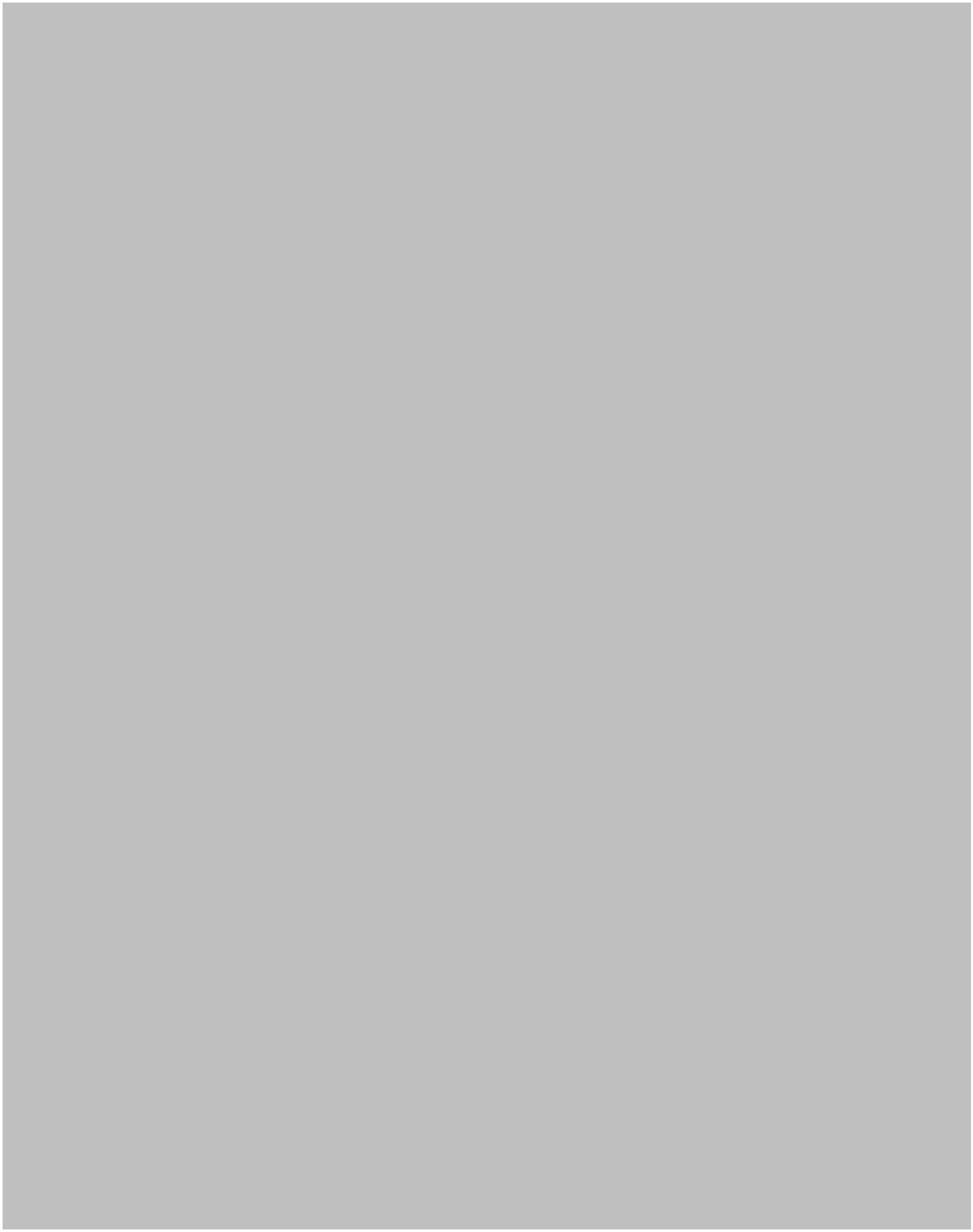
Signature [paper copies only]: _____Date: _____

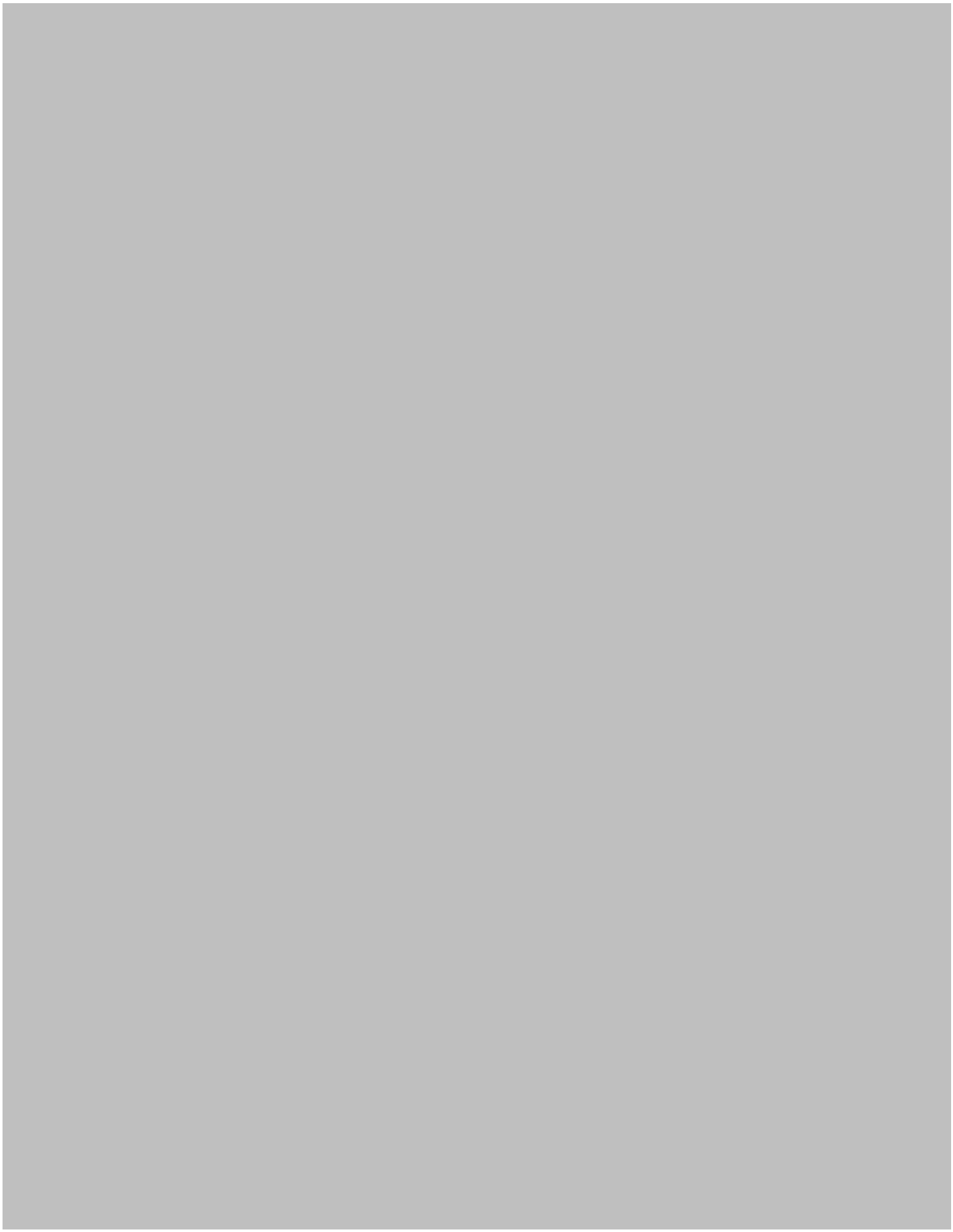
Appendix D: Picture Information Sheet

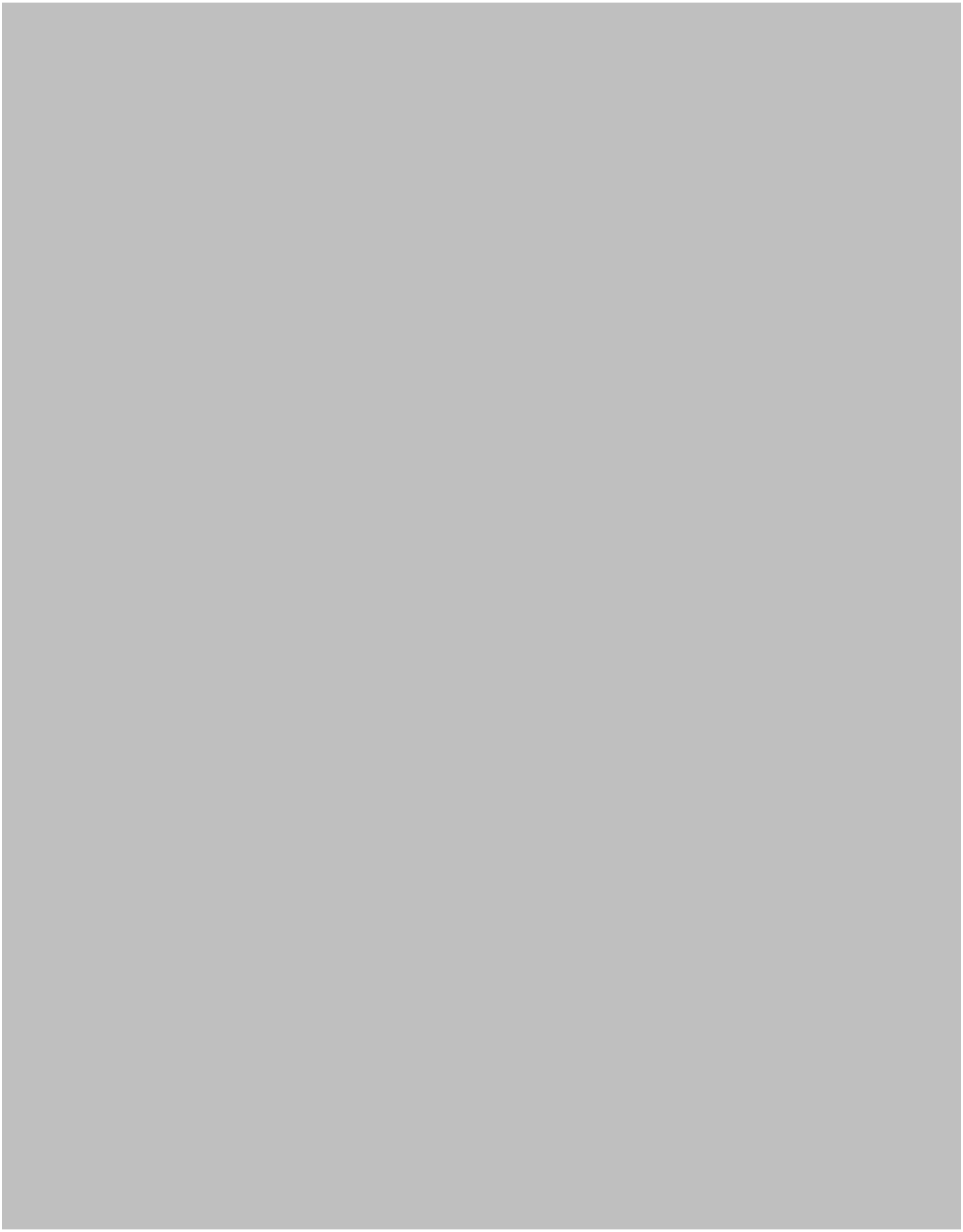
	Hello, our names are Jane Waite, Alicia Kutsch and Helen Cressey.
	We are doing some research about people with Lowe syndrome. We want to visit you at home and do some tasks with you. You can also visit us at the University.
	These tasks are just like games and games on the computer
	We also want to speak with your parents or carers about you.
	They may answer questions on the phone, computer or by meeting with us.
	The things we want to ask them about are: • How old you are • Things you are good at and things you are not so good at. • What you do on a day to day basis

	When we have finished, everything we learn about you will be kept in a safe place.
	Remember that you do not have to say yes. If you do not want to take part, then say no.
	If you do say yes, and then change your mind, that is ok. Just tell us no. You do not need to say why you said no.
	Thank you!

Appendix E: Research Ethics Committee Approval Letter







Wales
REC 4

Attendance at Sub-Committee of the REC meeting on 28 May 2015

Committee Members:

Name	Profession	Present	Notes
Dr Kath Clarke	Senior Investigations Manager	Yes	
Mr John Gittins	Coroner	Yes	
Mr Philip Richards Sub-Committee Chair	Associate Specialist - Surgery	Yes	

Also in attendance:

Name	Position (or reason for attending)
Mrs Tracy Biggs	Research Ethics Committee Manager

Appendix F: Lowe Syndrome Temper Outburst Interview Schedule
(adapted from Tunnicliffe, 2012)

The interview schedule has been removed from the electronic copy of this thesis.

A copy can be made available on written request to the senior author

c/o Professor Chris Oliver,

The Cerebra Centre for Neurodevelopmental Disorders,

School of Psychology,

University of Birmingham,

Edgbaston,

Birmingham, B15 2TT.

Email: cndd-enquiries@contacts.bham.ac.uk

Appendix G: Coding scheme for Lowe Syndrome Interviews

(adapted from Tunnicliffe et al., 2014)

Question	Item	Coding Instructions	Example
1	Next outburst	Taken from CBI (Oliver et al. 2003). Fixed response categories	By this time tomorrow
2	Longest outburst		More than an hour
3	Typical length of outburst		More than an hour
4	Longest over 1hr	Report duration	3 hours
5	What keeps an outburst going for longer periods?	Report reason or state unable to identify	OCD - "gets in the wheel" "Mind has to click in – can take hours"
6, 10, 11	Description of last temper outburst, sequence and predictability	Listen to account Report antecedent Is the sequence typical? Report each behaviour in sequence of occurrence Report frequency of each behaviour	Antecedent: Pacing, pulling lip, twisting fingers (always). Typical sequence: Smacking himself (sometimes), shouting (often). Hitting furniture/walls (often). Hitting others (rarely). Crying (sometimes). Verbal and physical threats (rarely) Lip pulling (always) Predictable sequence? - Yes
7	Precursors	State yes or no Report precursor if yes	Yes Asking for attention
8	Intervention at time of precursor	Report intervention Report success out of 10.	Agreeing to what he wants 8/10
9	Critical point?	State yes or no Report critical point if yes	Yes Invasion of personal space or uncontrollable crying
12	Emotion during outburst	Report emotion	Confusion/frustration
13	Emotion after outburst	Report emotion	Reconciled/exhausted
14	Behaviour after outburst	Report behaviour	Goes to his room
15	Point of intervention – when which behaviour is seen?	State behaviour State intervention	As start. Talk about positive things.
16	How often intervention used?	Item taken from CBI (Oliver et al., 2003). Fixed response categories	Always
17	Other strategies	Report other strategies	Ignore Time out Withdraw from situation Social stories Explain decisions Write down reasons
18	Reasons for other strategies	State reason if given	Depends on topic, and who else is around. Does not have outbursts in public.
19	Different response at different stage?	Report different strategies and when used.	At start might remind him of strategies suggested by therapist; then ask him to think about how his grandfather would respond in the same situation.
20	One thing most likely to stop an outburst	Report strategy	Giving in. Undivided attention.

21	Success rate for above	Report success out of 10.	Not answered.
22	Antecedent to last outburst	Report antecedent	Saying “no”.
23	Most common antecedent	Report antecedent	Frustrated goals (saying “no”)
24	Proportion of temper outbursts triggered by most common antecedent	Report proportion out of 10.	8/10
25	Does most common antecedent always result in an outburst?	State yes or no	No
26	Out of 10, how often does antecedent result in temper outburst?	Report how often out of 10.	Not answered.
27	Information on times when outburst not antecedented	Report reasons given or state unable to identify.	Negotiation – flexibility – offer alternatives.
28	Other antecedents	Report other antecedents	Relationship difficulties
29	More likely to occur at certain times of day? Setting events (motivational states)	State yes or no and report time of day. Report other setting events	No None identified.
30	List of other antecedents	Fixed list of 21 antecedents. Tick those which apply.	Change in routine. Change in expectation. Conflicting information. Told off about food. Imperfection in belonging. Making mistakes. Losing item. Thinks he has lost something. Might lose something. Thinks something might have been stolen. Asked to do what does not want to do. Told he cannot have something (non-food). Following disagreement. After being teased.
31	Anything else you would like to mention about temper outbursts?	Record any information given	Deterioration over time – worsening outbursts as he has got older.
32	Any questions about the research?	If unable to answer record question for later feedback	No questions asked.