Summary of research study: Temper outbursts in Lowe Syndrome

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Why was the study completed?

This summary will outline an interview study that focused on temper outbursts that are observed in individuals with Lowe Syndrome. This interview study is part of a broader study aiming to identify behaviours that are common in this genetic syndrome.

The interview study was completed as approximately 70-80% of individuals with Lowe Syndrome display behavioural difficulties such as self-injury, aggression and temper outbursts.

There have been factors that have been identified that are associated with the development and maintenance of these behavioural difficulties in other groups of individuals but they have never been explored in individuals with Lowe Syndrome.

Exploring these factors may allow us to develop more effective interventions and improve quality of life for individuals with Lowe Syndrome and their families.

What the study included?

To further explore particularly, temper outbursts, interviews were conducted with parents/carers of individuals with Lowe Syndrome.

Interviews were completed with parents/carers of 17 individuals with a diagnosis of Lowe Syndrome:

- 9 adults (18 years or older)
- 8 children (under 18 years)

The interview questions aimed to identify what occurs during a typical temper outburst including what happens before, during and after and also the strategies that parents have used to manage outbursts. All parents/carers were asked the same questions.

The study was similar to another study that focused on temper outbursts that are common in Prader-Willi Syndrome. This allowed comparison between the studies and these two genetic syndromes.

The findings from the interview were broken down into 5 sections:

- Frequency and duration of temper outbursts
- Factors that increase the likelihood of a temper outburst occurring
- Behaviours that are observed during an outburst
- What happens following an outburst
- The strategies parents/carers had used to manage outbursts

What the study found?

Frequency and duration of temper outbursts

All parents/carers that were interviewed had already reported that the person they cared for had frequent temper outbursts.

- Parents/carers reported that temper outbursts happen at least once a day with some indicating that one would be observed within the next hour
- Typically, it was reported that temper outbursts last between 1 and 15 minutes
- However, 41% of parents/carers had experienced an outburst that lasted longer than an hour and the longest outbursts could last between 2 and 4 hours

Factors that increase the likelihood of a temper outburst occurring

- Tiredness, thirst and hunger
- Physical pain/discomfort
- General low mood
- Time pressure
- Changes to routine/changes in noise level e.g. blender starting
- Most common trigger: “being told no”, “not getting his own way”, “doing something he doesn’t want to do”, “not being able to do something he wants to do”
- This common trigger was also associated with extended temper outbursts
- Other triggers for extended outbursts were: anxiety, frustration, needing attention and not being able to “let go” of an issue

Behaviours that are observed during an outburst

- All parents/carers indicated that there are clear warning signs that precede an outburst which include repetitive questioning, shouting, non-compliance, self-injury and lashing out at others
- During outbursts, emotional vocalisations and physical aggression were the most common behaviours which included shouting, screaming, cursing, hitting, kicking and biting
- Approximately 71% of parents/carers also reported aggression towards property such as kicking or hitting furniture, walls or windows and throwing objects
- The emotions identified by parents/carers as being experienced by the person they care for were mostly anger and frustration

What happens following an outburst

- The majority of parents/carers reported that following an outburst, the person they care for apologises, asks for a hug, seeks reassurance or makes a cup of tea
- Some reported that the person they care for may go back to what they were doing as if nothing has happened

Strategies parents/carers had used to manage outbursts

- When parents/carers observed the warning signs that precede a temper outburst, 41% reported the most successful strategy seemed to be distraction or redirection to another activity
- This strategy was successful in preventing an outburst between 50-90% of the time
- Other strategies that parents/carers reported were calm reasoning, removal of choice, giving attention, offering help and reiterating clear routine or instructions. These strategies were successful between 40-80% of the time
- One parent/carer reported that nothing seemed to work even in the early stages of a temper outburst
- Once an outburst has progressed, the above strategies become less effective and may reduce the length of the outburst between 0-60% of the time
- However, at this stage, most parents/carers reported that they felt the most successful strategy was to remove the individual to a quiet and safe environment to allow them to calm down in their own time. Although this strategy did not necessarily stop an outburst immediately, it reduced the risk of harm to the individual themselves and others around them
- Approximately 24% of parents/carers reported that the only thing that would stop an outburst was giving in to requests but even this, was not successful all of the time

Temper outbursts in Lowe syndrome compared to Prader-Willi syndrome

Temper outbursts seem to be similar in Lowe syndrome and Prader-Willi syndrome but there were some differences that were identified.

- Temper outbursts appear to be more frequent in Lowe syndrome
- Temper outbursts can last longer in Prader-Willi syndrome e.g. up to 24 hours
- In Lowe syndrome, the most common trigger for temper outbursts was being thwarted and the individual being denied something they want when they want it
- Additionally, change in routine or expectation was found to be a trigger in Lowe syndrome and this is the most common trigger in Prader-Willi syndrome
- Individuals with Lowe syndrome appear to show higher levels of physical aggression and aggression to property compared to individuals with Prader-Willi syndrome
- Parents/carers in the Lowe syndrome group also reported behaviours such as urinating or smearing which were not reported at all in Prader-Willi syndrome

What do these research findings mean?

The behaviours that have been reported during an outburst in Lowe syndrome are similar to that of typically developing children (2-4 years old) and individuals with other genetic syndromes e.g. Prader-Willi syndrome. This is important to indicate as this suggests there may be similar processes that are driving these behaviours in different groups of individuals.

For example, in young children temper outbursts are thought to be associated with under-developed thinking processes which are involved in emotion regulation, planning and controlling actions/behaviours. These thinking processes are often referred to as "*executive functions*". There is new evidence to suggest that individuals with Lowe syndrome may have difficulty with some of these planning and control tasks.

The comparison between Lowe syndrome and Prader-Willi syndrome was particularly important as it was highlighted that changes in routine and expectation are a common trigger for temper outbursts in both genetic syndromes.

In Prader-Willi syndrome change in routine has been associated with the cognitive (thinking) difficulty of switching between different tasks. From this, there has been the development of interventions that support task switching for individuals with Prader-Willi syndrome and there has been evidence to suggest that these interventions have helped to reduce the number and length of temper outbursts in this group.

The similarity in the behaviour patterns of temper outbursts between the groups may mean that individuals with Lowe syndrome also experience this cognitive difficulty but further research is needed to explore the exact nature of that difficulty and how interventions could be developed for individuals with Lowe syndrome.