

A RESEARCH UPDATE FOR PARENTS AND CARERS

PRELIMINARY RESULTS: THE FACTORS THAT MAY LEAD TO TEMPER OUTBURSTS IN CHILDREN AND ADULTS WITH LOWE SYNDROME

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WHAT IS A TEMPER OUTBURST?

A temper outburst is a cluster of behaviours that often occur together and may include behaviours such as crying, shouting, stamping, face flushing and arguing.

When parents describe temper outbursts they often describe these outbursts as being emotional in nature with the person experiencing a range of emotions from frustration to rage.



TEMPER OUTBURSTS ARE VERY COMMON IN CHILDREN AND ADULTS WITH LOWE SYNDROME

Low syndrome is a very rare genetic syndrome that occurs in 1 in 500,000 individuals and mainly affects males. It is caused by a change in a single gene on the X chromosome. To date, most of the research with children and adults with Low syndrome has focused on the health difficulties associated with the syndrome; however, parents and carers often report that one of the greatest perplexities they face on a regular basis is trying to understand why particular behaviours are shown by their children with Low syndrome. Everyone with Low syndrome is an individual with their unique preferences and personality; however, there are certain behaviours that appear to occur more often in children with Low syndrome and some of these behaviours are particularly challenging for families. One cluster of behaviours that appears to occur in children and adults with Low syndrome are temper outbursts, which are reported to be shown by as many as 8 out of 10 people.

What causes a temper outburst?

Researchers at the University of Birmingham have been conducting a study over the last three years to learn more about why temper outbursts occur in people with Low syndrome. A better understanding of the factors that lead to temper outbursts is

important because it will inform the work of health professionals working with children and adults with Low syndrome, as well as help family members decide on the best intervention strategies to try with their children. Parents have reported that temper outbursts can have a significant impact on the quality of life for people with Low syndrome as well as the day to day functioning of the family. Therefore, it is important to understand these behaviours to reduce the impact on the person with Low syndrome as well as on parents and carers. Over thirty-five families of children with Low syndrome from the UK, USA and Australia supported the researchers from the University of Birmingham to conduct the research project.

What is the purpose of this research update?

This research update will describe what the researchers have learnt during their study, what the findings mean for people with Lowe syndrome and their families, and what the researchers hope to find out more about in the future. The results described below are preliminary, as some of the findings are not published yet in scientific journals; this can take several months after a study ends. Therefore, the results should be interpreted with caution until they are published. The research team anticipate that the majority of the findings will be published within the next 18 months.

The precise nature of temper outbursts in Lowe Syndrome

One of the research team, Dr Helen Cressey, spoke to families and asked them about the characteristics of the temper outbursts that they observe in their children. Some of the key findings from this interview study were:

- Temper outbursts typically last between one to fifteen minutes but may last for an hour or longer
- Aggression is often a core feature of temper outbursts in Lowe syndrome
- Other behaviours such as self-injury, hitting windows or deliberate urination/defecation may occur
- The most common triggers reported by parents are changes in routine and unmet desires e.g. wanting something that is not available
- Some strategies that parents report are helpful during outbursts are distraction, redirection or allowing an individual to calm themselves down in a safe and quiet environment
- Temper outbursts may be associated with difficulties in thinking processes such as emotion regulation, planning and controlling behaviour. More research is needed to further understand this potential association

“There is often not much we can do once the outburst starts. He usually takes himself off to his room to calm down but this might take some time” – Parent of an adult with Lowe syndrome

HEALTH RELATED DIFFICULTIES AND BEHAVIOURAL DIFFICULTIES IN LOWE SYNDROME

When there is a sudden change in a person’s behaviour, such as an increase in temper outbursts or behaviours that pose a challenge to families, it is important to rule out whether a physical health difficulty might be driving



the behaviour. Research has shown that if a person with intellectual disability is in pain, this can increase the likelihood of this individual showing challenging behaviour.

Common health difficulties such as middle ear infections, reflux or constipation have all been associated with challenging behaviours in people with intellectual disability. Given this previous research, one of the

hypotheses that the researchers at the University of Birmingham had was that physical health difficulties might explain the heightened presence of temper outbursts in children and adults with Lowe syndrome, particularly given that health difficulties are particularly common in the syndrome.

To date, the researchers found no association between the presence of 16 health difficulties, including seizures, and temper outbursts. They also observed children and adults with Lowe syndrome and watched for signs that they were in pain. The researchers observed very few behavioural signs of pain in the individuals they observed. Furthermore, very few parents reported that they thought behaviours were associated with pain in the group, which fitted with what the researchers had observed. While pain was not associated with behaviour in the current study, this is not to say that for an individual child with Lowe syndrome, pain and health related difficulties will not be contributing to behavioural difficulties. For example, the research team conducted an assessment with a boy with Lowe syndrome whose challenging behaviour and temper outbursts reduced considerably once he had dental work carried out. However, overall pain and health difficulties did not appear to explain why more temper outbursts occur across the whole group of individuals with Lowe syndrome.

ACTION BOX 1.

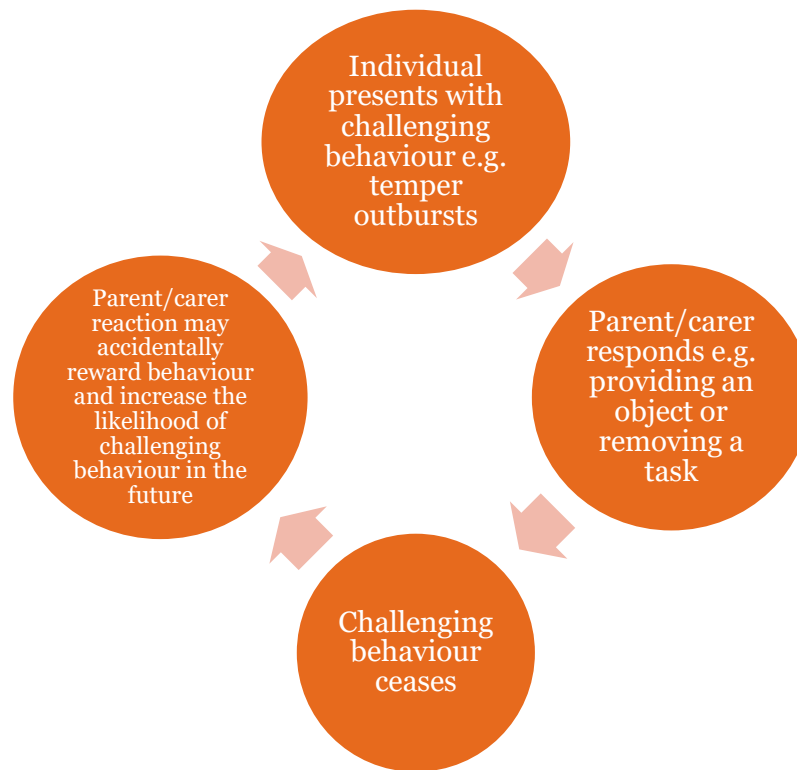
Ensure you rule out pain before you consider any other cause of behaviour.

Even if you are not concerned about pain in the person you care for it is often worth familiarising yourself with the detection of pain in people with intellectual disability. Sometimes pain is not easy to spot. The Cerebra Pain Guide is a useful resource and can be downloaded <https://tinyurl.com/ycoxoldl> and gives comprehensive information about some of the signs that a person is experiencing pain. A pain assessment tool accompanies the guide, which is useful when ruling out whether a person is in pain. It is always wise to see your GP if there is a sudden escalation in behaviours that challenge or behaviours appear to come 'out of the blue'.

THE TRADITIONAL BEHAVIOURAL MODEL IS CENTRAL TO UNDERSTANDING BEHAVIOURS THAT CHALLENGE IN PEOPLE WITH INTELLECTUAL DISABILITIES

Behaviours that challenge often include behaviours such as self-injurious behaviour, aggression towards others and temper outbursts. Typically, when parents and carers observe these types of behaviours and are asked to think about some of the potential causes of the behaviour, they tend to focus on what happened just before the behaviour – they try to find what is triggering it. This makes sense because as human beings we tend to think in 'cause and effect' terms, whereby the cause usually proceeds the effect. While what happens before the behaviour is important to consider, equally important is what happens immediately after a behaviour occurs. Years of research has indicated that the consequences that follow a behaviour can make the behaviour more likely to happen again in the future, particularly when a child or adult has an intellectual disability and communication impairments.

To illustrate this, consider the following interaction that often occurs between a parent and a child:



In this cycle, the consequence that follows after the behaviour is the removal of something (a task); however, this cycle can also occur when something is 'added' rather than being taken away after a behaviour is shown. For example, if a child hits their head and a parent responds to this with concern, the behaviour might be strengthened over time because the concern a parent shows is rewarding to the child, and strengthens the behaviour. Similarly, if a child pushes a sibling and this is followed by the sibling giving the child their iPad, this may increase the likelihood of the child showing pushing behaviour again in the future because it has led to being given the iPad in the past. The purpose the behaviour is serving for a child is often called the *function* of the behaviour. There are two important things to remember about this though:

BOX 1. It's not the parents' fault

A parent may find challenging behaviour e.g. self-injury unpleasant or aversive and consequently act to do anything they can to stop the challenging behaviour from occurring.

This is a natural reaction and a parent may feel relief after the challenging behaviour has stopped, increasing the likelihood that they may respond this way in the future. Parents are only responding as anyone would in the situation and it is not the parents fault that challenging behaviour occurs.

BOX 2. It's not the person with Lowe syndrome's fault

The person with Lowe syndrome may engage in challenging behaviour because it has been followed by removal of a task or in the above example, the availability of the iPad. Once this process has occurred many times, an individual may quickly learn to engage in behaviours because it leads to these outcomes.

This does not mean that an individual intentionally hurts themselves or others in order to gain the desirable outcome. It is a completely natural process whereby behaviours that challenge evoke reactions from parents/carers and these reactions are rewarding for the individual with Lowe Syndrome. Over time the behaviour and consequence are paired.

A member of the research team at the University of Birmingham, Alicia Kutsch, attempted to explore whether particular triggers occurred before an outburst in Lowe syndrome, as well as the consequences that followed after outbursts. Alicia found that temper outbursts were most likely to be triggered by the removal of a preferred item, or were followed by a preferred item being given. She also found that temper outbursts were often followed by individuals not having to follow through with non-preferred activities.

Typically, psychologists who work with families to try and understand why behaviour is occurring might examine these patterns by asking parents to keep a record of what happens before, during and after the behaviour is shown. You can find out more about these principles by downloading the self-injurious behaviour guide by following the link below. While this guide focuses on self-injury and not temper outbursts or aggression, many of the behavioural ideas that are described here have been argued to be applicable to a range of behaviours.

Self-injurious behaviour guide: <https://tinyurl.com/y82tkctf>

You can also work through the Lowe syndrome challenging behaviour resources on FIND (www.findresources.co.uk) that explain many of the principles of this traditional behavioural approach to understanding behaviour. These resources will help you to develop a good understanding of the basics of why behaviour occurs and are a good starting point. In addition, the Challenging Behaviour Foundation (<http://www.challengingbehaviour.org.uk/>) have a number of resources that may be useful. There are also some useful hints and tips for managing behaviour in these resources. It is important not to change how you respond to behaviours that challenge before working through these resources carefully and discussing changes you may make with a health professional, such as a clinical psychologist.

BEYOND THE BEHAVIOURAL MODEL IN LOWE SYNDROME

Despite the traditional behavioural model being very important for understanding why behaviours might occur in Lowe syndrome, research is now suggesting this might not be the whole picture. During Dr Helen Cressey's temper outburst interviews with families, she learnt some additional interesting things about temper outbursts in Lowe syndrome.

You may remember from above that Alicia Kutsch found that behaviour was most likely to be followed by something a person wants, usually in the form of an object, or by the removal of a non-preferred task. The research team wondered whether this might indicate that people with Lowe syndrome have difficulty coping with having their desires thwarted by others. The research team hypothesised that maybe individuals with Lowe syndrome had difficulty being able to *delay gratification*.

BOX 3. The traditional snack delay task



Waiting longer obtains more rewards

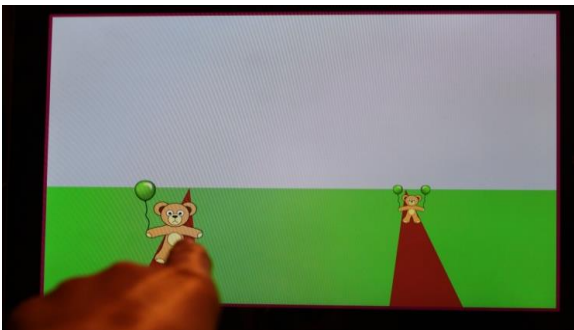
We delay gratification whenever we are asked to wait for something we really want. In very young children this is often assessed using a marshmallow task. In the marshmallow task children are left alone with a marshmallow and are told they can eat the one marshmallow now or wait until the researcher returns and get two marshmallows. Children that have difficulty waiting usually eat the one marshmallow and these are the children that often have more difficult time regulating their emotions when they are upset. The task relies on brain based mental abilities that help us control and regulate our behaviour. To pass the marshmallow task the child has to put the brakes on 'or inhibit' their behaviour

in a context where they are very motivated to obtain the marshmallow that is in front of them. Therefore, the task has an emotional component because of the child's desire to obtain the marshmallow.

To test the hypothesis that children and adults with Lowe syndrome might have difficulties delaying gratification the researchers used two assessments. The first assessment required parents to complete a questionnaire about their child's ability to inhibit behaviour, regulate emotions, and be flexible. The answers on this questionnaire showed parents who reported that their children had the poorest inhibition and emotional control were more likely to be the children that had the most severe temper outbursts. However, a limitation of this questionnaire assessment was that some of the questions on the questionnaire and the assessment of temper outbursts

overlap, which could explain this result.

BOX 4. The Bear Task



The second assessment was a computer assessment with individuals with Lowe syndrome. Children and adults were asked to play a game to win as many stickers as possible. The rules of the game were straightforward: there were two bears on a computer screen and the person with Lowe syndrome could choose one of them on each go by touching its tummy. When a bear was chosen, it walked towards the front of the screen and let go of a balloon. One of the bears walked quickly and held one balloon, which indicated that the child would win one sticker.

The other bear walked very slowly and held two balloons. If the

person with Lowe syndrome selected the slow bear they won two stickers once the bear reached the front of the screen. Children and adults with Lowe syndrome were asked to select a bear on each go (20 goes in total) and had to try and win as many stickers as possible by the end of the game. To succeed on this game (by choosing the slow bear) the person with Lowe syndrome had to wait and delay gratification.

The results from the bear task showed that people with Lowe syndrome who had more difficulty waiting on this task were more likely to be the individuals that showed temper outbursts. This result was not explained by visual impairment or ability level. These results need to be replicated but they are the first to point to emotional regulation/delay of gratification difficulties in Lowe syndrome. These results also fit with existing research that has found temper outbursts are associated with lower self-control in typically developing children.

WHAT DOES THIS ALL MEAN FOR UNDERSTANDING TEMPER OUTBURSTS?

The traditional model for understanding behaviours via the consequences that follow them is a robust model with many years of research behind it; it is likely to still be applicable to people with Lowe syndrome. However, research suggests that individuals with Lowe syndrome may have difficulty controlling and regulating their behaviour when they are thwarted. This lack of control may be because of differences in cognitive brain-based processes in Lowe syndrome. Essentially, individuals with Lowe syndrome may find it very difficult not to react in situations when they are thwarted, and once behaviours are shown it may be very difficult for the person to calm down.

STRATEGIES AND TIPS TO MANAGE BEHAVIOUR IN LOWE SYNDROME

Carers and professionals working with Lowe syndrome should assess the function of the behaviour using the traditional methods that are used for understanding behaviour. More information can be found in the resources described earlier in this guide. A good understanding of any function of behaviour will inform interventions.

However, it is important to not overlook the contribution of potential emotional regulation difficulties in Lowe syndrome. There are no evidence-based strategies for improving emotional control in children with Lowe syndrome but some strategies that have been tried with children with intellectual disabilities who have moderate to mild intellectual disability and young typically developing children may be useful for parents to try with their children, particularly if their children are verbal. These strategies may not be useful for all individuals with Lowe syndrome as it will depend on the person's ability level and language skills. We would always recommend that parents and carers seek advice from their paediatrician, GP or behavioural specialist before implementing any new strategy and that they get input from a clinical psychologist if possible.

BOX 5. Strategies to promote emotional regulation in children with intellectual disabilities

- **Self-monitoring interventions.** e.g. introduce a self-monitoring diary where the person with Lowe syndrome is supported to record how they feel each day. This can be a simple scale with three pictures of faces (happy, neutral, angry) or an anger thermometer. Self-monitoring is often an early step in being able to regulate emotions, so while it may not help immediately during an outburst, it may help in the long term. The person may need support to help them learn to identify emotions in themselves and others first.
- **Relaxation** (counting & breathing). Teaching a person a strategy to relax can be very useful. This is usually taught when the person is not experiencing emotions associated with an outburst and is practiced many times. The person can then be prompted to use their relaxation strategy when they experience a difficult emotion. These sorts of strategies work best if a person is prompted to use them when they are showing very early signs of a temper outburst.
- **Turn taking games** may help a person to improve self-control and inhibition.
- **Role play.** More able individuals may be able to practice regulating their emotions using role play. Once a person with Lowe syndrome has learnt an emotional regulation strategy such as counting and breathing, it may be possible to role play scenarios where this strategy may need to be applied. Start with scenarios that do not evoke a lot of emotion in the person, followed by more challenging role-plays as the person masters the use of the relaxation strategy.

Key References

- Cerebra pain guide: <https://tinyurl.com/ycoxoldl>
 - Cerebra self-injurious behaviour guide: <https://tinyurl.com/y82tkctf>
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