

Screaming Episodes in Rett syndrome

Dr Hilary Cass, Neurodisability Consultant

Evelina London Children's Hospital St Thomas, UK

Through adolescence and into adulthood, a proportion of people with Rett syndrome may develop screaming episodes by day or sometimes by night. Even when short, they can be stressful for everyone concerned, and longer episodes can be particularly distressing and difficult.

Sometimes the person with Rett syndrome may have outbursts of laughter. Parents and carers often feel quite powerless to deal with these, and it is very upsetting when the person with Rett syndrome appears to be unhappy or in pain. You may find the following suggestions helpful:

- Keep a diary of the attacks, including how long they last and what was happening immediately before and at the outset. Also note any factors that help terminate the episode.
- Check out possible causes of pain – period pain, a urine infection, dental problems, dislocated hip, pressure points on back, feet or buttocks, gastro-oesophageal reflux (which causes acid regurgitation and heartburn).

Sometimes a trial of regular (3–4 per day) paracetamol or ibuprofen can be helpful. If the person with Rett syndrome settles on this it is likely that there is a source of pain that you need to track down!

- Check out environmental problems – changes of teachers, changes of carers, upsetting noises etc.
- Some people with Rett syndrome can get very frustrated when unable to communicate their needs or if bored during periods of inactivity. It is important to offer choices whenever possible. Music can be helpful during quiet periods when there are no other activities going on. Some girls and women have a favourite video that regularly stops the screaming. Advice from a speech and language therapist in supporting communication may be helpful.
- Check out emotional problems – depression is common in all adolescents, and more so in those with a neurological disability. You should particularly think about this if the person with Rett syndrome is tearful, off her food and / or waking at night.

Sometimes a trial of anti-depressant medication can be helpful. This should be prescribed and monitored by a consultant with relevant experience.

- Many people with Rett syndrome enjoy other forms of therapy such as aromatherapy, music therapy etc. – all such options should be explored.
- Finally, there is the residual group of people with Rett syndrome in whom no answer can be found. With the indications we have of widespread dysfunction in the brain it is possible that periodic mental distress is part of the disorder itself.

If you are sure that nothing important is being masked, this very small group of people with Rett syndrome may respond to various sedative or mood stabilising medications. However, these should only be prescribed by someone with expertise in this area and should be very carefully monitored.