Sex chromosomal trisomies (SCT) are quite common: the most well known is Klinefelter syndrome, 47,XXY, the other two are 47,XY and 47,XXX (Trisomy X or Triple X). People with an extra X or Y chromosome have a higher risk of neurodevelopmental difficulties, for instance socio-emotional problems. There is a higher risk of emotional outbursts, affective problems and diagnosis of an autism spectrum disorder (ASD). We do not know much about how the extra chromosome contributes to the developmental issues. So far, studies have mainly focused on the behaviour of SCT children and did not measure the responses of the body (e.g. heart rate). By studying how children with SCT react emotionally and express themselves after a scary event, we learn more about how these skills relate to daily life problems. By studying how they differ from children with a typical chromosome pattern (46,XX or 46,XY), it can help us recognize children with a higher risk for future problems and to develop interventions to help them.

It is important that when a child is emotionally moved by a situation (e.g., their body responds to the situation with increased heart rate, increased breath rate), that child also needs to effectively express their emotion. When a child shows fear, people around the child will come to help. When it shows anger, people may distance themselves. These interactions will help the child to learn how to cope with emotions. When children respond in an unusual way, other people will respond accordingly. That may result in a higher risk of problems in the interaction with other people and the development of coping skills.

In this study, 90 SCT children and 97 (no SCT) children from the local communities took part. Most of the SCA children were diagnosed prenatally, so the risk of selecting children with more severe developmental issues is relatively low. The number of children who participated was quite high, which helps to find a wider range of functioning and which gives a representative sample of the group we study. The heart rate and the expression of negative emotions of the children were registered in a direct, objective way. The children had heart rate monitors on and sat in a car seat. They were videotaped from two angles. All children first watched a video of a fish tank, to measure their resting heart rates. Then they were exposed to a frightening situation: a robot toy approaching them with a scientist who wore a lab coat, did not speak and had no eye contact. This is a stress inducing event that reflects a ‘threatening situation’ in daily life (for example a barking dog) which causes the emotional system to respond fast. After meeting the robot, the caregiver of the child was allowed to come to the child and comfort it if necessary while the child still sat in a car seat. In this way, we measured recovery.

The SCT children reacted differently compared to the average children. They showed less physical response to the situation but it took them longer to recover. They also expressed less emotions in their face. Another finding was that the heart rate and the emotional expression of the children did not always match within one child. In the no-SCT children, there was a stronger relation between heart rate and expression of emotions within one child. So a child with a higher heart rate would also show more emotions. An SCT child could have a high heart rate and less expression of emotions or a
relatively low heart rate and much expression. This has been seen before in a similar experiment with ASD children.

This is an important finding because emotions have an impact on daily functioning. So differences in processing emotions likely has an effect on the development of children. The observations in this study also help us to understand the huge difference we see within the group of SCA children. The results are similar to results from other studies. So the combination of the studies, contributes to the knowledge of the neuropsychological development and risk factors in SCA children.

What parents and caregivers can learn from this study:
- children with SCT can benefit from guiding their attention to relevant information on emotions: ‘look, your friend is happy, he smiles’
- mirroring their emotional response can help: copying their expression and saying ‘I see you are afraid of ...’
- It is not always visible when your child needs more help in a difficult situation. Even when you do not see any signs of emotions, the child might need you. You can help by saying 'It is okay when you are scared of the dog' or 'Maybe you are upset because Nana isn't coming for your birthday'.
- SCA children may need a longer recovery time after a challenging situation and this need is not always visible on their faces