



RESEARCH SUMMARY

Social Behaviour in Rubinstein-Taybi Syndrome



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Why study social behaviour in RTS?

You may remember that back in 2008 the Cerebra team attended an RTS weekend meeting in Weymouth. The research team asked parents whether there were any behaviours that they were particularly concerned about in the people they cared for. A number of parents spoke about 'repetitive behaviour' such as repetitive questioning and a strong desire to adhere to particular routines. Another thing parents spoke about was social behaviour. Parents explained that although the people they cared for were 'very sociable and loving', they had concerns that they were 'overfriendly', and may be 'vulnerable' with no 'stranger danger' awareness. Laurie Powis was recruited to conduct research that would gather more information about these behaviours and to explore the possible causes.

What did the research involve?

Laurie conducted two main studies. The first was a questionnaire study that aimed to confirm the descriptions you had given us. Although you had told us about repetitive behaviour and social behaviour, very little research had been conducted before, so we needed to do research that systematically showed that these behaviours were present in RTS. In the second study Laurie looked more specifically at a brain process called 'Theory of Mind'. Theory of Mind is known to be very important for effective social interaction and communication, so we wanted to find out how this brain process related to social behaviour in RTS. To keep things nice and clear, the report will go through the findings of each study separately.

Special points of interest:

- Parents have described the presence of 'repetitive behaviours' in the people they care for.
- Parents have also expressed concern about the people they care for being 'overfriendly' and lacking 'stranger danger' awareness.
- There is very little research around that documents these characteristics in RTS.
- There is evidence to suggest that 'Theory of Mind' is very important for effective social interaction and communication. This means it may play an important role in the social behavior in RTS



"...She's so fun, loving and caring...but she would just go off with anyone...she has no concept of stranger danger..."

Parent talking about her daughter.

Nathan completing the Appearance-Reality task.

Study 1: Questionnaire study.



Those of you who completed the questionnaire study will know that there were lots and lots of questionnaires that asked about different behaviours. Laurie looked specifically at the questionnaire that asked about Autism Spectrum Disorder (ASD) behaviours. The reason Laurie focused on this questionnaire is that the questions specifically ask about repetitive behaviour, social interaction, and social communication – the behaviours you spoke to us about. We also thought it would be helpful to know how many individuals with RTS reached criteria for ASD.

The Cerebra team would like to extend a huge thank you to all the families who took part in the project!



Special points of interest:

- 87 parents completed questionnaires.
- Data showed 64.9% of the RTS group met 'cut off' criteria for Autism Spectrum Disorder (ASD).
- However, closer examination showed that individuals with RTS do not show the 'same pattern' of behaviours as in ASD.
- Similar to ASD, individuals with RTS **do** show repetitive behaviours.
- However, differently to ASD, individuals with RTS show less impaired social interaction and social communication skills.

Study 1: Questionnaire findings

We collected data from 87 participants with RTS. Our findings showed that quite a high proportion (64.9%) of participants met the 'cut off' score for ASD. The prevalence of ASD in RTS has not been studied before so this is an important new finding. However, when we looked at the data more closely we realised that it wasn't quite this simple - there were some important differences between ASD and RTS.

We compared the RTS group to a 'just ASD' group and found that although many individuals with RTS reached the 'cut off' for ASD, they did not show the same level of difficulty across **all** the areas that individuals with ASD do. Individuals with ASD show repetitive behaviour and impairments in social interaction and social communication. Our results showed that similar to ASD, individuals with RTS showed high levels of repetitive behaviour. However, we found that individuals with RTS were much less impaired than the ASD group in the areas of social interaction and social communication. These findings therefore confirmed what you told us about in Weymouth – individuals with RTS show ASD 'like' repetitive behaviours, but more different, more 'sociable' behaviour.

Study 2: Theory of Mind

Although we found that individuals with RTS did not show the same kind of social impairments as seen in ASD, we knew that you had concerns that the people you care for seemed to lack ‘stranger danger awareness’ and so might be socially vulnerable. Because of this Laurie carried out a second study to look at a specific brain process known as ‘Theory of Mind’.

What is Theory of Mind?

Theory of Mind (ToM) is the ability to reason about other people’s thoughts and feelings. We use this ability every-day without even realising. It helps us explain, predict, and make sense of other’s behaviour. Without this ability, social interaction can be tricky and confusing. Research has shown that ToM plays a fundamental role in the social behaviour of many individuals with social difficulties and therefore we predicted that it might also be important in RTS.

How was Theory of Mind assessed?

ToM is something that develops progressively through childhood. Children begin life with very limited abilities and then get better and more skilled as they get older. To test the development of this skill in people with RTS Laurie put together a number of tests that formed a developmental scale (Figure 1). Tests at the beginning of the scale tapped early ToM abilities: the ability to help in simple scenarios; to play cooperatively; and to understand the communicative intent of point and gaze gestures. An example task is shown in Figure 2.



Figure 1: Developmental scale of ToM tasks

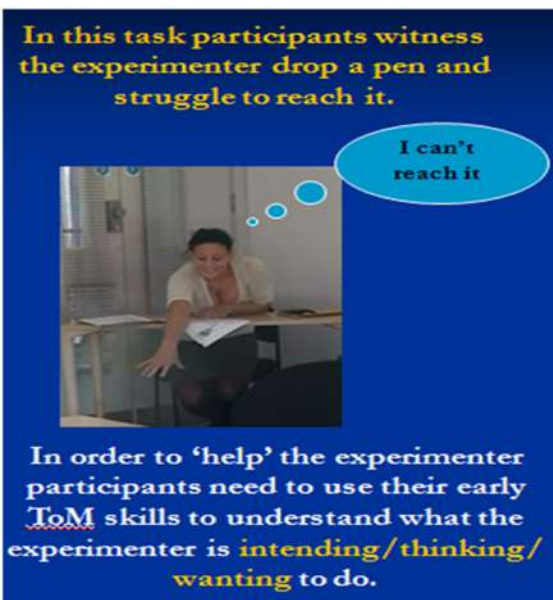


Figure 2 'Early' ToM task (Helping)

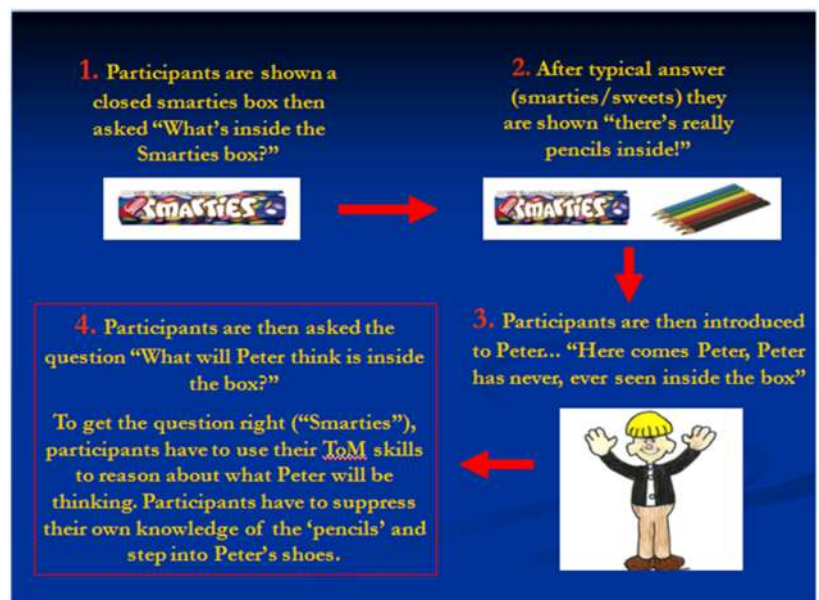


Figure 3 'Later' ToM task (False Belief)

Tests at the latter end of the scale tapped harder skills: the ability to judge that someone may think something different to your own thoughts; and the ability to judge that someone may try and hide what they are thinking and feeling. An example task is shown in Figure 3.

In typical development children pass these tasks in order and by specific ages. Laurie wanted to see whether individuals with RTS showed this same developmental progression and whether they showed any difficulties or delays in their ToM.

Study 2: Findings

We collected data from 32 participants with RTS. When we compared the development of ToM in RTS to typically developing individuals, findings showed that individuals with RTS showed a different developmental progression – with a unique profile of strengths and weaknesses.

1. Early abilities

When we looked at tasks at the beginning of the scale (those assessing early ToM abilities) we found that for most of these tasks, individuals with RTS were performing very well. Virtually all individuals with RTS passed the tasks that assessed ‘helping’, ‘cooperation’, and ‘understanding pointing gestures’. Some individuals even passed these tasks earlier than we would have expected given their mental age. These findings suggest that these early ToM skills could be ‘spared’ or even ‘advanced’ in RTS relative to mental age. However, our data showed that individuals with RTS found the ‘understanding gaze’ task more difficult than expected. Fewer individuals passed this task and so it might be that gaze understanding is a specific area of weakness in RTS. More research is needed to investigate why this might be.

2. Later abilities

When we looked at tasks at the latter end of the scale (those assessing later ToM abilities) it was clear that individuals with RTS found these tasks much more

difficult. Many individuals struggled to reason about other’s thoughts, feelings and beliefs. When we looked at the reasons for this, it seemed likely that these difficulties were underpinned by working memory problems. More specifically, to reason about other’s mental states, individuals have to remember critical facts about the story or event. Our findings suggest that individuals with RTS struggle to remember the information that they need to go on and develop later ToM abilities.



Henry completing the cooperation task.

What does all this mean?

The ability to understand mental states is crucial for social interaction as it helps us understand humour and sarcasm, to work out if/when a person may not want to talk to us, and judge whether a person might be trying to ‘trick’ or deceive us. The finding that a large majority of individuals with RTS struggled with their later ToM skills provides one possible explanation for the social difficulties that you described in Weymouth. The finding that these ToM difficulties may be due to memory impairments would suggest that intervention strategies may be best to focus on training working memory first. More research is needed to work out which intervention strategies would be most helpful for people with RTS.



If you would like any details of ongoing research at the Cerebra Centre for Neurodevelopmental Disorders or would like more details of anything reported in this article please contact:

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Thank you!