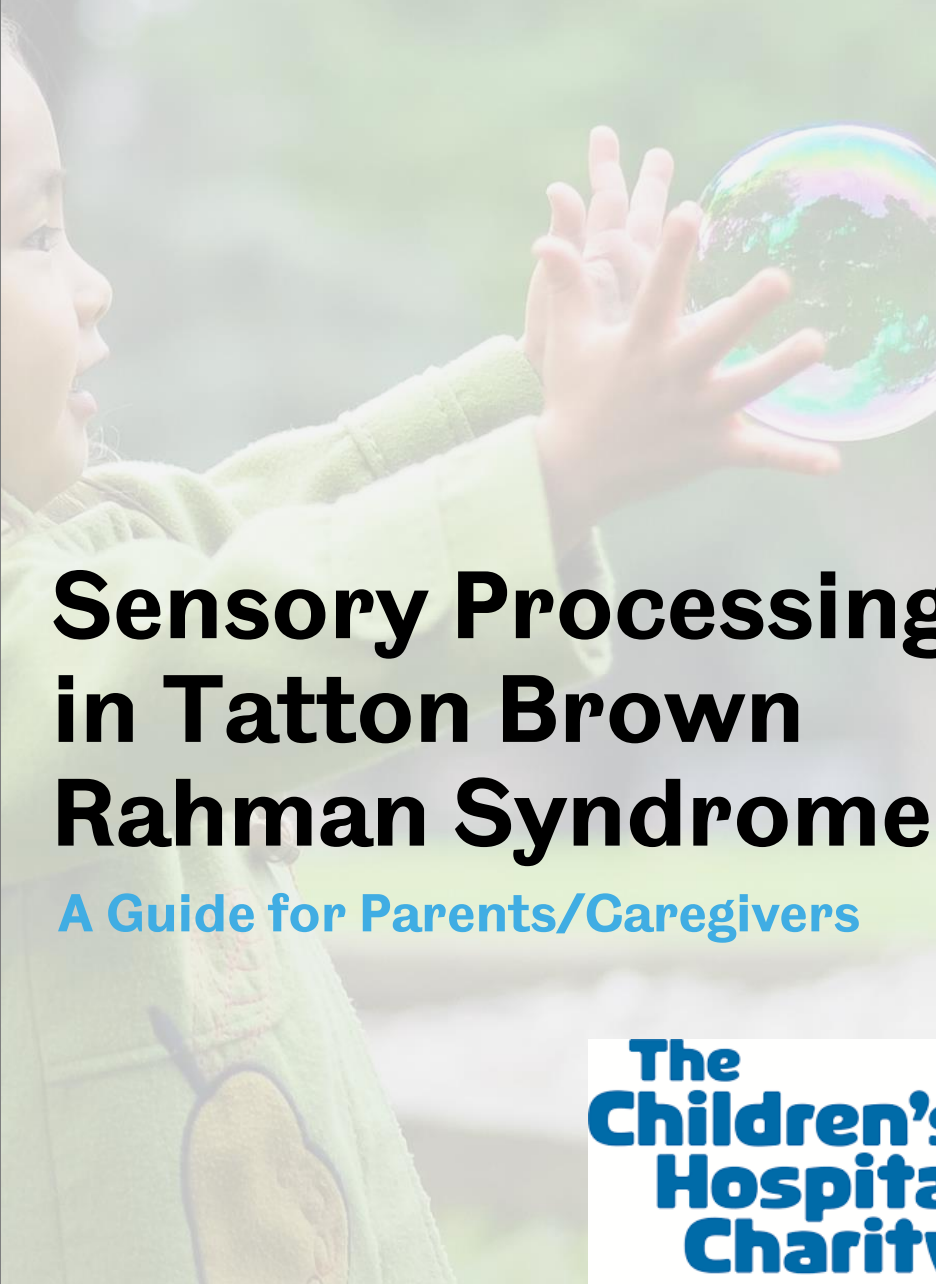




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A young child in a green sweater is shown in profile, reaching out with both hands towards a large, colorful, iridescent bubble. The background is a soft, out-of-focus green, suggesting an outdoor setting. The child's expression is one of curiosity and wonder.

Sensory Processing in Tatton Brown Rahman Syndrome

A Guide for Parents/Caregivers

**The
Children's
Hospital
Charity.**

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What is sensory processing?

Sensory processing refers to how our brain receives information from the senses.

There are eight key senses. Five of these are our **core senses** and three of these are our **internal senses**. These are:

Core senses:

- Touch
- Hearing
- Vision
- Taste
- Smell

Internal senses:

- Proprioception - *lets us know where our limbs are and how they are moving*
- Vestibular - *maintains our balance, posture and eye movement*
- Interoception – *detects information from our internal organs. For example, hunger, tiredness, and headaches*

Our brains often need to interpret and use information from multiple different senses at the same time.



Sensory processing is essential to every experience that we have. Sensory information is sent to our brains via our nerves, our brains register this input and process it in order to organise a response.

For example, you may stub your toe on the corner of your bed. The nerves in your toe, which detect pain, carry a signal to your brain which processes the experience as painful. This results in you grabbing your toe and avoiding the corner of the bed in the future.

What are sensory processing differences?

Some children process sensory information differently. This means they experience the world differently to others. Sensory processing differences can make everyday activities or certain environments challenging for a child. This can result in difficulties with learning, sleeping, eating, or challenging behaviour.

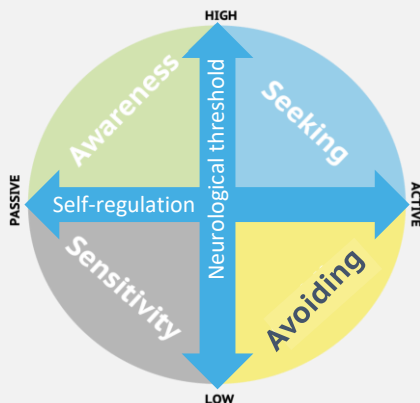
By understanding a child's sensory processing differences, changes can be made to their environment or routines to help reduce the negative impact of sensory processing problems.

For example, a child may show difficult behaviour that later is found to be caused by an uncomfortable label on their t-shirt or a tight seam on their socks.

Sensory Processing Model

Health professionals and researchers use models to explain complex processes that involve the body and/or the brain. In 1997 an Occupational Therapist, Dr. Winnie Dunn, proposed a model to explain children's processing of sensory information.

The model proposes that sensory processing in children can be explained by the relationship between two factors: "neurological threshold" (detection of sensory information) and "self-regulation" (response to sensory information). This interaction results in four categories (or "quadrants") describing different patterns of sensory processing:



Awareness (Registration) – children who miss or seem to have low awareness of sensory information.

Examples:

- Needs help to find objects obvious to others
- Doesn't notice messy hands or injury
- Gets tired easily or is clumsy
- Is comfortable in different sensory environments

Seeking – children who are more interested in sensory information than others.

Examples:

- Touches items with interesting textures
- Puts objects in mouth
- Gets excited during movement activities
- Watches people as they move around the room

Sensitivity - children who detect more sensory information than others.

Examples:

- Is picky about food textures
- Becomes distressed during face washing
- Is easily distracted
- Notices things that other people miss

Avoiding - children who actively avoid or become overwhelmed by sensory input.

Examples:

- Likes "sameness" or is good at routines (provides predictable sensory experiences)
- Has emotional outburst when being touched
- Puts hands over ears in a noisy environment

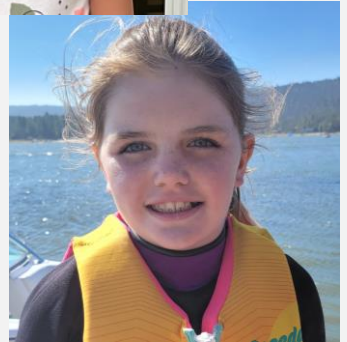
Sensory processing differences can be highly individual and varied. A child is unlikely to fall into just one of the sensory quadrants and different patterns can occur across, and within, sensory modalities. Some children may show sensory patterns from all four quadrants.

For example, a child may be sensitive to sounds but seek out activities involving touch.

Tatton Brown Rahman Syndrome

Tatton Brown Rahman Syndrome is a rare genetic disorder which is present from birth.

Children with Tatton Brown Rahman Syndrome usually present with overgrowth characteristics e.g., have large heads, are overweight and/or are tall for their age. Individuals with Tatton Brown Rahman Syndrome may have distinctive facial features such as low-set heavy horizontal eyebrows.



Individuals may also experience sensory processing differences, behavioural challenges, learning difficulties, or autism spectrum disorder symptoms. Tatton Brown Rahman Syndrome is caused by a range of different variants in the DNMT3A gene. The majority of cases are due spontaneous mutation.

The Sensory Profiles in Rare Genetic Syndromes Study

Research has shown many children with rare genetic syndromes (including TBRS) experience symptoms associated with autistic traits, which may increase the likelihood of sensory processing differences. So far, there has been no research that has investigated sensory processing in children with TBRS.

The aim of this study was to investigate the sensory profile of children aged 3-14 years old with TBRS.



A **sensory profile** is a measure of how a child processes sensory information and can help inform understanding of sensory differences

Parents and caregivers were asked to complete questionnaires about their child's sensory processing and other behaviours.

Children's sensory processing was measured according to six sensory modalities (hearing, visual, touch, movement, body position, and oral) and according to the four sensory quadrants (awareness, seeking, sensitivity, avoiding). The findings were compared to a population of typically developing children aged 3-14 years old.

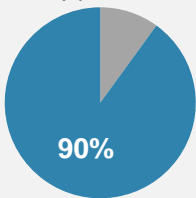
Who took part?

Twenty parents/caregivers of children with TBRS aged 3-14 years old took part. There were 13 male and 7 female children. Participants took part from the UK, USA, Australia, German, and Sweden.

How common are sensory differences in TBRs?

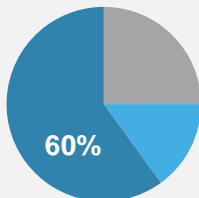
Research Findings

Body position



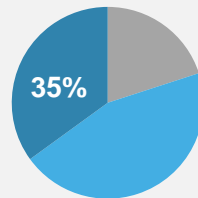
90% of children with TBRs were **much more** likely to process **body position** information differently compared to the majority of children of a similar age (e.g., get tired easily or walk as if feet are heavy)

Touch



60% were **much more** likely to process **touch** information differently (e.g., more sensitive or less sensitive to touch than others)

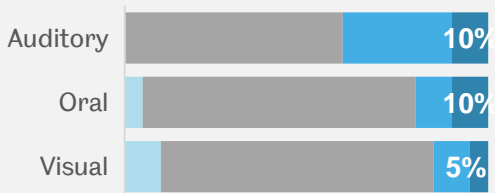
Movement



35% were **much more** likely to process **movement** information differently (e.g., constantly on the go or lose their balance easily)

■ Much less than the majority of others
 ■ Less than the majority of others
 ■ Just like the majority of others
 ■ More than the majority of others
 ■ Much more than the majority of others

Differences were also seen in the processing of oral and auditory information.



Low Awareness (Registration)



85% were much more likely to **miss sensory cues** compared to others

Sensitive



15% were much more **sensitive** to sensory cues

Seeking



20% were much more **interested** in sensory experiences

Avoiding



15% were much more likely to **avoid** sensory experiences

■ Much less than the majority of others
 ■ Less than the majority of others
 ■ Just like the majority of others
 ■ More than the majority of others
 ■ Much more than the majority of others

Quotes

"She is usually the one making the noise but doesn't like it when others talk too much. She also has a verbal stim when she's very happy or very irritated." - Mother of 8 year-old daughter with TBRS

"He broke his arm and badly burned it on another occasion and it didn't bother him. However he is hypersensitive on his hands and the back of his neck. If he gets a small nick he goes on about it for ages." - Mother of 14 year-old son with TBRS

"He is a sensory seeker. He craves compression, hugs, etc." - Mother of 7 year-old son with TBRS

"He is a toe walker to the point that he physically can't plant his feet on the ground due to the tightness in his muscles. This is why he is unbalanced." Mother of 7 year-old son with TBRS

"She gets upset when bumped into or brushed against. She can have a spiked face, hands, or clothes and is not worried about it. Ever..." - Mother of 8 year-old daughter with TBRS

"She grinds her jaw and spits often." - Mother of 5 year-old daughter with TBRS

"He has learnt to be careful with his footing as he knows he has poor balance...he takes it carefully or asks for assistance. He is also more careful about bumping into people now he is older and knows more social rules" Mother of 14 year-old son with TBRS

Useful Strategies

The below tables contain strategies that may support your child’s sensory processing differences. Use of strategies such as these may help to reduce sensory-related behavioural problems.

Not all of these strategies will be useful to your child. When selecting strategies **you should be guided by your child’s individual needs and think carefully about what is right for them** (e.g., your child may have low awareness of body position but is sensitive to touch). All children will be different and it is important not to force activities on your child. Advice and tailored support can be provided by a professional with training in ‘Sensory Integration’.

Seeking	More than others	<ul style="list-style-type: none"> • Incorporate novelty into child's routine • Choose activities that incorporate touch with others (e.g., dancing, massage) or with the environment (e.g., gardening, cooking) • Select clothes, furniture, or utensils with a variety of textures • Help the child engage in physical activity before a task (e.g., handing out books in the classroom, setting the table before dinner) • Introduce new tastes and smells (e.g., scented soaps, spices) • Chew gum, or eat mints when feeling restless • Talk to the child about what they smell or see
	Less than others	<ul style="list-style-type: none"> • Send the child on errands throughout the home and classroom • Play in parks on different equipment
Avoiding	More than others	<ul style="list-style-type: none"> • Avoid high traffic areas, crowds, and/or busy times • Establish consistent routines that are comforting and supportive (e.g., a daily walk) • Limit large group exposure; find opportunities for small groups or one-on-one interaction • Provide quiet places for the child to have time alone • Explain personal distance needs to others • When choosing clothing, select fabrics that don’t irritate and styles that are not constricting (e.g., seamless socks) • Warm wipes or towels before using them
	Less than others	<ul style="list-style-type: none"> • Incorporate breaks when doing tasks • Plan home and school schedules ahead of time and talk about changes in advance • Develop routines for outings to familiar places

Sensitivity	More than others	<ul style="list-style-type: none"> • Incorporate breaks and time-outs • Help the child break down tasks into smaller parts • Go to places where the sensory environment is known/predictable (e.g., the park, museums) • Use deep pressure touch rather than light touch • Provide a buffer space for the child to play without getting easily bumped • Use lots of blankets or heavy comforters on the bed • Reduce the volume or number of auditory stimuli • Provide written instructions or pictures alongside verbal information • Simplify complex visual information or environments (e.g., organise drawers so the child can find things easily) • Serve foods with spaces between them • Remove all light sources at bedtime • Introduce new foods and smells gradually
	Less than others	<ul style="list-style-type: none"> • Talk to the child about sensory features in daily activities (e.g., listen to the birds) • Vary routines
Low Awareness	More than others	<ul style="list-style-type: none"> • Visit places that offer a variety of sensory experiences (e.g., parks, interactive exhibits) • Vary bathing or dressing routines (e.g., try a shower instead of a bath) • Use heavy blankets to sleep • Provide opportunities to carry/ move heavy objects (e.g., carry groceries) • Add texture to objects to help with detection (e.g., writing materials for school, switches on toys) • Teach the child to use visual cues to notice when people or objects touch them (e.g., look to see if hands are messy) • Set water heaters at a lower temperature to prevent burns • Use extra care when drinking hot liquids and eating hot food • Use visual cues (e.g., watch where you are going) to support movement activities • Clear pathways of objects that could be tripped over • Have the child repeat or explain information back to you to make sure they have processed what was said • Make visual cues clearer (e.g., underline, bold, highlight)
	Less than others	<ul style="list-style-type: none"> • Identify familiar routines to return to when the child seems overwhelmed • Provide familiarity in settings, people, and experiences

Getting Support

Where to seek support for sensory processing in TBRS:

- TBRS social media groups (e.g., Facebook support groups)
- Tatton Brown Rahman Syndrome Community <https://tbrsyndrome.org>
- The Child Growth Foundation www.childgrowthfoundation.org
- Visit your child's GP
- Discuss your child's sensory processing differences with other professionals (e.g., Occupational Therapist with postgraduate training in Sensory Processing)

Further Reading

Books

- Can I Tell You about Sensory Processing Difficulties? A Guide for Friends, Family and Professionals – by Sue Allen
- The Out-of-Sync Child – by Carol Stock Kranowitz
- Living Sensationally – by Winnie Dunn

K. Tatton-Brown et al., "The Tatton-Brown Rahman Syndrome: A clinical study of 55 individuals with de novo constitutive DNMT3A variants," Wellcome Open Research, 3:46, 2018. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5964628/>

Dunn, W. "Supporting Children to Participate Successfully in Everyday Life by Using Sensory Processing Knowledge," *Infants & Young Children* 20(2) 84-101 (2007). <https://doi.org/10.1097/01.IYC.0000264477.05076.5d>

National Autistic Society. "Sensory Differences - a Guide for All Audiences." *Autism.org.uk*, 2021, <https://www.autism.org.uk/advice-and-guidance/topics/sensory-differences/sensory-differences/all-audiences>

At Home Sensory Diary

Use this page to note down your child's sensory behaviours.

Sensations your child...	
Tends to avoid or is overwhelmed by	
Is interested in	
Tends to notice easily or is sensitive to	
Tends to miss or is unaware of	

About The Authors

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Poppy is a Student Researcher at the University of Sheffield working in collaboration with Harriet Smith and Dr Megan Freeth. Poppy created this booklet as part of a funded research placement provided by the University. Poppy has an aspiration to become a Clinical Psychologist in the future and is beginning to gain experience in specific interests she is passionate about; one being rare genetic syndromes.

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Harriet Smith is a Research Associate at the University of Sheffield and member of the Sheffield Autism Research Lab. She is lead researcher on the "Sensory Profiles in Rare Genetic Syndromes" project. Her research experience includes projects in genetic syndromes, autism spectrum conditions, tinnitus, and hearing health. Harriet is also in the final stages of her PhD at the University of Nottingham which investigated the impact of tinnitus in children and has developed a child-specific questionnaire measure of tinnitus.

Dr Megan Freeth

Dr Megan Freeth is Principal Investigator on the project "Sensory profiles in genetic syndromes". She is Senior Lecturer in Psychology at the University of Sheffield and Director of the Sheffield Autism Research Lab (ShARL). The goal of ShARL is to have a positive impact on the lives of individuals diagnosed with autism spectrum conditions (ASC) and genetic syndromes via advancements in research. The scope of ShARL's research is broad and includes neuroimaging studies aimed at gaining insight into neural and cognitive divergence and applied studies aimed at improving understanding of the lived experience of ASC and genetic syndromes

Acknowledgments

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