

Sotos Syndrome: An Educational Guide

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What is Sotos Syndrome?

Sotos syndrome is an overgrowth disorder which is present from birth. Children with Sotos syndrome are usually tall for their age and have large heads. Increased height often becomes less apparent after puberty but adults will usually still have large heads. Individuals with Sotos syndrome have characteristic facial features which include a prominent forehead, downslanting eyes and a prominent chin. Medical problems that are common in Sotos syndrome are neonatal jaundice, difficulty with feeding, heart and kidney problems, seizures and scoliosis. The severity of these issues is variable and not all individuals with Sotos syndrome will be affected.



What is Sotos Syndrome?

Sotos syndrome is caused by abnormality of a single gene (NSD1) and in most cases, the abnormality occurs spontaneously. The syndrome is not usually inherited from a parent. However, if an individual with Sotos syndrome has a child, the child will have a 50% chance of inheriting the syndrome.



Intellectual Ability

The majority of individuals with Sotos syndrome have mild – moderate or borderline intellectual disability. This means that, in general, children will have delayed development and will often be behind their peers in terms of learning. However, there is a broad spectrum of ability associated with Sotos syndrome. A few individuals will have average intellectual ability whilst some may have more severe intellectual disability.

For children with Sotos syndrome, approximately 50% attend special educational needs schools and the remaining 50% attend mainstream schools but often require additional support. Determining the appropriate school for a child with Sotos syndrome will often depend on the provisions available within each local area.

Learning Profile

Broadly, cognition refers to the mental processes used to acquire knowledge and interpret the environment. Cognitive abilities include skills such as language, memory, attention and reasoning.

Individuals with Sotos syndrome have a consistent profile of relative strengths and difficulties for different cognitive abilities. This broad profile is characterised by relative strength in verbal skills and relative difficulty with non-verbal reasoning skills. Although the majority of individuals with Sotos syndrome will have difficulty with both of these skills when compared with their peers, this profile indicates that for individuals with Sotos syndrome, their verbal skills are much better than their non-verbal reasoning skills.

Learning Profile

As individuals with Sotos syndrome have a relative strength in verbal skills, this means that their understanding of language and their vocabulary is generally quite good. However, this may not always be apparent during conversations and social interactions as they often struggle to communicate appropriately.

Non-verbal reasoning skills are usually an area of difficulty for individuals with Sotos syndrome and this means that they struggle with logic and problem-solving. In practical terms, they will often find it difficult to integrate and process lots of information and particularly struggle when in a busy environment.

Memory

In terms of more specific cognitive skills, individuals with Sotos syndrome tend to have a relative strength in visuospatial memory. This means that they tend to process and remember information well if the information is presented visually, rather than just listening to information or instructions. Therefore, using pictures, diagrams and demonstrations will be particularly beneficial for learning.



Memory

Individuals with Sotos syndrome often have good attention to detail. So, for example, when entering a room, they may notice if an ornament has been moved. Once again, this means that they are very good at remembering what they have seen.



Numeracy

The majority of individuals with Sotos syndrome have difficulty with numeracy and struggle with simple arithmetic and understanding relationships between numbers. Children with Sotos syndrome will therefore require lots of support with numeracy skills. As they tend to learn information well if it is presented visually, the use of physical aids and demonstrations may be helpful when teaching mathematical concepts.

In practical terms, individuals with Sotos syndrome often struggle with daily skills requiring numeracy, such as dealing with money and telling the time.

Language and Communication

Although individuals with Sotos syndrome tend to have a good understanding of language and reasonable vocabulary, they often struggle with use of language. This means that they may have difficulty with structuring their language (e.g. getting words in the correct order) and with using language appropriately (e.g. using appropriate language in different contexts).

As developmental delay is common in Sotos syndrome, language development is likely to be delayed for young children with Sotos syndrome. However, the majority of children with Sotos syndrome will develop language. Speech and language therapy may be helpful for language development, as well as support with communication skills.

Social Skills

Individuals with Sotos syndrome are often motivated to engage in social interactions but may have difficulty with understanding social norms and how to behave appropriately within a social situation. Individuals may also struggle to understand what other people are thinking or feeling. For example, they may not be able to recognise when someone is not particularly interested in the conversation.

Individuals with Sotos syndrome like repetition and may enjoy talking about the same thing over and over. They may also repeatedly ask the same questions. Difficulty relating to peers is common for children with Sotos syndrome and they often prefer the company of adults.

Behaviour

A number of behavioural issues have been associated with Sotos syndrome. These include autism, ADHD, anxiety and aggression/tantrums.

For most of these issues, it is not clear how many people with Sotos syndrome display these behaviours. However, it seems that the majority of individuals with Sotos syndrome display behavioural characteristics that are typically associated with autism. Specifically, these include difficulty with social skills and social interaction, as well as restricted interests and repetitive behaviours.

In some cases, a behavioural assessment may be appropriate in order to establish whether an individual has significant difficulty with any of these behavioural issues and whether a clinical diagnosis would be beneficial.

Strategies

- Verbal instructions and demonstrations should be given sequentially, not at the same time. This will reduce the amount of information that the individual has to focus on and process.
- Chunking - use simple, clear, step-by-step instructions to break down tasks into manageable chunks.
- A consistent format should be used when presenting information (e.g. using the same picture when referring to an object).
- Using visual/spatial learning strategies (e.g. objects, demonstrations, videos and pictures) will assist learning.
- Repetition is useful for reinforcement and can help to alleviate anxiety.
- A visual daily planner can be used to visualise daily routines and to provide structure.
- Technology (such as iPads and tablets) can be a useful tool to support learning as information is presented visually.

Further Reading

Overview of Sotos syndrome

<https://www.ncbi.nlm.nih.gov/books/NBK1479/>

Cognition and behaviour in Sotos syndrome

Lane, C., Milne, E., & Freeth, M. (2016). Cognition and behaviour in Sotos syndrome: A systematic review. *Plos One*, 11(2).

<http://journals.plos.org/plosone/article?id=10.1371/journal.pone.0149189>

Sotos syndrome and ASD

Lane, C., Milne, E., & Freeth, M. (2017). Characteristics of autism spectrum disorder in Sotos syndrome. *Journal of Autism and Developmental Disorders*, 47(1), 135 - 143.

<https://link.springer.com/article/10.1007/s10803-016-2941-z>

Further Reading

Cognitive profile of Sotos syndrome

Lane, C., Milne, E., & Freeth, M. (2018). The cognitive profile of Sotos syndrome. *Journal of Neuropsychology*.

<http://onlinelibrary.wiley.com/doi/10.1111/jnp.12146/full>

Child Growth Foundation

<http://www.childgrowthfoundation.org/>

