Sotos Syndrome: A Summary of the Research

Introduction
Sotos syndrome was first recognised as a syndrome in 1964. Since this time, several studies have investigated cognitive and behavioural features of the syndrome. Cognition has been reported in terms of level of intellectual functioning, as well as more specific abilities such as language. A number of behavioural issues have been identified in some individuals with Sotos syndrome. These include autism spectrum disorder, anxiety and ADHD. The purpose of this review was to identify and summarise all of the research that has reported information about cognition and behaviour in individuals with Sotos. A summary of all of the existing research is helpful in providing an overview of what is currently known about these aspects of the syndrome.

Method
Academic databases were systematically searched to identify all of the research that has reported information about Sotos syndrome. The search yielded 917 results and these were checked in order to identify research that provided information about cognition or behaviour in Sotos syndrome. After screening all of these articles, 34 papers were considered relevant for the review. Below is a brief summary of each of the main findings from the review.

Intelligence Quotient (IQ)
Several studies (25) have measured IQ in individuals with Sotos syndrome. IQ scores are obtained by administering a series of tasks that measure cognitive abilities such as memory, language and reasoning. IQ scores are calculated by taking into account performance on each of the individual tasks. The scores therefore provide an indication of the general level of intellectual functioning of an individual. An IQ score of 100 is the average score for the general population. Scores in the range of 70 – 84 are considered to be in the borderline range and scores below 70 indicate that an individual has a learning disability. The existing research suggests that the majority of individuals with Sotos syndrome have a mild learning disability or are in the borderline range. However, scores that have been reported within these
studies have ranged from 21 – 113, indicating that intellectual ability in individuals with Sotos can range from severe learning disability to above average cognitive ability.

**Language**

Language abilities have been investigated in 13 studies. The findings from these studies suggest that language abilities are consistent with general level of intellectual functioning. This means that if a child with Sotos has a mild learning disability, their language skills will likely be at the same level as would be expected from any other child with a mild learning disability. Children with Sotos may display speech and language delays and experience greater difficulty with expressive, compared to receptive language. This means that they are likely to have a good understanding of language but have difficulty communicating and expressing ideas.

**Aggression and Tantrums**

Aggressive behaviour and tantrums have been reported in a small number of studies (6). All of the participants in these studies were children with Sotos syndrome. Although these issues have been reported in several children with Sotos, it is not clear whether these issues are specifically linked to Sotos. One suggestion is that children with Sotos may become frustrated as they are often larger than their peers and therefore mistaken as being older or more able than their actual developmental level.

**Autistic Features**

Four studies have reported individuals with Sotos syndrome who also have a diagnosis of autism spectrum disorder (ASD). This suggests that there may be an association between Sotos and ASD but this has not been explored in detail.

**Attention Deficit Hyperactivity Disorder (ADHD)**

ADHD may be another common behavioural issue associated with Sotos syndrome. Eight studies have investigated ADHD in Sotos syndrome and several individuals were reported to meet criteria for a diagnosis of ADHD. In addition, a small number of individuals were described as being inattentive, hyperactive or demonstrating a lack of inhibition. These are all symptoms that are commonly associated with ADHD.
Anxiety

Two studies have investigated anxiety in individuals with Sotos syndrome. The existing research suggests that anxiety may be a common issue associated with Sotos syndrome. Specifically, it seems that individuals with Sotos are likely to display separation anxiety (anxiety provoked by separation from a parent or caregiver), be anxious in new situations and have some form of phobia.

Conclusion

To date, a total of 34 studies have provided insight into the cognitive and behavioural features of Sotos syndrome. The common themes arising from the review are explained above. As much of the existing research is based on small samples of children with Sotos, it will be important for future research to investigate cognition and behaviour in larger groups of individuals with Sotos. This will be beneficial in identifying what to expect from a child with Sotos. In addition, it will be important to investigate cognition and behaviour in adults with Sotos as very little is currently known about cognitive abilities and behavioural issues in adults with Sotos.

For the full paper, please see:

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