Psychological Aspects of Shwachman Diamond Syndrome

A research project at GOSH from 2008-2009 aimed to identify and understand some of the psychological implications of this condition, in terms of its cognitive, behavioural and learning impact. As this research is now complete in the UK and has managed to include 89% of all the children and young people in the UK identified with SDS, we are now in a position to outline some of the factors that might be pertinent in an educational context.

A central point identified by this research is that each child with SDS is different and although the research identified certain psychological issues that appear to be associated with SDS, not all issues will be seen in each child. It is very important that this is borne in mind when considering the results of this research so as not to over-interpret the effects regarding each individual child. Additionally, this letter does not consider the medical and physical implications of SDS, hence questions regarding any of these issues would need to be directed to the child's medical team.

Educational Impact:

- Children with SDS have an overall IQ norm that is significantly lower than the norm of the general population.

However, as with the normal population, these scores are within a range that allows for IQ scores both significantly above the average, as well as below. The general trend of scores is *shifted* slightly further down than those of the normal population.

- Difficulties in academic subjects were noted within both literacy and numeracy areas.

Academic difficulties in literacy areas may be noted in most areas particularly in spelling, reading, listening and reading comprehension. Numeracy skills based on mathematical reasoning abilities were also found to be significantly lower than that of the normal population.

- Executive functioning was one of the more significant areas that showed a potential detrimental effect.

Executive functions are those skills that describe how we approach cognitive tasks, including domains such as working memory, planning/organisation, self-monitoring, emotional control and task initiation or ability to self-start. Hence, children that have difficulties in one or some of these or related areas may be described as having broader executive functioning difficulties.

Suggestions for intervention:

- Good communication and a close working relationship between the child's school and the child's parent/carers can really support a child with SDS within a school in terms of their educational development.

- A detailed individual profile of the child's cognitive abilities and area of need is essential to help the teacher and school provide an appropriate education package and ensure the child's future learning potential.

- Although no significant difference between an SDS child's numeracy and literacy abilities were identified, anecdotal data still suggests that children and their teachers/parents *feel* that they find literacy areas more challenging. Whatever the basis of this, the child's confidence in literacy tasks is likely to be affected and thus, affect their further attainment. Children with SDS therefore require confidence-building literacy tasks that aim to gradually stretch their ability without being too difficult. They may also benefit from additional individual or small group support regarding literacy tasks.

- Children with SDS appear to have more executive functioning deficits, therefore strategies that aim to support those areas would be positive, e.g., giving clear, limited instructions for each tasks that emphasise structure and support some limited, structured decision making; ensuring tasks are time-limited with the time increasing as a child's ability to concentrate and focus increases, keeping tasks within the general scope of a child's ability whilst still stretching them a little to reduce frustration; allowing different working styles to be used depending on how a child works best, e.g., instructions using visual or colour-based diagrams, or verbal, written etc. Routines can be very important and helpful; teachers can emphasise existing class routines by noting them and repeating this to the child. Using teaching cues can also help, both to highlight current issues of importance and to highlight any forthcoming changes to routine or task, e.g., noting key issues or important information "Now this is particularly important to remember…" or any upcoming changes to the current task "In 2 minutes we will finish this task".

  If you feel that a child with SDS presents with some particular executive functioning difficulties it may be helpful for them to receive an assessment of these by an educational or clinical psychologist in order to gauge the strength of any difficulties, the particular area/s affected and specific strategies that might help, as there are many strategies that can be integrated into a child's routine.

- Depending on the Inclusion and educational needs policies of the school and LEA, a referral by the school to an educational psychologist may be the most appropriate next stage for an SDS child presenting with cognitive or learning challenges. This would clarify the specific educational abilities and challenges of the individual child and assist in developing a learning strategy or plan for the child.

Behavioural Impact:

- Children with SDS may find the behavioural and learning demands of a classroom environment challenging to follow at times.

Children with SDS were found to score more significantly across the domains of Inattention, Hyperactivity and Aggression than either the normal population or a control population of children with another medical condition. Not all children with SDS achieved clinically significant scores in all or some of these domains but the proportion of those that did was greater than within a normal population. These findings suggest that some children with SDS can find the classroom environment and its behavioural demands quite challenging.

- It may be that with the support of an educational or clinical psychologist a clear, specific behavioural support programme can be developed to facilitate the child's development of more appropriate behaviours using strategies

appropriate to the setting, such as reward charts and highlighting appropriate behaviour from peer models.

- Similar strategies as might be used with other children who may find attentional demands more difficult, such as limiting the time of each individual task, changing the task slightly at frequent intervals to give the sense of a new task, and gradually increasing the time a child continues at one task using rewards and praise to enable them to gradually become more able to remain on-task for longer periods at a time.

- Where appropriate a school will also have to follow its own policies regarding inappropriate and/or aggressive behaviour but where possible support regarding this should be sought from a local clinical psychologist with the involvement of the family in order to try to develop an appropriate behavioural intervention programme aiming to reduce unwanted behaviours and encourage positive behaviours.

Social Impact:

- Children with SDS may find it more challenging to make and maintain close peer relationships.

It is unclear exactly why this might occur, and may be partially attributable to lower self-esteem and confidence. However, there seems to be greater difficulties in peer interactions and relations in the context of close, personal relationships. This may also relate to the difference between close friendships as opposed to more casual acquaintances, which has been suggested as being harder for children with SDS to develop, manage and maintain.

Additionally, children with SDS may be more likely to have a smaller stature. This can potentially lead to a variety of difficulties related to poorer confidence, reduced self-esteem and an increased possibility of being bullied or ostracised.

- It may therefore be useful to review periodically the social integration of a child with SDS and where possible, support their social development, such as with the use of group work in class, restructuring the composition of groups where necessary or sensitively observing to ensure the child is integrated into class and peer activities.

- Children with SDS may benefit from support around developing confidence generally, such as having a small position of responsibility in the class where appropriate, that can enable their confidence in social settings to develop.

- Children with SDS who are not integrating well or who start to develop difficulties that may be related to their condition, such as being picked on due to their height, social confidence or class absence may require a referral to a local therapy/support service via the school Inclusion Manager.

Physical Impact

- Children with SDS may be more prone to additional fluctuating health issues and complications

As well as the core, chronic physical issues that may affect children with SDS, they are also more likely to be affected by acute health difficulties, such as infections, that can impact on their attendance in class and long-term educational attainment. They are also more likely to have periodic medical appointments which may require absence from school and these can occur more or less frequently at different times.

- Absences from class can affect a child's educational and social development over time. It is therefore helpful if a child is not penalised for such absence, and that the teacher can provide 'catch-up' work, either to be done at home with the co-operation of parents or in class.

- If a child has had a prolonged absence due to an illness or series of medical appointments, their social skills and confidence may be affected and they may need support in re-integrating into their social group, such as with structured classroom activities and temporary observation.

The information that has formed the basis of this letter is research carried out within Great Ormond Street Hospital and previous research carried out in Canada by Kerr et al (2010). What did become apparent through this research was the variation in abilities, strengths and areas of challenge that can occur for children and young people with SDS. This needs to be considered when trying to support the individual. Furthermore, each school and teacher is different and many of the strategies and suggestions above may be a normal part of the school's routine. Additionally, these findings and strategies are themselves quite general and may need adaptation depending on the age and ability of the child, the type of education environment and the potential provision of the environment and LEA. For the most effective support and intervention an individual assessment of the child with a clinical or educational psychologist would be most appropriate, with further liaison with the school regarding the child's specific needs.

We hope that this letter and its information regarding educational issues and SDS, is of use. Please feel welcome to contact the team at Great Ormond Street Hospital in London if you require any further information or assistance.