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**INVESTIGATION OF ADAPTIVE AND  
MALADAPTIVE BEHAVIOUR IN PEOPLE  
WITH WOLF HIRSCHHORN SYNDROME**

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**BACKGROUND** Wolf Hirschhorn syndrome (WHS) is caused by a deletion to chromosome 4p16. A number of studies have described the physical characteristics associated with the syndrome; however, with only a handful of exceptions, the behavioural profile of WHS has to date escaped systematic attention. The current study aimed to provide an overview of the behavioural profile associated with WHS.

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**AIMS AND OBJECTIVES** The project aimed to explore four areas:

- ◆ The adaptive behaviour profiles of people with WHS;
- ◆ The prevalence of autistic characteristics in people with WHS;
- ◆ The extent to which people with WHS display challenging behaviour and the type, severity and context of any behaviours displayed;
- ◆ Caregivers' experiences of support from local services.

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**METHOD** Information was collected by survey and interview on 49 people with a reported diagnosis of WHS (aged 2-37 years). Interviews were conducted over the telephone with parents/caregivers and included questions examining: the form, severity and communicative function of any challenging behaviours displayed by the person with WHS; social impairment (i.e. symptoms of autistic spectrum disorder (ASD)); and levels of adaptive behaviour across communication, daily living skills, socialization and motor skill domains. Questions were also asked regarding family experiences of the support provided by local services.

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**OUTCOMES**

**Adaptive behaviour**

- People with WHS, generally, had severe deficits in adaptive behaviour. The average developmental age equivalent was just over 2 years and the majority of the sample scored in the profoundly intellectually disabled range;
- Deficits in adaptive behaviour were, on average, more severe than those of comparison groups consisting of people with Fragile X Syndrome, Smith-Magenis Syndrome and for people with intellectual disabilities of a mixed etiology;
- A minority of the sample presented much less severe deficits in adaptive behaviour more consistent with a moderate or mild intellectual disability;

- The extent of variability in adaptive behaviour was greater in the sample of people with WHS than in the comparison groups. That is, in a number of areas the WHS group included both the least able and the most able of all those for whom scores were available;
- Significant within-group differences were found in the profile of scores for the WHS group. Scores on the socialization subscale were significantly higher than other adaptive behaviour domains.

#### **Social impairment**

- Detecting the presence of ASD in people with WHS is more difficult with respect to the more disabled individuals. At a minimum, it appeared that close to a third of participants met criteria for possible ASD including a smaller number meeting the criteria for Autism itself.

#### **Challenging behaviour**

- On average, people with WHS were reported to display less, and less serious, challenging behaviour than people with Prader-Willi, Smith-Magenis and Cri du Chat Syndromes;
- There was considerable variability in scores relating to challenging behaviour. Almost a third sometimes displayed aggressive behaviour and over a fifth sometimes behaved destructively;
- Nearly half of the sample displayed at least 1 form of self-injury (SIB). The most common topographies were teeth grinding, self-biting and head banging;
- In relation to behavioural function, significant within-group differences were found for all three forms of challenging behaviour. It appeared that people with WHS presented with relatively high levels of attention-maintained challenging behaviours and relatively low levels of challenging behaviour maintained by other functions such as escaping from demands or gaining access to preferred objects or activities;
- Physical discomfort/pain may be a relatively frequent contributing factor to the self-injurious and aggressive behaviour of people with WHS;
- Overall, challenging behaviour was more likely to be reported in people with WHS who had lower levels of adaptive behaviour and higher levels of social impairment;
- Those displaying SIB were substantially more disabled with poorer daily living, socialization and motor skills and higher levels of social impairment than those who did not display SIB.

#### **Family support**

- The most commonly provided form of support was 'therapy' (e.g., physiotherapy, occupational therapy). The least common type of support came from psychologists;
- The most 'helpful' source of support was from therapists, the least helpful was psychology;
- Families reported relatively high levels of satisfaction with all kinds of support other than psychological input though a number of families reported having had to fight to obtain support.

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#### **IMPLICATIONS**

- Very little is known about the developmental trajectory of people with WHS. The current study could only provide a snapshot of behavioural characteristics at one point in time. It would be very useful to follow up the participants looking at changes over the course of time. Such follow-ups would focus on both development during childhood and change during adulthood (e.g., the extent to

which anecdotal reports of “premature aging” are supported). A longitudinal programme of research would also help to establish a large, well-described sample from which smaller samples could be drawn for the more detailed investigation of particular issues;

- It is clear from the current study that a significant number of people with WHS present challenging behaviours, perhaps especially of self-injurious and stereotypic kinds. More research is needed into the severity and impact of the behaviours they present and the causes of these behaviours. It might be particularly useful to investigate the possible relationship between challenging behaviour and physical health issues;
- Previous research has suggested that some of the characteristics of WHS are related to the size of the chromosomal deletion. It would clearly be useful for future research to consider this issue in respect of adaptive and maladaptive behaviour;
- The relatively high prevalence of ASD found in the sample suggests that it would be useful to investigate this with more detailed assessment measures.
- The measures used in the current study were not the most sensitive to the severity and impact of challenging behaviours presented. There was some suggestion, however, that self-injury was relatively frequent and something of a “hidden” problem, not always identified immediately as “challenging”. When coupled with the apparently limited psychological input to families this carries the possible implication for some families that more professional input should be sought. Psychological or behaviour support services should, routinely, investigate the possibility of a link to physical health issues;
- Many families probably already appreciate their son/daughter’s relatively good social skills and liking for their and others’ attention. While deficits in other areas should not be ignored it is obviously also important to build on strengths. This would imply (amongst other things) using (e.g. as a reward) the liking for attention to aid in the development of other skills, encouraging participation in activities which allow the use of social skills and encouraging the greatest development possible in the area of social skills;
- It is often forgotten that different disorders can co-exist and the “problems” an individual has are attributed to his or her “primary” disorder. Should families of someone with WHS suspect that their family member may also have Autism or a related disorder they should not hesitate from asking for a diagnostic evaluation.

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