Research in Developmental Disabilities 34 (2013) 4355–4365

Contents lists available at ScienceDirect

Research in Developmental Disabilities

The nature of social preference and interactions in Smith–Magenis syndrome

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ARTICLE INFO

Article history:
Received 31 July 2013
Accepted 9 September 2013
Available online 9 October 2013

Keywords:
Smith–Magenis syndrome
Social behaviour
Attention-seeking
Down syndrome
Natural observation

ABSTRACT

This natural observation study was designed to evaluate hypothesized elevated 'attention-seeking' and preference for adult attention in Smith–Magenis syndrome. Ten children with Smith–Magenis syndrome were observed across one school day, together with an age matched sample of 10 children with Down syndrome. Levels of attention given to, and vigilance for, adults and peers were recorded and compared. Sequences of behaviour were analyzed to evaluate the temporal relationships between giving and receiving attention during adult-child interactions. Compared to children with Down syndrome, children with Smith–Magenis syndrome gave preferential attention to adults and looked towards adults significantly more than they looked towards peers. Sequential analyses revealed that while children with Smith–Magenis syndrome did not initiate interactions with adults more than children with Down syndrome did, reciprocity between child and adult social behaviours in Smith–Magenis syndrome within interactions was compromised. This less synchronous sequence of child and adult interactions in Smith–Magenis syndrome may be the result of children with Smith–Magenis syndrome attempting to initiate interaction at times when it is unavailable. The marked preference for interacting with adults over peers in Smith–Magenis syndrome indicates atypicality of social interaction in this syndrome.

1. Introduction

Smith–Magenis syndrome (SMS) is a rare genetic syndrome with a prevalence rate of approximately 1/25,000 to 1/15,000 births (Greenberg et al., 1991; Laje et al., 2010). It is caused by a deletion on chromosome 17 p11.2 (Greenberg et al., 1991; Smith et al., 1986) or, more rarely, by a mutation of the retinoic acid-induced 1 (RAI1) gene located on this chromosome (Slager, Newton, Vlangos, Finucane, & Elsea, 2003). SMS is characterized by distinctive facial appearance, a range of health problems and increased likelihood of behavioural problems including sleep disturbance, stereotyped behaviours, challenging behaviour, impulsivity and attention-seeking (De Leersnyder et al., 2001; Dykens & Smith, 1998; Martin, Wolters, & Smith, 2006; Smith, Dykens, & Greenberg, 1998).

Sleep disorder and challenging behaviour have been the focus of behavioural phenotype research in SMS (e.g. De Leersnyder et al., 2001, 2003; Finucane, Dirrigl, & Simon 2001; Martin et al., 2006). Other areas, including attention-seeking and impulsivity, are less well researched despite their potentially critical role in influencing challenging behaviours (Oliver et al., 2013; Sloneem, Oliver, Udwin, & Woodcock, 2011; Taylor & Oliver, 2008). Research reporting high levels of attention-seeking behaviour has primarily employed caregiver report behavioural checklists, for example the Reiss Screen for Maladaptive Behavior and the Child Behavior Checklist (Dykens, Finucane, & Gayley, 1997; Dykens & Smith, 1998). While the

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http://dx.doi.org/10.1016/j.ridd.2013.09.014

findings of this research are striking (attention-seeking reported in over 80% of individuals), use of behavioural checklists limits both the scope of behaviours examined and detail. Other descriptions of attention-seeking come from anecdotal accounts. For example Haas-Givler (1994) describes individuals with SMS as attention-seeking at school and 'very adult-oriented' with little interest in interacting with peers. They are further described as demanding an 'inordinate amount of' and having a 'sometimes insatiable' need for individualized attention from adults, with aggression resulting if availability of attention is restricted. While this description gives valuable insight into the nature of attention-seeking, it is limited by its anecdotal nature.

High rates of attention-seeking behaviour are likely to have implications both for those with the syndrome and those who care for and work with them. In addition to being reported by caregivers to be problematic in and of itself, links have also been made between attention-seeking and challenging behaviour in SMS. In addition to Haas-Givler's description (1994), suggesting aggression results from lack of adult attention, Sarimski (2004) found aggressive behaviours were reported by caregivers to be motivated by desire for social attention. Two subsequent studies have carried out indirect assessments of functions of challenging behaviour using the Questions About Behavioral Function scale (Matson & Vollmer, 1995) and found that attention maintained challenging behaviour is a feature of SMS. Sloneem et al. (2011) found that for aggressive behaviour the attention sub-scale of the QABF was scored highest by caregivers (above escape, tangible, pain and discomfort and self-stimulation). Subsequent analyses revealed that this function was significantly more common than self-stimulation or pain and discomfort. Langthorne and McGill (2012) found that more children with SMS met the criteria for attention maintained challenging behaviour than children with Fragile X syndrome. Although the authors did not find a within groups difference in function, suggesting that gaining attention is one of a number of functions of challenging behaviour, this finding does indicate that attention is likely to be one function of the difficult behaviours shown by those with SMS. The only study employing direct observations of social behaviour in SMS also supports this association. Taylor and Oliver (2008) observed children at school and found that challenging behaviour was reliably preceded by reduced availability of adult attention. Whilst this study was an important step in examining attention-seeking behaviour in SMS, lack of a contrast group limits whether this behaviour can be considered to be related to SMS. Furthermore, the role of peers, a key aspect of school environments, was not examined.

In summary, problematic attention-seeking and difficult behaviour in conditions of reduced attention together with preference for adult attention, are purported to be characteristic of SMS. Alongside causal models of self-injury and aggression in SMS (Oliver et al., 2013; Sloneem et al., 2011; Taylor & Oliver, 2008) such findings indicate a need to further investigate this behaviour, using objective and reliable methods. While excessive social drive in genetic syndromes is studied infrequently (in contrast to widely studied social deficits, for example in Autism Spectrum Disorder and Fragile X syndrome, Carter, Davis, Klin, & Volkmar, 2005; Constantino et al., 2003; Cornish, Turk, & Levitas, 2007; Turk & Graham, 1997), there is an increasing body of research specifically examining strong social drive in genetic syndromes which may inform the study of attention-seeking in SMS.

A similar strong drive for attention is evident in Angleman syndrome (AS) caused by the loss of genetic information on the maternal chromosome 15 (Brown & Consedine, 2004; Oliver, Demetriades, & Hall, 2002; Oliver et al., 2007; Strachan et al., 2009). Those with this syndrome are described as highly sociable, actively seeking and maintaining adult interaction more successfully than those without the syndrome (Oliver et al., 2007). They also demonstrate a range of prosocial behaviours including laughing and smiling (Horsler & Oliver, 2006). Oliver et al. (2007) used natural observations of children with AS at school in order to examine social drive in AS, recording child and adult social behaviours and the amount of attention children received. Analyses indicated children with AS smiled more when receiving adult attention and initiated interaction more than a contrast group. Sequential analyses revealed that the child smiled first and adult attention, smiling and eye-contact were more likely to occur after a child with AS smiled than by chance, a pattern not found for the contrast group. This supported assertions that children with AS are more likely to initiate social interactions with 'prosocial' behaviours to solicit attentional resources from caregivers (Brown & Consedine, 2004). This methodology is applicable to the study of attention-seeking in SMS as it enables examination of sequences of behaviour and identification of who initiates interaction. In SMS, children reportedly *seek* attention, thus in sequential analyses it would be anticipated that they would initiate interactions more than a contrast group.

In the current study, natural observation methods will be used to examine behaviour of children with SMS at school. This setting was chosen as it is the context in which both adults and peers are present, it has social validity and there is competition for social resources. Levels of attention (verbal and non-verbal social initiations) and looking (which can be considered an index of allocation of attentional resources, e.g. Riby and Hancock (2009) report prolonged face looking in Williams syndrome suggesting "atypical allocation of attention" in this syndrome) directed towards adults and peers, can then be examined to evaluate preference for adult attention. Examination of sequences of behaviour will also be performed, using lag sequential analyses, to evaluate the nature of reciprocal interactions between adults and children and also the association between adult attention and child behaviours. Results will be contrasted to a Down syndrome (DS) sample, selected because of the similar profile of intellectual disability and expressive language deficits (Greenberg et al., 1996; Chapman, Seung, Schwartz, & Kay-Raining Bird, 1998).

The following hypotheses were tested:

(1) Social preference: Children with SMS will have greater preference than children with DS for interacting with and attending to adults versus peers. Differences between *child to adult attention* (verbal and non-verbal initiations from the