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

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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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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

child towards the adult) and *child to peer attention* (verbal and non-verbal initiations from the child towards the peer) and between *child to adult looking* and *child to peer looking*, will be greater in SMS than in DS.

(2) Sequences of adult–child interactions
   (i) Children with SMS will initiate and maintain interaction to a greater extent than those with DS. This will be reflected by different patterns of reciprocal social interaction such that *child to adult attention* is shown earlier in sequences of interaction, i.e. before *adult to child attention* (verbal and non-verbal initiations from the adult towards the child) occurs, and *child to adult attention* is shown for longer after *adult to child attention* has ceased. Additionally, sequence of interactions between adults and children will be examined to explore how adults behave prior to, during and after *child to adult attention* occurring.
   (ii) Attentional bias: Children with SMS will show greater vigilance for adult attention as evidenced by looking at adults earlier in an interaction (i.e. before *adult to child attention* occurs) than those with DS and will continue to look at the adult for longer after *adult to child attention* has occurred (thus maintaining interaction).

2. Methods

2.1. Participants

2.1.1. Recruitment

Expressions of interest in participation were gained from caregivers involved in an existing research project (citation withheld for blind review). Nineteen of 21 caregivers of children with SMS expressed interest, 11 of whom met the inclusion criteria of being school age (over four years old and under 16) and having completed the cognitive assessment in the previous study which enabled matching by mental age. Ten caregivers confirmed participation. Of the 17 caregivers of children with DS who met inclusion criteria and indicated that they would like to part in this study, caregivers were contacted in the order which reflected the best mental age matches for the SMS sample until 10 participants had been recruited. Following initial expressions of interest, consent for participation was obtained from children's caregivers. Children's schools were then contacted in order to gain their consent for researchers to visit.

2.1.2. Demographic information

Ten participants with SMS and 10 with DS participated. Demographic details are shown in Table 1. All participants with DS were reported to have confirmed diagnosis of trisomy 21, nine participants with SMS had chromosome 17p11.2 deletions, one had a gene RAI1 mutation.

Participants were comparable based on estimates of mental age (assessed using the Wechsler Abbreviated Scale of Intelligence, Wechsler, 1999, or the Mullen Scales of Early Learning, Mullen, 1995) and also current raw receptive language scores derived using the Vineland Adaptive Behavior Scales II: classroom edition (Sparrow, Cicchetti, & Balla, 2005). Raw scores were used as confirming that ability level was similar was of interest rather than ability normed to chronological age (the latter was well matched).

2.1.3. Type of school

Nine of 10 children with DS attended mainstream schools, the remaining child attended a special school. In the SMS sample eight children attended special schools, one was at a mainstream school and one had a mixed placement (special school in the morning, mainstream in the afternoon).

Table 1
Gender and mean chronological and mental age (standard deviation and range) and Vineland Adaptive Behavior Scale receptive language scores (standard deviation) in each group.

	SMS	DS
N	10	10
Gender		
Number of males	4	5
Age (Years)		
Mean (SD)	9.98 (3.11)	9.10 (2.99)
Range	5–15	4–13
Mental age[a] (Months)		
Mean (SD)	48.13 (21.91)	52.32 (20.00)
Receptive language scores[b]		
Mean (SD)	18.33 (5.59)	17 (6.08)

[a] Mental age estimates obtained from participation in previous study using either the *Wechsler Abbreviated Scale of Intelligence* or the *Mullen Scales of Early Learning* depending on ability.
[b] Vineland Adaptive Behavior Scale II: classroom edition raw scores.

2.2. Measures

2.2.1. Vineland Adaptive Behavior Scales II – Classroom Edition (VABS, Sparrow et al., 2005)

The Vineland Adaptive Behavior Scales II – Classroom Edition is an informant report rating measure completed by teachers. It assesses four domains of adaptive behaviours; communication, daily-living skills, socialization, and motor skills. Scores for each of the four domains can be combined to provide an Adaptive Behavior Composite score. The rating form is suitable for use with children aged over 3 years.

2.3. Procedure

2.3.1. Testing and ethical considerations

The study was reviewed and approved by the ethics committee of the University of Birmingham. Children were observed at school for the duration of one school day, typically from 9 am to 3 pm. The total observation period ranged from 3.5 to 5 h. Observations were carried out during classroom teaching (group and one-to-one sessions), outside classroom activities, transition periods and break times in 10 min blocks and stopped if there was potential for intrusion into children's privacy (e.g. during toileting). Teachers and support staff were asked to interact with children as they would typically. Researchers aimed to be as unobtrusive as possible.

Live coding was used and researchers coded behaviour on a ASUS EeePC 900 Netbook, using ObsWin32 software (Martin, Oliver, & Hall, 2001), which enables real time coding of frequency and duration of operationally defined behaviours and environmental events. Researchers also recorded the ratio of adults to children for each 10 min observation block.

2.3.2. Real time coding procedure

Participant responses included behaviours directed towards adults and peers including child speech (words and utterances, e.g. umm), non-verbal gestures (e.g. pointing, nodding) and child initiated touching. These behaviours were combined during analysis resulting in variables reflecting *child to adult attention* and *child to peer attention*. Additional child variables included child looking (*child to adult looking* and *child to peer looking*). Environmental events included adult behaviours towards the child including speech, non-verbal gestures and adult initiated touching. These were combined during analysis resulting in the variable of *adult to child attention*. Three settings were also recorded and are shown in Table 2.

2.4. Data analysis

An alpha level of $p \leq .05$ was used throughout.

2.4.1. Between-groups comparisons of attention and looking

Percentages of time that target behaviours and events occurred were calculated using ObsWin32 software. Difference scores were calculated for *child to adult attention* and *child to adult looking* versus *child to peer attention* and *child to peer looking* in order to obtain a score for attention and looking representing preference (higher scores denoting greater preference towards adults). Difference scores for attention and looking were then analyzed using 3×2 mixed ANCOVAs, with setting (one-to-one attention, shared attention and free play) and syndrome (SMS and DS) as independent variables and adult:child ratio as the covariate, to control for variability in these ratios.

2.4.2. Adult–child interactions

Data were analyzed to assess associations between child and adult behaviour, enabling examination of reciprocal interactions between adult and child, i.e. how likely it is for a child to be giving attention to an adult when the child is receiving attention from the adult, and vice versa, and how likely it is for a child to be looking at the adult when the adult gives them attention. Yule's *Q* values, which describe strength of association between a criterion and target event, were calculated using ObsWin32 (Martin et al., 2001). Yule's *Q* is analogous to a correlation coefficient, a value of 0 denotes no relationship between the criterion and target behaviours, +1 a strong positive relationship and −1 a strong negative relationship. Yule's *Q* values therefore represent the extent to which two variables (a criterion behaviour and a target behaviour) co-occur within a given time window (lag). All variables were lagged from presence of the criterion variable (where every interval that the criterion variable occurs is used to initiate lagging, Martin et al., 2001) except for looking which was lagged from onset of the target variable to identify the point at which it started.

Table 2
Social contexts recorded in natural observations.

Setting	Description
One-to-one attention	The child is receiving attention from an adult with no other child present
Shared attention	The child is receiving adult attention within a group setting where other children are present and also receiving attention
Free play	The child is not receiving direct attention from adult and is free to play without expectations from adults

Time based lag sequential analysis was used (Bakeman & Gottman, 1997), analysing sequential relationships for each participant by calculating the probability of an environmental event within a 20 s epoch for a behaviour for 60 s before and 60 s after each event of interest (lag −3 to +3 respectively). Lag 0 represents the point at which the criterion variable occurs. Analyses split sequences into three stages, *before* (−60 s to −20 s; lag −3 to −1), *during* (lag 0) and *after* (+20 s to +60 s; lag 1–3) the criterion variable occurrence.

Mean Yule's Q values for all participants were calculated for each 20 s epoch. Mixed 3 × 2 Analyses of Variance (ANOVAs) were then used to compare strength of association (denoted by mean Yule's Q values) between child and adult behaviour across syndrome groups and over time. The onset of attention allocation was defined as the criterion variable; one ANOVA was conducted for epochs before this criterion variable occurred (lag −3 to lag −1) and the second for after the criterion variable occurred (lag 1 to lag 3). Where assumptions of sphericity were violated Greenhouse–Geisser corrections were used. A *t*-test assessed whether there were significant differences between the two syndrome groups in the strength of association between child and adult behaviours at the point at which the criterion variable occurred (lag 0).

A significant main effect of lag in the ANOVAs would demonstrate an effect of temporal proximity to the point at which attention is given. A significant main effect of syndrome group in either ANOVA, or in the *t*-test, would indicate an overall difference, during the relevant time period, in how strongly child and adult behaviours are associated with one another. Finally, an interaction between syndrome group and lag in either of the ANOVAs would suggest that increased temporal proximity to the criterion variable has a different effect depending on the syndrome group. Broadly, this would reflect different patterns of social reciprocity between children and adults for the two syndromes groups such that there is an effect of syndrome on timing of social behaviours.

Analyses of the association between *adult to child attention* (as the criterion variable) and *child to adult attention* (as the target variable) will provide insight into whether children with SMS and DS differ in the timing of their initiation and maintenance of social interaction. Analyses of the time period prior to *adult to child attention* occurring will identify differences in the timings at which children with SMS and DS initiate directing their attention to the adult (before that adult gives them attention). Analysis of the point at which *adult to child attention* occurred will identify differences in the strength of association between these child and adult behaviours at the point at which the adult gives attention to the child. Analyses of the time period after *adult to child attention* occurs will highlight differences in how long *child to adult attention* continues after attention from the adult has ceased, providing insight into whether children with SMS and DS *maintain* social interaction to the same extent. Finally, analyses of the association between *child to adult attention* (as the criterion variable) and *adult to child attention* (as the target variable) will provide insight into adult behaviour directed to the child before, during and after the child directs attention to the adult.

Examination of patterns of association between *adult to child attention* (as the criterion variable) and *child to adult looking* (as the target variable) will identify whether children with SMS show an attention bias towards adults (denoted by looking behaviour), compared to those with DS. Analysis of the time period prior to *adult to child attention* occurring will identify differences in the timing at which children with SMS and DS start to look towards the adult, with early looking potentially reflecting greater vigilance for adult attention. Analysis of the point when *adult to child attention* is given will identify any differences in the strength of association between this attention being provided to the child and the child's looking towards the adult. Finally, analysis of the time period after *adult to child attention* occurred will demonstrate whether there are differences in how long children continue to look at the adult after *adult to child attention* occurs, with prolonged looking potentially serving to maintain interaction.

2.4.3. Inter-rater reliability

Inter-rater reliability was calculated for approximately 30% of observations (24 h in total). A second researcher was present for 6 of 20 visits and reliability observation periods were conducted live. Cohen's Kappa (Cohen, 1960) was calculated to control for influence of chance levels of agreement. Agreement was calculated using 10 s epochs for presence of the target variable. Mean reliability of each variable was over .6, thus was 'good' Fleiss (1981).

3. Results

3.1. Differences in environmental context

As noted more children with DS attended mainstream schools than those with SMS ($\chi^2(2) = 12.80$, $p < .001$), potentially undermining internal validity. Consequently, to ensure that the time the children spent in each social context within the school did not differ, this and the ratios of adults to children throughout the school day were compared across syndromes. Mean times spent in each social context are presented in Table 3.

There were no between-group differences in amount of time spent in one-to-one attention ($t(18) = -1.21$, $p = .24$), shared attention ($t(18) = -.18$, $p = .86$) and free-play ($t(18) = 1.22$, $p = .24$), suggesting groups were exposed to social contexts that were comparable on critical variables despite differences in schooling. Analyses of ratios of adults to children throughout the school day suggested significantly smaller overall ratio of adults to children for SMS (mean adult child ratio .54) compared to DS (mean adult child ratio .23) ($t(18) = 5.53$, $p = .002$). Adult:child ratio was therefore entered into comparisons of child attention and looking to adult and peers as a covariate in order to control for variability in these ratios.

Table 3
Percentage time spent by children with SMS and DS in one-to-one attention, shared attention and free-play social contexts.

	One-to-one attention	Shared attention	Free play
SMS			
Mean (SD)	24.42 (16.33)	49.64 (20.03)	27.21 (14.41)
DS			
Mean (SD)	24.57 (10.98)	40.83 (11.04)	34.82 (13.77)

3.2. Between-groups differences in child attention and looking

3.2.1. Attention

SMS and DS were compared on differences between *child to adult attention and looking* and *child to peer attention and looking* in each social context (one-to-one attention, shared attention and free play). Fig. 1 demonstrates the similarities of patterns of results for attention and looking preferences.

For attention a main effect of social context was evident ($F(2, 16) = 25.62$, $p < .001$). Contrasts indicated that, as expected, preferential attention directed towards adults was greater in one-to-one attention than shared attention ($F(1, 17) = 24.71$, $p < .001$) and greater in shared attention than in free-play ($F(1, 17) = 5.84$, $p = .03$). There was also a main effect of syndrome ($F(1, 16) = 8.87$, $p = .008$), whereby SMS values were consistency higher, denoting greater preferential attention directed towards adults than peers compared to DS, when overall adult child ratio is controlled for. There was no interaction between syndrome and social context ($F(2, 17) = .004$, $p = .996$), thus pattern of differences between SMS and DS was independent of setting.

3.2.2. Vigilance for adults and peers

For *child to adult and child to peer looking* a main effect of social context was found ($F(2, 16) = 23.68$, $p < .001$), again contrasts showed preferential looking towards adults was greater in one-to-one attention than shared attention ($F(1, 17) = 21.32$, $p < .001$) and greater in shared attention than in free play ($F(1, 17) = 7.07$, $p = .017$). There was also a main effect of syndrome ($F(1, 16) = 7.58$, $p = .014$), with SMS having consistently higher scores, denoting greater preferential looking to adults, compared to DS. Lack of interaction between syndrome and social context ($F(2, 17) = 12.98$, $p = .7$) suggests pattern of differences between SMS and DS did not differ across the settings.

These analyses indicate that the SMS group showed higher levels of *child to adult attention and looking* (over peers) across different social contexts than the DS group. For both syndromes preference towards adults was greatest in one to one attention and least in shared attention.

3.3. Sequences of child–adult interactions

3.3.1. Association between adult to child attention and child to adult attention

In order to evaluate whether children with SMS initiate and maintain interaction to a greater extent than those with DS the association between *child to adult attention* and *adult to child attention* was compared across syndromes. Time based lag sequential analysis was used to examine sequential relationships between *adult to child attention* and *child to adult attention*, before (−60 s to −20 s; lag −3 to −1), during (lag 0) and after (+20 s to +60 s; lag 1–3) *adult to child attention* occurred. Two mixed 3 × 2 ANOVAs were used to compare strength of association (denoted by mean Yule's *Q* values) between child and adult behaviour across syndrome groups and over time, one ANOVA leading up to when *adult to child attention* occurred and one after it occurred. A *t*-test was used to compare the strength of association between child and adult behaviours at the point at which *adult to child attention* occurred.

Fig. 2 (left panel) shows that for both syndromes the association between *adult to child attention* and *child to adult attention* increased leading up to when the adult gave the child attention and then decreased following this point. The

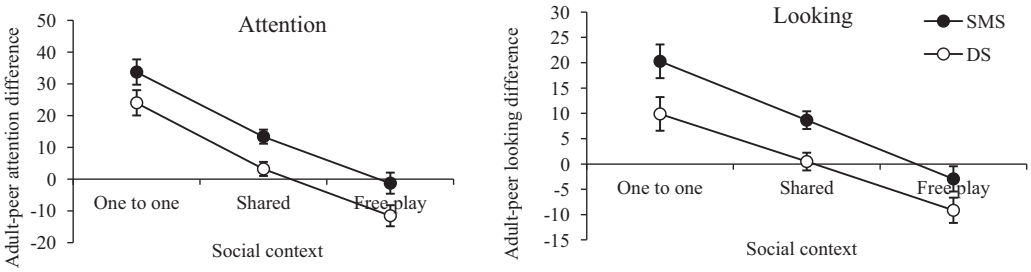

Fig. 1. Adult–peer difference for adult and peer directed attention (left panel) and looking (right panel) across social contexts for SMS and DS (positive values represent adult preference, negative values indicate peer preference) (±1 standard error).

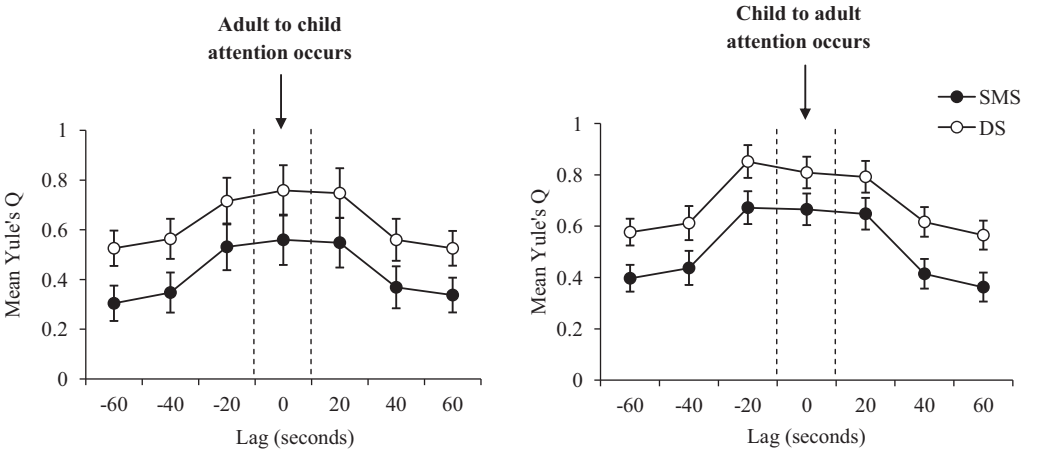

Fig. 2. Association between *adult to child attention* and *child to adult attention* (left panel) and association between *child to adult attention* and *adult to child attention* (right panel) for SMS and DS (±1 standard error).

ANOVA performed for the time period before *adult to child attention* occurred indicated a significantly stronger association between *adult to child attention* and *child to adult attention* overall for DS than SMS, indicated by the main effect of syndrome ($F(1, 18) = 5.46$, $p = .031$) and across syndromes the association increased as occurrence of adult attention being given to the child became closer, demonstrated by the main effect of lag ($F(1.25, 22.43) = 83.04$, $p < .001$). Syndrome groups did not show different patterns of association over time, only magnitude of association, denoted by lack of interaction between syndrome and lag ($F(1.23, 22.43) = .7$, $p = .44$).

At the point at which *adult to child attention* occurred, groups did not differ in strength of association, $t(10.86) = -1.87$, $p = .09$, equal variances not assumed). However, for the period after *adult to child attention* occurred the ANOVA again showed a stronger association overall for DS than SMS, indicated by the main effect of syndrome ($F(1, 18) = 4.41$, $p = .05$) and across syndromes the association between *adult to child attention* and *child to adult attention* decreased the further away from the occurrence of the adult giving the child attention, illustrated by the main effect of lag ($F(1.24, 22.26) = 57.07$, $p < .001$). There was no interaction between syndrome and lag ($F(1.24, 22.26) = .04$, $p = .90$).

The lack of a significant interaction in the ANOVAs of the period prior to and after the adult giving the child attention indicates that that although there was a generally weaker association between *adult to child attention* and *child to adult attention* in SMS compared to DS there was not a difference in the timings at which the association between these child and adult behaviours changed. Therefore in the period prior to the adult giving the child attention the association between child and adult behaviour increased at a similar rate as the point at which the adult directed their attention to the child became closer, i.e. children with SMS did not start directing their attention to the adult earlier in the sequence, thus children with SMS did not initiate interaction earlier than those with DS. Similarly in the period after the adult gave the child attention the association between adult and child behaviour decreased at a similar rate after the adult gave attention to the child, i.e. children with SMS did not continue directing attention to the adult after attention from the adult has ceased to a greater extent than those with DS, thus children with SMS and DS appear to *maintain* social interaction to a similar extent.

3.3.2. Association between child to adult attention and adult to child attention

As an exploratory analysis the association between *child to adult attention* and *adult to child attention* was examined, evaluating adult behaviour directed to the child before, during and after the child directing attention to the adult. Fig. 2 (right panel) shows that the association between *child to adult attention* and *adult to child attention* increased up to the point that the child gave attention to the adult and decreased after this point. This association was consistently stronger for DS compared to SMS.

The ANOVA of the time period prior to the occurrence of *child to adult attention* indicated that the association between *child to adult attention* and *adult to child attention* was stronger for DS than SMS ($F(1, 18) = 4.87$, $p = .04$). Across groups this association increased as the point of *child to adult attention* became closer ($F(1.53, 27.46) = 61.33$, $p < .001$) and lack of interaction between lag and syndrome indicates that the pattern of this increase did not differ between groups ($F(1.53, 27.46) = .006$, $p = .995$). This shows that while there was a generally weaker association between *child to adult attention* and *adult to child attention* in SMS compared to DS there was no difference in the timings at which this association increased approaching the point at which the child gave attention to the adult.

When *child to adult attention* occurred no difference in strength of association was found between groups ($t(18) = -1.65$, $p = .12$). In the period after the occurrence of *child to adult attention*, the ANOVA showed that the association between *child to adult attention* and *adult to child attention* decreased across groups ($F(1.33, 23.85) = 96.9$, $p < .001$) and this association was stronger overall for DS than SMS ($F(1, 18) = 5.24$, $p = .03$). Rate of decrease was the same across syndromes, indicated by lack

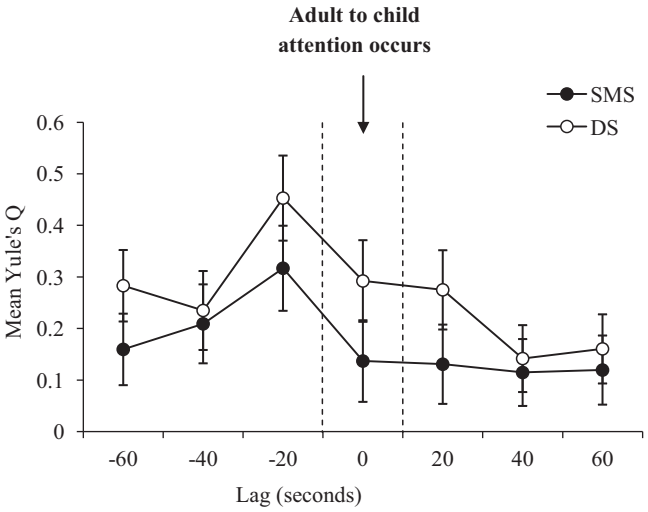

Fig. 3. Association between *adult to child attention* and onset of *child to adult looking* for SMS and DS (±1 standard error).

of interaction between syndrome and lag ($F(1.33, 23.85) = 1.49$, $p = .24$), again suggesting that groups showed similar patterns of results, with no difference in the timings at which the association between these behaviours decreased after the child gave attention to the adult.

3.3.3. Association between adult to child attention and child to adult looking

To identify whether children with SMS show increased vigilance for adult attention (demonstrated by looking at adults earlier in an interaction) than children with DS, and whether they look at the adult for longer after the adult has given them attention (potentially maintaining interaction), the association between *adult to child attention* and *child to adult looking* was compared across groups before, during and after the adult gave attention to the child. The results of this analysis are shown in Fig. 3.

Fig. 3 shows that whilst there was a stronger association between *adult to child attention* and *child to adult looking* in DS than in SMS, there is variability in this association. For the ANOVA of the period before *adult to child attention* occurred a main effect of lag demonstrated increases in the strength of association between onset of *child to adult looking* and *adult to child attention* across the groups as the point at which adult attention is given to the child became closer ($F(2, 36) = 23.16$, $p < .001$). Groups did not differ in the magnitude of this association, as indicated by the lack of main effect of syndrome ($F(1, 18) = .85$, $p = .37$) and a non-significant interaction suggests that the timings at which the association increased in the lead up to the adult giving attention to the child did not differ between syndromes ($F(2, 36) = 2.37$, $p = .11$), i.e. children with SMS did not look at the adult earlier in the sequence.

At the point when *adult to child attention* occurred the *t*-test indicated that groups did not differ in strength of association between *adult to child attention* and *child to adult looking* ($t(18) = -1.39$, $p = .18$). The ANOVA of the period after *adult to child attention* occurred also found no overall between syndrome differences in the strength of association between *adult to child attention* and *child to adult looking*, demonstrated by the lack of a main effect of syndrome ($F(1, 18) = .56$, $p = .46$). A main effect of lag suggests that across groups the association decreased over time after the adult gave attention to the child ($F(2, 36) = 5.53$, $p = 0.008$). A significant interaction between syndrome and lag ($F(2, 36) = 3.52$, $p = 0.04$) suggests that the groups showed different changes in their looking behaviour over time after *adult to child attention* occurred. Post hoc analysis of simple effects found no significant difference between groups at any lags; lag 1 ($t(18) = -1.33$, $p = .2$), lag 2 ($t(18) = -.29$, $p = .77$) or lag 3 ($t(18) = -.43$, $p = .67$). Within-groups analysis showed a significant decrease in DS between lags 1 and 2 ($t(9) = 4.21$, $p = .002$), but no difference between lags 2–3 ($t(9) = -.82$, $p = .44$). For SMS no significant differences were found between lags 1–2 ($t(9) = .5$, $p = .63$) or 2–3 ($t(9) = -.16$, $p = .88$). These analyses therefore indicate that there were early decreases in strength of association between *adult to child attention* and *child to adult looking* in DS which were not evident in SMS, suggesting that those with SMS continued to look at the adult after *adult to child attention* occurs for longer than those with DS.

4. Discussion

This is the first study to systematically observe social behaviour of children with SMS in schools, when interacting with both adults and peers and to contrast social behaviour directed towards adults and peers to evaluate preference. Use of a comparable contrast group, recruited based on their close mental age matches and comparable on both age and estimates of ability, suggests differences are unlikely to be artefacts of level of intellectual disability. The study is also the first to examine

sequences of behaviour in SMS to evaluate patterns of reciprocal social initiation, which are independent of absolute levels of attention given or received by children. Use of operationally defined, reliably coded behaviours and natural observation in schools, means that findings are likely to accurately reflect children's behaviour in their natural environment.

Reports that children with SMS have a strong preference for adult attention, over that of peers (e.g. Haas-Givler, 1994) were supported in analyses of children's preferential attention and looking towards adults and peers. As hypothesized, children with SMS showed greater preference for directing attention to adults versus peers than children with DS did and they also showed more preferential looking directed towards adults. Using looking as an index of vigilance or attention is common in research investigating social preferences, for example eye tracking is commonly used as an implicit measure of attention to social stimuli (Klin, Jones, Schultz, Volkmar, & Cohen, 2002; Riby & Hancock, 2008; Speer, Cook, McMahon, & Clark, 2007). In William's syndrome eye tracking has revealed prolonged face gaze (Riby & Hancock, 2009) which has been linked to the social phenotype which includes prolonged face looking during social interactions (Mervis et al., 2003) and hypersociability (Jones et al., 2000). Together, the increased preference for interacting with and looking towards adults shown by children with SMS suggest strong preference for interactions with adults over peers. This finding takes into account the higher adult child ratio found in SMS classrooms, the result of the majority of children with SMS attending special schools, whereas most children with DS attended mainstream schools. This is unlikely to reflect difference in intellectual ability between groups, particularly as DS was a developmentally matched contrast group. It has been noted previously (Udwin, Webber, & Horn, 2001) that high rates of challenging behaviour shown in SMS may compromise adaptive behaviour which, together with social and emotional difficulties, can result in the requirement for special education.

The importance of this preference for adult attention, particularly in an educational context, has been outlined previously (Haas-Givler, 1994; Haas-Givler & Finucane, 1996) in the advice distributed by the American SMS support organization (Parents and Researchers Interested in Smith–Magenis Syndrome; PRISMS). It is noted that attention-seeking from adults shown by children with SMS can interfere with classroom activities (Haas-Givler, 1994). Furthermore, challenging behaviour can arise when attention is unavailable (Langthorne & McGill, 2012; Sloneem et al., 2011; Taylor & Oliver, 2008). This is illustrated by the following quote, "If I could provide constant, unwavering, one-on-one attention to this child throughout the day, she might never have another tantrum." (Teacher of a child with SMS, cited by Haas-Givler, 1994, p. 36). It is notable in the only published direct observation study of behaviour in schools to date, by Taylor and Oliver (2008), all children received one-to-one adult support throughout the school day, yet despite this reductions in adult attention resulted in challenging behaviour. Clearly, maintaining constant attention is not achievable, either at school or home. Strategies for dealing with this elevated drive for adult attention may address challenging behaviour associated with reduced attention and the burden it places on those who care for and work with individuals with SMS. Proof of principle has been established for interventions which aim to reduce difficult behaviour related to excessive drive for social interaction in AS. For example Heald, Allen, Villa, and Oliver (2013) demonstrated that it is possible to reduce excessively high rates of social approach behaviour in children with AS using discrimination training to teach children to discriminate times of adult availability. It has also been shown that Functional Communication Training is effective in reducing the frequency of challenging behaviours associated with accessing attention (Radstaake, Didden, Oliver, Allen, & Curfs, 2012). These types of intervention may also be effective in reducing the problem behaviours associated with strong drive for adult attention in SMS, both the approaches from children which may in themselves be difficult to manage and also the challenging behaviour which can result if adult attention is unavailable.

The statistical analyses employed catered for inevitable differences in classroom environment (greater number of adults and fewer children) in special needs schools. However, there may be other differences in the classroom and in adult interactions with children that cannot be controlled for statistically. For example the typically developing peers present in a mainstream school environment may be more responsive to other children's interactions than those at a special school. Therefore, it is important to remain cautious when interpreting differences found in preference between children with SMS and DS. Future research employing developmentally matched contrast groups also matched on schooling may address this potential confound. However, given the need to match in terms of ability, whether this is achievable is doubtful given children with SMS are likely to be in special school settings due to their behaviour rather than their ability.

In addition to differences in overall levels of behaviour between the groups, differences in *sequences* of behaviour were found. This finding is independent of absolute levels of attention received by children from adults in their environment and as such this finding is unlikely to be an artefact of differences in schooling. Stronger associations between *adult to child attention* and *child to adult attention* and vice versa were found in DS compared to SMS, both prior to and after attention was given. This difference suggests behaviours of adults and children are more closely aligned or synchronized in DS, such that when *adult to child attention* is occurring then *child to adult attention* is also taking place and vice versa (i.e. there is greater reciprocity between adults and children in DS than SMS). During interaction no difference was found between groups, possibly because cues become stronger immediately before and immediately after a person gives attention, prompting responses.

The weaker association in SMS between adult and child attention before and after an adult gives the child attention suggests that children were showing adult directed behaviours at times other than when the adult was giving them attention. Children with SMS may be less effective at responding to the cues for initiating, maintaining and terminating interaction and they may be making overtures towards the adults at times when the adult is unavailable. Thus, they may show adult directed behaviours at times that do not synchronize with adult behaviours towards them, explaining the lower associations found. Physical constraints on communication and interaction may be an additional explanation, as those with

SMS may be more likely to fail to detect an adult interacting with them and reciprocate due to hearing impairment (Chen, Potocki, & Lupski, 1996; Greenberg et al., 1991, 1996). That the overall patterns of interactions were very similar across groups suggests the syndromes showed similar patterns of association between *adult to child attention* and *child to adult attention* across the sequence, failing to support the hypothesis that children with SMS would initiate interaction earlier and attempt to maintain it for longer than those with DS.

It is somewhat surprising that adults also showed similar patterns of reduced association between *adult to child attention* before and after *child to adult attention* occurring. This suggests that adults respond less to cues that the child is initiating interaction, such that the adult makes initiations towards the child at times when the child does not respond. This may be linked to the previous explanation regarding children with SMS directing attention to adults at times when they cannot or do not respond, potentially reflecting deliberate classroom strategies for dealing with attention-seeking behaviour (i.e. ignoring child initiations made at inappropriate times, an approach recommended by professionals working with those with SMS, Haas-Givler & Finucane, 1996).

No evidence was found supporting the hypothesis of an attentional bias in terms of increased vigilance towards adult attention as indexed by earlier looking towards adults. In Riby and Hancock's (2009) study of face gaze in Williams syndrome it was also found that there was not an attentional bias in terms of social stimuli (faces) capturing attention *earlier* than they do in typically developing individuals, only prolonged looking once the faces caught participants' attention. This finding demonstrates that looking is a rich source of information about attention to social stimuli, including vigilance and sustained attention. It is evident from both the current findings and those of Riby and Hancock (2009) that a behavioural phenotype associated with strong social motivation to interact with social targets may be associated with unusual patterns of looking which may be atypical at different stages of interaction. In both SMS and Williams syndrome findings suggest that early attention to salient social stimuli may not be atypical. In WS it has been suggested that there is a difficulty with disengaging attention which may account for the prolonged face looking (Riby & Hancock, 2009), however in SMS no study has yet looked at patterns of looking in such detail. Future research employing eye tracking methodologies in this population could provide more robust evidence for atypicalities in visual attention to social stimuli.

There was some evidence suggesting children with SMS may show a less rapid decrease in looking after *adult to child attention* ceases, than those with DS. Interestingly Mount, Oliver, Berg, and Horsler (2011) also found children with AS did not try to *initiate* eye contact with their mother more than a stranger, but that once it was established they were more likely to try to *maintain* it with their mother than the unfamiliar adult. Therefore, AS and SMS potentially demonstrate similar patterns of maintaining, but not eliciting, eye contact with a preferred person (a familiar person in AS, an adult in SMS). It is unclear what function this eye contact has in either syndrome, whether it is an index of interest or functions to maintain interaction (for example after *adult to child attention* has ceased in SMS). Alternatively, it may reflect lack of responsiveness to indicators of termination of attention from adults, such that children continue to look at them.

Overall findings relating to sequences of behaviour fail to support the hypothesis that children with SMS initiate interaction earlier than other children, or that they are more vigilant towards adult cues that they are going to provide attention, although they may maintain eye contact more than DS. This contrasts with suggestions that children with SMS *seek* more attention than other children (Dykens et al., 1997; Dykens & Smith, 1998; Feinstein & Singh, 2007; Haas-Givler, 1994), but does not directly contradict these. It suggests rather that any attention-seeking behaviours do not consistently start earlier in sequences of adult child interaction. Furthermore, findings that interactions between children with SMS and adults are less synchronized suggest children make initiations at times when adult attention is unavailable, adults fail to respond to children's initiations or a combination of the two. This, combined with initial findings of greater preference for adults over peers, may support suggestions that children with SMS do seek attention from adults but possibly not always at times when attention is available.

As noted previously a key limitation of the current study is differences in schooling between the syndromes. While steps were taken to control for difference in numbers of adults and children in the classroom in analyses it is possible that other differences in schooling may have affected findings. Additionally, whilst the current study doubled the sample size of the only previous study to directly examine social behaviour in SMS, the sample size was still relatively small which may have reduced power for some comparisons. Future research would therefore benefit from a larger sample.

In summary, results indicated that children with SMS show greater preference in terms of giving attention to and looking towards adults (compared to peers) than those with DS. Furthermore, children with SMS appear to show a weaker association between adult and child behaviours leading up to and after the point that attention is given than those with DS. This could indicate a less organized relationship between child and adult behaviours, possibly a result of children with SMS showing social behaviours that initiate social interaction at times when it is unavailable.

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