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Atypical development of language and social communication in toddlers with Williams syndrome

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Abstract

Williams syndrome (WS) is a genetic disorder which results in an uneven cognitive profile. Despite superior language compared to other syndromes in the phenotypic outcome, toddlers with WS are as delayed in their language onset and early linguistic development as are toddlers with other syndromes. The cause of this delay in WS is as yet unknown. In a series of experiments, we examined whether atypical socio-interactive precursors to language could contribute to the explanation of the late language onset and atypical developmental pathways observed in WS. Experiment 1 showed that despite superficially good social skills, toddlers with WS were only proficient at dyadic interaction. They were impaired in triadic interaction, essential for the referential uses of language, and showed none of the correlations between socio-interactive markers and language seen in the typical controls. Experiment 2 focused on the comprehension and production of referential pointing. Again, the WS group was impaired, despite vocabulary levels higher than those of typically developing controls. Finally, Experiment 3 examined fine motor skills. The WS lack of pointing could not be explained in terms of motor impairments, since the WS toddlers were proficient at fine motor control, such as the pincer grip. Overall, our data indicate that the early stages of WS language follow an atypical pathway. The findings challenge the frequent claims in the literature that individuals with Williams syndrome have preserved linguistic and social skills.

Williams syndrome (WS) is a rare genetic disorder occurring in 1 in 20,000 live births (Beuren, 1972; Greenberg, 1990). It is caused by a hemizygous sub-microscopic deletion of some 17 contiguous genes on chromosome 7q11.23 (Ewart *et al.*, 1993; Tassabehji *et al.*, 1996). Phenotypically, Williams syndrome is characterized by relatively low IQ, facial dysmorphology and a number of other physical anomalies, including supra-valvular aortic stenosis, hyperacusis and dental hypoplasia (Jones & Smith, 1975; McKusick, 1988).

In the phenotypic outcome, Williams syndrome presents an uneven cognitive profile. Some aspects of language performance and social interaction seem to be relatively good, although many non-linguistic functions, such as spatial cognition, number, planning and problem solving, are impaired (Arnold, Yule & Martin, 1985; Bellugi, Bihrle, Jernigan, Trauner & Doherty, 1990). The unevenness of the profile is particularly striking in that while IQ in this population is generally in the 50–65 range, language is often surprisingly proficient in comparison.

However, despite its relative strength in the WS phenotypic outcome, the onset of language is severely delayed in early childhood (Paterson, Brown, Gsödl, Johnson & Karmiloff-Smith, 1999; Singer-Harris, Bellugi, Bates, Jones & Rossen, 1997). Furthermore, there is some evidence to suggest that language acquisition in WS follows a different developmental pathway from that seen in typical development. In a longitudinal study of children with WS and children with Down's syndrome (DS), Mervis, Morris, Bertrand and Robinson (1999) showed that some of the relationships between language and cognitive milestones that obtain for typical development as well as for DS are not apparent in WS. For example, in contrast to the two other groups, the WS group had already undergone the vocabulary spurt prior to engaging in spontaneous exhaustive sorting. These occur simultaneously

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both in typical and in the albeit delayed DS development. In the WS toddlers, speech production preceded pointing, whereas it followed pointing in the typically developing and DS toddlers. Furthermore, Stevens and Karmiloff-Smith (1997) demonstrated that while young children with WS abide by the fast mapping and mutual exclusivity constraints when learning new words, the whole object or taxonomic constraints were considerably weaker. These studies suggest that in spite of relatively good vocabulary scores, children with WS go about the process of acquiring words differently. They seem to rely on strategies which place more weight on phonological short-term memory than semantics when acquiring novel words (Vicari, Brizzolara, Carlesimo, Pezzini & Volterra, 1996; Vicari, Carlesimo, Brizzolara & Pezzini, 1996; Grant, Karmiloff-Smith, Gathercole, Paterson, Howlin, Davis & Udwin, 1997).

The delay in the onset of WS language and the subsequent atypical pathway remain to be explained. It is thus crucial to examine development at an earlier stage than speech production. Speech production delays, such as those found in DS, are sometimes due to articulation difficulties. However, in the case of WS where articulation problems are not apparent, even the comprehension abilities of toddlers are delayed (Paterson *et al.*, 1999). Levels are at about half their chronological age and as delayed as toddlers with DS, despite the fact that in later life WS vocabulary levels clearly surpass those of people with DS (Paterson, 2000). The present study aims to explore the precursors that have been argued to support language acquisition in normal infants and toddlers (Bates, Benigni, Bretherton, Camaioni & Volterra, 1979) i.e. their preverbal communication and joint attention skills.

Joint attention refers to the ability to engage in triadic co-ordination between a child, another person (usually the parent/carer) and an object external to space of the face-to-face interaction of the dyad. This skill develops pre-linguistically. It follows a shift from the primarily face-to-face dyadic interaction of typically developing infants aged 0–5 months to the triadic exchanges (the triangle of child–interlocutor–object) of children between the ages of 6–18 months (Bakeman & Adamson, 1984). Once capable of triadic exchange, the infant begins to use and respond to non-verbal acts that go beyond face-to-face interaction (i.e. direction of gaze towards external objects, attention shifting, reaching, pointing with vocalizing). These acts are used to co-ordinate attention with others towards a given object or event outside the confines of the dyad. A distinction is made between declarative joint attention and instrumental joint attention. The former refers to exchanges in which the child seeks to share or comment on an event or object with the interlocutor (e.g. a point or vocalization meaning 'look, a nice toy'). By contrast, the function of instrumental joint attention is to request or obtain an object out of reach (e.g. a point or vocalization meaning, 'I want you to give me that toy').

A number of studies have demonstrated that the ability to engage in both instrumental and declarative joint attention is important for subsequent language development (Bates *et al.*, 1979). In particular, the emergence of triadic communication has been claimed to underlie crucial developments in social cognition (Butterworth & Cochran, 1980) and in representational skills (Werner & Kaplan, 1963), as well as providing an important way in which reference is established (Baldwin, 1991). Individual variability in such non-verbal skills has been shown to be related to subsequent language acquisition (Bates *et al.*, 1979; Ulvand & Smith, 1996). Moreover, Mundy and Gomes (1998) argued that aspects of joint attention are related differentially to expressive and receptive language. They demonstrated that 'initiating joint attention' (i.e. alternating eye contact between an object and a person, using declarative pointing, and showing) is a significant predictor of expressive language, while responding to joint attention (following the adults' gaze or point) is related to both expressive and receptive language.

Within joint attention, a special case has been made for the role of pointing. Pointing is a species-specific gesture that normally emerges around 10 months of age. It has been shown to be strongly related to language development. For example, Camaioni, Castelli, Longobardi and Volterra (1991) demonstrated that pointing at 12 months strongly predicts later speech production. Similarly, Harris, Barlow-Brown and Chasin (1995) showed that pointing onset is related to the comprehension of object names. Pointing is obviously one way in which the typically developing child establishes reference between an object and its label. It is also an important means by which the caregiver provides labels in reaction to the child's pointing (Kessler-Shaw, 1992). Finally, Franco and Butterworth (1996) maintain that true pointing is distinct from requesting in that it has a specialized declarative/referential function in relation to language.

If the development of joint attention skills is a crucial precursor to language acquisition, then impairments in these skills may provide important clues as to why language in WS is delayed and why it follows an atypical developmental trajectory. Furthermore, if children turn out to be impaired in their use of the pointing gesture, this is likely to have implications for the pattern of their interactions with parents and carers.

A few studies have examined the role of joint attention skills in atypical development. Mundy, Sigman, Kasari and Yirmiya (1988) investigated the joint attention skills of two groups of children with Down's syndrome: one with mental age less than 21 months (mean chronological age = 22.9 months) and one with mental age above 21 months (mean chronological age = 43 months). Compared with two groups of mental-age matched controls (typically developing and general learning disability), the DS children showed higher levels of social interaction but a lower frequency of requests for objects. Furthermore, the children with DS displayed a unique pattern of association between language and aspects of joint attention: instrumental joint attention was significantly related to expressive language, whereas in typical development it was declarative joint attention that was correlated with expressive language. Franco and Wishart (1995) also investigated the requesting and pointing gestures of young children with DS aged between 21 and 47 months. They found that these children actually made more declarative gestures and fewer instrumental gestures than typically developing children. This is a surprising finding until one notes that declarative gestures rather than oral language are used for longer in development of children with DS, because non-verbal communication temporarily replaces their very delayed spoken language. These findings have led to the suggestion that aspects of spoken language may be dissociated in DS, with the social/communicative bases to language being intact and the development of morphosyntax impaired (Fowler, 1990; Franco & Wishart, 1995).

A different pattern of impairments in joint attention has been observed in young children with autism. They have been shown to have good instrumental joint attention but poor declarative joint attention (Mundy & Gomes, 1998; Baron-Cohen, 1989a). This holds for both understanding and production of pointing (Baron-Cohen, 1989b). This syndrome-specific pattern of impairments in joint attention has been linked to the general problems in autism with social communication (Mundy & Sigman, 1989).

With respect to WS, some preliminary observational evidence suggests that joint attention may also be impaired. Mervis *et al.* (1999) showed that in a free-play situation, toddlers with WS did not spontaneously use the pointing gesture. Furthermore, as mentioned above, while in normal development children point referentially before using referential language (Fenson *et al.*, 1993), the opposite pattern was observed in WS. In another observational study, Bertrand, Mervis, Rice and Adamson (1993) investigated the joint attention skills of one girl with WS whom they observed longitudinally from age 1 year 8 months to 2 years 8 months. They report that this child did not engage in co-ordinated joint attention until well after the vocabulary spurt. Furthermore, the child showed little interest in objects and made few requests, preferring to focus on the faces of those with whom she was interacting. These observational data point to the possibility that children with WS use different strategies to establish reference. Moreover, the fact that children with varying developmental disorders have been shown to display different patterns of joint attention suggests that this skill may offer clues as to the nature of the early language delay in WS.

The present experimental study aimed to replicate and extend the observational case study of Bertrand *et al.* (1993) by using a standardized experimental procedure for measuring joint attention on a larger sample of children with WS. In Experiment 1, the Early Social Communication Scales (Mundy & Hogan, 1996) was administered. Experiment 2 investigated pointing in an experimentally constrained situation rather than in spontaneous play. Finally, given evidence to suggest a relationship between the development of fine motor skills and pointing as well as between such skills and language (Butterworth & Morrisette, 1996), Experiment 3 investigated the fine motor control in toddlers with Williams syndrome.

Experiment 1

Method

Participants

A sample of 13 toddlers with Williams syndrome was recruited through the Williams Syndrome Foundation, a UK-based parent support group. All of these children had been diagnosed clinically as well as by means of the fluorescence in situ hybridization (FISH) genetic test for deletion of the elastin gene. The mean chronological age of the children was 31 months (range: 17 months to 55 months). Thirteen mental-age matched control children were also tested. Both groups comprised 7 boys and 6 girls.

Background measures

Mental ages were calculated using the Bayley Scales of Infant Development (Bayley, 1993). The mean mental age of the WS group was 14 months (range: 6 months to 23 months). The children were individually matched on sex and mental age to a group of typically developing toddlers. The mean chronological and mental ages of the control group were, respectively, 13 months (range: 5 months to 21 months) and 13 months (range: 5 months to 22 months).

Parents were asked to complete the MacArthur Communicative Development Inventories (Fenson *et al.*, 1993) to obtain a measure of language development. All

Table 1 *The chronological, mental ages and language scores of participants in Experiment 1*

	WS	Control
Chronological age[a]*	30.9 (11.5)	13.5 (5.04)
Mental age[a]	13.9 (5.04)	13.5 (4.96)
Language comprehension[b]	116.2 (101.6)	110.3 (89.5)
Language production*[b]	56 (83.3)	31.5 (53.2)
Gestures[b]	5.85 (3.6)	6.77 (2.92)
Games[b]	20.1 (15.1)	19.6 (13.5)

*p = <0.05
Notes: [a]in months; [b]raw scores from the MacArthur Inventory.

parents were given the same form (Words and Gestures) to complete. Although some of the participants were, strictly speaking, too old to be scored on this form, for comparison purposes it was decided that all participants should receive the same form. This decision turned out to be justified since no child was at, or close to, ceiling on the CDI. Performance on these background measures is shown in Table 1. While the groups did not differ in terms of their mental age, there were some differences in terms of their language production although these differences were not significant ($t = 0.90$, $p = 0.38$). There was also no significant difference in language comprehension ($t = 0.16$, $p = 0.88$). We, therefore, consider the groups to be well matched in terms of both mental age and language.

Procedure

All the children were visited at home where the above background measures were administered. The remainder of the testing took place at Sussex University. Joint attention skills were assessed using the Early Social Communication Scales (ESCS, Mundy & Hogan, 1996). The ESCS has been shown to give reliable and valid indices of early social communicative development both in typically developing children (Mundy & Gomes, 1998) and in children with developmental disorders (Mundy *et al.*, 1988).

The testing took place in a room measuring 3.2 m by 2.5 m; the walls were completely covered with plain curtains. Testing sessions lasted about 25 minutes and were carried out by a single experimenter. During the assessment, the child and experimenter sat facing one another across a small table, with the child's parent seated to the right of the child. The experimenter presented the child with a sequence of toys: 3 trials with 3 different activated mechanical toys (e.g. wind-up animals) and 3 trials with hand-operated toys (e.g. squeaky toys). The toys were presented and/or activated on the table one at a time. Verbal communication during this time was kept at a minimum. The experimenter also engaged the child in a tickle game (2 trials) and a turn-taking game in which either a ball or a small car was rolled between the child and the experimenter. The child was also given a hat, a comb and glasses, and invited to play with them and to put them on the experimenter. Children were also presented with a tightly fixed screw jar containing an attractive wind-up toy to ascertain whether they would ask for help in opening the jar. During the session the experimenter verbally and gesturally requested to the child to give toys to the experimenter. Comprehension of pointing was also assessed in the course of the session by the experimenter drawing the child's attention to a number of posters around the room. The posters were placed on the walls 90 degrees to the child's left and right as well as 180 degrees behind the child. The tester visually fixated on and pointed to the posters in each position twice and called the child's name 3 times with increasing emphasis. The tester also pointed to pictures in a book on the table. The extent to which the child followed the points was measured. Table 2 lists each of the behaviour categories which were coded during the session.

Some of these behaviours may be considered more central to joint attention than others. Joint attention is usually taken to mean triadic attention between child, interlocutor and objects/events. In this sense, the social interaction category is an exception in that it primarily assesses dyadic turn-taking and maintenance of interaction between child and adult, without considering events or objects outside the dyad. Nevertheless, dyadic interaction is known to be a precursor of triadic attention in typical development and is therefore also included here.

Child–experimenter interaction was videotaped to record the front upper body view of the child and the upper body profile of the experimenter.

Results

Observations of the interaction yielded frequency of behaviour scores in 5 categories: Initiating Joint Attention (IJA); Responding to Joint Attention (RJA); Initiating Requesting (IR); Responding to Requesting (RR) and Social Interaction (SI). The behaviours observed within these categories are listed and defined in Table 2. Each child received a frequency score for the behaviours, except on the RJA trial where the percentage of points followed for the book and poster trials was calculated. This reflected the proportion of correct head-turn responses to the experimenter's points. In addition, the number of times the child turned to the parent in a socially referential way was recorded. Social referencing was said to occur when the child looked first at the object with interest or surprise and then at the parent.

Table 2 *The behaviour categories of the Early Social Communication Scales*

Variable	Behaviours observed
Initiates Joint Attention	Makes eye contact while manipulating a toy Alternates eye contact between an active toy and the tester Points to a toy or distal object in the room Shows objects to the tester or to the parent
Responds to Joint Attention	% trials child follows tester's point to the poster % trials child follows tester's point to the book
Initiating Requesting	Makes eye contact when object is moved out of reach Makes eye contact before tester manipulates toy Reaches for toys Combines eye contact with a reach Gives object to tester/parent Points to indicate desired object Asks for help with getting toys from a screw jar
Social Interaction	Initiates turn-taking Teases Initiates tickle Eye contact after tickle Eye contact during tickle Combines eye contact with appeal after tickle Turn-taking with a ball and car Responds to invitation to play with hat, glasses and comb
Responding to Requesting	% responses to tester's request for the toy

Inter-rater reliability was assessed with videotaped data from a random selection of four children from the WS group and four children from the control group scored by two independent raters. All Pearson correlations between paired ratings ranged from $r = 0.88$ to 0.98 for the WS group and from 0.82 to 0.98 for the control group. A series of Mann-Whitney U tests revealed no significant differences between the two coders.

Comparing group performance on the ESCS

Mean performance (and standard deviations) on each of the behavioural variables is shown in Table 3.

As is frequently the case with atypical groups, the data showed large standard deviations. These data were therefore analysed using non-parametric tests. A series of Mann-Whitney U tests revealed significant group differences on the behavioural category of Initiating Requesting ($p = <0.05$). The group differences on the overall Responding to Joint Attention ($p = 0.07$) and Social Interaction ($p = 0.07$) categories approached significance. It can be seen from Table 3 that while the control group was better at initiating requesting and responding to joint attention, the WS group scored higher in terms of dyadic social interaction behaviours.

Table 3 *Performance on the ESCS*

Variable	WS group	Control group
Initiating Joint Attention*	11.1 (8.6)	15.2 (6.9)
Responding to JA (%)	50.8 (34.8)	65.4 (27.6)
Initiating Requesting**	10.5 (4.3)	23.1 (8.2)
Responding to Requesting	30.8 (43.5)	41.6 (34.5)
Social Interaction*	13 (4.5)	9.3 (6.2)
Social Referencing	3.0 (4.78)	5.2 (5.5)
Social Behaviour*	4.2 (4.6)	8.5 (5.4)

**$p = <0.05$, *$p = 0.07$

In order to consider group differences on the individual behaviours coded, a series of Mann-Whitney U tests was conducted. The mean frequency scores (and standard deviations) for each of these behaviours as well as significant group differences are shown in Table 4. It should be noted that within Social Interaction, no child produced behaviour in the initiating tickle, initiating turn-taking or teasing categories; these behaviours were, therefore, excluded from further analyses.

It can be seen from Table 4 that the groups differ on a number of the behavioural measures. The WS group produced far less pointing behaviour during the interaction than the control group. This was the case both in terms of referential/declarative pointing (initiating joint attention) and in terms of pointing as a form of request (initiating/instrumental requesting). There was also a trend for the children with WS to reach for the toys less often than the control group, although this difference failed to reach significance ($p = 0.1$). The children with WS also made less eye contact with the tester just before the tester manipulated the toy than did the control children. This kind of eye contact can be considered another form of requesting behaviour. The data show that the WS group was less interested in objects and did not request to play with objects as often as the children in the control group.

Interestingly, the WS group scored higher on the social interaction scale than the control group. The WS group was better at turn-taking, was more interactive during the tickle game and requested more tickles than the control group. While the WS group made more eye contact during the dyadic social interaction part of the assessment, this eye contact cannot be considered to be true joint attention as it was mainly not done with reference to an object outside the dyad. By contrast, the control group produced more triadic eye contact. They combined eye contact with reaching for the toy, made more eye contact while the tester manipulated the toy, and made more eye contact while manipulating the toy themselves.

Table 4 *Performance on the individual behaviour categories of the ESCS*

Behaviours observed	WS	Control
Initiates Joint Attention		
Makes eye contact while manipulating a toy	2.5 (4.2)	5.0 (3.1)
Alternates eye contact between an active toy and the tester	8.3 (6.8)	7.2 (5.7)
Points to a toy or distal object in the room**	0.08 (0.28)	2.5 (2.9)
Shows objects to the tester or to the parent	0.15 (0.55)	0.38 (0.65)
Responds to Joint Attention		
% trials child follows tester's point to the poster	46.4 (41.8)	62 (38.9)
% trials child follows tester's point to the book	55.3 (36.2)	68.8 (24.3)
Initiating Requesting		
Makes eye contact when object is moved out of reach	0.77 (1.2)	1.5 (1.66)
Makes eye contact before tester manipulates the toy*	0.31 (0.48)	1.69 (1.9)
Reaches for toys	6.5 (3.9)	9.2 (3.98)
Combines eye contact with a reach*	0.46 (0.88)	1.62 (1.9)
Gives object to tester/parent	1.1 (1.5)	3.0 (3.9)
Points to indicate desired object**	0.38 (0.87)	5.7 (5.75)
Asks for help with getting toys from a screw jar	0.62 (0.65)	0.38 (0.51)
Social Interaction		
Eye contact after tickle*	0.08 (0.28)	0.63 (0.87)
Eye contact during tickle	2.9 (1.6)	2.1 (1.3)
Combines eye contact with appeal after tickle**	0.62 (0.65)	0.31 (0.75)
Turn-taking with a ball and car**	7.7 (6.98)	4.8 (3.7)
Responds to invitation to play with hat, glasses and comb	1.7 (1.2)	1.5 (1.3)
Responding to Requesting		
% responses to tester's request for the toy	30.8 (43.5)	41.6 (34.5)

**p = <0.01; *p = <0.05

The frequency of social referencing to the parent during the whole assessment was recorded for both groups. The children in the control group made a mean number of 5.15 (SD = 5.5) references to the parents while being presented with the novel toys, while the children in the WS group made a mean of only 3.0 (SD = 4.78) references. While the difference between the groups was not significant ($p = 0.1$), there was a clear trend in this direction. When social behaviour also included the object, group differences were greater. To demonstrate this, a composite measure was created of socially referential behaviour, including both social referencing as well as giving and showing an object to the adult. The WS group and control groups scored means of 4.2 (SD = 4.6) and 8.5 (SD = 5.4), respectively. The difference between these two means was significant ($p < 0.01$).

The relationship between joint attention and performance on the language inventory

Correlations between language comprehension and language production (as measured by the MacArthur) and the 5 behavioural variables in the ESCS were calculated. The only behavioural category which was significantly related to language was the Responding to Joint Attention variable. This was significantly related to WS language comprehension ($r = 0.88$, $p < 0.05$), WS language production ($r = 0.81$, $p < 0.05$), as well as to control group language comprehension ($r = 0.64$, $p < 0.05$) and control group language production ($r = 0.55$, $p < 0.05$). There was also a strong relationship between mental age and Responding to Joint Attention for both the WS group ($r = 0.86$, $p < 0.05$) and the control group ($r = 0.79$, $p < 0.05$). It is therefore possible that the relationship between language and this aspect of joint attention is mediated by differences in mental age. The above correlations remain significant when a more conservative alpha level is adopted (0.05/7 = 0.01) with the exception of the correlation between Responding to Joint Attention and language production for the control group.

None of the other ESCS measures were significantly correlated with the MacArthur language scores. However, a number of interesting relationships between aspects of the ESCS can be noted. The correlation between Social Interaction and Responding to Joint Attention was significant for the control group ($r = 0.61$, $p < 0.05$) but not for the WS group ($r = 0.38$, $p = 0.20$). Similarly, the correlation between Social Interaction and Initiating Requesting was significant for the control group ($r = 0.61$, $p < 0.05$) but not for the WS group ($r = -0.02$, $p = 0.14$), although this correlation did not reach significance when the more conservative alpha level was applied. It would seem that although the WS group produced slightly more behaviours from the Social Interaction category, these behaviours were less well integrated with other aspects of joint attention than for the control group. In other words, the behaviour of the WS group was predominantly dyadic, whereas that of the control group was predominantly triadic. Furthermore, while there was no significant difference in the frequency of social referencing for the two groups, this social referencing behaviour was found to be significantly related (using both alpha levels) to the language measures for the control group (production: $r = 0.69$, $p < 0.05$; comprehension: $r = 0.41$, $p = 0.1$) but not for the WS group (production: $r = 0.16$, $p = 0.60$; comprehension: $r = 0.27$, $p = 0.37$).

Discussion

Experiment 1 demonstrated that children with WS perform atypically on a number of measures of joint attention. The WS group was impaired in both initiating and responding to joint attention bids in comparison to the group of typically developing children with the same mental age. In particular, the children with WS produced very few pointing gestures, either in an instrumental or declarative function. This lack of pointing confirms experimentally the observational findings of Mervis *et al.* (1999) and suggests that this is an area of genuine impairment for children with WS. Similarly, the WS children reached for the toys less often and combined eye contact with reaching significantly less often than the control group.

Bertrand *et al.* (1993) have argued that children with WS may be less interested in objects and more interested in faces than typically developing children. The findings of the present study provide some support for this suggestion. The WS group performed equally well or better than the controls most often when objects were not involved, as in the dyadic social interaction behaviours where they produced a significantly higher frequency of behaviours than the control group. This is not necessarily a positive finding, however. Such behaviours are not, strictly speaking, joint attention as they lack the triadic structure of true joint attention which involves the triangle of persons and objects outside the dyad. Indeed, it is important to distinguish between true triadic joint attention involving drawing someone's attention to an object outside the dyad and those cases where objects are involved but as part of the direct dyadic interaction.

A crucial finding from the present study is that the association between social interaction skills, social referencing and language is different for the WS group than it is for the typically developing children. Although the children with WS produced an equal amount, or in some cases more, of these behaviours, they were not related to their language skills, in contrast to the control group. These behaviours clearly do not have the same function in the development of children with WS as they do in normal development.

The children with WS in the present study differ considerably from children with DS and autism. Children with DS also make fewer non-verbal requests for toys, and in this sense, are comparable to the children with WS examined here. However, while children with DS have been shown to make fewer instrumental gestures, they do make more declarative gestures (Franco & Wishart, 1995). It has been claimed that delays in language production due to immature articulation in children with DS are compensated for by the use of gestures. In other words, gestures play a strong communicative function for children with DS. Our data suggest that this does not hold for children with WS. Although children with autism are poor at declarative gesturing, they are relatively unimpaired in their use of instrumental gesturing. We have shown that children with WS are impaired in their use of both. A possible cause for the poor declarative gesturing in autism resides in the problem these children have with aspects of social communication. It is interesting to note, therefore, that children with WS who, on the surface at least, appear to be relatively good at social communication are actually also impaired in their use of the declarative gesture.

The results of Experiment 1 suggest that reference is not established in the same way in WS as it is for typically developing children. The question of establishing reference is considered in Experiment 2, in which the lack of pointing in children with WS is explored experimentally in more detail.

Experiment 2

Introduction

Among other interesting findings, Experiment 1 showed that children with WS point less than typically developing children at a similar level of general cognitive development. One limitation with respect to the pointing items, however, was that the conditions of the experiment offered relatively few opportunities for initiating pointing. Most of the toys were only just out of reach, a situation far more likely to encourage use of the reaching gesture than the declarative pointing gesture (Franco & Butterworth, 1996). Despite this, it is noteworthy that the WS group used significantly less reaching and instrumental pointing. Not only were the occasions to produce pointing relatively limited, there were also few opportunities to follow other's pointing. Some of the poster points were to locations behind the child, and it is known that the ability of children to follow such points appears relatively late even in normal development (Butterworth & Jarrett, 1991). The aim of Experiment 2, therefore, was to examine the production and comprehension of the pointing gesture in a situation more conducive to producing such behaviour.

Experiment 2 used a procedure similar to that of Franco and Butterworth (1996) and Franco and Wishart (1995). These studies employed large dolls that move by remote control and are placed further from the child than the toys in Experiment 1. Previous studies with both typically developing children and children with DS demonstrate that they are more likely to produce declarative than instrumental gestures in response to these dolls, and more likely to produce instrumental than declarative gestures in a situation with toys placed just out of reach.

While the above experiments were concerned primarily with the production of the pointing gesture, we were also interested in the ability of children with WS to comprehend the pointing gesture. Previous research has suggested that typically developing children begin to comprehend pointing at about 10 months of age and that the comprehension precedes that of production of pointing (Butterworth & Grover, 1989; Messer, 1994). Prior to fully comprehending the function of pointing, younger children often become fixated on the pointer's hand and do not attend to the actual object or event being pointed at (Morissette, Ricard & Gouin-Decarie, 1995).

In Bertrand *et al.*'s (1993) observational study, they noted that their small group of children with WS seemed impaired at both the production and comprehension of pointing. In order to investigate this more fully, we used an experimental design that included both production and comprehension trials.

Method

Participants

Eleven of the mental-age matched pairs from Experiment 1 also took part in this second experiment. The mean chronological age, mental age and performance on the MacArthur Communicative Development Inventory of the two groups are shown in Table 5.

While the groups did not differ in mental age, there were some differences in performance on the MacArthur. While these differences were not significant, it is worth noting that the WS group had higher scores in terms of both language comprehension and production. In principle, this should give them an advantage over the typically developing group.

Table 5 *The chronological, mental ages and language scores of participants in Experiment 2*

	WS	Control
Chronological age*	29.6 (12.1)	13.1 (5.4)
Mental age	13.5 (5.4)	13.1 (5.3)
Language comprehension	101.5 (103.7)	98.6 (92.9)
Language production	55.6 (89.5)	34.5 (57.7)
Gestures	5.2 (3.5)	6.6 (3.2)
Games	17.9 (15.5)	19.5 (14.7)

*p = <0.05

Procedure

The background measures were administered in each child's home as part of Experiment 1. The pointing study took place in a curtained room measuring 5.17 m × 3.17 m. The child sat in a high chair in the middle of one wall. In front of the child, in a semi-circular formation, were 6 remotely controlled dolls 0.43 m high on stands 1.20 m tall. Two video cameras were positioned 2.25 m apart between dolls 1 and 2 and between dolls 5 and 6. A chair was placed either side of the child to enable the experimenter to sit on either the left or the right-hand side of the child. The layout of the room is shown in Figure 1.

A second experimenter sat in an adjacent room and watched the child on a split screen video. A light box behind the child indicated which doll was active. This second experimenter activated the dolls from this adjacent room. In all but one of the cases (a control toddler who sat on the mother's lap), the parent was not present in the testing room but watched the procedure on the video screen with the second experimenter.

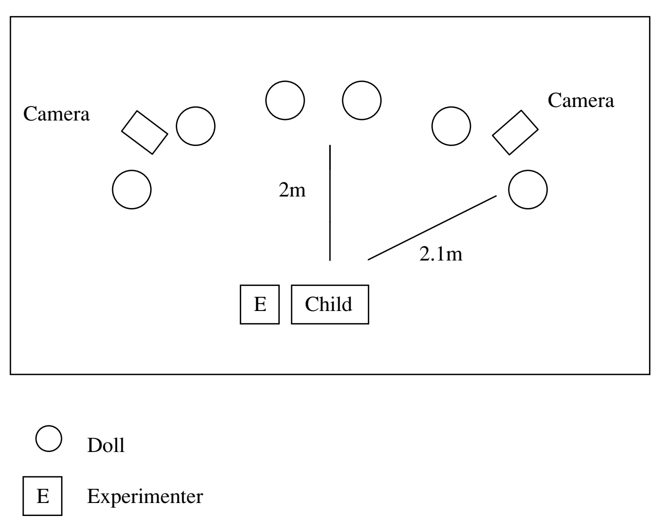

Figure 1 *The room layout in Experiment 2.*

The experiment consisted of 2 conditions: a set of production trials and a set of comprehension trials. There were 6 trials in each condition. The production and comprehension trials were alternated, with half of the participants starting with a production trial and half starting with a comprehension trial. The position of the experimenter was also counter-balanced: for one set of 6 trials (3 production and 3 comprehension) the experimenter sat on the left of the child, and in the other half the experimenter sat on the right of the child. Each of the trials was preceded by a bell presented at the child's midline.

In the production trials the experimenter stared straight ahead. The doll was activated and its arms and legs moved in a repetitive cycle lasting 7 seconds. In this condition, the doll also squeaked and 'talked' to get the child's attention. The experimenter was instructed to respond to the child, but not to lead their direction of gaze in any way. In the comprehension trials, the experimenter pointed to one of the dolls using an index finger point. This pointing was done either with the left or right hand, depending on which doll was activated in each item, with the pointing never crossing the body. The doll was only activated (without sound effects in the comprehension condition) when the second experimenter could see on the video screen that the child had turned his or her head to follow the experimenter's point.

The videotapes of this interaction were examined and the following behaviours coded.

Behaviours coded in the production trial

Index pointing: Both arm and index finger were extended in a conventional pointing posture.

Arm pointing: Arm was extended towards the target while the hand assumed various postures (e.g. hand held with all fingers extended, or fingers tightly clenched). These gestures lack the pointing index finger posture, but they have sometimes been considered to be functionally equivalent to pointing (Lock, Young, Service & Chandler, 1990).

Behaviours coded in the comprehension trials

Following of pointing: Head was turned towards the doll to which the experimenter was pointing.

Pointing: The number of child's index finger points produced after the experimenter had stopped pointing were recorded during the comprehension trials. These points were distinguished from children's points that occurred at the same time as the experimenter's, as these may have been purely imitative in nature.

Behaviours coded across both trial types

Contingent gesture: Included gestures such as hand banging, clapping, etc. which were judged to be in response to, or contingent upon, the movement of the dolls.

Social referencing: Child looked at the particular doll that was moving, then looked at the experimenter and then returned gaze to the doll. This sequence could occur while the doll was still moving or immediately after the doll had stopped moving.

The reliability of the coding scheme was assessed. A random selection of four of the tapes was fully coded by a second, independent person. A series of Mann-Whitney U tests revealed no significant differences between the two coders.

Results

The mean frequency of pointing (and standard deviations) in each of the categories is shown in Table 6. As previously mentioned, it is frequently the case that atypical groups are not normally distributed and display large standard deviations. Because of this, the raw scores were subjected to logarithmic transformation before being used in multivariate analyses of variance. These analyses showed that the groups differed in terms of index finger pointing ($F(1, 20) = 4.4$, $p < 0.05$) and in terms of ability to follow the experimenter's point ($F(1, 20) = 84.1$, $p = 0.05$). All other group differences failed to reach significance. The children with WS produced very few points and were less able to follow the experimenter's point than the group of typically developing children. These findings confirm those of Experiment 1.

As in Experiment 1, the groups did not differ in the amount of socially referential behaviour they produced. However, while there was a relationship between social referencing and pointing for the control group ($r = 0.63$, $p = 0.06$), there was no such relationship for the WS

Table 6 *Performance on the production and comprehension trials*

	WS group	Control group
Index finger pointing*	0.91 (2.4)	1.7 (2.2)
Arm pointing	0.18 (0.40)	0 (0)
Follows point*	4.1 (1.6)	5.3 (1.0)
Pointing in comp. trial	0.45 (0.93)	0.73 (1.2)
Contingent gesture	0.55 (0.69)	0.73 (1.6)
Social reference	2.3 (2.8)	2.3 (2.7)
Vocalization	2.0 (2.2)	2.7 (2.2)
Total index points[a]	0.73 (1.3)	2.5 (3.2)

*$p = <0.05$
Note: [a] = total number of points across production and comprehension trials.

group. This lack of relationship is interesting but unsurprising given that most of the children in the WS group did not produce any pointing.

The relationship between the comprehension of pointing and social referencing was significant for the control group ($r = 0.70$, $p < 0.05$) but not for the WS group. This finding supports that of Experiment 1 which demonstrated that socially referential behaviour was related to language skill for the control group but not for the WS group.

The relationship between amount of pointing, comprehension of pointing and the MacArthur language measures was also examined. None of the correlations between these variables was significant for either group.

Discussion

The findings of Experiment 2 further support those of Experiment 1 in showing that children with WS produce pointing significantly less than typically developing children of the same mental age, and also show a tendency to follow pointing less than controls. Furthermore, the lack of pointing behaviour was observed in an experimentally controlled situation in which typically developing children and children with DS have been shown to display pointing behaviour. While few of the children with WS produced even a single point across all trials, all were already producing referential language. This experimental finding provides support for Mervis *et al.*'s observations (1999) that children with WS use referential language before pointing referentially, a pattern not displayed in typical or DS development. Our experimental study showed that children with WS also had problems with the comprehension of pointing. However, unlike the Mervis *et al.* (1999) observations of WS toddlers, it was not the case that our children never followed pointing. But they did so significantly less frequently than the control group.

As in Experiment 1, the children with WS produced a similar amount of social referencing as the control group. Again, however, it appears not to have the same function. While social referencing behaviour was significantly related to comprehension of pointing for the control group, it was not for the WS group. This finding provides further support for the suggestion that social referencing is deviant in function for children with WS.

Mervis *et al.* (1999) argue that because the children with WS in their study had problems with both the comprehension and production of pointing, their lack of pointing cannot be explained by problems of gross motor control. However, it could be caused by problems of fine motor control because evidence closely relates the latter to the onset of pointing. Furthermore, the laterality of fine motor movements has been linked to both pointing and language development (Butterworth & Morissette, 1996). It therefore remains possible that poor fine motor control contributes to both limited pointing and delayed language in children with WS. Experiment 3 investigated this possibility.

Experiment 3

Introduction

Experiments 1 and 2 considered two important precursors to language development: joint attention and pointing. Experiment 3 investigated a third, related precursor. As mentioned above, a close relationship has been demonstrated between the development of manual and linguistic skills (Molfese & Betz, 1986). In a longitudinal study, Butterworth and Morissette (1996) showed that fine motor control preceded pointing. None of the infants in their study who could point lacked the pincer grip. While the Butterworth and Morissette study demonstrated a relationship between the pincer grip and pointing, the incidence of the pincer grip did not directly predict language production or comprehension. This suggests that the acquisition of fine motor skills *per se* is not sufficient alone to explain the beginnings of speech production but it may contribute to it.

In a subsequent study, Butterworth, Verweij and Hopkins (1997) demonstrated that children as young as 6–8 months produce both precision and power grips, together with a developmental trend towards increased use of the precision grip. If there were a relationship between fine motor and linguistic skills in typically developing children, how would this relationship manifest itself in children with WS? It is therefore important to consider whether the poor use of gesture among children with WS is related to impaired fine motor skills.

Experiment 3 aimed to investigate the extent to which the lack of pointing behaviour in Williams syndrome could be attributed to poor fine motor control.

Method

Participants

The same participants that had taken part in Experiment 1 also took part in Experiment 3. For all participants Experiment 3 was presented directly after Experiment 1. The mental and chronological ages of the participants are shown in Table 1.

Procedure

A test for the development of the precision grip, similar to that devised by Butterworth and Morissette (1996), was administered. The child was seated in a high chair in front of a specially designed semi-circular high-chair tray 26 cm wide. Two cubes and 2 spheres, 0.5 cm and 1 cm edge/diameter, were mounted securely on string. The string was threaded through a hole 16 cm from the inner edge of the semi-circular tray. Each object was presented in random order at the midline once, to give 4 trials in all. Each trial lasted 30 seconds. The child was videotaped during this interaction. The number of precision grips was coded according to the classification schemes devised by Gesell and Halverson (1936) and Touwen (1976). Precision grips were those in which the object is grasped between the index finger and the thumb, either at the tip, or before the last articulation of either or both the finger and thumb. Other grip types were not separately classified, but the hand used for all grasping of objects was noted. In order to check the reliability of the coding scheme, a random selection of 4 of the videotapes of the WS group and 4 of the control group was re-coded by a second person. The agreement for the WS group was 84% and for the control group 80%. A series of Mann-Whitney U tests revealed no significant differences between the two coders.

Results

The WS group produced fewer total grips (regardless of type) across all 4 trials, yielding 20.2 (SD = 5.5) grips as compared to the control group's 25.4 (SD = 13.2). This difference was not, however, significant ($F(1, 25) = 1.69$, $p = 0.21$). The proportion of these grips that were precision grips was higher for the WS group (50.3%; SD = 14.6) than for the control group (32.2%; SD = 20.1). This difference was significant ($F(1, 25) = 6.9$, $p < 0.05$), suggesting that, relative to typically developing children at a similar level of overall cognitive development, the children with WS had no problem producing precision grips.

Discussion

The results of Experiment 3 point to some interesting trends and lay the foundation for an important avenue to be explored in future studies. First, despite their lack of pointing, children with WS did produce precision grips. In fact, they produced more precision grips than the control group. Yet, Butterworth and Morissette's (1996) study of normal children demonstrated that precision grips were already in the repertoire of children when they began to point. The present study indicates that the relationship is not the same for the children with WS. While the WS group produced a higher proportion of precision grips than the control group, they rarely displayed pointing behaviour. This suggests that the lack of pointing shown by the children with WS is not caused by any of the motor components of the task. These data also raise doubts as to the universality of the timing of the relationship between the development of the pincer grip and the onset of pointing.

General discussion

The results of this series of experiments reveal intriguing differences in pre-verbal communication skills of young children with Williams syndrome. Particularly striking was the marked difference between controls and clinical group in terms of dyadic and triadic interaction, with the WS children displaying significant impairment in the latter. Experiment 1 showed that children with WS both initiate and respond to joint attention bids less than typically developing children at the same level of general cognitive ability. Further, the WS group produced both fewer instrumental and declarative gestures. Experiment 2 demonstrated that children with WS produce less pointing behaviour and fail to understand its referential function. The lack of pointing is not, as Experiment 3 indicated, the result of the motoric demands of the gesture. The fact that the children with WS were impaired in various other aspects of joint attention suggests that the lack of pointing must be considered within a general deficit in non-verbal communication rather than as a separate, unrelated impairment. Importantly, our experiments indicate that despite significantly less pointing, the WS children were already using considerable referential language, supporting experimentally Mervis *et al.*'s (1999) observational data that the developmental relations between these behaviours are different in WS.

Compared to the control group, children with WS also produced less instrumental gestures, reached for toys less often and used a different pattern of eye contact, implying less desire to obtain objects. These findings, together with the relatively good performance of the WS group on the social interaction scale, provide experimental support for Bertrand *et al.* (1993) who argued that young children with WS are more interested in people than in objects.

Not only was the frequency of behaviours observed in the children with WS different, but relations between non-verbal communication behaviours and language also differed. In Experiment 1, although the WS group produced more behaviours in the dyadic social interaction category, these did not relate to other aspects of joint attention in the same way as they did for the typically developing children. Similarly, in both Experiments 1 and 2, social referencing behaviour was differentially related to language for the two groups, with a stronger relationship obtaining for the control group than for the WS group. These findings suggest that in spite of similar levels of dyadic social interaction in the two groups, these skills may be deviant in function in Williams syndrome.

Some might think that the findings for the participants with Williams syndrome could be explained away in terms of visual processing difficulties. While it is true that some youngsters with WS have visual problems, all the participants with WS tested had normal or corrected to normal eyesight. Furthermore, they all oriented visually to the dolls when they started to move. The difference between the clinical group and the controls lies not in visually locating test items, but that they did not accompany their visual orientation with pointing or with social referencing to the adult in the room with them.

The finding that similar overt behaviours like dyadic interaction may differ in function across normal and atypical development is important. It suggests that equivalent behaviours may result from different brain processes (Karmiloff-Smith, 1998). For example, in studies of face processing, individuals with WS have been shown to reach behavioural scores equivalent to those of normal controls. However, they solve face processing tasks via different cognitive processes (Karmiloff-Smith, 1997; Grice, Spratling, Karmiloff-Smith *et al.*, 2001). The present series of experiments suggests that many aspects of non-linguistic communication differ in function for the WS toddlers compared to the normal controls. It is thus crucial to explore alternative developmental pathways, instead of focusing on behavioural outcome alone (Karmiloff-Smith, 1998).

The finding that the WS group had relatively good dyadic social interaction skills can usefully be related to the results of Tager-Flusberg and Sullivan (2000) with older children and adults with WS. These authors argue for two distinct components of social communication: a socio-cognitive component and a socio-perceptual component. The socio-cognitive component refers to the representational understanding of minds, is related to language acquisition and is thought to be dependent on pre-frontal cortex. The socio-perceptual component is considered less related to other cognitive and language abilities, appears earlier than the socio-cognitive component and is thought to be dependent on the limbic system, particularly the amygdala. In a series of tasks related to theory of mind, Tager-Flusberg and Sullivan demonstrated that children with WS are impaired on the socio-cognitive but not on the socio-perceptual component. The present study's focus on the distinction between dyadic and triadic interaction also suggests that the socio-perceptual component (rooted in dyadic interaction) is a relative strength in WS, whereas those aspects of joint attention which require representational skill are particularly problematic for this clinical group. Interestingly, Franco and Butterworth (1996) suggest that pointing is dependent on social representational skills and that it is this that underpins the relationship between language development and pointing found in typical development. The lack of such a relationship in WS gives support for this hypothesis.

The results of the present study provide important clues as to the possible causes of delay in language onset in children with WS. Impairments in triadic joint attention may slow down language acquisition by making it harder to establish reference (Baldwin, 1991). What remains unclear is whether children with WS establish reference by alternative means or whether their language production is simply less referential initially. The latter would suggest that early on toddlers with WS are merely repeating phonological wholes with less attention to meaning. Moreover, if children with WS are indeed less interested in objects, then caregivers would have fewer opportunities to follow their child's direction of interest. Yet it is the latter that turns out to be more successful for learning labels than relying on adult-directed labelling (Tomasello & Farrar, 1986).

The present set of experiments has demonstrated experimentally for the first time that despite relatively good language skills and superficially good social skills in later childhood and adulthood, young children with WS are impaired in several aspects of early communication that are normally related to language. Our series of experiments show that, in particular, two aspects of prelinguistic development – pointing and triadic joint attention – are impaired in toddlers with WS and are likely to contribute to the delay in their language. A third contributor resides in impairments in their speech segmentation abilities (Nazzi, Paterson & Karmiloff-Smith, 2002). Future research will chart processes of hemispheric lateralization in this clinical group, as this might be a fourth contributor to the serious delay in WS language onset. Furthermore, the roots of the WS focus on dyadic interaction as opposed to triadic interaction could lie in their tendency for sticky fixation and their impaired eye movement planning, compared to both DS and normal controls (Brown, Johnson, Paterson, Gilmore, Gsödl & Karmiloff-Smith, 2002). Of particular interest is the fact that our results provide a different explanation for the language delay in Williams syndrome, compared to the similar level of delay seen in Down's syndrome or in autism.

Our study has provided further experimental evidence that the language acquisition of children with WS may follow an atypical developmental trajectory, in that relations between referential language and gesture are different from those observed in typical development. Future research will ascertain whether reference is established by an alternative route or whether language acquisition is initially less referential in this clinical group. Finally, our experiments suggest that although some aspects of dyadic social interaction are relatively good in WS, the function of these social skills and the development of triadic joint attention differ from those observed in typical development. It is time to bury the myth still frequently found in the literature (e.g. Pinker, 1999) that Williams syndrome is a prime example of intact language and intact social skills.

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