

*Running headline: Functional play and language in Williams syndrome*

**NON-VERBAL COMMUNICATION, FUNCTIONAL PLAY AND LANGUAGE IN  
GREEK TODDLERS WITH WILLIAMS SYNDROME**

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**Abstract**

This study investigated the relationship between non-verbal communicative abilities, functional play and receptive and expressive language in 11 toddlers with Williams syndrome during spontaneous communication. Results demonstrated that children with Williams syndrome did not differ from typically developing children in vocabulary size. Nevertheless, children with Williams syndrome showed deficiencies in functional play, language comprehension and language production. Moreover, in the Williams syndrome group measures of vocabulary size, language comprehension, language production and functional play were not inter-correlated. These findings are interpreted through the theory of intersubjectivity, which suggests that the ability to generate and convey meanings as well as to use objects functionally, are based on innate motives for cooperation and mutual understanding.

## **Introduction**

Williams syndrome (also Williams-Beuren syndrome) is a genetic disorder caused by the deletion of about 25 genes from the region q11.23 of chromosome 7 (Hillier et al, 2003). Earlier studies report that Williams syndrome occurs in about 1 in 20,000 live births (Beuren, 1972; Greenberg, 1990). However, more recently it is estimated that the prevalence of Williams syndrome is higher, occurring in about 1 in 7,500 (Strømme, Bjørnstad, & Ramstad, 2002). Phenotypically Williams syndrome shows a specific physical, cognitive and social profile. In the physical domain Williams syndrome is characterised by facial dysmorphology, supravalvular aortic stenosis, and transient hypercalcaemia (Morris, 2006). In the cognitive domain Williams syndrome is characterised by mild to moderate mental retardation as well as deficiencies in visuo-spatial cognition, planning and problem solving, as well as numbering skills, but good verbal short-term memory and a particular interest in music (Arnold, Yule, & Martin, 1985; Bellugi, Bihrlé, Jernigan, Tranner, & Deherty, 1990; Levitin et al., 2004). There also appears to be a higher prevalence of left-handedness (van Strien et al., 2005). In the social domain Williams syndrome is characterised by overfriendliness with strangers, unusual cheerfulness, intense interest in others' faces, even from a very early age, but also unpredictable negative outbursts (Mervis & Klein-Tasman, 2000; Udwin & Yule, 1991).

However, the most striking finding about individuals with Williams syndrome is that their performance in assessments of lexical and grammatical abilities is higher than would be expected for the degree of mental retardation. This pattern held for both comprehension and production (Bellugi, Sabo, & Vaid, 1988; Volterra et al., 2003; Robinson, Mervis, & Robinson, 2003).

Despite relative strengths in certain domains of language and social interaction, individuals with Williams syndrome show particular difficulties with pragmatic abilities such as turn-taking, conversational and topic maintenance (Mervis, 2006; Volterra et al., 1995), and appropriate use of eye gaze (Mervis et al., 2003), while they have great difficulty in understanding the difference between lies and jokes (Sullivan, Winner, & Tager-Flusberg, 2003). The language of individuals with Williams syndrome has often been described as "cocktail party" speech, which is filled with stereotypic social phrases and little content, and is not well tuned to their conversational partner (Mervis, 2006; Stojanovik, 2006; Udwin & Yule, 1991).

These findings imply that individuals with Williams syndrome may show deficits in communicative abilities related to the conscious sharing of motives, intentions and emotions about topics in the surrounding environment with another person. These abilities are described as Secondary Intersubjectivity (Trevarthen & Hubley, 1978). Secondary intersubjectivity is a species specific process, which appears around 9 months and is based on the developing awareness that the other is a source of new ideas concerning objects, an intentional agent, and a carrier of feelings. Before nine months infants perceive interactions with others from their own perspective. However, after nine months infants become capable of perceiving interactions from the other's perspective as well. The primary manifestation of Secondary Intersubjectivity is joint attention (Hubley & Trevarthen, 1979; Trevarthen & Hubley, 1978). Joint attention refers to the mutual focus of attention and communication between the child and another person about a target external to the space of the dyad. Joint attention marks a shift from the primarily dyadic interactions between the infant and another person about emotions. Infants of this age attempt to direct the other's attention towards a topic of their own interest by using gestures such as pointing or showing, or by alternating their gaze between the communicative partner and an object. Moreover, infants now follow the other's focus of attention by looking where the other is looking, imitating what the other is doing with an object, or following arbitrary verbal and vocal instructions.

Toddlers' ability to coordinate their intentions and emotions about a topic in the environment with those of another person develop further during the second year and are revealed in functional play (Trevvarthen, 1994). Functional play is the primary manifestation of pretend play and is defined as the use of an object or the association of two or more objects in a conventional manner (Ungerer & Sigman, 1981). Functional play emerges as infants move from indiscriminant stereotypical play to relating objects in a non functional idiosyncratic manner to using objects in an appropriate way (Bigelow, McLean, & Proctor, 2004). The meaningful use of objects depends heavily on an extensive understanding of shared reality, which is understood because it is shared. At this age comprehension of what the mother says about or does with objects is much more evident than it was a few months before, but it is dependent on the interpersonal context. In other words, children's understanding of the meanings carried in linguistic utterances as well as of the way objects function are abut on a richer conception which has been accomplished through mutually coordinated interactions with significant others in the previous months (Trevvarthen, 1994).

A number of studies in typical and atypical populations have well documented that joint attention facilitates language development. Tomasello and Farrar (1986) showed that inside, as opposed to outside, episodes of joint attention children produced more utterances, more words and more words referring to objects. Moreover, inside joint attention episodes maternal references to objects that were already the child's focus of attention were positively correlated with the child's vocabulary. It has also been shown that joint attention measures contribute to multiple regression equations predicting individual differences in language development in normally developing children (Mundy, Fox, & Card, 2003) as well as in children with Down syndrome (Mundy, Kasari, Sigman, & Ruskin, 1995; Yoder & Warren, 2004). Mundy and Gomes (1998) argued that aspects of joint attention are related differentially to expressive and receptive language. In particular, in typically developing children initiating joint attention is a significant predictor of expressive language, while responding to joint attention is related to both expressive and receptive language. Similarly, in children with autism the ability to respond to the joint attention bids of others was positively correlated with receptive language scores and mean length of utterance, while better joint attention skills were associated with greater language development (Bono, Daley, & Sigman, 2004; Murray et al. 2008). As regards the relation between joint attention and functional play, Bigelow and her colleagues (2004) observed that typically developing infants displayed more functional play during joint attention episodes with the mother compared to outside joint attention episodes. Functional play within joint attention, but not outside joint attention, correlated with functional play when alone. The authors conclude that mothers' ability to scaffold infants' activities within joint attention may facilitate infants' advanced play.

Moreover, there is abundant evidence of parallel developments in play and language which are explained from a Piagetian perspective as deriving from a common underlying capacity for cognitive representation. However, there are relatively few studies focusing on the relation between functional play, preverbal communicative abilities and language development. Lewis and her colleagues (Lewis, Boucher, Lupton, & Watson, 2000) examined 40 typically developing children aged 1 to 6 years and found significant correlations between functional play and expressive language, but not with receptive language. The authors argue that both functional acts on particular objects (e.g. pretending to give Teddy a biscuit) and the understanding and production of the corresponding words (e.g. 'Teddy', 'biscuit', 'eating') require the formation of relevant concepts. A significant relationship between expressive language and functional play has also been identified in children with Down syndrome (Cunningham, Glenn, Wilkinson, & Sloper, 1985).

Mundy and his colleagues (Mundy, Sigman, Kasari, & Yirmiya, 1988) investigated the relation between non verbal communicative abilities, play and language in children with

Down syndrome aged 18 – 48 months. The results demonstrated that in children with Down syndrome expressive and receptive language skills were significantly correlated with request behaviours, while in normally developing children expressive and receptive language skills were significantly correlated with indicative behaviours. Moreover, it was observed that children with Down syndrome exhibited significantly more functional play than did the normal group. Also, functional play was significantly correlated with indicating and requesting behaviours as well as with receptive language scores in children with Down syndrome, but not in the normal group. More recent studies also demonstrate that children with Down syndrome spent the same amount of total play time in functional play and perform the same kind of elaborated functional acts compared to typically developing children matched for general cognitive and language development (Wright, Lewis, & Collis, 2006).

The investigation of the relation between abilities appertaining to joint attention, functional play and language in typical and atypical populations may contribute to a more thorough understanding of the role of intersubjective communication in the generation and sharing of meanings. Nevertheless, there are very few studies addressing this issue in children with Williams syndrome. This line of research is based on the premise that Williams syndrome is a developmental disorder and thus the most appropriate research models should be developmental ones (Mervis, 2003).

Laing and her colleagues (2002) compared a group of young children with Williams syndrome (mean chronological age 31 months, mean mental age 14 months) with a mentally – matched group of typically developing infants and toddlers. Children's social skills were assessed from video-tapes of child – experimenter interactions in experimental situations in the laboratory, during which verbal communication was kept at a minimum, using the Early Social Communication Scales (ESCS) (Mundy & Hogan, 1996). The ESCS is based on a cognitive developmental model which assumes that successful interactional skills depend on adaptive capacities that entail successful accommodation and assimilation to the social environment. This model derives from the cognitive developmental theories of Piaget and Werner and Kaplan. The results of this study showed that children with Williams syndrome produced less pointing and less behaviours included in the category Initiating Requesting (e.g. combining eye contact with a reach, pointing to indicate desired object). Moreover, while the Williams syndrome group produced more behaviours from the Social Interaction category than the control group (e.g. eye contact after tickle, turn-taking with a ball and a car), these behaviours were not well integrated with other aspects of joint attention. The authors argue that the behaviour of the Williams syndrome group was predominantly dyadic, whereas the behaviour of the control group was predominantly triadic.

Klein-Tasman and her colleagues (Klein-Tasman, Mervis, Lord, & Phillips, 2006) investigated the socio-communicative skills of 29 children with Williams syndrome aged 30 to 63 months (mean age 41.59 months) who had very limited language abilities. The average age equivalent ranged from 12 to 35.5 months (mean 22.9 months). Socio-communicative skills were assessed using the Autism Diagnostic Observation Schedule (ADOS Module 1) (Lord, Rutter, DiLavore, & Risi, 1999) on the basis of recent findings demonstrating that the types of communicative problems observed in Williams syndrome overlap with the difficulties exhibited in autism spectrum disorders (Mervis & Becerra, 2007). The ADOS assesses difficulties in the domains of verbal and non verbal communication, integration of various communicative behaviours, social reciprocity, functional play, creativity and imagination. Children are observed during play-like interactions with an examiner where predetermined materials are used in structured tasks (e.g. a pretend birthday party). Results demonstrated that children with Williams syndrome show abnormalities in Pointing, Giving, Showing and Eye Contact. Also, individuals with Williams syndrome very commonly showed abnormalities in their functional play behaviour. Initiation and Response to Joint Attention

showed some abnormality, but for less than half of the participants, while very few participants showed any abnormality in Shared Enjoyment or Requesting.

Taking into account the above considerations the present study aimed to examine the relation between joint attention, functional play and language abilities during spontaneous interactions in toddlers with Williams syndrome, in comparison with mentally matched typically developing children.

## Methods

### *Participants*

In the present study participated 11 children with Williams syndrome, 6 boys and 5 girls, and 11 mental-age matched typically developing toddlers 6 boys and 5 girls. The children with Williams syndrome were recruited from the Genetic Syndromes Association, a Greek-based parent support group, and had been diagnosed clinically as well as by means of the fluorescence in situ hybridization (FISH) genetic test for deletion of the elastin gene. All participants came from middle class Greek speaking families. Mothers' age in the Williams syndrome group ranged from 29 – 48 years (mean age 36,6 years), while mothers' age in the typically developing group ranged from 29 – 38 years (mean age 34.4 years). The vast majority of mothers in both groups had received Technological or University education.

None of the participants exhibited severe sensory or motor deficiencies or had been hospitalised within the previous 6 months. Among children with Williams syndrome 8 attended a mainstream private preschool, 1 child attended mainstream public preschool, 1 child attended mainstream primary school, and 1 child did not attend school. The majority (64%) of children with Williams syndrome received early intervention services by a speech therapist and an occupational therapist approximately 3 times a week. None of the participants in the control group attended any preschool. Written parental permission was attained before the children's participation in the study.

### *Materials*

Measures were obtained on children's expressive and receptive language, vocabulary production, communicative abilities and play performance. Participants' mental ages were assessed using the Cognitive Scale of the Bayley Scales of Infant and Toddler Development (Bayley III) (Bayley, 2006). This scale includes items which measure perception, memory, concept grouping, information processing, number concepts and counting, and problem solving in children aged 1 to 42 months. Age equivalent data are provided. Studies with clinical groups, such as children with Down syndrome or developmental delay, provide evidence for good discriminate validity of the Bayley III (Bayley, 2006). Chronological and mental ages of the participants are presented in Table 1.

**Table 1:** The chronological and mental ages of the participants

	WS	TD
<b>Chronological age (in months)***</b>		
Mean	67.4	26.5
Range	38 – 89	16 – 39
Sd	16.4	7.6
<b>Mental age (in months)</b>		
Mean	29.4	25.2
range	18 – 50	14 – 36
Sd	9.6	6.7

\*\*\*p<0.001

Expressive and receptive language were assessed using the Mullen Scales of Early Learning (MSEL) (Mullen, 1995). This test is administered in infants and preschool children from birth to 68 months and is commonly used in studies of toddlers and young preschoolers with developmental disorders, Williams syndrome included (Klein-Tasman et al., 2006). Receptive Language Scale measures the child's understanding of spoken language, knowledge of propositional and spatial concepts, ability to follow oral instructions, auditory short and long-term memory, auditory organization sequencing, auditory-visual memory, retrieval of facts, and general knowledge. Failure in the Receptive Language Scale often is due to inability to derive linguistic meaning from language. Expressive Language Scale assesses productive use of language i.e. the child's ability to use speech to communicate and express ideas, vocabulary, abstract thinking and reasoning, auditory short and long term memory, and comprehension of auditory information. In scoring not only it is marked whether a child has passed an item or not, but also the degree of her performance in that item, providing thus a more detailed and accurate account of the child's developmental level in expressive and receptive language.

Productive vocabulary was measured using the Language Development Survey (LDS) (Rescorla, 1989). The LDS is a checklist consisting of 310 words arranged into 14 semantic categories namely: food, toys, outdoors, animals, body parts, places, actions, household, personal, people, clothes, vehicles, modifiers and others. The LDS assesses spontaneous word production in children aged 18 to 35 months. The LDS has good concurrent validity with object and picture naming on various standardized instruments (Rescorla, 1989; Rescorla, Hadicke-Wiley, & Escarce, 1993; Rescorla & Alley, 2001). Although some of the participants were too old to be scored in this form, it was decided for comparison purposes that all participants should receive the same form. Mothers were asked to mark on the list the words their child says spontaneously, even if they are pronounced in an idiosyncratic way. The form provides also the opportunity for mothers to write in additional words, but these words were not included in any of the analyses described below.

Non-verbal communicative abilities and play performance were assessed from video recordings of spontaneous mother – child interactions in a semi-structured situation taking place at home.

### ***Procedure***

All the children were visited at their home three times during a month. Home environment was considerate to be more appropriate for eliciting a representative sample of the child's spontaneous behaviours, compared to the laboratory setting (Papaeliou, Minadakis, & Cavouras, 2002). In the first visit each child was administered the Bayley III Cognitive Scale, while the mother completed the LDS as well as a questionnaire on demographic information and the child's medical background. In the second and third visit children were administered the Receptive and Expressive Language Scales of the MSEL and were videorecorded while playing with their mother in a semi-structured situation with toys provided by the researcher. The set of toys included two different sized dolls, doll furniture, a tea set, a telephone, a brush and a mirror, a school-bus with little people in it, blocks, toy animals, a book, and a wind-up mechanical toy. Mothers were asked to play with their child as they would normally do introducing all the toys provided. Each play session lasted approximately 30 minutes. This process yielded a total of 1 hour videorecording for each child.

The coding scheme for the behaviour analysis was based on previous schemes (Laing et al., 2002; Papaeliou & Trevarthen, 2006) and was further expanded from an inductive analysis of the video recordings. Non-verbal communicative behaviours were grouped into three main

categories: Joint Attention (JA), Request (R), and Interpersonal (IP). Within the category Joint Attention there were the subcategories Joint Gaze and Communicative Gestures. Functional Play (FP) included the following acts: (a) conventional acts on toys directed to self e.g. brushing one's hair, (b) conventional acts on toys directed to mother e.g. holding telephone to the mother's ear, (c) conventional acts on toys directed to a doll e.g. placing a spoon to a doll's mouth, (d) conventional use of two or more objects in combination e.g. stirring a spoon in a pot. However, during spontaneous communication a particular behaviour may not be initiated by the child; rather, it may be directed by the communicative partner, usually the mother. In directive communicative exchanges mothers require verbally or non-verbally the child to attend to, or to produce an action, or to provide information (Kay & Chorney, 1980; Holtzman, 1972). Indeed it has been demonstrated that mothers of typically developing children very often use directives, in order to enhance the communicative performance of their child. Moreover, it has been shown that mothers of mentally retarded toddlers are more directive than the mothers of typically developing toddlers, while mentally retarded toddlers are more compliant than their non-retarded peers (Mahoney, Fors, & Wood, 1990; Roach, Barratt, Miller, & Leavitt, 1998; Henzlik & Stevenson, 1986). Thus, the behaviour of Following Mother's Interest, the Communicative Gestures as well as the behaviours included in the category Functional Play were further characterized as spontaneous, if they were initiated by the child, or as directed, if they were produced as a compliance to a mother's request. The coding scheme for non-verbal behaviours is described in Table 2.

Twenty minutes of continuous footage from each play session for each child were analysed and the frequency of each codified behaviour was noted down. The first five minutes of each session were not analysed so as to counter adaptation effects. Times where play was interrupted by irrelevant stimuli (e.g. bell ringing) or where it was difficult to see clearly what the mother and the child were doing, were excluded from the analysis.

**Table 2:** Description of the coding scheme for non-verbal behaviours.

Categories/ Subcategories & Behaviours	Description of behaviours
<b>JOINT ATTENTION</b>	
<b>Joint gaze</b>	
Converging Looking	Child and mother look at the same toy
Alternating Gaze	Child alternates gaze between mother and a toy
Eye Contact with Toy	Child makes eye contact while manipulating a toy
Following Interest	The infant looks at the same direction with the mother or at the direction indicated by the mother's pointing, giving or showing
<b>Communicative gestures</b>	
Pointing	Infant's index finger is extended in direction of a toy
Showing	Infant holds out a toy to mother, but does not permit her to take it
Giving	Infant puts a toy into mother's hand and permits her to take it
<b>REQUEST</b>	
Searching for Mother with Toy	Child looks at mother while she looks away as he/she manipulates a toy
Requesting Toy	Child points to a toy that is out of reach
Requesting Action with Toy	Child shows or gives a toy to the mother that he/she cannot use properly
<b>INTERPERSONAL</b>	
Eye Contact	Child makes eye contact. No toy manipulation
Teasing Mother	Child teases mother
Responding to Teasing	Child responds positively to mother's teasing

## Results

As it is frequently the case with atypical groups, the data showed large standard deviations. Therefore, group differences were calculated using the non-parametric test Mann-Whitney U. Results demonstrated that children with Williams syndrome did not differ significantly from typically developing children in Vocabulary Production. However, children with Williams

syndrome received significantly lower scores in Expressive Language and Receptive Language measures compared to the control group (Table 3).

**Table 3:** Performance on measures of Expressive Language, Receptive Language, & Vocabulary Production.

	WS	TD
<b>Expressive Language**</b>		
Mean	33.6	55.7
Sd	5.5	12.0
Range	20 – 39	34 – 67
<b>Receptive Language* *</b>		
Mean	28.4	52.9
Sd	6.9	9.9
Range	24 – 41	41 – 75
<b>Vocabulary Production</b>		
Mean	227.3	200.7
Sd	109.1	116.4
Range	15 – 310	2 – 310

\* p<0.05, \*\*p<0.01

The mean performance and standard deviations on each non-verbal communicative and functional play behaviour are presented in Table 4. According to the findings, children with Williams syndrome did not differ significantly from typically developing children in the overall category Joint Attention ( $p=0.82$ ). However, contrarily to typically developing children, children with Williams syndrome did not exhibit any alternating gaze at all. The two groups did not differ either in any of the other behaviours included in the subcategory Joint Gaze or in the overall subcategory. One interesting finding was that children with Williams syndrome exhibited significantly more Directed Showing, while they also received significantly higher scores in the overall subcategory Directed Communicative Gestures, compared to typically developing children ( $p=0.03$  and  $p=0.04$  respectively). Children with Williams syndrome did not differ significantly from the typically developing children either in any of the other behaviours included in the subcategory Communicative Gestures or in the overall subcategory.

As regards the overall category Functional Play, it was demonstrated that children with Williams syndrome received significantly lower scores compared to typically developing children ( $p=0.05$ ). Moreover, it was found that children with Williams syndrome exhibited significantly less Spontaneous Functional Play than typically developing children ( $p=0.03$ ). On the other hand, no significant differences were observed between the two groups in the subcategory Directed Functional Play ( $p=0.76$ ).

On the other hand, children with Williams syndrome exhibited significantly more positive reactions to mothers' teasing compared to typically developing children ( $p=0.04$ ). In addition, the clinical group received higher score than the control group in the overall category Interpersonal, although this difference was marginally significant ( $p=0.08$ ). The group differences in the category Child Teases Mother were not significant ( $p=0.19$ ). Also, children with Williams syndrome did not differ significantly from typically developing children either on any of the behaviours included in the category Request or in the overall category. Correlations between measures of language development, play performance and non-verbal communication were also calculated for each group using the non-parametric Spearman rho coefficient. Among the typically developing children expressive language skills showed significant positive correlations with receptive language skills ( $r=0.74$ ,  $p<0.05$ ) and

vocabulary production ( $r=0.66$ ,  $p<0.05$ ). The correlation between receptive language skills and vocabulary production was not significant in this group ( $r=0.42$ ,  $p=0.23$ ).

**Table 4:** Performance on non-verbal communicative behaviours and functional play

	WS		TD	
	mean	sd	mean	sd
<b>JOINT ATTENTION</b>	41.7	2.8	40.0	5.7
<b>Joint gaze</b>	35.7	1.9	34.8	5.5
Converging Looking	14.9	1.4	17.2	3.1
Alternating Gaze	0.0	0.0	0.5	1.4
Eye Contact with Toy	7.1	1.9	6.9	2.9
Spontaneous Following Interest	11.1	2.1	8.0	5.5
Directed Following Interest	2.6	1.4	2.1	1.7
<b>Communicative gestures</b>	5.9	2.5	5.2	3.4
Spontaneous Communicative Gestures	4.6	2.3	4.7	2.9
Directed Communicative Gestures*	1.4	1.1	0.5	0.8
Spontaneous Pointing	1.9	1.1	2.5	1.5
Directed Pointing	0.3	0.4	0.1	0.1
Spontaneous Showing	1.5	1.1	1.4	1.7
Directed Showing*	0.4	0.5	0.1	0.1
Spontaneous Giving	1.2	0.9	0.7	0.7
Directed Giving	0.7	0.4	0.4	0.7
<b>FUNCTIONAL PLAY*</b>	9.7	2.7	13.7	6.1
Spontaneous Functional play*	6.1	1.7	9.1	3.2
Directed Functional play	3.5	1.6	4.7	3.2
<b>REQUEST</b>	2.5	1.6	1.7	1.7
Request for Toy	2.1	1.1	1.6	1.8
Request for Action with Toy	1.9	0.9	1.4	1.8
Searching for Mother with Toy	0.4	0.8	0.1	0.3
<b>INTERPERSONAL</b>	0.7	0.6	0.4	0.8
Eye Contact	0.0	0.0	0.0	0.0
Teasing Mother	0.2	0.3	0.3	0.8
Responding to Teasing*	0.5	0.4	0.1	0.3

\* $p<0.05$ , \*\* $p<0.01$

Correlations of language measures with functional play and non-verbal communicative behaviours for the control group are presented in Table 5. As it is observed, expressive language skills showed significant positive correlation with Directed Functional Play ( $p<0.05$ ). The overall category Functional Play and the sub-category Spontaneous Functional Play did not correlate with any of the language measures, i.e. Expressive Language ( $p=0.20$  and  $p=0.33$  respectively), Receptive Language ( $p=0.26$  and  $p=0.13$  respectively) and Vocabulary Production ( $p=0.63$  and  $p=0.63$  respectively). A striking finding was that in typically developing children of this age there was a significant negative correlation between Joint Attention and Receptive Language ( $p=0.02$ ). Moreover, relatively high but non-significant negative correlations were observed between Joint Attention and Expressive Language ( $p=0.07$ ), Joint Attention and Vocabulary Production ( $p=0.07$ ), Joint Gaze and Receptive Language ( $p=0.07$ ), Directed Communicative Gestures and Expressive Language ( $p=0.06$ ), and Directed Communicative Gestures and Receptive Language ( $p=0.08$ ). It seems that around the age of 2 years typically developing children rely more on language rather than non-verbal behaviours for achieving cooperative communication.

**Table 5:** Correlations of language measures with functional play and non-verbal communicative behaviours for the control group.

	Expressive language	Receptive language	Vocabulary production
Functional play	0.44	0.39	0.18
Spontaneous Functional play	0.35	0.51	0.22
Directed Functional play	0.65*	0.46	0.35
Joint attention	-0.59	-0.72*	-0.60
Joint gaze	-0.41	-0.60	-0.55
Communicative gestures	-0.43	-0.09	-0.20
Spontaneous Communicative Gestures	-0.39	-0.03	-0.20
Directed Communicative Gestures	-0.61	-0.58	-0.21
Request	0.03	0.38	0.21
Interpersonal	0.29	0.07	0.25

\*p<0.05

Among children with Williams syndrome the correlations between Expressive Language, Receptive Language, and Vocabulary Production were not significant ( $r=-0.01$ ,  $p=0.98$ ,  $r=0.12$ ,  $p=0.74$  and  $r=0.23$ ,  $p=0.52$ ). Correlations of language measures with functional play and non-verbal communicative behaviours for the Williams syndrome group are presented in Table 6. According to the findings, the overall category Functional Play showed significant negative correlation with Vocabulary Production ( $p<0.05$ ). None of the other measures of functional play and non-verbal communication was correlated with any of the language measures i.e. Expressive Language, Receptive Language, or Vocabulary Production.

**Table 6:** Correlations of language measures with functional play and non-verbal communicative behaviours for the Williams syndrome group.

	Expressive language	Receptive language	Vocabulary production
Functional play	-0.05	0.07	-0.64*
Spontaneous Functional play	-0.27	-0.39	-0.42
Directed Functional play	0.07	0.45	-0.55
Joint attention	0.51	-0.25	0.37
Joint gaze	-0.04	-0.37	-0.20
Communicative gestures	0.45	-0.34	0.32
Spontaneous Communicative Gestures	-0.42	-0.24	-0.41
Directed Communicative Gestures	0.15	-0.09	0.20
Request	-0.35	0.39	-0.32
Interpersonal	0.35	0.04	-0.17

\*p<0.05

## Discussion

The present study revealed that toddlers with Williams syndrome with mental age around 2 years show intriguing differences from typically developing children in functional play. In particular, the clinical group exhibited less conventional acts with toys towards the self, the mother, a doll, or another toy during spontaneous communication, compared to the control group. Similarly, Klein-Tasman et al. (2006) demonstrated that toddlers with Williams syndrome usually show deficiencies in functional play. On the other hand, children with Williams syndrome in this sample did not differ from typically developing toddlers in behaviours related to joint attention, such as joint gaze or communicative gestures. This finding contrasts with findings of other studies which demonstrate that children with Williams syndrome with mental age around one year initiate and respond spontaneously to joint attention bids less than typically developing children at the same level of general cognitive ability (Laing et al, 2002; Mervis & Bertrand, 1997). This discrepancy between the results of different studies may be explained by differences between the samples in mental age.

Children with Williams syndrome of mental age around one year may not be able to produce age appropriate behaviours, such as joint attention. On the other hand, children with Williams syndrome of mental age around 2 years may be more capable of producing joint attention behaviours corresponding to a younger age, but they exhibit deficiencies in age appropriate cooperative behaviours, such as functional play. Nevertheless, this hypothesis deserves further investigation by a longitudinal study which would examine the development of cooperative behaviours in children with Williams syndrome.

A striking finding of the present study was that children with Williams syndrome demonstrate more communicative gestures such as pointing, showing or giving under their parents' guidance, compared to typically developing children. This result implies that parents of children with Williams syndrome may intuitively recognise both the critical importance of episodes of joint attention in language acquisition and their children's deficiency in this domain, and they attempt to elicit relevant behaviours from their children. Mervis and Bertrand (1997) also observed that parents of children with Williams syndrome direct episodes of joint attention by using the children's eye gaze to determine what they are looking at or by deliberately placing an object in front of the children's eyes before labelling it for them. Thus, it seems that in toddlers with Williams syndrome the early stages of lexical development are more dependent on adults determining the contexts in which new words are acquired, compared to typically developing toddlers.

Also it was demonstrated that although children with Williams syndrome did not differ from typically developing children in vocabulary size, they received lower scores in measures of language comprehension and language production. Also, in contrast to the control group, the vocabulary size was not integrated with language comprehension and language production. This finding implies that while children with Williams syndrome acquire an adequate number of words, they do not seem to be able to use them productively in order to create meanings and convey verbally thoughts, intentions and feelings to a communicative partner.

Moreover, in accordance with other work (Laing et al., 2002), the present study showed that children with Williams syndrome have relatively good performance in behaviours related to dyadic social interactions. This result supports further the argument that children with Williams syndrome are more interested in people than in objects (Bertrand, Mervis, Rice, & Adamson, 1993).

The finding that children with Williams syndrome have relatively good skills for interpersonal interactions but they show deficiencies in cooperative interactions has been explained by the model of Theory of Mind (ToM) proposed by Tager-Flusberg and Sullivan (2000). According to these authors, social intelligence comprises of two distinct components: a socio-cognitive component and a socio-perceptual component. The socio-cognitive component incorporates what has traditionally been referred to as ToM and it entails reasoning about the content of others' minds, it is closely related to language acquisition, it is thought to be dependent on the prefrontal cortex, and it begins to emerge around the age of 3 years. The socio-perceptual component is considered as less related to other cognitive and language abilities and more closely connected to emotions and intentions, it appears earlier and is thought to be dependent on the limbic system, particularly the amygdala and associated regions of the medial temporal cortex. Although several theorists argue that the social-cognitive component of ToM builds on social-perceptual knowledge (Baron-Cohen, 1994; Hobson, 1993), it is strongly emphasized that the two components of social intelligence depend on distinct underlying neuro-cognitive mechanisms and have a different developmental time course (Tager-Flusberg & Sullivan, 2000).

In contrast to the ToM, the Theory of Intersubjectivity provides empirical support to the view that there is a continuity from the prelinguistic to the linguistic period in the development of the infant's ability to understand the other's emotional reactions, intentions, interests and thoughts. The infant's ability to understand the other's emotionality changes from a simple interest in expression at birth to a sensitivity to the reciprocity of emotions at 2 months to a more complex management of affects at 6 months and then at 9 months, to a more pronounced interest in exploring specific emotional reactions and relating them to their targets. Similarly, an understanding of others' communicative intentions is changing from a recognition of communicativeness and its absence or appropriateness at 2 months, to a recognition of invitations to games at 6 months, and a recognition of commands and prohibitions at 9 months (Reddy, 1999; Trevarthen, 1994). At this age an infant shows a new readiness to tune in with the intentions and interests of a partner in joint exploration and use of objects. This ability forms the basis for a creative imagination of roles, actions and "tools" that are arbitrary or symbolic. This peculiarly human intelligence prepared for learning cooperatively culture and language, depends on affection for familiar people and equally on a discriminating caution or timidity for strangers (Trevarthen, 1994). In support of this view the present study demonstrated that in typically developing children, but not in children with Williams syndrome, certain aspects of functional play are integrated with expressive language abilities.

Data obtained from neuro-anatomical studies also advocate the Theory of Intersubjectivity. These studies provide evidence that in typically developing individuals the expression of coherent emotions in coordination with the expressions of other persons is regulated by the Intrinsic Motive Formation (IMF), which involves the brain stem, the basal ganglia, and limbic structures. The IMF is present at birth (Trevarthen & Aitken, 1994). The later maturing of neocortical circuits, which regulate conscious activity emerge in reciprocal, dynamic and increasing involvement with IMF. In particular, the integration of emotional expressions with sensory processing, motivation, intention, attention, learning, planned and purposeful action as well as understanding of the other's state of mind is mediated through robust bidirectional pathways between the structures involved in the IMF and the amygdala as well as the temporal and frontal cortex, especially the orbitofrontal cortex and the temporal mesocortex (Bush, Luu, & Posner, 2000; Morris et al., 1998; Trevarthen & Aitken, 1994).

On the other hand, the picture that emerges if one synthesizes the data from the present and previous studies illustrate that children with Williams syndrome exhibit deficiencies in all forms of communication. Specifically, the extreme social interest observed in this group even before the first year, seems quite inappropriate in both quality and quantity (Bertrand et al., 1993; Mervis et al., 2003). As regards cooperative communication, it is well documented that infants with Williams syndrome are delayed in the onset of joint attention (Laing et al., 2002; Mervis & Bertrand, 1997), while the present study revealed that toddlers with Williams syndrome show marked deficiencies in functional play. At the neuroanatomical level individuals with Williams syndrome show abnormalities in neural systems that underlie emotional sharing, cooperative communication and language development. In particular, they have disproportionately large volumes and increased gray matter density in the amygdala and the orbital and medial pre-frontal cortex. The neuroanatomical abnormalities observed in individuals with Williams syndrome may explain the deficits exhibited by this group in cooperative communication with the others as well as the creation and sharing of meanings (Reiss et al., 2004).

In order words and phrases to be meaningful to more than one person, the things they refer to must already be shared. In other words, meanings for things and phenomena of the world are created and acquired during interpersonal exchanges which are modulated by dynamic affects and expressions of interests, intentions and feelings (Reddy, 1999; Trevarthen, 1994). The

deficiencies exhibited by individuals with Williams syndrome in cooperative understanding which are reflected in their poor functional play, may explain the difficulty of this group in using language productively. In the same vein, it has been observed that children with autism, whose major problem focuses on deficits in social communication, face severe problems in language use, while they spent significantly less time in functional play and their functional acts were less varied, less elaborated and less integrated compared with those of the control group (Williams, Reddy, & Costall, 2001).

The finding that children with Williams syndrome acquire a large number of words and syntactic structures, which they cannot use productively, while they are able to respond in adults' initiatives of creating episodes of joint attention, is of particular importance for speech therapists and other specialists. Intervention programs for children with Williams syndrome should put particular emphasis not only in the acquisition of linguistic structures, but also in their functional use.

## References

- Arnold, R., Yule, W., & Martin, N. (1985). The psychological characteristics of infantile hypercalcaemia: a preliminary investigation. *Developmental Medicine and Child Neurology*, 27, 49-59.
- Baron-Cohen, S. (1994). How to build a baby that can read minds: cognitive mechanisms in mindreading. *Current Psychology of Cognition*, 13, 513-552.
- Bellugi, U., Bihlre, A., Jernigan, T., Trauner, D., & Doherty, S. (1990). Neuropsychological, neurological and neuroanatomical profile of Williams syndrome. *American Journal of Medical Genetics, Supplement*, 6, 115-125.
- Bellugi, U., Sabo, H., & Vaid, J. (1988). Spatial deficits in children with Williams syndrome. In J. Stiles-Davis, M. Kritchevsky, & U. Bellugi, *Spatial cognition: brain bases and development* (pp. 273-298). Hillsdale, NJ: Lawrence Erlbaum Associates.
- Bertrand, J., Mervis, C.B., Rice, C., & Adamson, L. (1993). *Development of joint attention by a toddler with Williams syndrome*. Gatlinburg Conference on Research and Theory in Mental Retardation and Developmental Disabilities., Gatlinburg, TN.
- Beuren, A.J. (1972). Supravalvular aortic stenosis: a complex syndrome with and without mental retardation. *Birth Defects*, 8, 45-46.
- Bigelow, A.E., MacLean, K., & Proctor, J. (2004). The role of joint attention in the development of infants' play with objects. *Developmental Science*, 7(5), 518-526.
- Bono, M.A., Daley, T., & Sigman, M. (2004). Relations among joint attention, amount of intervention, and language gain in autism. *Journal of Autism and Developmental Disorders*, 34(5), 495-505.
- Bush, G., Luu, P., & Posner, M.I. (2000). Cognitive and emotional influences in anterior cingulate cortex. *Trends in Cognitive Science*, 4, 215-222.
- Cunningham, C.C., Glenn, S.M., Wilkinson, P., & Sloper, P. (1985). Mental ability, symbolic play and receptive and expressive language of young children with Down syndrome. *Journal of Child Psychology and Psychiatry*, 26, 255-265.
- Hillier, L.W., Fulton, R.S., Fulton, L.A. et al. (2003). The DNA sequence of chromosome 7. *Nature*, 424, 157-164.
- Greenberg, E. (1990). Introduction to special issue on Williams syndrome. *American Journal of Medical Genetics Supplement*, 6, 85-88.
- Hobson, R.P. (1993). Understanding persons: the role of affect. In S. Baron-Cohen, H. Tager Flusberg, & D.J. Cohen (eds.), *Understanding other minds: perspectives from autism* (pp. 204-227). Oxford: Oxford University Press.
- Hubley, P., & Trevarthen, C. (1979). Sharing a task in infancy. In I.C. Uzgiris (ed.), *Social interaction during infancy* (pp. 38-52). San Francisco: Jossey-Bass.
- Klein-Tasman, B., Merivs, C.B., Lord, C., & Phillips, K.D. (2006). Socio-communicative deficits in children with Williams syndrome: Performance on the Autism Diagnostic Observation Schedule. *Child Neuropsychology*, 13(5), 444-467.
- Laing, E., Butterworth, G., Ansari, D., Gsödl, Longhi, E., Panagiotaki, G., Paterson, S., & Karmiloff-Smith, A. (2002). Atypical development of language and social communication in toddlers with Williams syndrome. *Developmental Science*, 5(2), 233-246.

- Levitin, D.J., Cole, K., Chiles, M., Lai, Z., Lincoln, A., & Bellugi, U. (2005). Characterizing the musical phenotype in individuals with Williams syndrome. *Child Neurology*, 10(4), 223-247.
- Lewis, V., Boucher, J., Lupton, L., & Watson, S. (2000). Relationships between symbolic play, functional play, verbal and non-verbal ability in young children. *International Journal of Language and Communication Disorders*, 35(1), 117-127.
- Mahoney, G., Fors, S., & Wood, S. (1990). Maternal directive behaviour revised. *American Journal of Mental Retardation*, 94(4), 398-406.
- Mervis, C.B. (2003). Williams syndrome: 15 years of psychological research. *Developmental Neuropsychology*, 23(1&2), 1-12.
- Mervis, C.B. (2006). Language abilities in Williams-Beuren syndrome. In C.A. Morris, H.M. Lenhoff, P.P. Wang (eds.), *Williams-Beuren syndrome: Research, evaluation and treatment* (pp. 3-17). Baltimore, MD: John Hopkins University Press.
- Mervis, C.B., & Becerra, A.M. (2007). Language and communicative development in Williams syndrome. *Mental Retardation and Developmental Disabilities Research Reviews*, 13, 3-15.
- Mervis, C.B., & Bertrand, J. (1997). Relations between cognition and language: A developmental perspective. In L.B. Adamson & M.A. Romski (eds.), *Research on communication and language disorders: Contributions to theories of language development* (pp. 75-106). New York: Brookes.
- Mervis, C.B., & Klein-Tasman, B.P. (2000). Williams syndrome: cognition, personality, and adaptive behaviour. *Mental Retardation and Developmental Disabilities Research Reviews*, 6, 148-158.
- Mervis, C.B., Morris, C.A., Klein-Tasman, B.P., Bertrand, J. Kwitny, S., Appelbaum, L., & Rice, C.E. (2003). Attentional characteristics of infants and toddlers with Williams syndrome during triadic interactions. *Developmental Neuropsychology*, 23 (1-2), 243-268.
- Morris, C.A. (2006). The dysmorphology, genetics and natural history of Williams-Beuren syndrome. In C.A. Morris, H.M. Lenhoff, P.P. Wang (eds.), *Williams-Beuren syndrome: Research, evaluation and treatment* (pp. 3-17). Baltimore, MD: John Hopkins University Press.
- Morris, J.S., Friston, K.J., Buchel, C., Frith, C.D., Young, A.W., Calder, A.J., & Dolan, R.J. (1998). A neuromodulatory role for the human amygdale in processing emotional facial expressions. *Brain*, 121, 47-57.
- Mundy, P., Fox, N., & Card, J. (2003). EEG coherence, joint attention and language development in the second year. *Developmental Science*, 6(1), 48-54.
- Mundy, P., & Gomes, A. (1998). Individual differences in joint attention skill development in the second year. *Infant Behaviour and Development*, 21(3), 469-482.
- Mundy, P., & Hogan, A. (1996). *A preliminary manual for the abridged Early Social Communication Scales (ESCS)*.
- Mundy, P., Kasari, C., Sigman, M., & Ruskin, E. (1995). Non verbal communication and early language acquisition in children with Down syndrome and in normally developing children. *Journal of Speech and Hearing Research*, 38(1), 157-167.

- Mundy, P., Sigman, M., Kasari, C., & Yirmiya, N. (1988). Non verbal communication skills in Down syndrome children. *Child Development*, 59, 235-249.
- Murray, D.S., Creeghead, N.A., Manning-Courtney, P., Shear, P.K., Bean, J., Prendeville, J-A. (2008). The relationship between joint attention and language in children with autism spectrum disorders. *Focus on Autism and Other Developmental Disabilities*, 23(1), 5-14.
- Papaeliou, C., Minadakis, G., & Cavouras, D. (2002). Acoustic patterns of infant vocalizations expressing emotions and communicative functions. *Journal of Speech, Language and Hearing Research*, 45(2), 311-317.
- Papaeliou, C., & Trevarthen, C. (2006). Pre-linguistic pitch patterns expressing 'communication; and 'apprehension'. *Journal of Child Language*, 33(1), 163-178.
- Reddy, V. (1999). Prelinguistic communication. In M. Barrett (ed.), *The development of language* (pp. 25-50). Hove: Psychology Press.
- Reiss, A.L., Eckert, M.A., Rose, F.E., Karchemskiy, A., Kesler, S., Chang, M., Reynolds, M.F., Kwon, H., & Galaburda, A. (2004). An experiment of nature: Brain anatomy parallels cognition and behavior in Williams syndrome. *The Journal of Neuroscience*, 24(21), 5009-5015.
- Rescorla, L. (1989). The Language Development Survey: A screening tool for delayed language in toddlers. *Journal of Speech and Hearing Disorders*, 54(4), 587-599.
- Rescorla, L., & Alley, A. (2001). Validation of the Language Development Survey (LDS): A parent report tool for identifying language delay in toddlers. *Journal of Speech, Language, and Hearing Research*, 44(3), 598-609.
- Rescorla, L., Hadicke-Wiley, M., & Escree, E. (1993). Epidemiological investigation of expressive language delay at age two. *First Language*, 13(37), 5-22.
- Roach, M.A., Barratt, M.S., Miller, J.F., & Leavitt, L.A. (1998). The structure of mother-child play: Young children with Down syndrome and typically developing children. *Developmental Psychology*, 34(1), 77-87.
- Robinson, B.F., Mervis, C.B., & Robinson, B.W. (2003). Roles of verbal short-term memory and working memory in the acquisition of grammar by children with Williams syndrome. *Developmental Neuropsychology*, 23, 13-31.
- Stojanovik, V. (2006). Social interaction deficits and conversational inadequacy in Williams syndrome. *Journal of Neurolinguistics*, 19, 157-173.
- Strømme, P., Bjørnstad, P.G., & Ramstad, K. (2002). Prevalence estimation of Williams syndrome. *Journal of Child Neurology*, 17, 269-271.
- Sullivan, K., Winner, E., & Tager-Flusberg, H. (2003). Can adolescents with Williams syndrome tell the difference between lies and jokes? *Developmental Neuropsychology*, 23(1-2), 85-103.
- Tager-Flusberg, H., Sullivan, K. (2000). A componential view of theory of mind: evidence from Williams syndrome. *Cognition*, 76, 59-89.

- Tomasello, M., & Farrar, J. (1986). Joint attention and early language. *Child Development*, 57, 1454-1463.
- Trevarthen, C. (1994). Infant semiosis. In W. Nöth (ed.), *Origins of semiosis* (pp. 219-252). Berlin: Mouton de Gruyter.
- Trevarthen, C., & Atiken, K. (1994). Brain development, infant communication and empathy disorders: intrinsic factors in child mental health. *Development and Psychopathology*, 6, 597-633.
- Trevarthen, C. & Hubley, P. (1978). Secondary intersubjectivity: Confidence, confiding and acts of meaning in the first year. In A. Lock (ed), *Action, gesture and symbol : The emergence of language*. London: Academic Press.
- Udwin, O., & Yule, W. (1991). A cognitive and behavioral phenotype in Williams syndrome. *Journal of Clinical and Experimental Neuropsychology*, 13, 232, 244.
- Ungerer, J.A., & Sigman, M. (1981). Symbolic play and language comprehension in autistic children. *Journal of the American Academy of Child Psychiatry*, 20, 318-337.
- Van Strien, J.W., Haselen, G.C., Van Hagen, J.M., De Coo, I.F.M., Frens, M.A., & Van Der Geest, J.N. (2005). Increased prevalence of left-handedness and left-eye sighting dominance in individuals with Williams-Beuren syndrome. *Journal of Clinical and Experimental Neuropsychology*, 27(8), 967-976.
- Volterra, V., Sabbadini, L., Capirci, O., Pezzini, G., & Ossella, T. (1995). Language development in Italian children with Williams syndrome. *Genetics Counseling*, 6, 137-138.
- Volterra, V., Caselli, C.M., & Capirci, O, Tonucci, F., & Vicari, S. (2003). Early linguistic abilities of Italian children with Williams syndrome. *Developmental Neuropsychology*, 23(1-2), 33-58.
- Williams, E., Reddy, V., & Costall, A. (2001). Taking a closer look at functional play in children with autism. *Journal of Autism and Developmental Disorders*, 31(1), 67-77.
- Wright, I., Lewis, V., & Collis, G. (2006). Imitation and representational development in young children with Down syndrome. *British Journal of Developmental Psychology*, 24(2), 429-450.
- Yoder, P.J., & Warren, S.F. (2004). Early predictors of language in children with and without Down syndrome. *American Journal of Mental Retardation*, 109(4), 285-300.

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