

THE FRAGILE X SYNDROME : WHAT ABOUT THE DEFICIT IN THE PRAGMATIC COMPONENT OF LANGUAGE ?

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ABSTRACT

Language profile of fragile-X syndrome individuals looks like the one of Down syndrome individuals, except for phonological and pragmatic abilities. If the pragmatic aspect of language is relatively preserved in Down syndrome, it is one of the most impaired language components in fragile-X syndrome. Discourse of fragile X persons generally contains of repetitions, perseverations and stereotypies. These behaviors are well documented in the literature but, we do not know, for now, the reasons of these repetitions and perseverations. Are they the consequence of a real pragmatic disorder or the consequence of motor difficulties ? Another aspect of the pragmatic component of language remains almost unexplored in this pathology : the common ground management and the organization of the old and the new information in conversation. It is this last point that we want to explore. We decided to conduct a preliminary study with four fragile-X boys aged from 6 to 12 years-old. Groups of two boys work together on referential communication tasks. We proposed traditional experimental situations in a non-eye contact condition between participants. We compare the results of our subjects with those of typically developing children matched on the lexical age. Fragile-X boys are less efficient than typically developing children when the message to give or receive contains spatial terms or "ordinal" attributes. They also manage less efficiently with an incomplete message especially when it is given by an adult.

INTRODUCTION

Fragile-X syndrome (FXS) is considered to be the most common inherited cause of mental retardation. Recent molecular studies suggest a prevalence of one affected individuals per 4,000 in males and 1 per 8,000 in females (Turner, Webb, Wake, & Robinson, 1996). The syndrome is caused by an inherited abnormal repetition of a trinucleotide sequence (CGG) on the distal end of the long arm of the X chromosome creating a fragile site or a break at the bottom of the chromosome (Verkerk, Pierretti, Sutcliffe, et al., 1991). In non-affected individuals, there is about 5 to 50 CGG sequences. In affected individuals, there are more than 230 repetitions. This is termed a full mutation (Imbert & Mandel, 1995). Between normality and full mutation that causes mental retardation, one can find a third situation : the premutation. In the case of premutation, there is no intellectual disability but there is a high risk of expansion when it is passed to the next generation through a female (Imbert & Mandel, 1995). This genetic abnormality inhibits a gene called FMR-1 gene (Fragile-X Mental Retardation-1) from forming the FRMP (FRM1 protein) which is thought to be important for the brain development (Verhey, Bakker, de Graaf, et al., 1993; Hagerman, 1996; Oostra, 1996).

The phenotype of FXS males is characterized by a wide range of physical, behavioral, cognitive and language problems. The ***main physical characteristics*** include long face, proeminent ears, and craniofacial asymmetry which are subtle features in childhood becoming more pronounced with advancing age. One can also note soft and smooth skin, flat feet, hyperextensible finger joint which reflect loose connective tissue. Scoliosis are often reported. Finally about 80% of postpubertal boys and men present macroorchidism. The most often mentioned ***behavioral features*** are the limited attention spans, hyperactivity, oversensitivity to tactile, auditory, olfactory and visual stimuli. Moreover, FXS individuals generally avoid eye contact and present autistic-like stereotypies (e.g., hand flapping and hand biting). Many of their atypical behaviors look like those found in anxiety disorders, especially social anxiety (cf. Bregman, Leckman, & Ort, 1988). Unlike males with autism, FXS males are sensitive to facial cues of others (Simon & Finucane, 1996, Turk & Cornish, 1998). Some individuals may be initially wary or avoidant with unfamiliar people. In these cases, one can observe what is called the "classic fragile-X syndrome handshake" (Wolff, Gardner, Paccia, & Lappen, 1989) characterized by turning the trunk, extending the hand, averting the gaze, mumbling of greeting and returning the gaze after the other person looks away. Let us note that FXS individuals tend to become increasingly comfortable with unfamiliar persons over time. The more frequent socially avoidant behaviors observed in FXS males are gaze aversion, tactile

defensiveness, overt turning of the body during face-to-face social interactions. Belser and Sudhalter (2001) think that the fundamental underlying cause of the apparent social anxiety is a deficit of the autonomic nervous system that interferes with the ability to regulate arousal. Briefly, neuroimaging studies show smaller posterior cerebellar vermis sizes in FXS males than in normal individuals (Mostofski et al. 1998; Reiss et al., 1991, 1995). This structure is involved with processing sensory stimuli and modulating motor activity. It seems that anomalies in its size may be associated with the FXS males hyperactivity and hypersensitivity to tactile and auditory stimuli. Furthermore, the caudate nucleus is enlarged in FXS males (Reiss et al. 1995). It seems that the caudate nucleus volume is inversely correlated with IQ. It is involved with several frontal subcortical circuits. Interruptions or lesions in these circuits produce problems in executive functions, in regulating affect and motor functioning, and in the ability to inhibit responses and to respond flexibly to environmental cues. So, it seems that anomalies in the caudate nucleus may be associated with particular FXS features as verbal perseveration, stereotypies, hyperactivity and attention impairments, impulsivity, emotional lability, inflexibility, and difficulties with organization, planning and sequencing information. Hyperarousal and subsequent inadequate behavioral responses are more likely seen in social situations with demands to perform and with changes in routine. Some authors (Fish, 1992, and Hagerman, 1996) think that in demanding situations, autism features in FXS boys may be best associated with anxiety and hyperarousal and less with profound social indifference as it is the case in autism. On the *cognitive level*, one can observe IQs ranging from normal to moderate mental retardation. Mental retardation occurs in about 85% of males with full mutation. Young boys with FXS may have an IQ above 75 and only show a decline in IQ by late childhood or adolescence. Males with a methylated full mutation have a mean IQ of 41. For males with a mosaic pattern, the IQ is around 60. Finally, males with an unmethylated or partially methylated full mutation have IQ around 88. Females are generally cognitively less affected than males. They typically have the full mutation on one of their two X chromosomes and the unaffected chromosome moderates the effects of the mutation. About 50 to 70% of the females with the full mutation have IQs in the borderline or mentally retarded range. Affected FXS males show particular patterns of cognitive strengths and weakness. Theobald, Hay and Judge (1987) mention relative strengths in verbal skills and visual learning. But as we will see further, verbal skills are not so preserved as initially mentioned and some aspects of language are really part of FXS males weakness. FXS children seem to have particular difficulties in tasks involving sequential processing, or short term memory in the recall or

reproduction of items in serial or temporal order (Turk, Hagerman, Barnicoat, & McEvoy, 1994; Dykens, Hodapp & Finucane, 2000).

SPEECH, LANGUAGE AND COMMUNICATION IN FXS

Speech production seems to be problematic for many individuals with FXS. Some are non-specific and may be found in people without this disorder and some are claimed to be specific to this syndrome. Conversational speech is characterized by omissions, distortions and substitutions of consonants and vowels (Hanson, Jackson, & Hagerman, 1986; Newell, Sanborn, & Hagerman, 1983; Vikman, Niemi, & Ikonen, 1988). These errors reflect the simplification processes observed in typically developing children and in other genetic etiologies of mental retardation (e.g., Down syndrome, see Van Borsel, 1988 for more details). What is more particular to FXS is the variability in speaking rate (Dykens, Hodapp, & Leckman, 1994). Actually, one can frequently observe unpredictable shifts from rapid to slower rates (Hanson, Jackson, & Hagerman, 1986). This variability is seen across all levels of IQ and seems to be specific to FXS boys as it does not occur in other mentally retarded people (e.g. Down syndrome) matched on the age and cognitive levels (Wolf-Schein, Sudhalter, & Cohen, 1987). According to Belser and Sudhalter (2001), speech of FXS individuals differs from those without FXS in the production of a specific type of dysfluency. By the expression "dysfluent speech", the authors refer to speech in which one's articulatory flow has been disrupted. This disruptions can take the form of stuttering-like dysfluencies, such as repetitions of sounds, words or phrases, or of non-repetitive dysfluencies such as self-corrections, reformulations or false starts. Intelligibility of speech is also lower than what is expected regarding the chronological age.

On the *lexical level*, FXS males perform well below chronological age expectations on both receptive and expressive measures of vocabulary (Madison, George, & Moeschler, 1986). However, data are contradictory. Some authors (see Fryns et al., 1984) reveal an important lexical deficit and some others (see Paul et al., 1987) suggest relatively good receptive and expressive lexical levels. The differences between results of these researches may be due to the differences in assessment tools used and/or to the characteristics of the FXS participants. So, for now, it is not yet clear whether receptive and expressive vocabularies are impaired to the same degree in FXS males. Furthermore, some questions about the lexical development of FXS individuals remain unsolved : which strategies do they use for new words learning ? Which are the semantic and the lexico-grammatical categories the best mastered ?

The ***morphosyntactic level*** is also impaired in FXS and is far below chronological age expectations on both receptive and expressive measures (Dyckens et al., 1994; Schopmeyer & Lowe, 1992). Data from Paul et al. (1984, 1987) suggest that receptive morphosyntax is commensurate with nonverbal mental age. Results for expressive morphosyntax are less clear. According to Paul et al. (1984), there is a deficit in the morphosyntax of the conversational language relative to the nonverbal mental age. These data contrast with those of Madison et al. (1986) whose suggest that mean length of utterance (MLU) is equivalent or higher to mental age expectations. Once more, differences in the results may be due to variations in participants characteristics, small sample size and assessment tools.

The ***pragmatic component*** of language is clearly impaired in FXS males. As we already mentioned, perseveration, echolalia and poor eye contact (gaze aversion) are often noticed in conversation and social interaction context. *Perseveration* on a word, a phrase, a sentence or a topic are well documented in FXS males and differ from echolalia. Perseveration consist in self-repetition whereas echolalia consist in repetitions of linguistic contribution of other people. FXS males who do not meet the criteria for autism frequently engage in perseveration but never in echolalia. Furthermore, FXS males are more likely to produce perseveration than are non-FXS males with autism or non-FXS male with other etiologies of mental retardation (Ferrier, Bashir, Meryash, Johnston, & Wolff, 1991). These observations suggest that perseveration in FXS males differ from a general tendency to repeat any previous behavior (Abbeduto & Hagerman, 1997). The causes of these perseverations are not very clear. Four hypothesis are proposed. The first one is the deficient expressive morphosyntax hypothesis which suggests that perseverations represent a strategy for participating in conversation when a failure to master morphosyntax makes meaningful contributions impossible (Sudhalter, Scarborough, & Cohen, 1991). The second one, the word-retrieval deficit hypothesis, considers that perseveration is a strategy emerging from the need to talk in the face of an inability to find the words needed to express an intended message (Sudhalter, Maranion, & Brooks, 1992). The hyperarousal hypothesis, the third one, claims that perseverations are the consequence of the arousal induced by various classes of stimuli, especially those including interpersonal component (Cohen, 1995; Belser & Sudhalter, 2001). Finally, the fourth hypothesis relates perseveration to executive function deficits and frontal lobe dysfunction (Mazzocco, Pennington, & Hagerman, 1993). The first hypothesis is not supported by data whereas the other can separately explain several dimensions of FXS male behavioral phenotype (see Abbeduto & Hagerman for more details).

Conversational roles relating to the ability of the speaker to initiate, maintain, or appropriately change a conversation topic; to elicit new information; or to answer questions are also problematic in FXS males. Relatively to Down syndrome individuals and males with autism, FXS males are more apt to engage in two types of conversational behavior : "eliciting utterances" and "continuing utterances" (Ferrier et al., 1991). They also engage in many more instances of conversational turn-taking compared to children with autism (Sudhalter, Cohen, Silverman & Wolff-Schein, 1990).

If pragmatic and communication disorders in FXS are well documented, it is important to note several limitations of the researches. First of all, there is only few researches on communication and pragmatic development in FXS females; probably because of the few number of affected females. Returning to FXS males, assessments are almost exclusively done within the conversational context and with a limited range of partners. Furthermore, no complete description of the ability of FXS male to fulfill the requirements of the listener's role is available. Finally, there are few studies on the emergence of the communicative problems of FXS individuals over the course of development.

REFERENTIAL COMMUNICATION

One aspect of pragmatic functioning which is believed to be problematic for pragmatically impaired children is that of providing too much or too little information for adequate processing by the listener to be possible (Mc Tear, 1985; Bishop & Adams, 1989). Referential communication tasks provide a way of examining children's sensitivity to the needs of the listener in the communication, and thus of examining the child's potential for successful exchange of information. By the way, this kind of tasks also allow to get information on the child's ability to be a good listener by adjusting the completeness of information given to him by the speaker. These tasks have two characteristics which make them particularly valuable for looking at the pragmatically impaired child (Leinonen & Letts, 1997). Firstly, they incorporate aspects of realistic communication in the sense that one participant must make information available to another one who cannot obtain that information from any other source. Secondly, referential communication tasks are highly structured. The experimenter can control both the nature of the barrier between participants and the nature of the verbal response needed for successful completion of the task. Generally, referential communication tasks are sets of pictures (usually between four and eight) of a single referent that varied on a number of dimensions (for example, color, shape and/or size).

In referential communication tasks, the *efficient speaker* is able to discriminate the features that uniquely identify the object, to present these features in a coherent message and to exclude unhelpful redundant information. Furthermore, good speakers engage and maintain the attention of the listener and are able to modify their message if the listener indicates that he is unable to select the target-item or exhibit a failure of comprehension. The *efficient listener* is one who can, following an accurate message, give the appropriate response. When messages are not complete (for example, they refer to more than one item), it is interesting to observe how children deal with this inadequacy. Can they frame the type of questions that are ordinarily used in telling another that one is unable to act on the received message ?

Speaker skills typically develop before listener skills in contrast to the normal pattern for language development where comprehension is said to precede production (Lloyd, 1994). It is also important to note that the correlation between speaker and listener skills is not high. Thus, different capacities are involved in the speaker and the listener roles (Higgins, Fondacaro & McCann, 1981).

The present study aims to assess and describe the ability of FXS males to fulfill the speaker and the listener roles in a set of referential communication tasks with three kind of partners. We decided to use the classical experimental paradigm design by Glucksberg et al. (1966). The situation is build so that there is a physical barrier which prevents one participant from seeing materials that is available to the other and, more particularly, which prevent FXS subjects to see the interlocutor during the verbal exchange. We are aware that, as Longhurst and Berry (1975) claimed it, conversational situations in which participants cannot see the eyes and the face of their interlocutors are not frequent in everyday life. However, as we mentioned previously, FXS males tend to avoid eye contact (Bregman et al., 1988). In fact, FXS males are more aroused by the maintenance of eye contact than are males with other etiologies of MR (e.g. Down syndrome) or with attention deficit hyperactivity disorder (Belser & Sudhalter, 1995). One direct consequence of eye contact maintenance is the perturbation of verbal behavior. Furthermore, as we note it previously, FXS individuals are sensitive to facial cues (Simon & Finucane, 1996; Turk & Cornish, 1998). So, we can suppose that seeing approbation or disapprobation signs on the interlocutor's face will be disturbing and will also lead to perturbation of verbal behavior. Actually, greater rates of tangential and perseverative language are observed when the conversational partner of FXS males are looking directly at them compared to when their conversational partner are looking away from them (Belser & Sudhalter, 2001). So, in order to avoid tangential and perseverative language,

we decided to use Glucksberg et al. (1966) experimental design without eye contact instead of the one of Longhurst and berry (1975) with eye contact.

We suppose that : (1) in the speaker role, FXS boys will be less performant than typically developing (TD) children matched on the lexical age; (2) in the listener role, FXS boys will be the most performant with a "complete" interlocutor; in the case of an "incomplete" interlocutor, we suppose that performances will be higher when fronting an adult than when fronting another child.

PROCEDURE

Four French-speaking non-autistic FXS males aged from 10;6 years-old to 12;7 years-old participate to the study. They were matched on the basis of lexical and chronological age with mentally retarded children of other etiologies and on basis of the lexical age with typically developing children. Not FXS mentally retarded children only participate as interlocutors in this study. Their results and performance are not taken into account in the analysis. The lexical age was measured with the "Echelle de Vocabulaire en Images Peabody" (EVIP, French version of the PPVT-R, 1993).

	<i>Chronological age in years</i>	<i>Lexical age in years</i>
<i>FXS subject 1</i>	12;7	}
<i>Matched MR interlocutor</i>	10;4	6;6
<i>Matched TD child</i>	6;0	}
<i>FXS subject 2</i>	11;10	}
<i>Matched MR interlocutor</i>	11;5	5;10
<i>Matched TD child</i>	5;5	}
<i>FXS subject 3</i>	11;1	}
<i>Matched MR interlocutor</i>	14;1	4;2
<i>Matched TD child</i>	4;5	}
<i>FXS subject 4</i>	10;6	}
<i>Matched MR interlocutor</i>	10;10	6;0
<i>Matched TD child</i>	6;2	}

Table 2. Description of the participants.

MR : Mentally retarded, TD : Typically developing

Children were placed in four situations. In the first situation, the FXS child is facing another MR child (not FXS). He is in a speaker role and has to describe objects or pictures in order to make them identify by the interlocutor who does not see them. In the three next situations, he is in a listener role and receives information and instruction from another MR child (not FXS) or an adult. In the case of the adult speaker, two conditions are proposed. In the first one, the

adult gives automatically a complete and accurate message to the child. In the second one, the message given by the adult is voluntarily incomplete and need the child to ask more information to the adult in order to identify the target-item.

	<i>Speaker</i>	<i>Listener</i>
Situation 1	FXS child	OE child
	TD child 1	TD child 2
Situation 2	OE child	FXS child
	TD child 2	TD child 1
Situation 3	Adult (complete)	FXS child
		TD child
Situation 4	Adult incomplete	FXS child
		TD child

Table 3. Description of the four situations.

OE : Other etiology, TD : Typically developing

In the four situations describe above, the participants are placed in the same room. They are separated by a screen which prevent them to see each other faces (cf. Gulcksberg et al., 1966, classical experimental situation).

Six tasks were build. Each of them was proposed to children both in the speaker condition and in the three listener conditions. Of course, in order to avoid a bias due to the familiarization with target-items and distractors, four parallel forms of the material were created. The tasks were distributed in two groups : 1] building tasks, and 2] combination tasks.

A. Building tasks.

1. **Task 1 - Tower of pearls** : the subject has to build (in the listener situation) or to describe (in the speaker situation) a tower made of colored pearls. The speaker has in front of him four pearls strung on a wooden stick. The listener has in front of him the wooden stick and the pearls. The speaker has to describe precisely his tower in order to help the listener to build the same one.
2. **Task 2 - Tower of "Legos"**: the subject has to build (in the listener situation) or to describe (in the speaker situation) a tower made of "legos" blocks differing by the shape (rectangle or square) and/or the color (red, yellow, blue and green). The speaker's task is to give to the listener precise information concerning the sequence, the shape and the color of the blocks to use in order to build a correct tower. More than the necessity to identify the different features of the blocks, the difficulties of this

task are to give : (1) the correct sequence of blocks from the bottom to the top of the tower, and (2) the accurate position of a block relatively to the previous one (e.g., "you put the little red square block in the middle of the green rectangle block"). In order to succeed in the task, the listener has to identify the blocks described by the speaker and to put them in the right sequence and the right position. This last success condition requires that the listener is able to understand instructions containing spatial vocabulary.

B. Combination tasks.

3. ***Task 3, 4, 5 and 6 – Combinations*** : the subject has to find (in the listener situation) or to describe (in the speaker situation) a target-item which differ from distractors by the color, the shape, the size or by a combination of two of those criteria. In all cases, two elements are at least necessary to carry out an accurate description of the target-item. Four sub-tasks corresponding to four possible combinations of criteria are proposed :

- "shape + color",
- "shape + size",
- "size + color"
- "shape + size + color".

Before we started the tasks, we ensure that subjects accurately name the four colors (red, yellow, blue and green), the four shapes (square, circle, triangle and star) and the three sizes (small, medium and large).

Each FXS subject is randomly placed in the speaker situation and in the three listener situations. The experimentator precisely write down the productions of the participants. In the "incomplete adult" situation, the speaker's interventions were previously constructed in order that each listener receives precisely the same quality of information

RESULTS

Because of the few number of subjects, we decided to conduct non-parametric Kruskal-Wallis tests in order to compared the performances of the two groups. As we will observe, difference between groups is often near statistic significance but are not despite of important differences in the number of responses. It seems to us that with more important samples, difference will become significant.

1. FXS boys as speakers.

For this preliminary study, we decide to only study spontaneous sufficient messages given by the subjects. Messages that follow experimentator stimulations are not analyzed and taken into account here. In this view, four kinds of messages were analyzed :

- *Spontaneous sufficient messages* (SSM) : They contain all the information needed and generally lead to a correct response of the interlocutor.
- *Spontaneous insufficient messages* (SIM) : They do not contain enough information. The listener has to question the speaker in order to find the correct response.
- *Spontaneous non-informative message* (SNIM) : No pertinent information is given concerning the target-item to describe (e.g., in task 4a : "it's a puppet").
- *Spontaneous incorrect message* (INCM) : The information given is incorrect (e.g., it may concern a distractor).

The results mainly indicate that FXS boys are poor communicators. Compared to typically developing children, they give much more spontaneous insufficient messages.

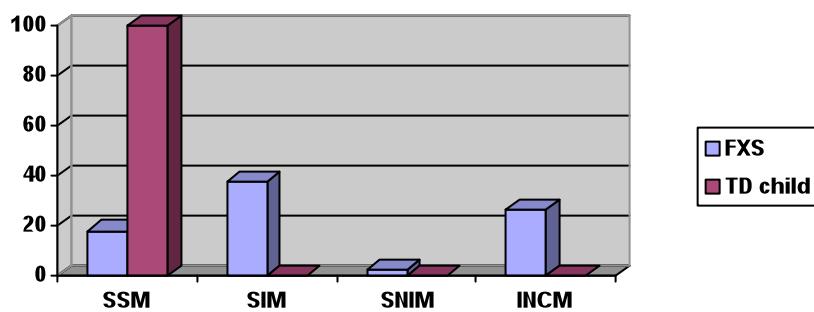


Figure 2. Global performances of FXS boys and TD children in the speaker condition.

SSM : Spontaneous Sufficient Message, SIM : Spontaneous Insufficient Message, Spontaneous non-informative Message, INCM : Spontaneous Incorrect Message.

We conducted a non-parametric Kruskal-Wallis test in order to compare the global performances of the two groups for the different kind of messages. TD children produce significantly more spontaneous sufficient messages than FXS boys [the means are respectively, 100% and 18.26 %; $H (1, 8) = 6.14$, $p < 0.05$]. As we will see, the superiority of TD children for producing spontaneous sufficient messages is present in all the tasks except in the first one (the tower of pearls). Generally, FXS boys produce significantly more spontaneous insufficient messages [FXS: 37.99% and TD children : 0.00%; $H (1, 8) = 6.05$, $p < 0.05$]. The differences between both groups are not statistically significant for spontaneous non-informative messages [FXS: 2.88% and TD children : 0.00%; $H (1, 8) = 2.28$, $p = 0.11$],

and spontaneous incorrect messages [FXS: 26,44% and TD children: 0.00 %; $H(1, 8) = 2.28$, $P=0.11$].

a) ***The tower of pearls*** : A total of 12 messages is necessary in order to accurately describe all the target-items. TD children produce the 12 expected spontaneous sufficient messages whereas FXS boys produce an average of 5.75 spontaneous sufficient messages and 4 incorrect messages. Despite this, there are no significant differences between the groups. The results of the Kruskal-Wallis tests are respectively $H(1, 8) = 3.15$ ($p=0.075$) and $H(1, 8) = 1.75$ ($p=0.19$). We can assume that with more subjects significant differences between groups will appear in the sufficient spontaneous message case.

b) ***Combinations*** : In the four sub-tasks, five messages are needed in order to describe the target items. TD children always produce the 5 expected sufficient messages while FXS boys produce a maximum of 1.25 accurate spontaneous sufficient messages (see Figure 3).

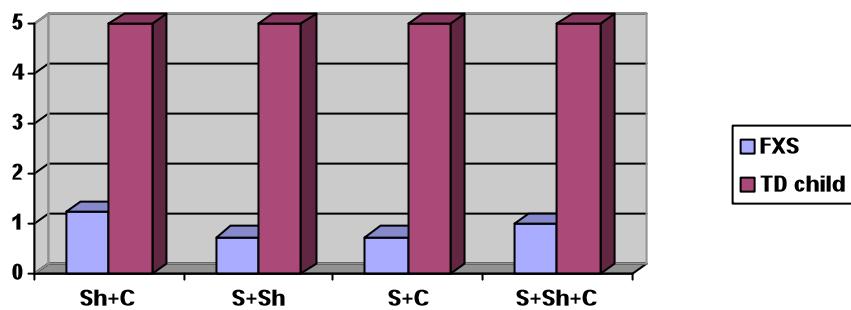


Figure 3. Task 2 : Combinations – Spontaneous sufficient messages produced by FXS boys and TD children.

In the first sub-task, FXS boys and TD children do not differ significantly in the production of spontaneous sufficient messages (SSM) whereas TD children produce significantly more SSM in the three other sub-tasks.

Sub-tasks	TD children mean	FXS boys mean	Kruskal-Wallis
1. Shape + Color	5.00	1.25	$H(1,8) = 3.75$, $p=0.066$
2. Size + Shape	5.00	0.75	$H(1,8) = 5.25$, $p<0.05$
3. Size + Color	5.00	0.75	$H(1,8) = 5.25$, $p<0.05$
4. Shape + Size + Color	5.00	1.00	$H(1,8) = 5.04$, $p<0.05$

Table 4. Spontaneous sufficient messages - Difference between TD children and FXS boys on the combination tasks.

Generally, FXS boys mainly produce spontaneous insufficient messages (SIM). Spontaneous non-informative messages (SNIM) and incorrect messages are not very frequent (INCM) (see Figure 4).

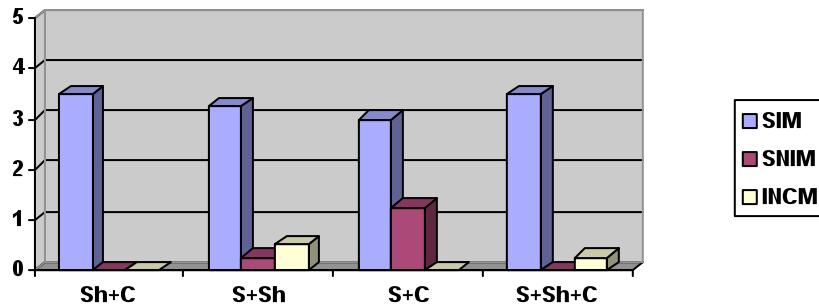


Figure 4. Task 2 : Combinations – FXS boys type of messages for the 4 sub-tasks.

In the sub-tasks "shape + color", "size + color" and "shape + size + color", FXS produce significantly more SIM than TDC children.

Sub-tasks	TD children mean	FXS boys mean	Kruskal-Wallis
1. Shape + Color	0.00	3.50	$H(1,8) = 4.85, p < 0.05$
2. Size + Shape	0.00	3.25	$H(1,8) = 4.94, p < 0.05$
3. Size + Color	0.00	3.00	$H(1,8) = 3.15, p = 0.076$
4. Shape + Size + Color	0.00	3.50	$H(1,8) = 4.85, p < 0.05$

Table 5. Spontaneous insufficient messages - Difference between TD children and FXS boys on the combination tasks.

As we note it before, SNIM and INCM are very rare and FXS boys do not differ from TD children on these levels.

c) **Legos** : A total of 20 messages is necessary in order to accurately describe the target-items. TD children give the 20 expected spontaneous sufficient messages whereas FXS boys do not produce any SSM. The difference between the groups is significant [$H(1,8) = 6.00, p < 0.04$]. Instead of SSM, FXS boys produce SIM (mean: 2.75) and INCM (mean: 9.00). Let's note than when we analyze the FXS subjects productions, we can observe the use of terms such as "big" and "small" instead of "rectangle" and "square". As we will see in the discussion, the use of the size (an ordinal dimension) instead of the shape (a nominal dimension) seems to have a negative influence on the performance.

2. FXS boys as listeners.

As we can observe on the following figure, FXS boys seem to be poor listeners but ... only in certain situations

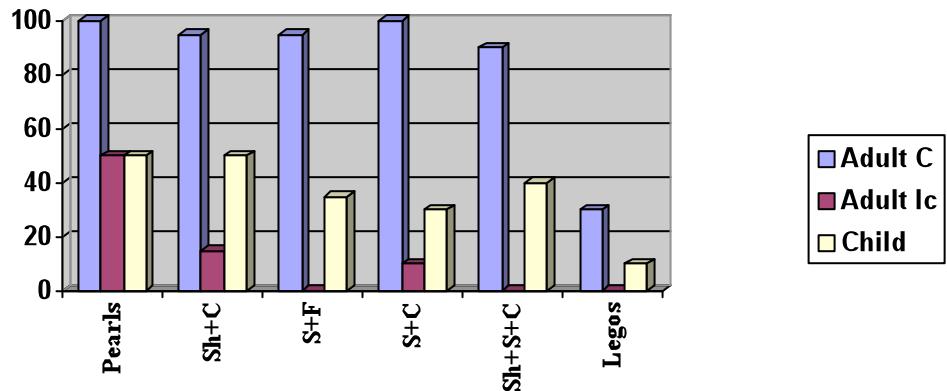


Figure 5. Performances of FXS boys in the three listener situations.

- a) **"Complete" adult** : In this situation, **FXS boys** tend to be very performant in decoding the verbal message as they give more than 80% of correct answers for the following tasks : tower of pearls (mean : 100% of correct responses); combinations (means of correct responses : shape + color : 95.00%, size + shape : 95.00%, size + color : 100%, shape + size + color : 90.00%). In the "legos" tasks, the mean percentage of success is far below 50% (mean of correct responses : 30.00%). Let's note that this task requires the correct understanding and manipulation of spatial concepts and spatial vocabulary. In the same situation, **TD children** succeed in all the tasks. The description made by the adult automatically lead to a correct response.
- b) **"Incomplete" adult** : As expected, the **FXS boys** performances decrease in this situation. But, surprisingly, we observe a dramatic decrease of the performance. Except in the "tower of pearls" task (mean : 50% of correct responses) and in the "shape + color" (mean : 15% of correct responses) and in the "size + color" sub-tasks (mean : 10% of correct responses), FXS boys fail in all the tasks. In the same situation, **TD children** performance decrease in the following comparatively to the previous situation : the tower of pearl task (100% of correct responses in the complete adult situation vs 83.33% in the incomplete adult situation) and in the "shape + color" combination sub-task (100% of correct responses in the complete adult situation vs

95.00% in the incomplete adult situation). The decrease is not significant and probably reflects lack of attention.

c) **Another child** : The **FXS boys** performance are, as expected, inferior to the one obtained in the complete adult situation but surprisingly are superior to the one observed in the incomplete adult situation. When they have another mentally retarded child as interlocutor, FXS boys give an average of 50.00% of correct responses in the following tasks : tower of pearls and "shape + color" combination task. The percentages of correct responses in the other tasks are respectively : 40.00% , 35.00% and 30.00% for the "shape + color + size" combination task, the size + shape combination task and the "size + color" combination task. The most difficult task in this situation seem to be the legos (mean : 10% of correct responses). When facing another child, **TD children** remain good listeners as they succeed in all the tasks.

DISCUSSION

Firstly, we want to stress the fact that this study represents a pilot stage in a more global work. This research aims to assess and describe the ability of FXS males to fulfill the speaker and the listener roles in referential communication situations.

Several tasks were proposed to a small group of FXS boys : 1] building tasks (the tower of pearls and the tower of legos) and 2] combination tasks ("shape + color", "color + size", "size + color" and "shape + size + color"). The results mainly indicate that FXS boys are poor communicators. Globally, they produce 18% of spontaneous sufficient messages whereas TD children produce 100%. FXS boys productions are mainly spontaneous insufficient messages (38%) and spontaneous incorrect messages (26%). It is interesting to note that 35% of the insufficient messages and 65% of the incorrect messages are produced in the "tower of legos" task. When we look closely at the insufficient messages, we can see that one or more information is missing : the shape (or size) and/or the position. The color is rarely missing as it constitutes a very salient characteristic of the blocks. In the case of incorrect messages, we can see that spatial terms used by FXS subjects are often the wrong ones. As we noticed it in the results, when they want to describe the shape of the items, subjects rather use the terms "big" and "small" used instead of "rectangle" and "square". However, this remains relatively rare. It appears that what seems to be an easier way to proceed is in fact more difficult for the subjects and can lead to incomplete messages. We will discuss further this assumption. But, for now, it is important, before discussion, to introduce the concepts of "nominal dimensions" (i.e., the color) and "ordinal dimensions" (i.e., the size). Ordinal dimensions can be view as intrinsically more demanding than nominal dimensions because

they are inherently relational (Johnston, Smith & Box, 1997). As Johnston et al. explain it, there is no direct mapping from a particular size to an attribute category such as "big" or "small" as an object can be small relatively to some objects and big relatively to others. It is clear than nominal dimensions do not have this relational character. Gentner and Rattermann (1991) have established a hierarchy between physical properties of the objects. Attributes like the color seems to be cognitively simpler than relations like the size and this partly because only one object must be considered at a time. So, the tower of legos task seems to be the more difficult for FXS boys as it implies the mastery of an ordinal dimension (such as the size) and specific vocabularies (the spatial terms) which seem to be problematic for these subjects. In other words, two kind of difficulties are embedded in the tower of legos task : 1] the use of spatial vocabulary and 2] the use of the ordinal dimension (the size) to describe the blocks.

The spatial vocabulary : In order to succeed in the legos tasks, the subject has to do more than communicating the blocks shape (rectangle or square) and color (blue, green, red or yellow), but he has to communicate the position of each blocks relatively to the others. In the case of FXS subjects, spatial information is not correctly given. Four explicative hypothesis can be proposed : 1] the perception of the blocks positions is inadequate, 2] adequate spatial terms are unknown, 3] adequate spatial terms are well known but are not available during the task, and 4] spatial terms are not considered as a pertinent information to give. Our data do not allow us to give a definitive explanation of the difficulties to communicate the right positions of the blocks. However, we can assume that if the "labels" of spatial positions are well known, they are often used in a wrong situation. Actually, terms to used are not the simplest one for children "in the middle", "on the right side" and "on the left side" which we know to be of early acquisition. Generally, subjects do not produce these terms and prefer using the word "on". But, when they try to give a more precise information, they often make mistakes and do not choose the right label for the right position. We can often notice confusions between "right" and "left". This confusion is not surprising and reflect previous data obtained by Comblain et al. (1993) with moderate mentally retarded subjects of mixed etiologies (Comblain, Fayasse, & Rondal, 1993; Fayasse, Comblain, & Rondal, 1993). Actually, in the experiment conducted by Comblain et al. with subjects aged from 10 years-old to 21 years old (mental age range : 5 years-old to 7 years-old), difficulties in the use and understanding of spatial terms such as "right" and "left" were mentioned. It seems that these spatial concepts remain problematic for mentally retarded people until late in adulthood. In addition to this problem of labeling, we can assume that FXS subjects considered the spatial information as non pertinent or, more exactly, non important. Color and shape (or size) seems

to be more salient features of the blocks. Subjects may consider that they only have to build a tower made off a succession of blocks of different colors and different shapes (or size).

The ordinal dimension. When we take in consideration the Gentner and Rattermann hierarchy, we can explain the apparition of insufficient messages in FXS descriptions. As we already mentioned it, objects attributes like color are supposed to be cognitively simpler than size which is inherently relational and implies that subjects take, at least, two objects into account. In a paper published in 1997, Johnston, Smith and Box used this contrast between nominal and ordinal attributes to explain performances of specific language impairment preschoolers in referential tasks. They point out that young SLI are generally unable to adequately use ordinal dimensions. Our FXS subjects seem to behave like Johnston et al. SLI preschoolers. They mainly use nominal attributes to describe items (the color and sometimes the shape). Even if, they are able to produce and adequately use labels as "big" and "small", they don't. Then, messages are incomplete as subjects only communicate the blocks color.

Difficulties of FXS boys with the size and the position of the blocks would indicate problem with the coordination of multiple components in a single description. The two points exposed before may explain the relatively good performances (48 % of success in the task and 60% of all the spontaneous sufficient messages) of FXS boys in the "tower of pearls" task. Once the general procedure has been explained ("you put the pearls on the wooden stick"), the speaker only have to produce the names of the color (all the pearls having the same shape and the same size). In this task, the difficulties are to correctly produce the general procedure and to communicate the right sequence of pearls (the first pearl to describe is the one which is at the bottom of the string, the last one is the one which is at the top).

Combination tasks need both information about nominal (color and shape) and ordinal (size) attributes of items. The coordination of these two kind of information makes the situation complex and explains the low percentage of success in the four combination tasks ("shape + color" : 25%, "size + shape" and "size + color" : 15%, "shape + size + color" : 20%). Each of these sub-tasks represents respectively 13, 8, 8 and 11 % of all the spontaneous sufficient messages. Furthermore, subjects have to communicate the right sequence of items : the first one to describe is the one located on the left size of the subject and the last one is the one located on the right side. Combination tasks seem to be more difficult to realize than the tower of pearls but simpler than the tower of legos (0% of spontaneous sufficient messages). This can be explain as follow. The tower of pearls only requires the communication of a nominal attribute (the color). Three of the combination tasks require, at least, the communication of nominal (color and/or shape) and ordinal (size) properties. Only one

combination task does not require this association of attributes : the "shape + color" task. In this case, only nominal attributes are useful for a correct description. This task is, however, more difficult than the tower of pearls as the it leads to 25% of success and the tower of pearls to 48% of success. This can be explain by the fact that the sequence to communicate in the combination task is more difficult than in the tower of pearls. In this last task, as we already mentioned it, the subject has to deal with a vertical sequence. In the "shape + color" task, subjects have to deal with an horizontal sequence oriented from the left to the right. It seems that it is more difficult for our subjects who are not still familiarized with this way of working (in the writing sense).

When we look at the results obtained in the listener situation, we can see that FXS boys are poor listeners in the "incomplete adult" and the "child" situations. In the "complete adult" situation, results are excellent in all the tasks proposed except in the "legos" tasks. In this case, even with precise and correct spatial information, FXS subjects, contrarily to TD children, build the right tower (only 30% of success).

In the "incomplete adult" situation, the performance of TD children decrease but not dramatically and probably reflect a lack of attention. The FXS boys decrease of performance is more dramatic than expected. On the six proposed tasks, three lead to a total absence of correct responses (no identification or building of the target-item) : "size + shape" and "size + shape + color" combinations and the "tower of legos". As in the other situations, the "tower of pearls" leads to the greater percentage of success (50%). Correct responses are also observed in the "shape + color" (15%) and in the "size + color" tasks. Let's note that the "shape + color" task involves two nominal attributes and the "size + color" task involves one nominal attribute. As we noticed it previously, it seems that FXS boys are likely to treat and understand the nominal attributes of items.

Finally, it is interesting to compare the performances of FXS boys with an "incomplete adult" interlocutor and with a "child" interlocutor. Contrarily to what happens in TD children, FXS boys seem to be more embarrassed when the incomplete information is given by an adult. In the case of the "child" interlocutor, it appears that the description given by the speaker is largely incomplete. What differs with the "incomplete adult situation" is the degree of incompleteness. In the "child situation" we cannot control the degree of incompleteness and the kind of information that we wanted to give to the listener. Furthermore, we cannot control the use of gestures. In the "incomplete adult situation" all these variables are controlled the description to give is precisely write down and all the children receive the same amount of verbal information without gesture help. We also observe that FXS boys are more

reluctant to ask information to the adult than to the other child. In general, the interaction with the adult seems to be more difficult. The analysis of FXS performance in the "child interlocutor" situation shows approximately the same hierarchy than previously. The "tower of pearls" and the "shape + color" combination task which only imply the comprehension of nominal attributes leads to the greater percentage of success (50%). The "tower of legos" task leads to the lower percentage of success (10%).

In summary, this pilot work has shown that in some situations FXS boys can be efficient speakers and listeners. The efficiency of the communication depends of the nature of the items attributes to communicate. Difficulties mainly appear with spatial and ordinal attributes. Spatial information implies the mastery of vocabulary and concepts which seems to be deficient in several etiologies of mental retardation including FXS boys. Ordinal information implies the manipulation of information that are intrinsically more demanding than nominal dimensions because they are inherently relational. Finally, FXS boys manage less efficiently with an incomplete message especially when it is given by an adult.

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