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COMMUNICATIVE PREVERBAL DEVELOPMENT IN A SAMPLE OF CHILDREN WITH WILLIAMS SYNDROME

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1. WHAT IS THE WILLIAMS SYNDROME?

The Williams-Beuren syndrome (Williams et al. 1961; Beuren et al. 1962) is included in the so called "rare diseases"[2]. It is a genetic disease caused by the micro-deletion in chromosome 7 in the band q.11.23 (Ewart et al. 1993). A map of 17 genes deleted in this region has been traced (Osborne et al, 2001). Its incidence is 1 per each 25.000 new borns.

Deletions in the same area seem to be present in 98% of the population with Williams syndrome, that is called the "clasical pattern" or "common Williams syndrome" (Mervis et al. in press); nevertheless, there is another 2% which present variations that can be explained not by a deletion in the genetic pattern, but by an inverted genetic pattern that results in an atypical Williams syndrome (Morris and mervis, 2000); atypical Williams syndrome share most of the particular characteristics the Williams syndrome present: mental retardation, attentional deficits, facial features and the pattern of behaviour; albite Tassabehji et al. (1999) have seen that there are patients with the chromosomical deletion who do not present the typical dismorphic features as the Williams syndrome subjects do.

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[2] In Spain there is an Institution called FEDER (Federación Española de Enfermedades Raras) in which the Williams syndrome is included together with other 6000 rare diseases. Although nowadays it is not the case for the Williams syndrome, these diseases are characterized by a low frequency of new borns and by the little information about them, owed, precisely, to this low frequency of appearance.

Until 1988, with the publication of Bellugi´s studies about Williams syndrome, this disease was mostly taken into account by cardiologists, because one of the clinical characteristics of this syndrome is supravalvular aortic stenosis, although not all Williams syndrome patients present this problem.

From 1988 a new tendency in the characterization of Williams syndrome was begun and gave way to the study of the syndrome from a neuropsychological point of view that considered different domains: linguistic, motor, visospatial, cognitive, etc.

From the neuropsychological point of view the traditional pattern describes a cognitive and behavioural outline of peaks and valleys.

Williams syndrome co-occur witn mental retardation (≤ 50). Though in general there are severe deficits in the cognitive domain, linguistic abilities are quite well preserved. That is an outstanding fact which cannot be found in other populations with mental retardation (Mervis, 2000). Nowadays, data show that although language is quite preserved, it is not as intact as it was initially said. Studies focus on the variablility of skills in this domain to know where strenght and weak points are in order to observe the dissociations of the different components of language.

2. SOME ASPECTS OF THE GENERAL DEVELOPMENT OF THE WILLIAMS SYNDROME

Considering development we must bear in mind the complete process of growth, not only language or cognitive aspects. It is wellknown that deficits in the general development of a person can affect to several domains.

From a clinical point of view, the presence of hight levels of calcium during infancy have been observed, leading to hypercalcaemia; although it is transitory it hinders growth. Data show that more than 50% of patients with Williams syndrome have a low stature, even though there are surprising exceptions. This growth delay might be also explained by the lack of *insulin-like growth factor* (IGF) which has a very important role in the process of growing in mammals in general acting on the growth hormone (Froesch et al, 1985).

Restrictions of growth is ever presents in most of the cases. Many studies have observed that from birth to puberty the relation bone age/chronological age is a year or two below and mature height is lower than the normal average. Pankau et al. (1992)

suppose that this low height in Williams syndrome can be due to a precocious puberty that means a quick skeletal maturation as well as a fast fussion of cartilages.

There are other alterations that affect the muscle-skeletal system that are to the detriment of laxity problems, articulation contractions, spinal column problems and low muscular tone.

It has also been observed that the brain is in general terms smaller than a typically developed brain, although the frontal and the temporal lobes as well as limbic areas and cerebellum seem to be enlarged (Gallaburda, et al. 1994; Wang et al. 1992). Frontal lobes, as well as limbic areas, have an important role in the control of emotions, but frontal lobes have also great importance in personality and social behaviour. These areas, when stilted, involve emotional inestability, disability to establish social relations and tendency to isolation. In the case of the Williams syndrome, the hyperdevelopment of frontal lobes leads to a hypersociability during childhood and pre-adolecent years, but a tendency to isolation in later ages.

Disproportionately enlarged temporal lobes might explain partly their love to music and the musical abilities they have in general (Levitin and Bellugi, 1998; Don, Schellenberg & Rourke, 1999; Lenhoff, Perales & Hickok, 2001), other recent studies suggest otherwise[3]. This development of the temporal lobes can also explain their sensitivity to high sounds which appear to be unbearable for them.

The cerebellum is also extremelly developed and has a prior role in motor and coordination aspects. It is also related to the comprehension of language, particularly to the analysis of sounds.

Children with Williams syndrome have a slow development in several areas, both cognitive and motor, that contrast with good linguistic abilities.

In respect to motor aspects, crawling and walking appear to be quite retarded in comparison with the typical development of children. It is quite infrequent to observe a child with Williams syndrome walking before the age of two, some parents also report a later development of this abilitiy. When they begin to walk or even as adults, their movements seem to be very awkward, with clear uncoordinated movements.

[3] Our experience after 7 years of organizing musical camps with people with Williams syndrome, shows that they are not so skillful at playing music, and at singing either. These musical camps are organized by the Spanish Williams Syndrome Association and there are musictherapist professionals that work for a week with them. Several of these children are having musical lessons during the year and the fact is that they do not stand out in this ability.
The wellknown case of Gloria Lenhoff (an american Williams syndrome who is 47 old) who can sing in at least 15 languages and can play the accordion, does not mean, from our point of view, that the WS are specially gifted for music (Martínez & Sotillo, 2004).

Reproducing or imitating basic movements is quite complicated for them, giving an overall impression of clumsiness. A possible explanation could rely on their physical build; in general terms they have short legs, hands and fingers, long arms and body, and, in the case of females, a low and protruding bottom. This physical build is partly similar to those who have dwarfism, and is much more outstanding in girls than in boys not during infancy but from the adolescence on.

Skillful motricity is also defficient in children as well as in adults; so they cannot cut a steak with a knife, they have problems sewing, or shuffling cards. The simple task of trying to pick up little pieces of whatever (i.e. pieces of paper, marbles) from the floor can be carried out with great difficulties when they are children.

Cognitive aspects are also delayed. As we have mentioned, they have mental retardation. Clasically the I.Q. was placed between 50/70; that is, they have a moderate or mild mental retardation. Nevertheless, everyday there is more data that supports the idea that the I.Q.s are lower than expected and it is frequent to find children with IQs of 40 or 35.

Cognitive aspects are more or less identified, although researches have not come to a unanimous conclusion. This disparity can be due to a particular phenomenom: a specific pattern of abilities in the frame of mental retardation mostly related to quite well preserved language skills, contrary to remarkably poor visual-spatial and motor abilites. This has lead to say that Williams syndrome shows an asymmetric cognitive profile of peaks and valleys (Bellugi et al. 1988).

Linguistic and communicative abilities are going to be developed in the following heading.

3. LINGUISTIC DEVELOPMENT IN THE WILLIAMS SYNDROME

The neuropsychological pattern of peaks and valleys proposed by Bellugi et al. (1988, 1996) and Wang et al. (1993) suggests that Williams syndrome linguistic skills are not as altered as other domains; that explains the interest this syndrome arouses in respect to the possible independence of language from cognition, and the consequent postulation of the existence of a language module totally independent from a cognitive module.

At the present moment this is a controverted aspect, since many authors have observed that language is not as preserved as it was believed, and that different levels of language are unequally altered (Gosch, Städing & Pankau,1994; Capirci et al, 1997; Howlin et al, 1998; Garayzábal & Sotillo, 2001a, b, c, d); thus phonetics and phonology seem to be undamaged; in relation to grammar, comprehension is better than production; vocabulary seems to be a strong and well developed aspect and, apparently, pragmatic, which, owed to their sociability, seems to be well preserved, eventhough communicative abilities are highly altered.

Linguistic skills are also delayed in the first stages of life (Paterson et al, 1999). As mentioned, language appears between the second and fourth years of age, and not only is language delayed, but it is also atypical, indeed there is some evidence that language acquisition in WS is quite different from that followed by typically developed children (Mervis et al, 1999). After a late onset of language, children with Williams syndrome normally start language development off by the age of four, coinciding with school age. In these early stages of language the fluency and grammatical use of language that seem to be well developed is surprising, in addition to the display of a quite sophisticated and rich vocabulary, that serves to engage people in conversations.

Due to their friendly personallity (Udwind & Yule, 1990), little children with Williams syndrome like to chat with others, above all with adults, but one can immediately see that they tend to use social clichés, while their comprehension is poor and their conversations are irrelevant, not aprropriate and sometimes lack semantic content (Lukàcs, 2003). When they are in the preadolescence, this social profile changes in many of the Williams syndrome subjects who tend to behave in a contrary manner. Parents use to refer behaviours of isolation: they are not as social as they used to be and they also have problems in establishing relations, this leads to depressive tendencies, to anxiety states or to unknown fears in many of them (Dilts et al, 1990; Hodapp, 1997). An explanation of this attitude could lie in the self-consciousness of being different to others, that is a fact not seen in other groups with mental retardation.

We have emphasized the main points regarding to language; but if we take a more detailed perspective of language development, there are several aspects we would like to point out, as their development is quite different to the development of typically developed children as well as to those who belong to other mentally retarded populations such as theDown syndrome children and this could highlight aspects related

to language acquisition and its function and importance in the general frame of language.

We have refered to a different way of acquiring language by children with Williams syndrome. When they are toddlers the fact that they produce speech before pointing is very outstanding (Laing et al, 2002). Adamson (1995) also underlined that for the acquisition of language the ability to refer by referential pointing was an important milestone that preceeded the lexical production. Mervis et al. (1997, 2003) studied the onset of referential pointing and the onset of referential language in a longitudinal research of 10 children with Williams syndrome whose ages ranged between 4 and 26 months. They concluded that, except for one, the rest of the sample produced referential labels before the comprehension or the production of referential pointing gestures; that is, they showed an opposite profile of language acquisition to that of normal children; that made the authors mistrust if referential pointing skills are really necessary for the onset of language and if joint attention requieres necessarily of the presence of referential pointing or if there are other ways to establish joint attention to objects that do not need to go through the possesion of referential pointing.

On the other hand, there is data that supports the impression that rhythmic banging is a predictor in the delayed onset of language in children with Williams syndrome. Masataka (2001) supports the idea that rhythmic hand banging correlates with the onset of cannonical babble, if rhythmic hand banging is delayed or does not apperar, cannonical babbling does not appear either, and, as it is well known, when canonical babbling does not appear, neither does the onset of language.

Several studies stressed on the relations between pre-verbal communication and joint attentions skills, which develop at the age of 10 months in typically developed children, and are precursors for the development and onset of language (Bates et al, 1979; Bakeman and Adamson, 1984), the absence of these in Williams syndrome language might explain the late onset of it. Laing et al. (2002) demonstrated through three different experiments with toddlers that although language abilities were relatively good, social skills were stilted, and they concluded that several aspects of early communication were impared as well. They emphasized two factors of the prelinguistic development that were especially lacking or sparse: pointing and joint attention. The authors suggest the idea that this lack of preverbal abilities are the explanation for the WS language delay.

4. QUESTIONARIES TO PARENTS ABOUT THE COMMUNICATIVE PREVERBAL DEVELOPMENT IN A SAMPLE OF CHILDREN WITH WILLIAMS SYNDROME

We would like to contribute with the data above described. In this sense we gave questionaries to parents with children affected by Williams syndrome. Questionaries were delivered by the Spanish Williams Syndrome Association (ASWE).

We divided the questionaries in three ranges of age, from 0 to 6 years of age, from 6 to 12 and from 12 further on. We will focus on the first stage.

As we have mentioned, the initial stage covers until the 6 years of age. We have done so because there was no data available for children under three years of age, to the moment we had the questionnaires delivered. Whereas for top age we considered that the period of time from 4 to 6 could provide relevant data about language onset once started. Our questionaries focus on communicative non verbal abilities development.

The sample is composed of five children genetically diagnosed, three boys and two girls. Ages ranging from 2'4 to 5'6 years of age (2'4, 3'9, 4'7, 4'8, 5'6) . We asked the parents to answer the same items not only for the present moment but making commentaries about the items when their children were around two years of age.

Parents had to complete the questionary at home together. The items parents were asked to fill in were the following:

1) Does your child point with the index finger to anything he/she is looking at?
2) Does your child use the index finger to show anything that surprises him/her?
3) Does your child emit any sound or word when pointing with the index finger?
4) Does your child show anyone any object he/she has in his/her hand?
5) Does your child look for an object of his /her interest to show it to others?
6) Does your child look at you to confirm you are looking at him/her?
7) Does your child push you to show you anything?
8) Does your child look at you when pushing you?
9) Does your child give you things?
10) Does your child look at you when giving anything to you?
11) Does your child look alternatively to you and an object?
12) Does your child give you an object to show a necessity (i.e. ball=play; glass= want to drink)?
13) Does your child look at you when doing the previous item?
14) What does your child do if he wants anything?

a) he /she asks for help using words
b) he/she emits sounds
c) he/she tries to get by him/herself what he /she wants
d) cries
e) he/she points out the object
f) he/she looks at the adult and object alternatively
g) he/she asks for help pushing you.

15) Does your child nod or smile to express happiness?
16) Does your child use basic gestures spontaneously to communicate basic necessities or moods (i.e. clapping hands to indicate approval, touching their zip to indicate his/her necessity to go to the toillet)?
17) Does your child express happiness, sadness, anger through facial expressions?
18) Does your child answer with gestures to gestures?

Items were arranged horizontally into a table of six columns:

Items	Very frequently	frequently	sometimes	never	commentaries

5. RESULTS

The small sample we have and the different ages of the children do no permit a statistical explanation. So descriptive results are given in the following table:

Table I

2'4

item	1	2	3	4	5	6	7	8	9	10	11	12	13	14a	14b	14c	14d	14e	14f	14g	15	16	17	18
Ans.	c	a	c	b	c	b	d	d	a	a	a	c	c	d	b	b	b	c	b	d	a	a	a	a

Ans: answers. a:very frequently; b: frequently; c: sometimes; d: never

Table II

3'9

Item	1	2	3	4	5	6	7	8	9	10	11	12	13	14a	14b	14c	14d	14e	14f	14g	15	16	17	18
Ans.	b	a	b	b	b	b	b	b	a	b	a	b	b	b	d	a	b	c	c	d	a	a	a	a

Ans: answers. a:very frequently; b: frequently; c: sometimes; d: never

Table III																								
4'7																								
item	1	2	3	4	5	6	7	8	9	10	11	12	13	14a	14b	14c	14d	14e	14f	14g	15	16	17	18
Ans.	a	a	a	a	a	b	a	a	a	a	a	a	a	a	c	a	d	c	a	a	a	b	a	a

Ans: answers. a:very frequently; b: frequently; c: sometimes; d: never

Table IV																								
4'8																								
item	1	2	3	4	5	6	7	8	9	10	11	12	13	14a	14b	14c	14d	14e	14f	14g	15	16	17	18
Ans.	a	a	a	a	b	a	a	a	a	a	c	d	a	a	d	c	a	a	b	a	a	b	a	a

Ans: answers. a:very frequently; b: frequently; c: sometimes; d: never

Table V																								
5'6																								
item	1	2	3	4	5	6	7	8	9	10	11	12	13	14a	14b	14c	14d	14e	14f	14g	15	16	17	18
Ans.	d	d	d	d	d	D	c	d	b	c	d	d	d	a	d	a	c	c	d	c	c	c	c	c

Ans: answers. a:very frequently; b: frequently; c: sometimes; d: never

Results were obtained only by the parents answers without any previous assessment of the children; for that reason some results can be surprising. There are two children in the poles of ages (2'4 and 5'6). The smaller one seems to be more able and not as delayed as the bigger one, who is more delayed than expected for his age.

For the child who is 2'4 (table I) years of age there are contradictory results. As we can see, items 10 and 11 show that he has achieved the the joint attention milestone, or at least he is on his way of progress; on the contrary, items 1, 8, 13, 14e, 14f seem to be quite delayed in relation to the others. In these last pointing is the milestone involved as well as joint atenttion but in relation to demanding responses, not in giving things.

If scanning the data of this child we can observe that some pieces of information seem contradictory. There appears to be great differences in items 1, 2 and 14e related to pointing seem to be quite different. So the child fluctuates from the occasional use of his index finger (1, 14e) to the quite frequent use of it (2).

In respect to the joint attention milestone there are also items which are not very stable, as can be seen in items 6, 8, 9, 10 and 13. So there are moments in which the child looks at the adult very often when giving something to him, and moments in which the child never looks at the adult when showing or wanting something from him.

In respect to the child of 3’9 years of age, item 14f is the only remarkable aspect. It has to do with joint attention when wanting something.

The child of 4’7 years of age does not have any problems with joint attention, but, for pointing, item 14e shows that sometimes he points to the object he wants, but not always; this is contradictory with item 1 in which he points with the index to show something he is interested in frequently.

Although the 4’8 year old child has no remarkable problems, he seems to have some difficulties in establishing joint attention when giving things and the fact that he does not use gestures spontaneously, as often as the other four, is outstanding and uncommon. Parents note that their child uses words more than gestures and when using gestures they are accompanied by words.

The answers of parents of the child of 5’6 years of age show a quite delayed pattern of their child. As we can see joint attention (10, 11, 13, 14f, 14g) as well as pointing (items1-8, 14e) are milestones not acquired yet. In general parents show a concerning picture of their child. Not only are these preverbal milestones mentioned not acquired, but other preverbal milestones are delayed as well, such as the emision of sounds, giving objects to make himself understood, the use of basic gestures for expression and the use of facial expresions of emotions. In general all of these items are achieved by WS children earlier than this child does, even by our youngest child who is 2’4 years of age.

As we said above, there was a section in which we let the parents make any commentaries they wanted with respect to the item they were answering; above all, they were asked to fill in each item bearing in mind earlier ages of development about the particular question.

Parents of the child of 2’4 years of age refer that their child does not say a word (item 3, or 14a) but he can communicate many things through gestures and facial expressions (items 15 to 18). For items 1 and 14f, they comment that their child has recently begun to do them.

For the child of 3’9 years of age parents did not give any answers refering to the onset of the items assessed.

Parents of the child 4’7 years of age made many commentaries. So for items 1-3 and 5, they emphasized their child had managed to do them at the age of two. For item 4 it was at the age of three when the child finally controlled got it. Items 7 to 18 were

achieved at the age of four. It is very remarkable that items which involve joint attention were very delayed in relation to the studies previouly mentioned.

Comments collected for the child of 4'8 years of age focus on the child's locuacity. Parents report that their child began to speak very early and used gestures very soon to serve words with.

For the eldest child of our sample, parents do not give any comments about early stages of acquisiton of the items we were assessing, which is not surprising as he seems greatly retarded.

All parents were asked if their children were able to speak at the age of two. All coincide in saying that their children began to speak, using structures, between 33 months (2'9 years of age) and 4 years of age.

6. DISCUSSION

Data were only analysed from the point of view of the children´s parents, therefore objectivity is relative. This is a handicap in order to obtain deffinitive results, but they give us an idea of what the general pattern for these preverbal milestones is in our sample.

We focussed on this particular study from the start, that is, we wanted to see if children with Williams syndrome below 6 years of age had achieved some of the preverbal milestones that are supposed to appear before the onset of language. This naïve approach meant children were not assessed previously we did not want to slope our study on the basis of a cognitive or linguistic assessment; we wanted to see how the matter at issue was in reality.

This principle on our study explains, for example, the great differences between the youngest child with respect to the eldest. A possible explanation could lie on their IQ´s, much lower than predicted for the WS in general (mild to moderate) for the eldest one. In the light of the data obtained about the youngest child, the fact that his I.Q. appears to be in the expected range (40/60) for the WS cognitive pattern might be relevantis very probable.

In general results reveal that preverbal milestones are very lately acquired while the onset of language is previous to it, which corroborates the studies reported previously. But there are two points we consider new and outstanding in this study.

On one hand data enhance the fact that preverbal milestones are developed later than supported by previous studies, whose participants were not older than 31 months of age. It is true that our sample is very small, and, as said before, we can not generalize the results for all the population with Williams syndrome. Actually we are trying to get a bigger sample to corroborate the results we have obtained.

On the other hand, answers mark a difference in the joint attention milestone. That is, we have seen that children have a clear difference in mastering joint attention when wanting or giving things. Three of the five children of our sample (2'4, 3'9, 4'7) seem to have more difficulties in joint attention when they want something (when they are the receptors), but not when they give something (when they are agents). One of the children (4'8) shows the opposite pattern, while the last child (5'6) has many difficulties in joint attention under all the situations. If we bear in mind that they are very interested in others and in establishing social relations, these results are quite comprehensible, giving can be a precursor for this social interaction, more than wanting or demanding. That is quite surprising due to the fact that in a typical development wanting or demanding appears earlier than giving. This has led us again to postulate the atypical development of language and preverbal skills in Williams syndrome.

Although our results are not conclusive, we think they are in the same line as other studies, but as happens with other skills results vary from one child to another, according to the parents' answers.

From our data we cannot conclude that the late and atypical development of these preverbal milestones is the explanation for the delay in the onset of language, as it appears before some preverbal milestones. Since parents report that their children developed language before preverbal abilities, we support the idea that they follow an atypical development of language rather than the idea of a delay. Perhaps the explanation for this late acquisition lies on other types of explanations that can be found in their cognitive development as happens in other groups with mental retardation.

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