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Research Article

Communicative and pragmatic skills: down syndrome vs williams syndrome

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Introduction

Down syndrome

Down Syndrome (DS, hereinafter) is a chromosomal alteration produced by changes in the DNA sequence of chromosome 21, in addition to being the main cause of intellectual disability in the world [1]. According to the World Health Organization [2], DS has a worldwide prevalence of 1 in every 1,000 live births, but these figures vary due to different factors such as prenatal diagnosis. Others say that DS occurs in about 1 in 700 births [3].

Language is one of the areas in which people with DS have more delays, problems, or difficulties, compared to people with Typical Development (TD). These difficulties or deficiencies are caused by neurobiological or cognitive deficits [4-6]. In general, the level of comprehension in people with TD is better preserved than the level of expression and the lexico-semantic area, as well as the pragmatic area, are considered stronger compared to the phonetic-phonological and syntax area [7,8].

In the case of people with DS, Abbeduto, et al. [9]. Show the existence of a delay in practically all facets of pragmatic performance in relation to TD children of the same age. However, Roberts, et al. [10], say that starting from how difficult this area is, as well as everything that constitutes it and, taking into account the capacity of these people, it is shown as a strengthened area. As a consequence, it can be concluded that pragmatics in people with DS has strong and weak points [10].

In people with DS, the ability to initiate a conversation is one of the weakest aspects [11]. As Abbeduto, Warren & Conners [12] state, children with TD elaborate a discourse through more complex contributions, while children with DS limit themselves to providing "simple answers", observing less significant content. The narrative capacity presents weaknesses [11], improving when visual information is presented that allows them to observe the course of the story [11,13]. Roberts, et al. [10], indicate that people with DS can hold conversations on a specific topic on a medium-high percentage of occasions.

In addition, Martín-Urda, Carchenilla & Moraleda [14] state that there is a delay in the appearance of intentional communication and the maturity of its statements. In addition, people with DS use pragmatic functions in the same way as children their age, although they appear delayed [15].

Williams syndrome

Williams Syndrome (SW, hereinafter) or Williams-Beuren syndrome (WBS) is a neurological and multisystemic development disorder, caused by a microdeletion on chromosome 7 [16,17]. This syndrome was officially described in 1961 by Williams, Barrat-Boyes & Lowe [18]. WS has an incidence of 1 per 7,500 people [19,20]. However, Garayzábal & Cuetos [16] state an incidence of 1 in 20,000 new births.

Taking into account the cognitive aspects, we can affirm that people with WS have a specific cognitive profile, presenting a great deficit that courses with moderate delay and their Total Intellectual Coefficient (CIT, hereinafter) may be between 40-70, coexisting with the possibility that there are attentional problems, in visual memory, motor skills, although with great strength in auditory memory [21], although other authors

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mention that some patients may have CIT scores between 75 and 85 [22].

People with WS very often produce narratives that lack coherence and content, even though they are grammatically correct [23,24]. Although people with WS manage to build and tell long stories, their narrations are deficient, accompanied by a lack of cohesion and coherence, as we have said, lack of plot, they do not establish cause-effect relationships or take the context into account [25]. Gonçalves, et al. [26]. Consider that their narration is not relevant and important data is lost in it.

As Garayzábal [25] indicates, these people show dependence on the context by manifesting themselves, speaking quickly to people, repeatedly about topics of no interest to the interlocutor, persisting in asking questions to which they have already obtained an answer, and losing the sense of a joke or irony. Some authors report not knowing how and when to start a conversation, or when and how to end it, not knowing how to maintain the topics of conversation, as well difficulties in managing and respecting turns [27-29].

On the contrary, we also find authors who report good pragmatics in children with WS, observing that they performed relatively well on pragmatic skills tests [30,31]. However, it is important to note that few studies have been carried out in this line.

Therefore, given the characteristics of both syndromes, the objective of this study is to compare, from a linguistic perspective, the pragmatic abilities of people with DS and WS.

Method

Participants

To obtain DS data, 55 relatives (parents, legal guardians, or another relative) of people with this genetic alteration participated, of whom 86.6% were parents, 11.9% were completed by other relatives and 1.5% were completed by a legal guardian. These people had children between 3 and 18 years old (mean age=11.4 years). 58.2% of the data collected were men, compared to 41.8% who were women.

The data obtained from WS were provided as part of another investigation previously carried out on Pragmatic Skills in SW. In this sample, 34 participants with children aged between 5 and 36 years were obtained, with an average of 12.17 years. 47.05% of the data provided are men, compared to 52.94% who were women.

Process

For the creation of this project, a Google Form questionnaire was carried out, in which the questions corresponding to the Pragmatic Awareness Questionnaire (PCC) were introduced (Rodríguez, 2012). In addition, some preliminary questions were added to the questionnaire to collect data on the age of the child with DS, sex, date of birth, and province where they reside. The same dynamic that had been created for the previous data collection on Pragmatic Skills in WS was followed. A question was included asking if you consent to the data being used for the research project. The link to the questionnaire created in Google Form, together with a brief explanation of the project, was sent to all Down Syndrome associations in Spain, in addition to being published on social networks, and sent to relatives and acquaintances to obtain maximum diffusion. This questionnaire was filled out by parents or legal guardians of people with DS individually and only one parent or guardian per person with DS. The responses were saved and analyzed later.

Instrument

The Pragmatic Awareness Questionnaire (PCC) (Rodríguez, 2012) is made up of 26 items in total, made up of 7 blocks, following a 5-point Likert-type scale to solve it, in which 1 = very bad, 2 = bad, 3 = regular, 4 = good and 5 = very good. Block I (corresponding to items 1-2) evaluates intelligibility, which refers to the understanding of the linguistic information by the interlocutor (1) and paralanguage (2), which is the intonation, pause, rhythm, and volume of the voice. In block II (corresponding to items 3-10) proxemics (3-4) are measured, which is the distance between the interlocutors while they communicate and the physical contact they maintain in communicative acts, and kinesics (corresponding to items 5-10), which is the perception that the subjects have of their body posture, gestures, facial expression, movements of their upper and lower extremities and gaze in conversations. Block III (corresponding to items 11-12) analyzes lexical cohesion (11) and competence (12), that is, the different linguistic forms to refer to the same meaning and the number of words that the subject knows of their language. Block IV (corresponding to items 13-15) evaluates the semantic-pragmatic deficit, including irony and humor. Block V (corresponding to items 16-18) analyzes the morphosyntactic relationships of discourse, such as the construction of words and sentences. Block VI (19) attends to the reflection on the stylistic variations of the subject based on its linguistic adaptation to the context of communication and its interlocutor.

This questionnaire has been considered one of the most reliable tools for evaluating pragmatic competence, especially in Williams Syndrome [25].

Results

Analyzing the results obtained in this study, we first find an overview of them, showing the corresponding comparison of the pragmatic abilities of DS and WS.

In the first item that evaluates the intelligibility of speech, we can observe that the response of the parents has been much better in people with DS, of which 77% responded very well or well, that in people with WS with percentages of 39 %, respectively. We also observed that no family member states that intelligibility is very bad in people with DS compared to 16% who say it is in WS.

In the item that evaluates suprasegmental aspects such as intonation, rhythm, etc., we found similar scores in both

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disabilities, although with a small difference, being better in DS, as can be seen in Table 1.

Analyzing the item that talks about the proximity that is maintained with the interlocutor in a conversation, it is observed that it is a little better in DS than in SW. It stands out that the parents have referred very well or well in 53% of DS compared to 38% in WS. We also found scores that refer very poorly or poorly with 5% in DS compared to 21% in WS. The rest of the scores look more or less similar.

Regarding the item that speaks of the use of physical contact in communication situations, better results are concluded in DS with 74% of responses that refer very well or well compared to 68% in WS. It stands out that no family member of people with DS reports that they use physical contact very badly or badly in front of WS with results of 12%.

Continuing the analysis with the next item, we find the body posture they maintain in a conversation. Better scores are observed in DS with 74% of relatives reporting scores of very good or good, compared to WS, with 53%. It stands out that nobody responded very badly in DS compared to 9% in WS.

In the item that evaluates how the body movements of the arms and hands seem to the relatives, we found very good scores in DS, with 82% of the parents reporting very well or well, compared to 56% in the case of people with WS.

When evaluating the item that refers to facial expression, there are no differences between both groups (89% vs 88% in DS and WS respectively).

The answers obtained by the relatives about the item that evaluates the look towards the interlocutors in a conversation show better results in DS, with 71% of answers that value that it is good or very good compared to 44% that refer to it in SW. It also highlights 9% who value it as bad or very bad in DS compared to 21% in SW.

As for the items referring to expression, the parents state that the ability to use synonyms is practically the same in WS as in DS.

In the item that evaluates the number of words that he knows and uses in his language, we observe similar results in general terms, although slightly better in SW compared to DS, as we can see in Table 2.

To finish, we analyze the last item of this block which includes the interpretation of ambiguous expressions and comments, such as set phrases or metaphors. Better results are observed in DS, with 17% of responses referring to very well or well, compared to 3% in WS. In both, there is the same percentage of answers referring to bad or very bad, with 56%.

We continue with another block that values understanding and reactions to irony and humor. In both, we observed better results in DS versus WS as shown in Table 3.

Analyzing the results reported by relatives on this block called morphosyntax, word construction, as well as phrases and sentences, we perceive better results in WS as we can see in Table 4.

In the item that evaluates the way of relating and connecting some ideas with others, we found slightly better results in DS, with 20% of family members reporting very well or well compared to 18% in WS. In addition, the results referring to very bad or bad are lower in DS, with 38% compared to 53%.

Relatives report much better results in DS about their changes in style or register and their degree of adaptation to the communicative situation and their interlocutor, with 49% indicating very well or well compared to 21% in WS. It is also observed that 11% indicate very poorly or poorly in SD compared to 41% in WS.

We grouped the last 6 items and made an overview of all of them, much better results are observed in DS than in WS, as we can see in Table 5.

Table 1: Suprasegmental aspects in DS vs. WS.

	VERY GOOD OR GOOD	REGULAR	VERY BAD OR BAD
DS	36%	47%	17%
WS	32%	53%	15%

Table 2: Number of words they know and use of their language in DS vs WS.

	VERY GOOD AND GOOD	REGULAR	VERY BAD AND BAD
DS	54%	24%	22%
WS	44%	41%	fifteen%

Table 3: Understanding and reactions to irony and humor in DS vs. SW.

		VERY GOOD AND GOOD	REGULAR	VERY BAD AND BAD
IRONY	DS	9%	31%	60%
	WS	3%	18%	79%
HUMOR	DS	40%	33%	27%
	WS	30%	29%	41%

Table 4: Construction of words, phrases, and sentences in DS vs. WS.

		VERY GOOD AND GOOD	REGULAR	VERY BAD AND BAD
Word construction	DS	24%	43%	33%
	WS	35%	29%	36%
Construction of phrases and sentences	DS	16%	49%	35%
	WS	24%	44%	32%

Table 5: Aspects of the conversation in DS vs. WS.

		VERY GOOD AND GOOD	REGULAR	VERY BAD AND BAD
Theme Settings	DS	34%	45%	21%
	WS	30%	29%	41%
Theme changes	DS	56%	27%	17%
	WS	29%	32%	39%
Maintenance and monitoring	DS	15%	45%	11%
	WS	21%	38%	41%
Wait time	DS	51%	40%	9%
	WS	26%	47%	27%
Interruptions	DS	42%	49%	9%
	WS	21%	38%	41%
Amount of information	DS	43%	38%	19%
	WS	29%	38%	33%

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Discussion

The results obtained in this research study have revealed how people with DS obtain better scores in pragmatic skills in relation to people with WS. However, it is important to highlight that these linguistic characteristics are essential when considering language intervention in these two populations.

With respect to the results found, consistency is observed in relation to other investigations. For example, in people with WS, there are low scores in understanding and reacting to jokes or irony, which confirms the studies carried out in this regard (Garayzábal, 2005). People with WS very often produce narratives that lack coherence, even though they are grammatically correct [23,25]. The results confirm that there is indeed a low level of coherence in their narratives, although, in this, lower results can be observed in DS. Along these same lines, we can affirm that the degree of adaptation to the communicative situation and the interlocutor is poor in WS, in accordance with Gonçalves, et al. [26].

In addition, the results obtained in this research follow the line of Garayzábal & Sotillo [27]. These authors consider one of the weak points in WS, the proper introduction and start of a conversation, as well as its end. Along the same lines, they also state that they have difficulties taking turns in conversations. The results of Udwin, Yule & Martin [29], who consider that people with WS do not give adequate information in the responses they generate, are also confirmed.

On the other hand, the results of our study question the idea of authors who report relatively good results in tests of pragmatic abilities in people with WS [31-56].

With regard to DS, the studies carried out by Martin, Klussek, Estigarribia & Roberts [11] and Roberts, et al. [10], in which in people with DS an acceptance of the adjustments to the subject is observed, in addition holding conversations on a specific topic on a medium-high percentage of occasions. However, Martin, Klusek, Estigarribia & Roberts [11], point out that there is a weakness in the ability to initiate conversations. Therefore, it seems that pragmatics has strong and weak points in people with DS [10].

Finally, regarding the amount of information they provide when communicating, we can affirm that the narratives of people with DS present more content than those of people with WS [24].

As a suggestion, we fundamentally consider two limitations of the study. For example, a larger population sample would be necessary to be able to generalize the results obtained. On the other hand, we believe it is convenient to continue expanding the research with other tools that complement the assessment of pragmatic competencies of people with intellectual disabilities.

In a conclusion, it should be noted that the results of this study have revealed some important points. Firstly, both people with DS and people with WS have pragmatic difficulties [9,25], which is important in order to continue working on the language throughout the entire evolutionary cycle. Secondly, after the research study carried out, it has been known that the pragmatic skills of people with WS are worse than those of people with DS, which makes it possible to further identify the linguistic profile of both syndromes. Therefore, it is necessary to continue focusing on linguistic research on rare diseases.

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