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Phonological Disorders and their Relationship to Agenesis of the Corpus Callosum in Children

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Abstract:

The subject of this study revolves around the phonological disorders recorded at a 5-and a half-year-old- child suffering from an uncommon type of agenesis of the corpus callosum. The results of the general clinical linguistic profile of this child show the various difficulties faced by this case in three types of phonological choices. Indeed, the results of the study show that pronunciation is characterized by the phonological deletion of the initial syllables of words, which reflect a defect in the cognitive and analytical skills of the different types of phonological elements, and the absence of maturity of the phonological representations that the child is gradually demonstrating. The findings of these studies also confirm what previous studies provided about the slow speed of cognitive information processing in children with agenesis of the corpus callosum.

Keywords: *Agenesis of the Corpus Callosum; Phonological Disorders; Oral Language ; Speech and Language Pathology; Verbal Memory.*

1. Introduction

The issue of congenital corpus callosum agenesis (CCA) is one of the most fundamental topics that have attracted researchers in neuropsychology, over the past twenty years. The cases suffering from this brain malformation represent a topic of great interest for speech - language pathology researchers who have been investigating their pathological and clinical linguistic characteristics and are still trying to develop the appropriate therapeutic principles.

Through this research work, we try to study the degree of phonological disorder in the case of a child suffering from isolated agenesis of corpus callosum (ACC).This was done based on the recorded data of the global clinical and cognitive features. Consequently, the focus of our attention is to identify the agenesis of corpus callosum syndrome from the anatomical and functional perspective, and to deduce the different psycholinguistic and psychopathological

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manifestations that were encountered in several previously conducted studies, especially in those that focused on the cognitive and auditory capacities to comprehend the different language elements of spoken speech.

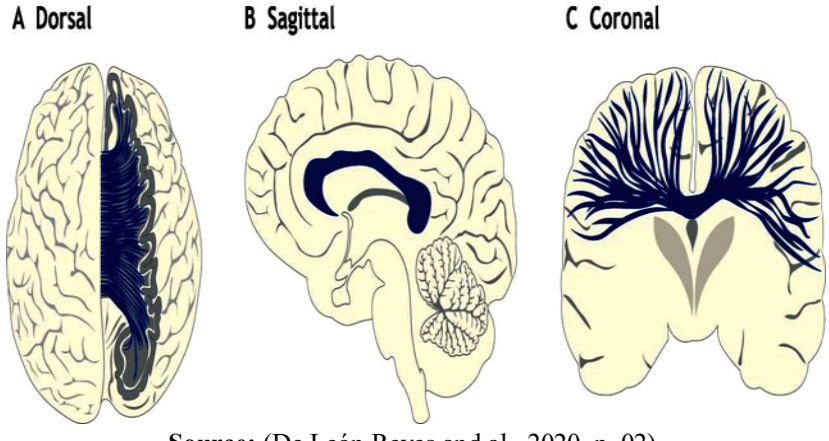
The purpose of this study is to address a main question related to the clinical linguistic dimension in a case with isolated agenesis of the corpus callosum: does the patient suffer from phonological disorders recorded in particular in the repetition of familiar and non-familiar polysyllabic words? The importance of our study lies in presenting the most important pathological linguistic characteristics that affect the phonological level of the Algerian Arabic dialect. This will improve the overall design of treatment protocols for Arabic dialect speaking children with isolated type agenesis of the corpus callosum.

2. The Problem of Congenital Agenesis of the Corpus Callosum and its Relationship to Linguistic, Cognitive and Phonological Disorders

It is worth knowing that the corpus callosum is the region of the brain that connects the left and right cerebral hemispheres; it contains a complex bundle of 200 to 800 million nerve fibers as represented in Figure 1 which illustrates the general structure of the corpus callosum. It is useful to know that the corpus callosum allows transferring and integrating various types of sensory, motor or connective information that is supplied by different regions of the cerebral hemispheres. In addition, the neural connections between the cerebral hemispheres facilitate the integration of complex sensory and motor information that is generated by both sides of the body. This would affect the high cognitive activities, abstract thinking, executive functioning, social interaction and language (Yeh H. R and al., p 634-635, 2018).

Figure 1:

General structure of the corpus callosum according to its three manifestations: dorsal - sagittal – coronal



Source: (De León Reyes and al., 2020, p. 02)

Moreover, one should know that the corpus callosum is divided into four different anatomical regions; these are the *rostrum*, genu, body, and *splenium*. The anterior part of the corpus callosum is connected to the anterior cortical regions (the prefrontal cortex); however, its posterior parts are linked to the different cortical regions, i.e. parietal, temporal and occipital areas. Its good medullary structure allows for the rapid transmission of information between the different lobes of the cerebral hemispheres within a time period ranging from 20 to 50 milliseconds. Further, the fibers that make up the corpus callosum can be homogeneous or different depending on the nature of the cortical regions that connect them (Dominguez J. V., 2020, p 262). The corpus callosum development begins between the tenth and eleventh week of pregnancy and continues in its front-to-rear part to reach its full shape starting from the twentieth week of pregnancy. The functional and spinal growth of nerve fibers continues until adolescence.

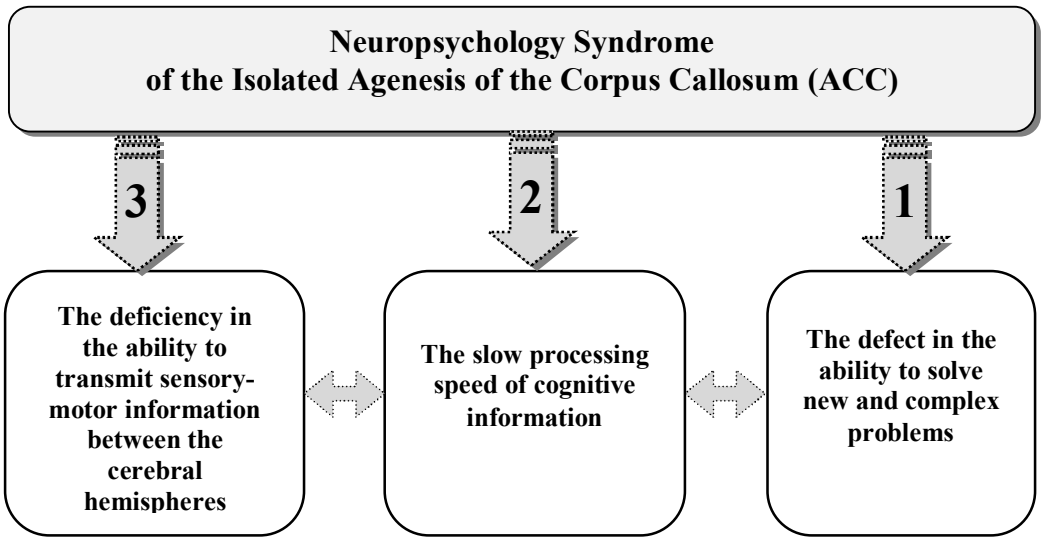
It is worth indicating that agenesis of the corpus callosum (ACC) is among the most common brain malformations, with an average of one case in 4000 to 5000 people. There are two types of ageneses of the corpus callosum (ACCs), i.e. the total ACC and the partial ACC which occurs when only one part of the corpus callosum grows. This early developmental defect occurs between the seventh (07^{th}) and the twentieth (20^{th}) week of fetal life. The causes of this defect are numerous; the main ones can be genetic, neurovascular and toxicological. It should also be mentioned that agenesis of the corpus callosum (ACC) may be isolated in the case of no injuries or comorbidities. Therefore, the prognosis would be better than in the case of the agenesis of the corpus callosum associated with other diseases (Dominguez, J. V, 2020, p 263).

The research progress and the studies that have previously been conducted on the different disorders observed in children suffering from isolated agenesis of the corpus callosum, in addition to the accumulated scientific knowledge emanating from these research studies over the past twenty years, prompted researchers to suggest the hypothesis of the "*Neuropsychology syndrome of corpus callosum agenesis*". Regarding, Brown and Paul (2019), they went on to describe the presence of this symptom in children suffering from total agenesis of the corpus callosum of the primary or isolated type without concomitant neurological or metabolic diseases and with a good level of knowledge, as illustrated in Figure 2. This same figure shows the triple structure of the isolated agenesis of the corpus calloussyndrome. This syndrome was described according to three basic symptoms. The first one concerns the deficiency in the ability to transmit sensory-motor information between the cerebral hemispheres, the second

one is the slow processing speed of cognitive information, and the third one is about the defect in the ability to solve new and complex problems. This primary pathological triad results in a series of cognitive disorders such as the cognitive deficit and cognitive flexibility disorders, visual processing disorders and verbal information impairments, disturbances in understanding the implicit meaning and deliberative dimension of language, in addition to some social difficulties like the difficulties in expressing and recognizing emotions, difficulties in interpreting and adapting to social situations, etc. All these impairments are observed in children with isolated agenesis of the corpus callosum.

Figure 2

Triple Structure of Agenesis of the Corpus Callosum (ACC) Syndrome according to Brown and Paul (2019)



Source: Prepared by the author

On the other hand, it was revealed that agenesis of the corpus callosum is a cerebral malformation that may occur alone or in association with other accompanying clinical signs. Previous studies have suggested the lack of heterogeneity in the neuropsychological manifestations in small samples of individuals who suffer from agenesis of the corpus callosum. In this regard, the study of Vanessa and al. (2013), which was an objective and critical reading of various previous studies (a systematic review) that focused on the topic under study, indicated that the neuropsychological and cognitive features of individuals suffering from agenesis of the corpus callosum are characterized by cognitive abilities at *below-average* and*average levels*. Indeed, the clinical examination showed that most of these cases suffer from disorders in several psychological,

cognitive and linguistic areas, in addition to difficulties in pragmatic language skills and arithmetic skills.

As previously asserted by Vanessa and al. (2013), and confirmed by preliminary methodological studies conducted on similar cases, and revealed that individuals suffering from agenesis of the corpus callosum exhibited limited expressive and receptive language abilities, with confined spatial and visual attentional skills. In addition, it was found that these disorders appear during childhood in varying proportions. Indeed, investigation established that 75% of them suffer from expressive and receptive language disorders, 43% from visual and spatial disorders, and 36% from attentional disorders. Furthermore, 46% of the cases with agenesis of the corpus callosum (ACC) may present difficulties of slow processing speed of cognitive information, in addition to difficulties with verbal short-term memory, long-term spatial visual memory, and lexical linguistic skills. Similar studies also confirmed the issue of the absence of heterogeneity in the neuropsychiatric clinical rules in individuals suffering from agenesis of the corpus callosum, which is certainly due to the high probability of neurological injuries (like epilepsy) that usually accompany the corpus callosum agenesis. In addition, so far, only a limited number of studies have been carried out on large samples of cases, which would allow establishing the relationship of this congenital malformation with various neuropsychological functions. It is deemed important to specify that 64% of these studies were conducted on samples involving only three cases or less.

Among the rare studies that were directly concerned with phonological skills in the agenesis of the corpus callosum, it is worth mentioning the paper of C.M. Temple and J. Ilsley that was published in 1993. In this study, entitled "*Phonemic discrimination in callosal agenesis*", the researchers used a set of phonological tests, i.e. phonological discrimination between similar and dissimilar pairs of phonemic units, and also between pairs of phonologically identical words (example: lack-lack, badge-badge). They also used words that differ according to one phonemic unit within the word (example: tub-tug) that is considering the length of words and their degree of familiarity to children. Pairs of phonologically similar and different non-words, pairs of repeated multisyllabic words, and multisyllabic non-words were also used; these were presented in a random order of verbal repetition to the child. This was done through repeated applications of longitudinal data in spaced time intervals for the same cases. The results of this study showed that the cases actually suffered from significant difficulties in achieving rhyme tasks and understanding the organization of vocal units, especially when it comes to distinguishing between repeated word pairs and

polysyllabic non-words. In this study, it was assumed that the cases under study suffered from disorders in the initial recording and analysis of verbal material, as the role of the corpus callosum in auditory processing remains unknown.

However, most researchers explained the aforementioned disorders by the possible existence of a type of temporal lobe interaction which is essential in achieving contrastive discriminations between audio materials. These findings were further supported by recent studies that elucidated the most important relationship existing between the corpus callosum function and the language proficiency level in children. In this context, it is worth recalling the study of Bartha-Doeringand al. (2020) in which various tests of linguistic competencies were carried out based on the functional magnetic resonance imaging (FMRI) method, with the aim of establishing the relationship between the corpus callosum size and language network connectivity in 38 normal children with ages between 06 and 12. The study showed that the large size of the posterior corpus callosum parts is significantly associated with verbal fluency and vocabulary. In addition, a positive correlation was also established between the anterior part of the corpus callosum and the verbal span. Moreover, the findings indicated that the children who were characterized by a larger corpus callosum size exhibited greater neural interaction between the two hemispheres of the brain, in the network of linguistic activities; these children also showed good abilities in various linguistic areas. It should be noted that this study provided the first evidence of the existence of a direct connection between the corpus callosum function and the linguistic network activities. It also highlighted the role that the corpus callosum plays in stimulating the process of integrating the information provided by the cerebral hemispheres, while taking into account the secondary role played by the temporal medial and lateral regions of the right cerebral hemisphere in the network of language activities.

Within the context of the neural basis for phonemic processing, Lacey (1985) referred to the poor usage of the brain-stem of (09) children with corpus callosum disorders; these children showed a central conduction delay in the medullar area. In addition, auditory information passes through the brainstem to the temporal lobes, and any slight defect can implicitly affect the processing of vocal units. These findings were also supported by those reported in the study of Temple and Ilsley (1993). Therefore, despite the disturbances recorded in some cases at the auditory discrimination level, it was found that two of them were normal on both language comprehension and production tests and on intelligence tests. However, some disorders of oral language development were detected in most of them, especially with respect to the morphological dimension. These

results prompted some researchers in the study to assume that auditory discrimination disorders are mostly present in children suffering from agenesis of corpus callosum; they therefore concluded that these disorders have a significant impact on oral language development in children and adults. Based on the aforementioned findings, one may appreciate the status of the degree of disruption of phonological manifestations in the agenesis of corpus callosum of the isolated type. Indeed, in this study, it is assumed that the clinical case under study suffers from recorded phonological disturbances, particularly in the repetition of familiar and unfamiliar polysyllabic words. These disturbances have an impact on the production and perception of sound units, which engenders speech incomprehension due to the severity of the phonological disorder.

3. Presentation of a Case (X) Suffering From a Complete Agenesis of Corpus Callosumof the Isolated Type

The five-and-a-half-year-old child (x), who was accompanied by her parents, came to the Language and speech assessment office that is located in the Polyclinic of *Harawa* which is affiliated with the Neighborhood Public Healthcare Center of Algiers (Algeria). The speech language pathologist discussed the steps to follow with the child's mother because the child presented a greater language delay compared to other children of her age. Then, through the initial clinical study, a case history was established. The medical history information obtained revealed that the child ranks last after 02 sisters in the family; the pregnancy conditions were normal with Caesarean delivery on time, without complications after delivery. Her mother tongue is the Arabic dialect that is currently practiced in the city of Algiers.

The child had some health problems; the most important one was the discovery, when she was two years old, of complete agenesis of corpus callosum (ACC) of the isolated type. In addition, magnetic resonance imaging (MRI) confirmed the absence of concomitant cerebral malformations. Moreover, other complementary tests indicated the absence of concomitant auditory or neurological disturbances. In addition to her medical history, it was found that her psychomotor development was normal and she did not present any particular behavioral or nonverbal communication disorders. However, the child suffered from a remarkable delay in the development of oral language skills, at various levels, especially at the lexical, phonological and syntactic-pragmatic levels.

4. Presentation of the Study Tools –Phonology Tests with regard to the General Clinical Linguistic Features

The phonology tests to perform were selected in accordance with the general clinical linguistic features to be assessed, based on the case study method

in speech and language pathology. The technique used is a part of the Chevrie-Muller language examination tests (EEL; Forme P: 04-05 years). Malek (1993) was first to develop the adapted Algerian version of these tests. Three types of phonological tests are presented below:

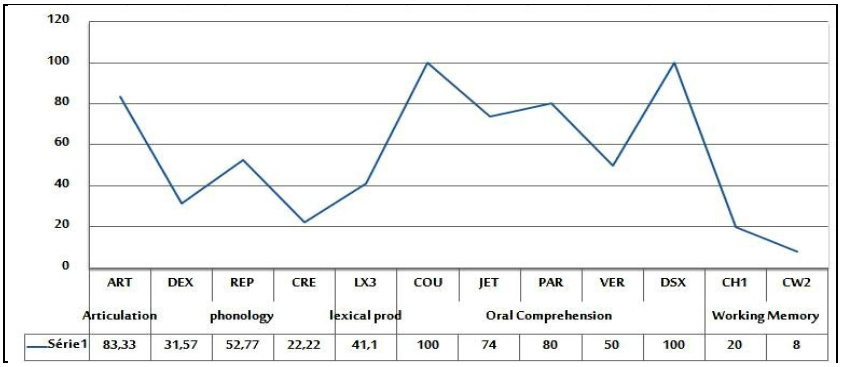
- ***The first test*** serves to assess the *phonological* awareness skills through the lexical nomenclature (DEX), using 06 pictures used by Chevrie-Muller, 08 pictures used by B. Ducarne in the field of aphasia, and 30 pictures previously selected by researcher Malek (1993); these last 30 pictures are part of the linguistic world of the Algerian child aged between 04 and 05.
- ***The second test*** deals with the repetition of easy words. This test, which was adapted to the Algerian socio-cultural environment, consisted in repeating a list of 46 easy words that were taken from the aphasia test developed by researcher B. Ducarne.
- ***The third test*** is about repeating difficult words. This test requires decoding the linguistic unit and involves the intervention of retention, coding and verbal achievement. However, this test differs from the previous one with respect to some problems related to the type of words used, as most of them were long syllable-words, i.e. 03 to 06 syllables in six consecutive words chosen according to the Algerian socio-cultural context. The silent units, i.e. /lm/, /mr/, /kc/, /mh/, /ks/, /bth/, appear in many words. Finally, most of the words used do not belong to the age-appropriate lexicon.

5. Results of the Phonological Awareness Tests for the Case under Study based on the General Clinical Linguistic Profile

The clinical linguistic profile represented in Figure 3 depicts all the results obtained for the case under study. These results, which are expressed in percentages, depend on the type of test used are based on the conversion tables from scaled scores to standard scores.

Figure 3

General clinical linguistic profile of case (X)



Source: Prepared by the author

At first glance, it is evident that the low percentages in the results of some tests, which were below the average. These were the two phonology tests DEX and CRE, as well as and the tests of memory of numbers and sentences. Moreover, the results of the lexical balance test (LX3) were also below the average by 41.1%, which reflects the limited development of the lexico-semantic level, either because the child did not use the appropriate words and did not diversify them, or because she used other words in place of the right ones. However, verbal comprehension (VER) was within the average 50%. In addition, the case tended to give very short answers that were often characterized by the lack of clarity while using compensatory means (for example: pointing the finger at pictures; using demonstrative pronouns). Unlike what was found with the verbal comprehension, the child passed in all items of the DSX test. Furthermore, the results of the concordance comprehension test (PAR) were estimated at 80%, and for the color comprehension test (COU) they were estimated at 100%.

Finally, those results were estimated at 74% for the test of understanding unit of words (JET). As for the retention or memory tests, the case under study exhibited considerable difficulties in repeating sequences of numbers (CH1) in an ordered form, and great difficulties in repeating simple and compound sentences (CW2).The results of the first lexical denomination phonology test (DEX) were below the average by 31.57%, while the case under study succeeded in producing 06 phonologically correct words out of 19 correct words at the lexical semantic level. Most of the phonologically incorrect words were characterized by the same feature, i.e. phonological omission, which mainly affected the beginning of multi-syllable words (two-syllable and three-syllable words), in contrast to the single-syllable words which were correctly pronounced. In most of the answers the last syllable of words was accurately pronounced (Table 1).

With regard to the second phonology test, the results showed that the case could repeat easy words (REP); this came with an average of 52.77%. Indeed, 19 words were repeated phonologically correct out of 36 words that were actually repeated. It turned out that the phonological disturbances affected especially monosyllabic words, with total ambient silence. Nevertheless, the case under study got the rate of 22.22% for the repetition of difficult words (CRE) test and showed a tendency to phonological deletion that mainly affected the initial syllables of difficult words. These findings are summarized in *Table 1* given below.

Table 1

Examples of Results in Phonological Tests between the Lexical Nomenclature and the Repetition of Easy and Difficult Words Tests

Phonological Test	Expected Word	Repeated Word
Phonology Test in Lexical Nomenclature (DEX)	/tobsi:/	/si:/
	/mašṭa/	/ṭa /
	/du:da/	/da/
	/mafta:h/	/ta:h/
	/ni:f/	/ni:f/
Easy Word Repetition Test (REP)	/kta:b/	/tab/
	/lba:s/	/'as/
	/skan/	/kan/
	/sma/	/ma/
Difficult Word Repetition Test (CRE)	/lmaɣṛab/	/ṛab/
	/ksabtha/	/ha/
	/lbarasyu:n/	/su:n/
	/kṛumbi:ṭ/	/bi:ṭ/

Source: Prepared by the author

6. Discussing the Results of the Case under Study in Phonology Tests based on the General Clinical Linguistic Feature

The phonological omission feature is viewed as the most important phonological disorder that characterized the phonology of the case. According to Brin& al. (2018), this feature depends on the phonological context within the word structure. In this case, the processes of phonological deletion were almost constant (quasi-systematic); they mainly affected the initial syllables of words. The phonological disorders occurred primarily in the repetition of difficult words, which can be attributed, from a psycholinguistic point of view, to two main factors. The first one concerns multiple silent structures within the syllables of these difficult words, while the second one is assigned to the fact that the case was not familiar with these words; these are unfamiliar words that do not belong to the linguistic context of the case under study. In addition, the phonological deletion characteristic appeared *secondarily* in the lexical naming task.

The phonological omission feature particularly affected two-syllable and three-syllable words, as opposed to monosyllable words that showed no phonological disturbance. The deletion came *tertiarily* in the repetition of easy words, which is due to their monosyllabic structure. This can be explained by the weak analytical phonological abilities of the case. In this context, the researchers De Weck and Marro (2011) indicated that the development of the phonological

system in the child is based on two types of skills. The first type concerns the set of cognitive and analytical skills that allow making the phonological distinction between different types of phonological elements (sound units and linguistic syllables), and the second one relates to the phonological cognitive representations that the child gradually develops.

The results of the present study, which suggest that the case under consideration suffers from a lack of understanding the organization of vocal units, are in good agreement with those of C.M. Temple and J. Ilsley (1993), especially with regard to polysyllabic words. Therefore, it can be concluded that case (X) actually suffers from disorders in the initial recording and processing of audio material; this is further confirmed by the quasi-constant deletion of primary linguistic elements in polysyllabic words. It is also interesting to cite the recent study of Bartha-Doering et al. (2020) who found a positive correlation between the anterior part of the corpus callosum, verbal span and lexical balance. It should be known that case (X) actually showed lexical poverty and severe difficulties in memorizing sentences and number sequences, which reflects the limited capacity of verbal memory and consequently hinders the achievement of the auditory discrimination activity of linguistic elements. Similarly, some recent studies, such as that of Siffrediand al. (2017), which further support this proposal, showed that the neural activation and memorizing strategies used by children suffering from agenesis of corpus callosum and normal children are different. This study is actually viewed as a first step in understanding the functional brain networks on which higher cognitive activities depend in children with corpus callosum agenesis (CCA).

The general clinical linguistic profile of case (X) presented a set of important data that are directly related to the level of phonological therapy. It should be noted that the most important data are those linked to functional memory and to the level of verbal comprehension and lexical expression as well in the case under study. Regarding Vanessa et al. (2013), they conducted a study in which they classified the different disorders in children with agenesis of the corpus callosum. Indeed, case (X) was found to belong to the first category which involves children with agenesis of the corpus callosum who suffer from expressive and receptive language disorders. Moreover, the results of the phonology tests prompt us to go back to the most important feature, i.e. the slow speed of processing cognitive information in children with agenesis of the corpus callosum that Brown and Paul (2019) mentioned in their description of the corpus callosum agenesis syndrome. It is believed that the above findings explain, to a large extent, the semi-constant phonological omissions occurring in polysyllabic

words, and the retention of the last syllable received by the case's ear. Moreover, the slow cognitive information processing speed also clearly explains the poor temporary retention and repetition of various verbal materials.

7. CONCLUSION

The present clinical study contributed to highlighting the features of a case presenting agenesis of the corpus callosum syndrome of the isolated type, while particularly focusing on the phonological side. The results obtained highlighted the main troubles faced by the case under study, especially those related to the repetition of multi-syllable and unfamiliar words. Indeed, it was found that the case presented some phonological deletions in most verbal productions, which reflects the limitations of the perceptual-analytical abilities of phonological components in the child. Moreover, the different lexical disorders recorded in the general clinical lingual profile also explain the insufficient capacities of the child in the temporary retention of verbal materials (types of sentences, number series). Consequently, based on the above mentioned findings, it is highly recommended to detect any speech–language pathology and select the most suitable orthophonic treatment, with a primary speech therapy assessment that includes various fields (linguistic, neuropsychological, communicative, as well as various sensory, motor and cognitive acquisitions) that can be affected by agenesis of the corpus callosum. Early evaluation allows the speech language pathologist (SLP) to have a clear idea and to develop an in-depth picture about the case's features and what kind of lifestyle her family is experiencing. Recall that the main purpose of the study was to develop a comprehensive program for the treatment of symptoms of the isolated agenesis of the corpus callosum.

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