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Disorders of speech and language in child with agenesis of the corpus callosum - case study

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Abstract

Introduction: The corpus callosum is the largest structure of the nerve fibers of the brain. Its main function is to coordinate the transfer of information between the right and left hemispheres of the brain. Agnesis of the corpus callosum is one of the most common defects in the CNS. ACC can be isolated as a defect (approx. 20%) or extend as a disorder co-morbid abnormalities of other systems and organs (approx. 80%). In patients who have a disorder of the corpus callosum work points to the plurality of delays and disturbances. They are related to physiological disorders, developmental, social and disorders of communication, speech and language. The aim of the study was to description of psychomotor impairment in a child with agnesis of the corpus callosum.

Material and method: The object of the study was a three-year-old girl diagnosed with agnesis of the corpus callosum. Pregnancy was complicated, in the study of prenatal suspicion of CNS

defects, threatening preterm delivery at 32 weeks gestation. In neonatal microcephaly found in ultrasound agnesis of the corpus callosum. EEG is abnormal. Made imaging CNS in sedation indicates holoprosencephaly. The current diagnosis of the child is also hypopituitarism, hypernatremia separable and short stature.

Conclusions: Agnesis of the corpus callosum is a congenital defect and now there are not known methods of treatment of this deficit. A child who has the disorder need to be taken care of multidisciplinary team: neurologist endocrinologist, genetics, optometrist, pediatrician, psychologist, a physiotherapist, and the speech therapist to compensate for developmental deficits.

Keywords: Agenesis of the Corpus callosum, Language Disorders

Introduction

The human brain is a fascinating organ for years researchers. It remains a mystery to the human way of knowing. One part of the brain is the corpus callosum (CC). It is the largest structure of the middle part of the brain, and lived in the eighteenth century, Emanuel Swedenborg spread belief that it is in the corpus callosum is the human soul [1].

The corpus callosum is the largest structure of the nerve fibers of the brain. Its main function is to coordinate the transfer of information between the right and left hemispheres of the brain [2]. It is a part of telencephalon with rostral commissure, transparent partition, vaulted ceiling and marginal badge. It consists of more than 200 million fibers associative in the nervous system, making it the best educated by commissural [3,4].

The corpus callosum develops from about 10.-11. week of pregnancy and most intensively evolving in fetal life. Later, the second year of life doubles its size. Of particular importance for the efficiency and effectiveness of the corpus callosum is aging period, when the rate increases considerably the interaction between the hemispheres. CC expands to 25 years of age, and then slowly decreases in size [5]. The corpus callosum in panel (splenium), trunk (truncus), elbow (gene) and the beak (rostrum) - extending into the plaque and thereafter the bow in the lamina terminalis. In the initial stage of embryonic development hemispheres ventral forebrain combine only the bow. Axons crossing the midline of the brain takes place about the 14th day of embryonic development.

Discussed structure is designed to coordinate the actions of cortical centers of both hemispheres, and also plays a role in brain lateralization. Typically, the left hemisphere of the brain is dominant for speech and the right is responsible for visual-spatial relationships and emotions. The rear part of the corpus callosum, i.e. istmus acts as a relay visual information, while the middle section is intended to convey somatosensory information.

There are three types of disorders of the structure of the corpus callosum [6,7]:

- Type I - agnesis;
- Type II - none back of the CC due to the presence lipoma or unknown causes;
- Type III - hypoplasia of the corpus callosum, which is accompanied by other abnormalities of the brain;

The goal of the work was to description of psychomotor impairment in a child with agnesis of the corpus callosum.

Case study

A girl born in 2015. Susan has complicated pregnancy, the study found prenatal suspicion of CNS defects, threatening preterm delivery at 32 weeks of pregnancy (mother had founded Pressarium). In the perinatal period due to a defect CNS TORCH infection were excluded. In neonatal said microcephaly (head circumference at birth 31 cm), ultrasound agnesis of the corpus callosum. EEG is abnormal with the registration activities of paroxysmal. Made imaging the CNS indicates sedation holoprosencephalię type II or III. The current diagnosis of the child also points to the pituitary, hypernatremia separable and short stature.

Currently Susan is 3 years old, attends a therapeutic point of kindergarten. It requires the presence of a supportive teacher. The girl is covered by the care of multiple specialists, including speech therapists, educators, physiotherapists. Participates in therapies, which goal is maximized receive stimuli from the world. Due to the Susan disability coupled and the difficulty in determining the causes of the most common symptoms it should be emphasized, the therapy is not simple.

Referring to the possible consequences of the AGCC, Susan can be seen at:

• physiological disorders:

- hypertonia (high muscle tone).
- gastric reflux, difficulty chewing and swallowing,
- facial dysmorphia,
- increased sensitivity to some sensory stimuli (touch, texture)
- convulsions,

- spasticity,
- genetic diseases (girl is in the diagnosis of genetic counselor);
• Developmental disorders:
- the global nature of the delay,
- a significant delay in the development of gross motor - multiple attempts seat does not move;
- delayed speech development - only the vocals,
- clumsiness coordination,
- lack of control of physiological functions.
• Social Characteristics:
- Susan likes to stay among people a positive attitude to the world, it is characterized by cheerfulness,
- no self-awareness,
- social immaturity,
- a result not understanding of speech, it is able to interact language,
- difficulty in understanding simple messages language like: "Give," "lie down", "throw", as well as non-linguistic type: gesture of finger-pointing, "bye bye", significantly increased spasticity within the motor small and large all the more difficult child interaction
- anxiety,
- fear, anxiety - manifested into tears when the caregiver does not establish eye contact with the child or out of sight of the child.
• Communication:
- delayed speech and language development - the girl creates vocals,
- mimics the movements of the speech organs therapist
- do not use expressions onomatopeic;

It should be emphasized that due to the occurring hypertonia girl barely grasps objects. Observed the desire to improve or maintain a toy in her hand, increased muscle tension makes, however, that fists are clenched. Despite the limitations in the development of motor girl interested the world around her. Susan follows the gaze of an adult, seeking to share with it a common attention. Also changes the behavior in response to the emotions of others.

Currently, speech therapy classes are geared mainly to the general development exercises and phonetic-phonological. It is also used massage articulation sphere streamline the organs of speech, reducing sensitivity within the sphere, and also reduces the tension to the joint-temporal mandibulofacial. Current therapy is in the process of stimulating processes right hemisphere.

Agenesis of the corpus callosum is probably not the sole cause of her developmental deficits. However, the implementation of strategies to improve Susan language competence and communication significantly slows down speech therapist from the obligation to focus exclusively on the reasons for the developmental abnormalities. The most important is the state of the child and the programming and implementation of speech therapy.

Discussion

Agnesis of the corpus callosum (ACC) is one of the most common defects in the CNS. The incidence of disorders in the general population of 3-7 per 1000, and in children who have problems with psychomotor 2-3 of 100 births [2], [8], [9]. No statistically significant differences were found in the incidence of ACC, depending on gender [10]. Today the corpus callosum lesions can be detected already in fetal life (over 20 weeks) with: prenatal ultrasound or prenatal MRI. After delivery, the child can be examined also by CT [1]. Usually agnesis of the corpus callosum occur occasionally. However, it happens that the current transfer is the tendency of the gene family by autosomal recessive, or X-coupled chrmosome. ACC may exist as isolated defect (approx. 20%) or extend as a disorder co-morbid abnormalities of other systems and organs (approx. 80%) [11], [12]. Among the drawbacks of the central nervous system of coexists ACC: hydrocephalus, microcephaly, encephalocele, porencephaly, holoprosencephaly, lissencefalia, Dandy-Walker syndrome, spina bifida as well. However, among the disadvantages of non CNS: hypertelorism, little mandible, cleft palate, malformation fingers dysplastic kidney, diaphragmatic hernia, and congenital heart disease [10], [13]. More than half of all patients noted the occurrence of epilepsy and mental retardation [14]. It needs to be highlighted that, that the agnesis of the corpus callosum symptoms likely depend on other brain malformations. It indicated that approximately 30-45% of the cases can be clearly specify the cause of the disorder of the corpus callosum. In 10% of cases are caused by chromosomal abnormalities, au 20-35% other genetic syndromes. Among the bands dysmorphic, characterized by the AGCC can be distinguished the following components: Mowat-Wilson CRASH Aicardiego, Seckel, Rubinstein-Taybi syndrome, Sotos, Opitz-Frias [1], [2], [15].

The International Organization for Disorders of the corpus callosum indicates delays in psychomotor development in patients with AGCC. Among the effects of the agnesis the corpus callosum distinguished [1], [16]:

• physiological disorders:

- blurred vision (near / far-sightedness, nystagmus, strabismus, problems with the assessment of distance)

- hypotonia (low muscle tone), hypertonia (high muscle tone).
- gastric reflux, difficulty chewing and swallowing,
- facial dysmorphia (abnormalities of the head and face)
- lowered pain threshold, but may be more sensitive to certain sensory stimuli, for example. Selected types of touch, texture,
- sleep disorders (difficulty in falling asleep, shallow sleep, nocturia).
- convulsions,
- spasticity,
- damage to hearing,
- problems with defecation,
- less: defects in the genitourinary system (e.g.,. Cryptorchidism), arrhythmia, skeletal defects, metabolic disorders, and genetic diseases.

• Developmental disorders:
- delaying the development of gross motor (sitting, walking, cycling, etc.).
- delayed speech development,
- clumsiness coordination,
- delayed control physiological functions manifested inability to use the toilet.

• Social Characteristics:
- cheerfulness, positive disposition,
- liking being with others,
- social immaturity,
- lack of self-awareness,
- inability to understand the perspective of others,
- difficulty within the meaning of language messages,
- difficulty in attention,
- restlessness or hyperactivity,
- fear, anxiety,
- obsessive-compulsive disorder.

• Communication:
- speech and language delays,
- misinterpretation of nonverbal communication,
- difficulties with complex tasks, abstract reasoning, ability to cope with difficult situations (managing money, planning)
- ignorance of their own behavior and limits the consequences of their own behavior,

- difficulty understanding slang, sarcasm and subtle humor,
- difficulty in understanding the emotions of others.

In the treatment of patients who have agnesis of the corpus callosum they proposed various forms of equalization deficits. One of them involves the need to use behavioral and cognitive training in early childhood (www.thriteen.org/curious/episodes/agenesis-of-the-corpus-callosum). Other authors [2] emphasize the need to build a system of language, with understanding of underlying messages. They propose to start speech therapy exercises right hemispheres, followed by stimulation of the left hemisphere. This is important because patients with AGCC both cerebral hemispheres are equivalent. Among the exercises right hemispheres stands out: recognizing, identifying and memorizing the vowels, simultaneous exercise of memory, repetition, pointing, calling onomatopeic expressions, repetition and automated playback sequences, as well as the identification of prosodic features. Exercise, which will be a bridge between therapy strategies right hemispheres to left hemispheres is to repetition of words other than onomatopoeic. This is important mainly due to activating thus both hemispheres simultaneously. In the left hemisphere speech centers it will therefore be essential in order to further stimulate its processes. You can do this by: training sequence sound and hearing, laying picture stories, dialogue exercises as well as exercises sequential memory.

In the treatment of patients with agnesis of the corpus callosum, remember that this is a congenital defect, and are now known treatments for this deficit. Child in whom the disorder is to be taken care of multidisciplinary team: neurologist endocrinologist, genetics, optometrist, pediatrician, psychologist, a physiotherapist and speech therapy [1].

Literature

1. Nowak K, Ogorzałek A. Agnesis of the corpus callosum - speech therapy point of view. In: Michalik M. (Ed.). Biological conditions of development and speech disorders. Kraków 2011; 2: 305-317.

2. Kaczan T, Śmigiel R. Children with defects of the corpus callosum - to support the development in the first years of life. In: Kaczan T, Śmigiel R. (Ed.). Early intervention and support the development of children with genetic diseases. Kraków 2012: 111-133.

3. Gaskill Sarah J, Marlin Arthur E. Neurology and neurosurgery of the children. Krakow 2000.

4. Dove KB. Functional Anatomy of the central nervous system. Warsaw 2000.

5. Nowicka A. Cooperation left and right hemispheres: the role of interhemispheric commissures. Psychology - Ethology - Genetics 2000; 1: 39-60.

6. Rauch RA, Jinkins JR. Magnetic Resonance Imaging of the Corpus callosum dysgenesis. In: Lassonde M, MA Jeeves. (Ed.). Callosal Agenesis. Advances in Behavioral Biology. Boston 1994; 42.

7. Bekiesińska-Figatowska M, Walecki J. Pathology corpus callosum in images computed tomography and magnetic resonance imaging. Polish Journal of Neurology and Neurosurgery 2001; 5: 829-840.

8. Bodensteiner J, Schaefer GB, Breeding L, Cowan L. Hypoplasia of the corpus callosum: a study of 445 consecutive MRI scans. J. Child. Neurol. 1994; 9: 47-49.

9. Wang LW, Huang CC, Yeh TF. Major brain lesions detected on sonographic screening of apparently normal term neonates. Neuroradiology 2004; 46: 368-373.

10. Janiak K, Respondek-Liberska M, Liberski PP. Agenesis of the corpus callosum - the importance of prenatal diagnosis. News Neurology 2009; 9 (3): 194-202.

11 Bedeschi MF, Bonaglia MC, Grasso R. Agenesis of the corpus callosum: clinical and genetic study in 63 young patients. Pediatr. Neurol. 2006; 34: 186-193.

12. Cavicchioni O, Gomes DM, Leroy B. et al. Prenatal Diagnosis of de novo (7; 19) (q11.2; q13.3) translocation associated with a thick corpus callosum and Wilms tumor of the kidneys. Prenat. Diagn. , 2005; 25: 876-878.

13. Lemke M, Pilarska E, Wierzba J, Balcerska A. Agenesis of the corpus callosum - clinical and genetic aspects. Annales Academiae Medicae Gedanensis 2007; 37: 71-79.

14. Dunin-Wąsowicz D, Chrzanowska KH. et al. Agenesis of the corpus callosum - neurological and genetic aspects. Children's Neurology 2001; 10: 53-61.

15. Kluczewska E, Golus T, Kluczewska-Zygan K. Congenital brain in children, In: Nowakowska-Kempna I, Pluta-Wojciechowska D. (Ed.). Studies in neurologopedy. Krakow, 2010: 59-75.

16. https://nodcc.org/corpus-callosum-disorders