CASE STUDY

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NEUROPSYCHOLOGICAL DIAGNOSIS OF A FEMALE PATIENT WITH ARNOLD-CHIARI MALFORMATION TYPE I

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SUMMARY

Background: Arnold–Chiari I malformation is a congenital malformation of the hindbrain characterized by displacement of the cerebellar tonsils into the foramen magnum, pressure on the fourth ventricle, and decreased fluid flow to the basal cisterns. It is a mild form of the neuro-cranio-vertebral syndrome (Arnold–Chiari syndrome) because in many cases it is asymptomatic or has few clinical signs and symptoms of mild severity.

Case study: The case study presented in the article concerns a 16.5-year-old patient with Arnold–Chiari I malformation following surgery. Based on the neuropsychological diagnosis, clinical symptoms that might result from the course of structural and functional changes in the hindbrain in Arnold–Chiari I syndrome were described and explained. The following were observed: selective difficulties in planning and concentration (maintaining the direction of action) and a weakening of visual perception (described by the patient as a subjective state of derealization).

Conclusions: The results of the neuropsychological diagnosis point to certain practical implications for the further therapeutic treatment of patients with a mild form of Arnold–Chiari I malformation, especially as there is a risk of clinical symptoms changing over time due to the posibility of progressive neurodegenerative changes in the hindbrain.

Key words: hindbrain dysfunction, derealization state, visual attention, perception, microgenetic theory, symptom formation

INTRODUCTION

The term“Arnold–Chiari malformation” (or “Chiari malformation”) comes from the names of the scientists who were the first to publish detailed case reports onhindbrain malformation consisting inan ectopia of the cerebellum into the occipital foramen magnum (Di Rocco, 2019; Ogryzlo, 1942) or on the presence of cerebellar hernia and a protrusion of the cerebellar vermis into the foramen magnum (Pruszewicz et al., 2007). According to Tubbs et al. (2016), it was Hans Chiari who first published illustrations of congenital abnormalities that are now referred to in the subject literature as Chiari malformation. In his publications (Chiari, 1891, 1895), he described birth defects of the cerebellum and distinguished three different types of these abnormalities. The first type is characterized by a degenerative change of the cerebellar tonsils without brainstem displacement (Pruszewicz &Wojnowski, 2007). There is an abnormal elongation of the cerebellar tonsils, which descend 5 mm below the foramen magnum into the upper part of the cervical spinal canal and may change with age (Blaser & Osborn, 2019). For this reason, there are some indications of mild neuropsychological symptoms (e.g., Greer, 2012; Houston et al., 2018).

Chiari malformations (CM) are classified based on their morphology and severity of anatomic defects, typically through imaging (or autopsy) (Kandeger et al. 2017; de Arruda et al. 2018), but also on the basis of effective surgery (Bhimani et al. 2018). It includes:

- **Chiari I** is the least severe and often found incidentally. It is characterized by one or both pointed (not rounded) cerebellar tonsils that project 5 mm below the foramen magnum, measured by a line drawn from the basion to the opisthion (McRae Line) (Arora et al. 2015).
- **Chiari II** consists of brainstem herniation and a towering cerebellum in addition to the herniated cerebellar tonsils and vermis due to an open distal spinal dysraphism/myelomeningocele (de Arruda et al. 2018);
- **Chiari III** involves herniation of the hindbrain (cerebellum with or without the brainstem) into a low occipital or high cervical meningoencephalocele (Bhimani et al. 2018).
- **Chiari IV** is now considered obsolete (Arora et al. 2015). Before it became an outdated diagnosis, it was already a more controversial and rare variant that showed severe cerebellar hypoplasia, similar to primary cerebellar agenesis. In older literature, one can find views that meningomyelocele herniation may be present (Cama 1995), while in more recent literature, based on neurosurgery performed (Hadley 2002) or the use of newer neurotechnologies for testing (Arora et al. 2015), it was concluded that the presence of meningomyelocele herniation should be classified as Chiari II with cerebellar atrophy.
- **Chiari V,** the most severe variant, represents cerebellar agenesis with occipital lobe descent and herniation through the foramen magnum (Kim et al. 2010; Tubbs et al. 2012: Giammattei et al. 2018; Markuna et al. 2014).

Patient with Chiari malformations may not have symptoms initially but may

develop them later in life (McClugage & Oakes, 2019). Depending on the type of Chiari malformation (types II, III, IV or V), we may observe a variety of neurological and neuropsychiatric symptoms involved over time (Blaser & Moore, 2019). It included headache, especially after sudden coughing, sneezing, or straining, neck pain, hearing or balance problems, muscle weakness or numbness, dizziness, difficulty swallowing, breathing, or speaking, excessive drooling, gagging, or vomiting, ringing or buzzing in the ears (tinnitus), curvature of the spine (scoliosis), and insomnia, depression, problems with hand coordination and fine motor skills and difficulty eating and an inability to gain weight. Symptoms are variable over time and we can observe even mental retardation (Pruszewicz et al., 2007; Bezuidenhout et al., 2019; Mangubat et al., 2014; Thunstedt et al., 2022; Mangubat et al., 2014; Kozubski&Liberski, 2020).

With reference to the presented case study, it will be important to bring the opinion of the McClugage et al (2019). These authors stated that as with many pathologies, the course of our understanding of the Chiari I malformation (CIM) has developed extensively over time. The early descriptions of the Chiari malformations by Hans Chiar, which is mention above, opened the door for future classification and research on this topic. However, even over a long timeframe, our understanding of the pathophysiology and, more importantly, treatment, remained in its infancy. As recently as the 1970s, CIM was not discussed in popular neurology textbooks. Syringomyelia is listed as a degenerative disorder with no satisfactory treatment. Radiation therapy was considered an option in treatment, and surgery was thought to play no role. During the last 40 years, equivalent to the duration of a neurosurgical career, our understanding of the pathophysiology and natural history of CIM, coupled with modern MRI, has improved the treatment paradigm for this patient population. More importantly, it has given us evidence confirming that CIM is a disorder responsive to surgical intervention, giving patients once thought to be destined for lifelong disability a comparatively normal life after treatment.

Rogers et al. (2018) conducted a meta-analysis using a protocol that included the keywords: cognitive function, socio-emotional, and/or psychiatric symptoms, language function, executive function in AMED, CINAHL, Cochrane Library, EMBASE, MEDLINE, PsycINFO, and Scopus databases. Articles meeting the following criteria were included in this review (i) examined children or adults with a clinically defined diagnosis of CMI, (ii) assessed cognitive function with a prospective examination, (iii) included at least one standardized instrument designed to measure general or specific domains of cognitive function, and (iv) were published in English in a peer-reviewed journal. Twelve articles were identified, including 783 cases aged 3 months to 64 years. General cognition, processing speed, and learning and memory appeared less affected, while language deficits appeared to diminish with age. Executive dysfunction was the most commonly reported cognitive impairment, while attention and working memory, and visuospatial and perceptual skills also appeared vulnerable. Numerous methodological limitations were identified that should be considered in interpreting the impact of

CMI and planning future investigations. Overall, there is currently insufficient evidence to describe a valid and reliable profile of cognitive impairment in CMI.

Mangubat (et al., 2014) pointed out that the clinical picture of neuropsychological functions is heterogeneous, varied across individuals,and determined by therange of structural and functional changes in the hindbrain without characteristic symptoms. Moreover, cognitive, socio-emotional, and/or psychiatric symptoms may occur as complications after surgery, even though surgical procedures often produce the expected and desired outcomes. These authors suggest that further research, including carefully designed case studies, is needed to gain a closer understanding of neuropsychological impairment in individuals with Chiari malformations.

The aim of this article is to present and explain the type and severity of the neuropsychological symptoms reported by the patient with a type I Arnold–Chiari malformation verified through detailed neuropsychological examination usingthe clinical-experimental and psychometrictesting approach.

CASE STUDY

Introductory information, research context, and documentation analysis

Patient Zofia A., aged 16.5 years, was referred by her parents for a neuropsychological examination to a private center called Gabinety Mindset (The "Mindset" Clinic Offices) in Poznań, Poland, due to her problems with concentration, learning problem, and a state of derealization. The parents reported that the decision to consult Zofia's difficulties in her mental functioning resulted from the neurological diagnosis and the upcoming follow-up visit in February 2022. Medical diagnoses: neuro-cranio-vertebral syndrome, Filum disease, a lowering of the cerebellar tonsils (Arnold–Chiari I syndrome), intramedullary cyst (idiopathic syringomyelia), and a curvature of the spine (idiopathic scoliosis). The examination was performed from November 22, 2021, to January 8, 2022. The patient was under the supervision of a neuropsychologist, neurologist, and neurosurgeon.

Observation and clinical interview

Clinical interview with the patient's parents conducted alongside observation

Cooperating parents, in agreement on the reported difficulties in the patient's mental functioning. Both are well-mannered, with narrative appropriate to the situation, balanced mood, and normal psychomotor drive. The conversation went smoothly, they discussed the main problems observed in their daughter, answered questions, and pointed no visible difficulties in cognitive and socio-emotional functioning. An interview included the girl's development in the early stages of life: she was born at term, after a spontaneous delivery, Apgar 10/10, without

complications during or immediately after delivery; infections, cerebral hypoxia, and other possible complications were excluded. The girl left hospital on the second day, everything proceeded normally.

In infancy, Zofia was breast-fed until 17 months of age; then she received bottled milk with lactose (no special diet was applied). The girl did not have any sleep problems and responded to verbal and non-verbal messages normally. Psychomotor development proceeded without deviations from the norm in all stages of early childhood (the parents reported no problems regarding the girl's speech, motor, cognitive and socio-emotional development). In middle childhood, the patient was a very good student at school (systematic and effective). At the age of 11, the first symptoms with pain in the lower limbs and spine began to appear. In early adolescence, a decision was made to perform surgery due to increasing health problems (difficulty walking, numbness of the lower limbs, pain in the neck area and the occipital region of the head). The parents reported that the patient had no vision or hearing impairment; there were no craniocerebral injuries or other neurological diseases, either. According to them, the girl had no mental or behavioral disorders (she caused no upbringing problems and required no psychiatric consultation). For approximately two years, the following symptoms have been present: problems with concentration, with effective learning, and the need to calm down when the surrounding world seems to be "hazy, as in a movie" (the patient called this state derealization).

Observational data and overall assessment of the patient's mental state

The patient is a well-proportioned person of average height. During all assessment meetings, she wore neat casual clothes sports outfit and had undyed hair carefully gathered into a ponytail; the makeup on her face was barely noticeable. She looked her age and as one would could be expected in anthe examination situation. She came to meetings on time, behaved politely (e.g., she said "good morning" and"goodbye," did not interrupt others, and gave more or less elaborate answers when asked). No signs of stress or nervousness were visible during any of the visits; the mood was balanced and cheerful. Emotional expression appropriate for the situation, normal psychomotor drive, eye contact maintained. Zofia showed no aggression or self-aggression; hallucinations and delusions were excluded. Throughout the clinical examination, she was autopsychically and allopsychically oriented, remained fully responsive, correctly established and maintained verbal and non-verbal contact, and adopted a cooperative attitude. There were no visible signs of agnosia, apraxia, and aphasia, nor were there any profound executive dysfunctions. The patient did not exhibit any difficulties in understanding instructions, and the course and length of her narration were appropriate for the topic discussed. An even train of thought (with no signs of slowing down or accelerating) and appropriate dynamics of thinking (no digressions, no features of inertia or lability) were noted. She has full legal capacity.

Clinical interview with the patient

Zofia lives in Poznań with her family – mother, father, and sister. She has an older brother, aged 20, who studies social sciences and left home after graduating from high school. The sister is 12- years- old and is a good student at primary school. The mother has completed secondary education and has a paid job, continues to improve her qualifications, and keeps house. The father has a university degree, runs a company, and is a family man. The patient adds that emotions and feelings is a topic she is reluctant to talk about. Her relationships with her parents and siblings are good. Her brother usually helps her in difficult situations, the younger sister has a different personality and different interests, which is why they havedid not always understooand each other well. They have had separate rooms for several years, which has also resulted in fewer quarrels and greater psychological comfort for the subject. The patient does not mention other significant people. She also denies using psychoactive substances, not mention metabolic diseases and excludes eating disorders. She has experienced no head injury, epileptic fit, or loss of consciousness, also not received psychiatric treatment and has not attended psychotherapy or pedagogical or speech therapy as well. During her early school years, she had problems with spine; scoliosis was diagnosed, and exercises were recommended; she complied with the recommendations to varying degrees. A few years ago, she was diagnosed with Arnold-Chiari I syndrome; her somatic complaints were so strong that she was unable to function properly for several weeks (she had difficulty falling asleep, problems with concentration, and changing mood). This translated into more frequent absences from school. Throughout school she was a good student and gained end-of-year diploma certificates with honors; in the last two years she has not shown any desire to learn and has not cared about her school grades; what mattered was simply a passing. Currently, she has no specific interests, her only clearly defined goal is to go abroad immediately after graduation. The patient talks very much about the difficulties in functioning that intensified after surgery for Arnold-Chiari I syndrome.

Although the surgery was performed in 2019, the patient still has selective problems with concentration, lower learning efficiency, and a sense of derealization, namely,“an unexpected state of haze lasting a few minutes; everything is gray, as if enveloped behind in a fog.” During the state of derealization, the patient is restless and annoyed with others and tries to calm down, for example by taking a walk. Problems with concentration and learning efficiency appear regardless of derealization, although during that state she has problems doing multistage tasks that require a detailed action plan. She reports that she has always liked to plan, but she is too meticulous, and it usually takes too long. She is reluctant to speak about emotions, only making the following remark: “I have always had a tendency to ignore emotions in other people, although I do not feel myself to be selfish.” The patient denies symptoms of low mood and stressful situations that she is unable to cope with. Even when nervous, she does not have headaches or any somatic symptoms. Additionally, the course of treatment

does not give her any stress or anxiety. Until the clinical signs of Arnold–Chiari syndrome became apparent, she had been in good health.

The specificity of the use of research methods used in the case report

Due to the complex clinical pictureof the reported difficulties in cognitive and socio-emotional functioning,and due to the possible complications resulting from structural and functional changes in the brain following a neurological disease – Arnold-Chiari syndrome, the research methods used included a detailed clinical interview with the patient's parents and the patient herself, observational data, the analysis of the submitted medical and psycho-pedagogical documentation as well as standardized neuropsychological tests and clinical trials verified in scientific publications.

THE CLINICAL PROBLEM – A NEUROPSYCHOLOGICAL APPROACH

Based on the data from clinical interviews, observations, and the analysis of documentation, health history, and treatment records of the patient with Arnold–Chiari I syndrome, the following clinical problem was formulated in neuropsychological terms: assessing the current clinical picture of the patient's cognitive and emotional functions, identifying the potential causes of the reported symptoms, and explaining themechanism of the disorders, with particular emphasis on the structural and functional changes inthe brain resulting from Arnold–Chiari I syndrome. The results of the MRI examination conducted before and after surgery for Chiari I malformation are presented in Figure 1.

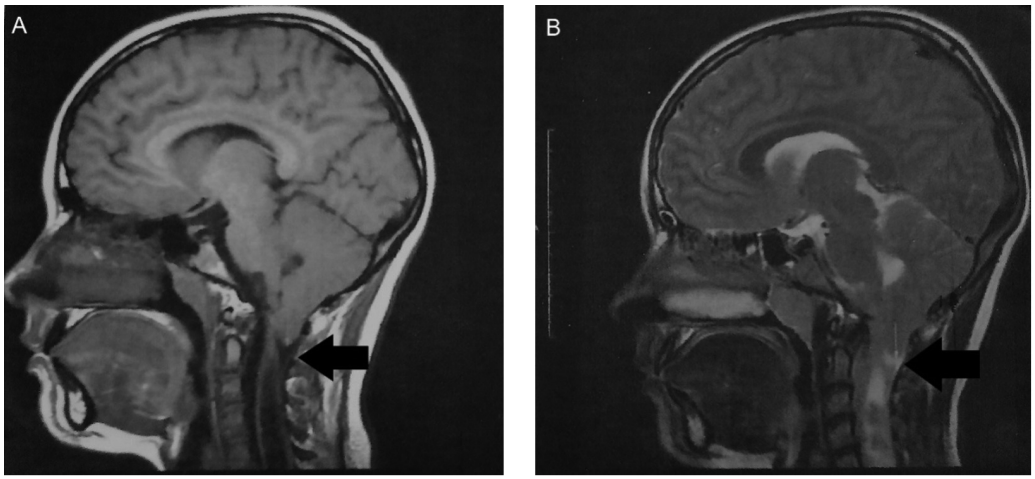

Fig. 1. Visualization of the patient's brain scan: A) after surgery: structural and functional changes inthe hindbrain: in 2020, elongated cerebellar tonsils and reduced syringomyelia (B) and before surgery: in 2019, elongated cerebellar tonsils and syringomyelia

Purpose, research questions, and hypotheses

The purpose of the neuropsychological study was to determine the clinical picture of cognitive and emotional functions with an indication of key resources and deficits in the patient's psychological functioning. Next, the study aimed to analyze the factors determining the appearance and severity of the reported clinical symptoms (cognitive and emotional) and to present the mechanism explaining the relations between the symptoms and the structural and functional changes in the patient's brain.

Research questions and hypotheses

The following questions and research hypotheses were formulated and subsequently verified in the course of the diagnostic procedure:

Research question 1: Do the reported clinical symptoms result from structural and functional changes in the central nervous system?

Hypothesis 1.1: The cognitive and emotional symptoms result from damage to the hindbrain.

Hypothesis 1.2: The cognitive and emotional symptoms result from derealization disorder.

Research question 2: Is there a likelihood of symptom variability?

Hypothesis 2.1: The clinical symptoms are due to brain changes in Arnold–Chiari I syndrome.

Research methods and tools

The neuropsychological study included research methods based on the experimental-clinical and psychometric testing models. Research methods were selected after a thorough analysis of the patient's medical and treatment history, after becoming getting acquainted with the results of medical tests and previous psychological and pedagogical opinions, and on the basis of patient observation and data from the clinical interview. A comprehensive neuropsychological diagnosis was carried out, using the research methods and tools presented below.

Mental state assessment

The general mental state assessment was made based on clinical interview data from the parents and from the patient herself, patient observation, and the analysis of the presented medical as well as psycho-pedagogical documentation.

Cognitive functions examination

Intellectual functions(thinking processes, language, visuospatial functions, and working memory).

Using the Stanford–Binet Intelligence Scale (SB-5; Roid et al., 2017), nonverbal and verbal intellectual functions were examined, namely: fluid reasoning (assessment of abstract thinking), general knowledge (assessment of conceptual thinking and language functions), quantitative reasoning (assessment of abstract

thinking and calculations), visuospatial processing (assessment of visuospatial functions), and working memory.

Examination of visual and auditory perception and attention processes.

The examination of visual and auditory gnosis was carried out using Włodzimierz Łucki's set of tests for the assessment of cognitive functions in patients with the brain damage (parts A and D; Łucki, 1995). Attention components (selectivity, divisibility, shifting) were tested using the Intelligence and Development Scales for Children and Adolescents (IDS-2)as adapted into Polish by Aleksandra Jaworowska, Anna Matczak and Diana Fecenec (2018) (subtests: "Divided Attention" and"Animal Colors") and the Attention and Perception Test (Polish acronym: TUS; the respondent's task is to select symbols among distractors and cross them out under time pressure; Ciechanowicz& Stańczak, 2006). Due to the already described state of "derealization," a clinical trial was carried out that consisted in drawing and explaining the symptom.

Examination of auditory-verbal and visual-spatial memory.

The individual components of memory and auditory-verbal learning were tested using the California Language Learning Test (CVLT; Delis et al., 1987, 1988; Łojek & Stańczak, 2010). The following were examined: encoding, reproduction, recall, recognition, learning verbal material, consolidation of memory traces in semantic memory, the number of errors in reproduction (cognitive control index), and the choice of learning strategy. The level of working visuospatial memory was assessedusing the Benton Visual Memory Test (method A, version C; Jaworowska et al., 2019).

Examination of executive functions

Selected executive functions were tested using the Intelligence and Development Scales for Children and Adolescents (IDS-2;subtests: "Listing Words,""Divided Attention,""Animal Colors,""Drawing Routes"), the Ruff Figural Fluency Test (RFFT; all indicators; Łojek & Stańczak, 2005; Ruff, 1996), and the California Verbal Learning Test (indicators: amount of perseveration and learning strategies). The following components of executive functions were examined: (1) fluency in generating patterns based on the figural criterion; (2) fluency in generating concepts based on the literaland categorical criteria jointly; (3) divisibility of attention, defined as the ability to control two tasks simultaneously; (4) switching attention, defined as the ability to choose and change action strategies and to inhibit reactions (the number of perseveration errors); (5) action planning.

Examination of motor functions and assessment of lateralization

Finger pose, spatial, oral, and dynamic praxis were assessedusing Włodzimierz Łucki'sset of tests for examining cognitive processes in patients with brain damage. Hand-eye coordination was assessed using the "Visual-Spatial Processing" subtest of the Stanford–Binet Intelligence Scale (SB-5). To assess lateralization, the researcher asked Zofia to write two arbitrary sentences (to determinehand dominance), look through a paper telescope (to determineeye

dominance), and move a piece of paper on the floor with her foot (to determine leg dominance).

Examination of emotional functions

The examination of emotional functions consisted in testing emotional intelligence components that mightreflect the reported clinical symptoms. The Two-Dimensional Emotional Intelligence Inventory (DINEMO; Matczak& Jaworowska, 2006) was used to verify the ability to recognize other people's emotions, control emotional reactions, express them appropriately,and take into account the importance of emotions in action. The ability to understand emotions was tested using the Emotional Understanding Test (Polish acronym: TRE; Matczak& Piekarska, 2011).

Neuropsychological examination results

Neuropsychological examination results, obtained using methods consistent withthe experimental-clinical and psychometrictesting models, were described and interpreted. Together with the data from the interview, observation, and documentation, these results constitute a full neuropsychological examination and the basis for the verification of the research hypotheses.

Results of the examinationof intellectual functions (thinking processes, language functions, visuospatial and working memory). The patient's intellectual functioning profile is in harmonious. There is a statistically significant difference between the non-verbal area (high level, II = 122 <113, 129>) and the verbal area (above-average level, II = 111 <103, 118>). Detailed analysis: **(1) fluid reasoning** is higher in the non-verbal area than in the verbal one; the patient correctly solves tasks that require inductive reasoning (from a single example to a universal rule) and deductive reasoning (inference based on general information), copes better with tasks presented in a symbolic form than with content-based tasks; **(2) the scope of general knowledge** accumulated in the course of formal and informal education indicates appropriate environmental stimulation (at school/home), preserved cognitive curiosity, and good development of language functions; **(3) visual-spatial processing results** indicate above-average efficiency of visual-spatial functions (correct orientation in space, very efficient arrangement of elements into figures); **(4) quantitative reasoning results** (abstract thinking and calculations) indicate efficient counting, correct solving of content-based tasksand tasks presented in the form of symbols requiring abstract thinking; **(5) verbal and non-verbal working memory** is at an average level (good memorization and processing of sentences/numbers).

Results of the examinationof visual and auditory perception and attention processes

Włodzimierz Łucki's clinical trials revealed normal visual and auditory gnosis, but it cannot be ruled out that visual perception and attention processes are impaired when derealization occurs. According to the patient, derealization is

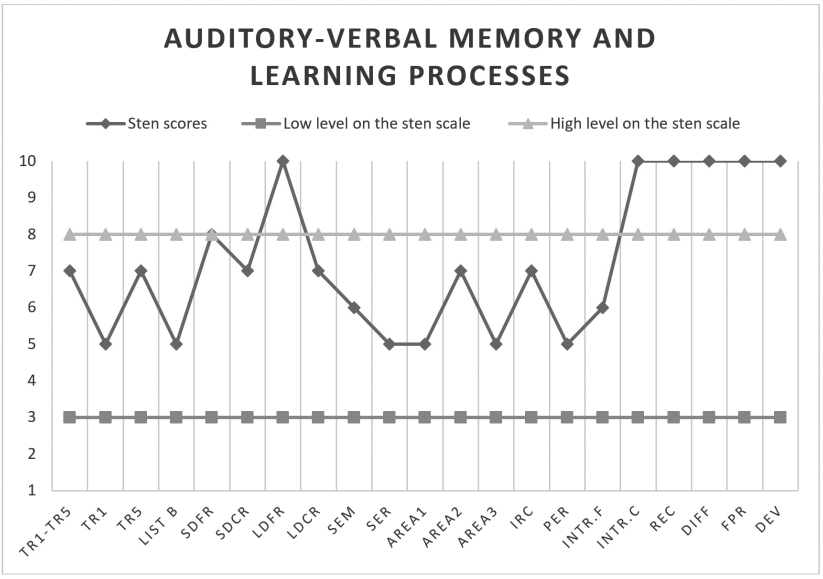

Fig. 2. California Verbal Learning Test results.
Legend: TR1-TR5 = correct recall on Trials 1-5 of List A(general measure of learning);TR1 = correct recall onthe first trial of List A;TR5 = correct recallonthe last trial of List A;List B = correct List B recall (distractor); SDFR= short delay free recall (correct recallimmediately after the distractor); SDCR = short delay cued recall (correct recallin individual categories immediately after the distractor; semantic memory range); LDFR = long delay free recall (correct recallafter a longer delay; long-term memory indicators); LDCR = long delay cued recall (correct recallin individual categories after a longer delay; semantic memory range); SEM= semantic clustering coefficient (a measure of semantic learning); SER= serial clustering coefficient (a measure of serial learning);AREA1= percent primacy recall (correct recallfrom the first four words of List A; primacy effect);AREA2= percent middle recall(correct recallfrom the middle area of List A);AREA3= percent recency recall (correct recallon the last four words of List A; recency effect);IRC= item recall consistency (a measure of effective learning); PER = perseveration errors; INTR.F= free recall intrusionerrors; INTR.C= cued recall intrusionerrors; REC= recognition hits (recognition of appropriate words among distractors); DIFF= differentiation (computed according to the following formula: [1−(FPR+ number of omissions/44)*100]);FPR –false-positive recognition errors; DEV= response deviation (computed according to the following formula: [FPR - number of omissions/ FPR + number of omissions])

a state “as if behind a fog”: gray, odorless, with noise. The patient drew and discussed this condition. The results of the “Divided Attention” and “Animal Colors” tests of the IDS-2 and the results of the TUS tests indicate the in harmonious efficiency of attention and visual perception processes, namely: good attention switching, low divided attention, weakened selectivity of attention (the faster the pace of work, the lower the accuracy and vice versa; some figures were “blurred”). Zofia reported that the tasks testing attention and perception were very interesting and motivating for her, particularly selecting symbols among distractors and crossing them out under time pressure.

Auditory-verbal and visuospatial memory test results. The results of the Benton Visual Memory Test indicate above-average short-term visuospatial memory (the patient made only one error and scored 9/10). The results of the

California Verbal Learning Test, including all indicators distinguished by Delis and colleagues (1987, 1988), Donders (2008), Łojek & Stańczak (2010), indicate: (1) correct verbal material learning (ascending curve, average level); (2) preserved ability to recall verbal information in the three phases of memory (encoding, storing, and recalling);(3) no difficulties in recognizing appropriate words among distractors; (4) maintaining cognitive control and skilful use of various learning and remembering strategies (few perseveration errors and intrusions). The results of the examination are presented in Figure 2.

Legend: TR1-TR5 = correct recall on Trials 1-5 of List A(general measure of learning);TR1 = correct recall onthe first trial of List A;TR5 = correct recallonthe last trial of List A;List B = correct List B recall (distractor); SDFR= short delay free recall (correct recallimmediately after the distractor); SDCR = short delay cued recall(correct recallin individual categories immediately after the distractor; semantic memory range); LDFR = long delay free recall (correct recallafter a longer delay; long-term memory indicators); LDCR = long delay cued recall (correct recallin individual categories after a longer delay; semantic memory range); SEM= semantic clustering coefficient (a measure of semantic learning); SER= serial clustering coefficient (a measure of serial learning);AREA1= percent primacy recall (correct recallfrom the first four words of List A; primacy effect);AREA2= percent middle recall(correct recallfrom the middle area of List A);AREA3= percent recency recall (correct recallon the last four words of List A; recency effect);IRC= item recall consistency (a measure of effective learning); PER = perseveration errors; INTR.F= free recall intrusionerrors; INTR.C= cued recall intrusionerrors; REC= recognition hits (recognition of appropriate words among distractors); DIFF= differentiation (computed according to the following formula: [1−(FPR+ number of omissions/44)*100]);FPR –false-positive recognition errors; DEV= response deviation (computed according to the following formula: [FPR - number of omissions/ FPR + number of omissions]).

Results of the examination of executive functions

The results indicate an in harmonious profile of executive functions. Resources and difficulties in functioning are specified. The patient's resources include **fluency in generating patterns** based on the figural criterion (the RFFT scoreis above average, the number of unique connections > 94th centile, 4 perseveration errors) and **attention switching as a change in the strategy of action and reaction inhibition** (average standard score on the IDS-2"Animal Colors" subtest = 10, few perseveration errorsin the RFFT and CVLT tests). The results indicate above-average non-verbal fluency, which is a measure of high cognitive flexibility associated with the appropriate choice and change of action strategy, with cognitive control maintained. No difficulties in initiating action and inhibiting the reaction have been observed, especially when there is a change of instructions. The patient does not respond impulsively, she thinks before she responds, the number of perseveration errors is at an average level. Difficulties concern the **fluency of generating concepts** based on the literal and categor-

ical criteria taken together (the result is near the lower limit of the average score on the "Listing Words" subtest of the IDS-2 scale); there may be problems with searching information in long-term memory – declarative and semantic. There was a decrease in **divisible attention and selective difficulties in action planning** (scores below average on the "Divided Attention" and "Drawing Routes" subtests of the IDS-2 scale; average error rates on the RFFT and the learning strategy of the CVLT).

Motor function test results and lateralization assessment

There were no disturbances in finger pose, spatial, oral, and dynamic praxis for the right orleft hands. The results indicate normal hand-eye coordination. There is a dominance of the right hand, leg, and eye.

Results of the examinationof emotional functions

Emotional Understanding Test score: sten 6 (average). DINEMO scores: overall score – sten 3 (low); score on the "Others" scale – sten 1 (very low); score on the "I" scale – sten 6 (average). The results of the study indicate in harmonious emotional intelligence development. The following were noted: correct recognition and understanding of emotions (average TRE overall scores and DINEMO "I" scale scores), partial ignoring of other people's feelings and emotions, expression of emotions inappropriate for the situation, or problems with controlling emotional reactions (low DINEMO overall scores and "Others" scale scores).

Neuropsychological therapy program

The patient took part in neuropsychological therapy program aimed at reducing the severity of clinical symptoms, strengthening resources, and developing social and emotional skills (cf. Pąchalska 2008). The therapy was aimed at inducing greater self-reflection in both the patient and her parents. These included training and support of the patient in:

- *engagement in activities* (e.g. walking, playing sports) to minimize focus on the state of derealization;
- *providing optimal learning conditions* through limiting access to distracting stimuli and incorporating relaxing music, and successively gradual learning in less favorable conditions (e.g., with access to distracting stimuli);
- *the ability to perceive the needs of others* (thinking about possible reasons for the appearance of certain emotional states in other people and what their point of view on a given situation might be);
- *the ability to pursuit of calmness and kindness* towards less well-liked people, as well as providing rest, relieving emotional tension and activating cognitive resources (readiness for action).

The therapist and the patient's family had also the goal to encourage and reward the patient for attendance in neuropsychological therapy, and to explain that it will strengthen her cognitive and emotional functions, improve visual perception and attention problem, as well as strengthen executive and emotional

functions.

DISCUSSION

We will present as the first in the discussion a verification of the research hypotheses and an explanation of the mechanism behind the reported symptoms, with an indication of patient resources and practical implications.

Verification of the first research question

Referring to the first research question, concerning the relationship between clinical symptoms and structural and functional changes in the nervous system, it can be concluded that hypothesis 1.1 was partially confirmed, while hypothesis 1.2 was rejected. Our study, as well as the studies of other authors (Brown & Pąchalska 2004), showed that, according to concepts concerning the microgenetic theory of the organization of mental functions, a neuropsychological symptom is a distortion of a given mental activity (cognitive, emotional or motor) resulting from the dysfunction of particular connections in the brain.

The results of the neuropsychological examination indicate the presence of neuropsychological symptoms in the areas generating visual perception and attention, as well as the components of emotional and executive functions examined. Compared to the results of individuals from the normalization group, the following were noted: weakened selective attention and selective problems with visual perception (blurred digits, slow pace with maintained concentration), lowered divisibility of attention, and difficulties with maintaining the direction of action when no action strategy was chosen (impaired verbal fluency, problems with effective planning). The patient experiences a state of derealization, resulting in a temporary sense of indifference and inappropriate expression of emotions. However she does not show any disorders in decision-making; her intellectual abilities, visuospatial memory, and cognitive flexibility are above average. Other examined functions are on the average level.

It should be stressed, that common manifestations of hindbrain damage include symptoms resulting from structural and functional changes in the cerebellum, which are responsible for neuropsychological disturbances such as a sensation of objects at rest vibrating or blurred contours of objects (Greer, 2012). This means that perception and attention disturbances (a blurred image of the surroundings during the state of derealization) may result from structural and functional changes in the central nervous system – a congenital malformation of the hindbrain involving pressure on the fourth ventricle and a decreased flow of fluid to the basal cisterns (Prockop & Murtagh, 2012). Although the damage to the cerebellum is not very extensive, the patient has isolated symptoms of the cerebellar cognitive-emotional disorder syndrome, such as problems with planning (Houston et al., 2018).

Lowered efficiency of some attention processes and executive functions may suggest dysfunction in the dorsolateral cortex of the frontal lobes (Herzyk, 2015).

However, neuroimaging examination results (see: Figure 1) do not indicate macrostructural brain damage. Based on all the results of the neuropsychological and neurological examinations and the patient's health history, it is possible to partially confirm hypothesis 1.1 – the patient's symptoms may stem from damage to the hindbrain, and selected connections of the hindbrain with other parts of the brain. However, this would require confirmation by methods that study the connections that condition brain function in milliseconds.

Referring to the second research question 1.2, cognitive and emotional symptoms stem from derealization disorder. Derealization disorder is classified in the ICD-11 as one of dissociative disorders (Gaebel et al., 2017) and results from structural and functional changes in various areas of the brain (Sterna & Sterna, 2018) responsible for a number of cognitive symptoms, including low resistance to distractors (Lemche et al., 2016). The results of the neuropsychological examination of the patient do not indicate a causal relationship between the appearance of derealization and cognitive and emotional disorders, namely, ignoring other people's emotional reactions or expressing emotions inappropriately for the situation while maintaining the ability to understand emotions. "Derealization" as described by the patient is an unexpected state of reduced contact with the environment, involving awareness, where the environment is blurred for several minutes. The patient has no symptoms of depersonalization, which is often an inseparable element of derealization disorder (cf. Tomalski& Pietkiewicz, 2022).

The results of the examination of emotional functions, the data from the interview, and the submitted psychological documentation do not indicate dissociative or neurotic disorders, and the clinical picture of the patient's mental functions is not consistent with the ICD-11 and ICD-10 diagnostic criteria for derealization disorder. Data from the medical history and records suggest that the clinical symptoms appeared in 2018 and worsened in 2019, after surgery for Arnold–Chiari syndrome. Based on other MRI and EEG results and the patient's health history, it can be concluded that there are reasons to reject hypothesis 1.2 – cognitive and emotional symptoms do not result from derealization disorder. This condition might be rather explained as a periodic decrease in the allocation of cerebral resources, which was strongly emphasized by the studies of Luria (1976), as well as Slansky & MacNeeal (1997) and Pąchalska (2007).

Verification of the second research question

Regarding the second research question of whether there is a probability of symptom variability, it was assumed that the clinical symptoms were a consequence of Arnold–Chiari I syndrome. The hypothesis was verified based on data from the clinical interview, observation, documentation (with particular emphasis on the results of neurological examinations), and neuropsychological examination. The patient's dominant symptoms include reduced divisibility of attention, selective difficulties with planning actions and processing emotions, and a state of derealization. The analysis of the subject literature on symptoms resulting from Chiari I malformation reveals a complex profile of cognitive and emotional disor-

ders (Blaser & Osborn, 2019; Greer, 2012; Prockop & Murtagh, 2012).

Research by James R. Houston and colleagues (2018) suggests the presence of two dominant neuropsychological symptoms: divided attention disorders and reduced emotional processing. Both symptoms are present to some degree in the patient, and the state of derealization she feels may result from impaired attention and emotion regulation processes. Patients with Chiari I malformation exhibitverbal working memory disorders, problems with direct visual memory, significant difficulties with memory and auditory-verbal learning, slower information processing, impaired strategy changes, and difficulties with recognizing famous people's faces (García et al., 2018). In the advanced stages of Chiari malformation, clinical symptoms are varied and more severe: there is cerebellar hypoplasia and mental retardation (Grazz& Andrasik 2012). Based on the results of the neurological and neuropsychological examinations, it can be concluded that the clinical symptoms do not indicate the presence of Chiari malformations type II, III, or IV (e.g., Greer, 2012) but stem from Arnold–Chiari I syndrome.

The patient's above-average intellectual abilities may have a compensatory function and minimize further development of symptoms –for example,reduce the sense of derealization. Based on clinical data and the subject literature, hypothesis 2.1 can be considered supported – the clinical symptoms probably stem from brain changes in Arnold–Chiari I syndrome. Regarding the research question, it can be asserted that there is a likelihood of symptom variability, what also microgenetic theory suggests (Brown & Pachalska 2003).

Indication of neuropsychological mechanisms of disorders

The starting point for determining the mechanism of disorders is the theoretical assumptions regarding the emergence and development of clinical symptoms in the context of the type and dynamics of Arnold–Chiari I syndrome, characterized by adisplacement of the cerebellar tonsils into the foramen magnum (Blaser& Osborn, 2019). In the dominant conceptions of the organization of mental functions, the course of diagnostic procedures is determined by the syndromological and/or individualized approach based on the microgenetic theory of symptom formation (Brown, 2003; Brown & Pąchalska, 2003), which suggests two mechanisms behind the reported symptoms. Firstly, reduced divisibility of attention, impaired selective attention and visual perception, selective difficulties with planning, and a sense of derealization probably stem from mild structural and functional changes in the central nervous system (elongated cerebellar tonsils ¾2019, 2020, and 2021, as wel as MRI results) in Arnold–Chiari I syndrome. Secondly, the state of derealization described by the patient causes anxiety and the need to calm down and may contribute to inappropriate reactions in particular social situations in the form of greater irritation with others. The development of emotional intelligence processes is inharmonious and, to some extent, independent of health condition. Additionally, emotional and cognitive symptoms intensified after the surgery that was performed due to brain dysfunction resulting from Arnold–Chiari I syndrome. Due to adolescence and the course of Arnold–

Chiari I malformation, there is a likelihood of clinical variability.

The patient's resources and the resources in her immediate environment

The patient has many cognitive resources, notably: above-average short-term visual-spatial memory, high efficiency of intellectual functions in the non-verbal area, normalmemory, attention, and auditory-verballearning ability, and retained efficiency of executive functions pertaining to reaction inhibition and the use of various action strategies. The patient is motivated to act and ready to undertake further activities. Apart from the health problems resulting from Arnold–Chiari I syndrome, she does not show any other somatic complaints or mood and behavior disorders. Based on the interview and observation, it can be concluded that the quality of bonds between her immediate family members is good; they show concern, are involved in the patient's recovery process, and provide various kinds of support: emotional (e.g., they give a sense of security), instrumental (e.g., they take care of the patient's health and education), information (e.g., they explain the chances of recovery), and financial (e.g., they finance treatment and education). Thanks to the healthy relationship between the parents and the support received, the patient will be able to continue treatment as well as start therapy and use other health interventions. She has a chance to preserve current resources and acquire new resources, which, according to Steven Hobfoll's conservation of resources theory, are an indicator of a better coping with stress (Hobfoll, 1989).

Modern treatments for Arnold–Chiari syndrome depend on the type of hindbrain malformation. The patient's neurological examinations indicate a mild form of the syndrome, which consists in a displacement of the cerebellar tonsils. Performing a standard surgical procedure involving the decompression of the posterior cranial fossa (Pruszewicz et al., 2007) contributed to reducing the range of neurological symptoms. Further observations and an annual MRI examination will allow for monitoring the course of Arnold–Chiari syndrome, and in the event of worsening it will be possible to decide on another surgery. Depending on the type of clinical symptoms, neuropsychological rehabilitation and pharmacological treatment can be included to minimize mood or behavioral disorders that are not specific to Arnold–Chiari syndrome but may co-occur with it (Almotairi et al., 2020). Psychoeducation offered by specialists who provide information about the symptoms, types, and methods of treatment of Arnold–Chiari syndrome (e.g., on the chiari.pl website) may contribute to reducing anxiety in the family, a better understanding of the specificity of hindbrain malformations, and distinguishing the symptoms resulting from the disease from those that may have a non-neurological basis. Additionally, the inclusion of an individualized learning path can contribute to the patient's more effective functioning.

Guidelines for intervention and/or psychological therapy

Standard therapeutic procedures in Arnold–Chiari syndrome include neurological, physiotherapeutic, and neuropsychological consultations and treatment. The scope of therapeutic interventions will depend on the course of hindbrain

dysfunction, particularly on the type of hindbrain malformation and comorbidities. The mild form of Arnold–Chiari syndrome is characterized by fewer symptoms and relatively low severity.

Comprehensive and individualized interventions aimed at alleviating the symptoms (Pąchalska, 2019) and improving neuropsychological functions (Brown & Pąchalska, 2003; Pąchalska 2022; Pąchalska et al., 2022) will be the most advisable.

The patient underwent such neuropsychological therapy program aimed at reducing the severity of clinical symptoms, strengthening resources, and developing social and emotional skills. It was possibile in the patient opinion to improve her visual perception and divided attention processes as well as executive and emotional functions was well received by the patient. However due to adolescence and the dynamic development of neuropsychological functions, further observation and follow-up examinations are required – neuropsychological (examination of cognitive and emotional functions) and neurological (physical examination, MRI, and EEG).

The literature explains why neuropsychological therapy can improve the functioning of the damaged brain through its neuroprotective potential (Pąchalska 2008; Prigatano 2008). It will be recalled, that healthy brain might have appropriate activity of the motor and somatosensory cortex and properely organized afferents and efferents connections (which makes possible the input of sensory information, and the output to other parts of the brain) as well as integrative functions (see: Fig. 3) chich can ensure an appropriate level of executive functioning (Pąchalska 2007; Kropotov 2009; Burgess et al. 2017).

In the patients with brain dysfunctions large individual differences in the activity of the brain, the motor and somatosensoty dysfunctions, and consequently

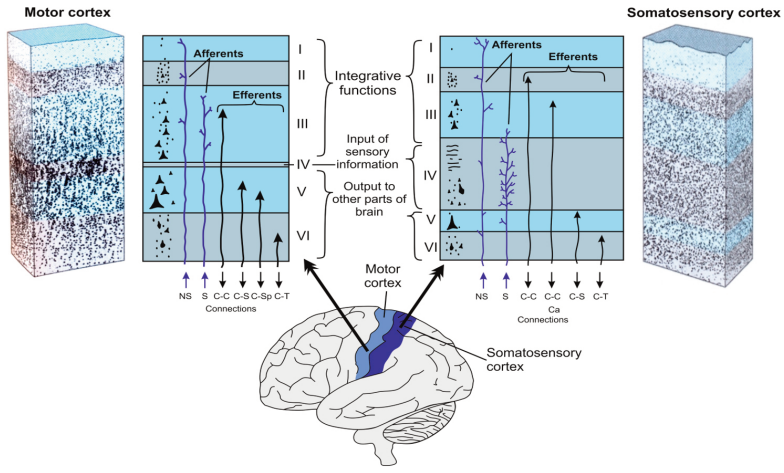

Fig. 3. Activity in the motor and somatosensory cortex
Source: M. Pachalska (2008), with permission

the organization of cognitive functions, as well as after the process of different therapeutic approaches are observed (Pąchalska 2008). From the above it follows that the microgenetic model of symptom formation perfectly explains the widespread making of the similar mistakes by people with brain dysfunction (Pąchalska & Góral-Półrola 2022). It May also explain the specificity of activity in the motor and somatosensory cortex in our patient with Arnold Chiari type I malformations.

It should be pointed out however, that Chiari malformations are relatively common and represent a spectrum of hindbrain anomalie. Therefore the prognosis for most patients with a Chiari I malformation is good, but depends alo on the initial neurological presentation. Those patients with mild neurological deficits tend to have good outcomes, but those with moderate to severe symptoms tend to have a guarded prognosis. The surgery is also associated with several complications, of which the most common are CSF leak and pseudomeningocele. A few individuals may have a persistent syrinx and may require a shunt (Lei et al. 2018).

To sum up, the diagnosis and treatment of this condition require an interprofessional team consisting of primary care, neurologists, radiologists, neurosurgeons, specialty trained nurses an neuropsychologists. Depending on the severity of the malformation, the individual may be asymptomatic or have severe neurological symptoms. While the patients are often managed with decompressive surgery, the nurses are responsible for looking after these individuals. Hence, the nurse must be aware of the potential post-surgical complications and their presentation (Bhimani et al. 2918).

Our clinical experience suggests that there is a need for an individualized and person-centered approach (especially in the case of a patient with Arnold Chiari malformation type I, with the inclusion of his family). This is because a person working with such a patient experiences a wide variety of difficulties (working together with the patient, with his environment, with other professionals). However, by using such interventions tailored to the patient's needs, we are able to improve his quality of life, which is the goal of modern medicine and neuropsychology.

CONCLUSIONS

Despite effective neurosurgical treatment methods, structural and functional changes in the hindbrain resulting from Arnold–Chiari type I malformation may give mild neuropsychological symptoms that should be carefully diagnosed and considered in further therapy and treatment.

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