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**Techniki plastyki opony twardej, komplikacje pooperacyjne oraz jakość życia u pacjentów z zespołem Chiariego typu I**

**Rozprawa na stopień doktora nauk medycznych** **i nauk o zdrowiu**

**w dyscyplinie nauki medyczne**

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# Streszczenie w języku angielskim

Chiari Malformations are rare congenital malformations characterised by hindbrain structures being displaced caudally through the foramen magnum. They were named after the Austrian pathologist, Hans Chiari, who was one of the first to describe this pathology in 1891. There are many types distinguished due to pathomorphological differences. Originally, types I through IV were described, and more recently types 0 and 1.5 have also been distinguished.

The most common of these is Chiari Malformation type I (CM-I), characterised by displacement of the cerebellar tonsils through the foramen magnum into the spinal canal, along with their distortion. This condition, over time, causes disruption of the natural cerebrospinal fluid flow between the intracranial space and the spinal canal, and compression of the brainstem. It can also lead to impaired function of the cerebellum itself. Coexisting syringomyelia is observed in 30-70% of patients. Despite many theories developed over the decades, the pathophysiology of the Chiari Malformation, as well as syringomyelia, still remains unclear. CM-I is diagnosed in adults mainly in the second and third decades of life, with a female predominance. MRI is the test of choice for diagnostic imaging. The criterion adopted was that in order to diagnose type I, the apex of one of the cerebellar tonsils must reach ≥5 mm below the level of the foramen magnum (McRea line).

Symptoms reported by patients primarily include severe headaches of the suboccipital region, characteristically exacerbated with Valsalva test (physical activity, sneezing, laughing or pushing) in this pathology. In addition to headaches, patients may also experience difficulty swallowing, expectoration, hiccups, and all types of sensory and motor disturbances as a result of direct pressure from the tonsils on the brainstem. Furthermore, there are symptoms of damage to the pathways leading to and from the cerebellum, as well as the cerebellum itself. Dissociated sensory disturbances are characteristic in case of coexisting syringomyelia.

In addition to deficit symptoms, examining neuropsychological and psychiatric aspects in patients with Chiari malformation has also recently received increasing attention. As far as observable cognitive deficits are concerned, depression was also found to be more frequently diagnosed among patients with Chiari malformations.

The treatment of choice in cases of symptomatic malformation is surgical decompression at the level of the atlanto-occipital junction. The technique involves performing a suboccipital craniectomy with removal of the posterior C1 arch, and sometimes the C2 and C3 vertebral arches. What is more, in order to widen the subdural space, a duraplasty is performed by making an appropriate incision and implanting an additional patch in this site. However, the details of the procedure itself, especially the technique of duraplasty, are still a topic of discussion and dispute among specialists.

The studies conducted as part of the publication series analysed the clinical and radiological results of the treatment and the observed complications due to the duraplasty technique being used. In order to holistically interpret the treatment results, the quality of life of both operated and non-operated patients was also analysed, with an assessment of depression severity and patients' attitudes toward the disease itself.

The following goals of the study were distinguished: 1) comparing long-term clinical and radiological results of CM-I surgery and analysis of complications, depending on the type of duraplasty material used and the method of its fixation, 2) determining the role of the occurrence and size of *pseudomeningocele* in long-term outcomes after atlanto-occipital decompression surgery with duraplasty in patients with CM-I, 3) assessing the severity of depressive symptoms and quality of life in patients with CM-I, both in operated and non-operated patients. Identifying and analysing the main determinants of these phenomena and comparing the results with a control group of healthy individuals.

The methodology included a retrospective analysis of patients diagnosed with CM-I who underwent surgery at the Department of Neurosurgery of the Medical University of Warsaw. All patients underwent suboccipital craniectomy with removal of the posterior arch of the C1 vertebra and, depending on the level of cerebellar tonsil decrease found, the C2 and C3 vertebral laminas as well. Since the analysis was single-centre, the differences were only related to duraplasty using grafts of different types of materials and different fixation methods. Therefore, it was possible to classify patients into a group where artificially produced materials (nonautologous: collagen matrix) or materials taken from the patient (autologous: e.g. fascia) were used. Differences in fixation included whether the graft was sutured in or glued in with fibrin glue. Because of that, a comparative analysis of the treatment results was conducted first, considering only the graft material used and the technique of its fixation. This was followed by further comparative analysis, which included four variants of duraplasty techniques that differed in the configuration of the material used and the type of graft fixation. Long-term clinical outcomes included subjective interpretation in *Gestalt* assessment (improvement, unchanged, and deterioration), and assessment using the *Chicago Chiari Outcome Scale* (CCOS) protocol. Radiological evaluation consisted of identifying the coexistence and thickness of the epidural fluid collection *(pseudomeningocele*) in the post-operative areas and also measuring the size of the intramedullary cavity compared to preoperative studies. Improvement or unchanged in the *Gestalt* assessment and a score of ≥12 on the CCOS were considered satisfactory. In the case of coexisting cavity, a decrease in cavity size was considered as such. Complications that required additional interventions occurred in 10% of patients.

Through a comparative analysis, it was found that both the use of autologous and nonautologous dural grafts lead to similar distant treatment results, regardless of fixation method (with sutures or glue). However, with the use of nonautologous grafts*,* the coexistence of *pseudomeningocele* was observed significantly more often, although the vast majority were asymptomatic.

Another study analysed the distant outcomes of patients who underwent atlanto-occipital decompression surgery and were diagnosed with coexisting *pseudomeningocele*  in follow-up MRI scans. The results were compared with those of patients in whom this phenomenon was not observed. Even small *pseudomeningocele*, previously treated as asymptomatic, recognised as hyperintense fluid collection in T2 sequences, were taken into account. The radiological changes observed in the posterior cranial cavity in both groups of patients were interpreted in detail. Clinical evaluation of the treatment was carried out again, taking into account the patient's subjective evaluation in the *Gestalt* assessment and using the CCOS protocol, as well as an analysis of changes in the intramedullary cavity size compared to the pre-surgery results. Detailed radiological analysis included the following measurements: the thickness of the *pseudomeningocele* and the distance between the graft and the cerebellar tonsils on postoperative examination. In addition, we analysed preoperative and postoperative differences in the distances between the superior cerebellar surface and the cerebellar tentorium, between the anterior border of the foramen magnum in the midline *(basion*), and the pontomedullary sulcus, as well as the level of the cerebellar tonsils herniation into the spinal canal in relation to the foramen magnum.

The study showed no significant differences in treatment outcomes with the known coexistence of *pseudomeningocele* regardless of their thickness. Nevertheless, *pseudomeningocele* caused significant deterioration in the patient's condition in rare cases shortly after surgery. Signs of cerebellar and brainstem lowering have been observed in patients with *pseudomeningocele*. Small but statistically significant differences were found between the groups.

Having considered the foregoing, the suspicion was put forward that the symptoms may have been partly due to the structures of the posterior cranial cavity lowering secondary to hypotension associated with CSF leakage, rather than to repeated tightness at the level of the foramen magnum.

In order to assess the quality of life, severity of depressive symptoms and patients' attitudes toward the disease, we conducted another study where patients diagnosed with CM-I, both post-operative and non-operative, were invited. Recruitment took place among members of Polish largest on-line group of patients affected by the condition. These patients were operated on and consulted at various centers in Poland and abroad. Each patient was asked to fill out an anonymous questionnaire. In addition to standard demographic and clinical questions, patients answered questions from *the* *Beck* *Depression Inventory* (BDI-II), *an abbreviated version of the WHO-proposed quality of life assessment* (WHOQOL-BREF), *the Acceptance of Illness Scale* (AIS) and *the Beliefs about Pain Control Questionnaire* (BPCQ). The control group consisted of healthy individuals who were demographically matched to the patient group.

In addition to confirming the increased prevalence of depression among CM-I patients, both surgical and non-operative patients had similar results on most questionnaires. Significant differences manifested themselves in lower pain levels in post-surgery patients on the VAS scale, both in terms of average and current pain levels. In assessing the quality of life of patients undergoing surgery, higher scores were observed in the environmental aspect category and in the perceived impact of the doctor's role on the level of pain experienced. In addition, higher levels of depression and lower quality of life in some aspects were observed in the post-surgery group compared to the non-operated group, despite the lower pain intensity found.

The observations from this study shed new light on the care of patients with CM-I, and highlight the important impact of the work of neuropsychologists and psychiatrists on treatment outcomes.