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Intrathecal CSF dynamic measurements in hydrocephalus associated with MMC – Experience of the Republican center of neurosurgery of Uzbekistan

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ABSTRACT

Purpose: We sought to determine methods of precise gradation of hydrocephalus in patients with spina bifida. Symptomatic hydrocephalus is a common condition associated with myelomeningocele (open spina bifida). Traditionally, hydrocephalus is treated with insertion of the ventriculo-peritoneal (VP) shunt. This has been the standard of treatment since the introduction of the Holter shunt valve for the VP shunt in the early 1960s.
Methods: We have analized the results of surgical treatment of 81 patients aged between 1 month and 1.5 year old with hydrocephalus and MMC. All patients underwent surgery in Republican center of neurosurgery of Uzbekistan for MMC with hydrocephalus in the period of 2013–2018. We suggest to use the ventricular index to determine the precise degree of hydrocephalus in patients with spina bifida and the method for selecting valve parameters.
Results: Patients with suspection of associated hydrocephalus, in order to arresting the risk of MMC rupture and prevention of possible leakage after the back closure a VP shunt was performed. According to above mentioned 52 (64.2%) patients for the 1st stage underwent VP shunt surgery with a low-pressure valve, 23 (28.4%) patients with medium pressure and 6 (7.4%) with high pressure valve. MMC repair was done in 1–3 month after VP shunt placement. To all patients we used regular valve shunts due to high cost of adjustable one and lack of official distributors (health insurance has not yet implemented in our country).
Conclusion: The implantable shunt systems parameters were chosen before surgery in the surgical management of hydrocephalus in children with MMC are essential. This is important in order to prevent under or over drainage states, CSF leakage from the MMC sac. Management of hydrocephalus should be performed by considering MMC affecting craniospinal balance.

1. Introduction

The 3 main signs of spina bifida – hydrocephalus, paraplegia and dysfunction of the pelvic organs by the type of incontinence have been known for many centuries, although these signs were not associated with myelomeningocele (MMC) until the 17th century [5,7,17].

The first researcher who came close to the understanding of connection between myelomeningocele and hydrocephalus was Frederick Ruysch (1638–1731), but only the Italian pathologist Giovanni Battista Morgagni (1672–1771) clearly described this relationship and that myelomeningocele can be accompanied both with hydrocephalus and without it [6,12,13,16,18].

Gardner advanced the theory that overgrowth of the neural tube could be the cause of hydrocephalomyelia, but this interesting theory found rebuttal in modern neuroimaging and embryology [12].

Hydrocephalus basically accompanies open forms of spina bifida – myelomeningocele. Before the introduction of cerebrospinal fluid (CSF) shunting surgery in the early 1960s, hydrocephalus was the leading cause of death and disability in patients with myelomeningocele [1–4,8,9,14,15].

The true frequency of hydrocephalus in patients with myelomeningocele is not known, although in the main multicenter studies the need for shunting procedures reaches the value 80–90% [10,11,19–23].

2. Methods

We have analized the results of surgical treatment of 81 patients aged between 1 month and 1.5 year old with hydrocephalus and MMC.

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All patients underwent surgery in Republican center of neurosurgery of Uzbekistan for MMC with hydrocephalus in the period of 2013–2018. From them, 45 (55.5%) were boys, 36 (44.5%) were girls. Complete laboratory and instrumental investigation were performed to all patients. The patients with associated hydrocephalus, in order to create favorable conditions for epithelization of the MMC, arresting the risk of rupture of the MMC and prevention of possible leakage after the back closure, a ventriculoperitoneal valve shunting was performed. The degree of hydrocephalus was determined by the posterior horn index. The parameter of the implantable shunting system was selected by the method of "Choose right shunt. In patients presenting with MMC and ventriculomegaly according to the standards of care to the patients with spina bifida in our country, we routinely do shunt surgery before MMC closure and monitor the MMC until ready for closure. If there is no ventriculomegaly, we certainly proceed to closure of the MMC first.

MMC repair was done in 1–3 month after VP shunt placement. To all patients we used regular valve shunts due to high cost of adjustable one and lack of official distributors (health insurance not implemented in our country).

Results. In our series all patients with MMC were associated with hydrocephalus. From them, 16 (19.8%) patients had MMC in thoracolumbar region, 19 (23.5%) in lumbar region, 31 (38.3%) in lumbosacral region, 5 (6.2%) in sacral and in 6 (7.4%) in thoracolumbosacral region, which is more correlated with the literature data (Figs.1 and 2).

The development of the pathological process in the overwhelming majority of cases (73.7%) was progressive. 64 (79.0%) patients had a type 2 Chiari malformation with a maximum ectopy of the cerebellar tonsils to 37 mm below the level of the foramen magnum. In 53 (65.4%) patients angiopathy of the retinal vessels was noted on the eye fundus, 19 (23.4%) pallor of the optic nerve disc, in 6 (7.4%) papilledema of 1st stage, in 2 (2.4%) papilledema 2nd stage. The low frequency of papilledema is due to the anatomical and physiological properties of the craniocerebral system in children, the compensatory capabilities of the skull, open cranial sutures. Craniometry index ranged from 36 to 53 cm. For these patients, hydrocephalic changes in the bones of the skull were characteristic. Out of 81 children, craniomegaly occurred in 23 (28.3), while in 14 it was markedly expressed in combination with the internal form of hydrocephalus. Eye set symptom was noted in 22 (27.1%) patients, which indicate the presence of intracranial hypertension. The manifestation of hypertensive syndrome was established in 32.5% of observations and in 22.5% of cases it was significant. In 15% of observations, there were some signs of herniation syndrome.

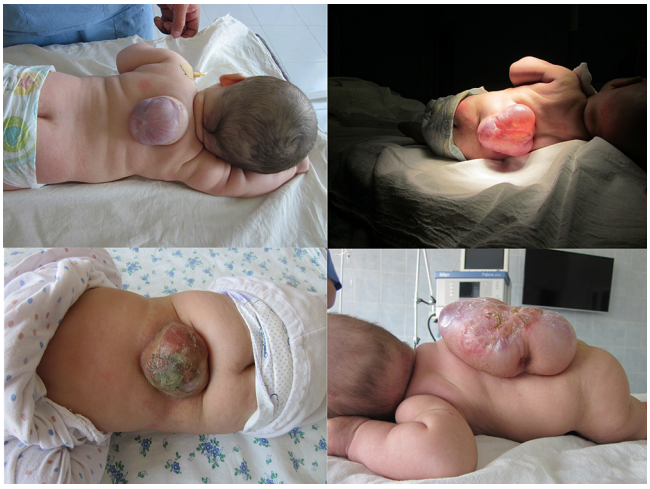

Fig.2. Different topical localization of MMC.

3. Discussion

According to McLone and Knepper's unified theory (1989) the small sizes of the posterior fossa, low location of torcular herophili, downward of the cerebellum and hindbrain, mostly enlarging of the posterior horns of the lateral ventricles with formation of colpocephaly is a common cranial changes in patients with spina bifida. As neurovisualization criteria of hydrocephalus we used the posterior horn index (PHI) – which corresponds to the ratio of the maximum distance between the outer walls of the posterior horns of the lateral ventricles (A) to the maximum bitemporal diameter of the skull (D), while the index 0.5–0.6 corresponds to hydrocephalus of mild degree, 0.6–0.7 of moderate degree, more than 0.7 severe degree. This method permits to determine the precise degree of hydrocephalus in children with MMC based on the ventriculo-cranial index with high accuracy (Figs. 3–5).

The posterior horn index (PHI) for determine the degree of hydrocephalus in children with MMC.

At present, hydrocephalus treatment in children with MMC is considered to the use of CSF-shunting devices, the meaning of which is the creation of an anastomosis between the CSF and extra-axial system (venous, abdominal cavity or other extracranial cavities) by the constant implantation of special drainage systems (shunts). The shunts provide a unidirectional CSF flow at a certain CSF pressure. CSF

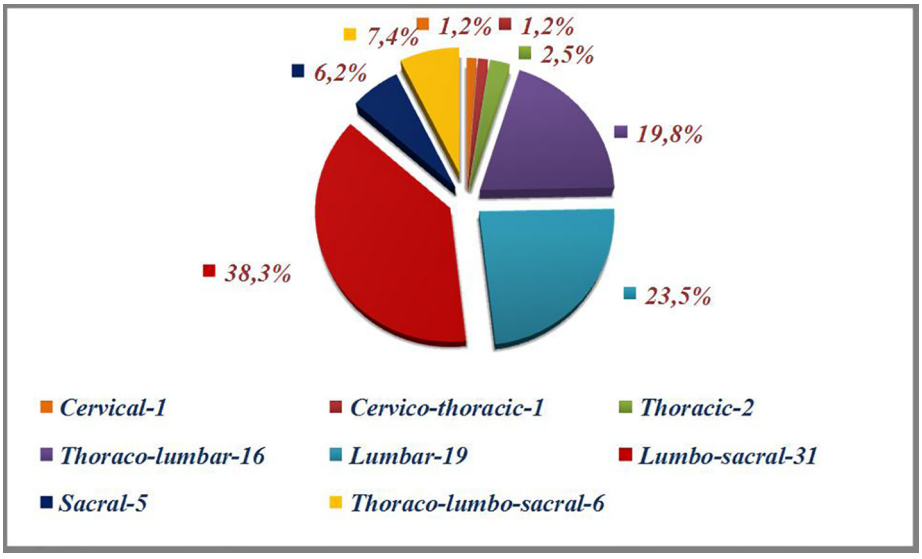

Fig.1. Distribution by topical localization of MMC.

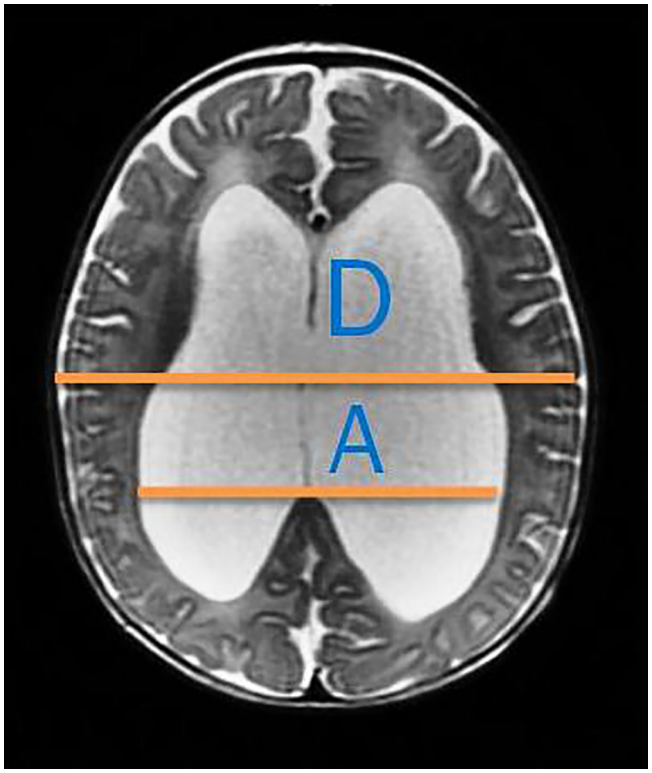

Fig.3. Measurement of posterior horn index.

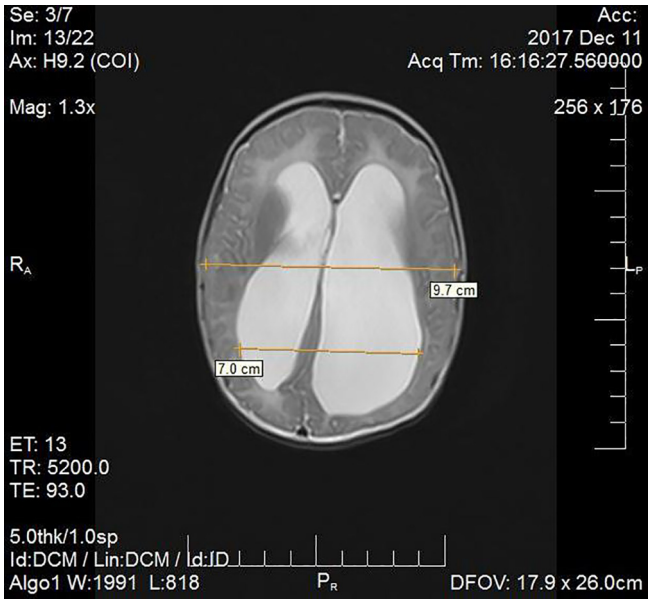

Fig.4. 2 month baby, before surgery on MRI is presented severe degree of hydrocephalus with PHI 0.72.

pressure, at which the shunt functions (throughput of the shunt), is an important indicator and largely determines the outcome of the treatment.

We have developed a method for selecting the throughput pressure of the implantable drainage system in the surgical treatment of hydrocephalus in children with MMC, including the determination of CSF pressure, determine the initial pressure of cerebrospinal fluid in the preoperative period by withdrawing 1 ml of CSFfrom MMC sac until constant value, the data is put into the computer software “Choose right shunt” and the value of the point of critical deformation of the

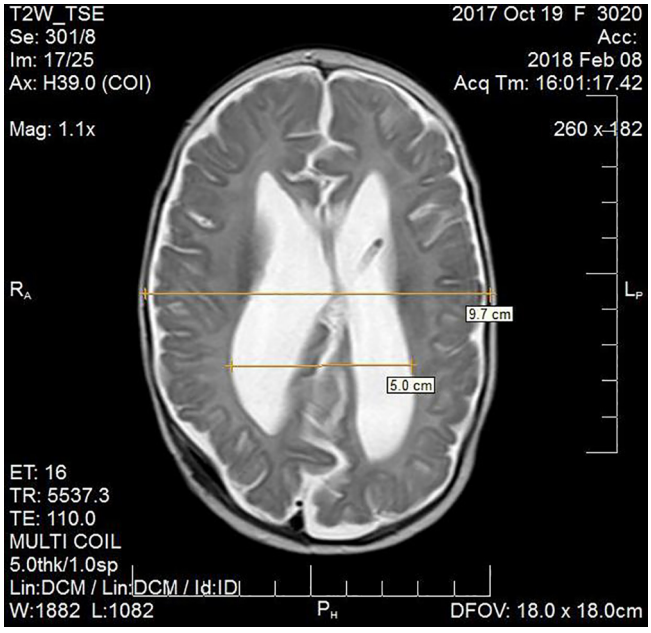

Fig.5. VP shunt with low pressure valve was performed, after 3 month on follow-up there is no signs of hydrocephalus, with PHI 0.51.

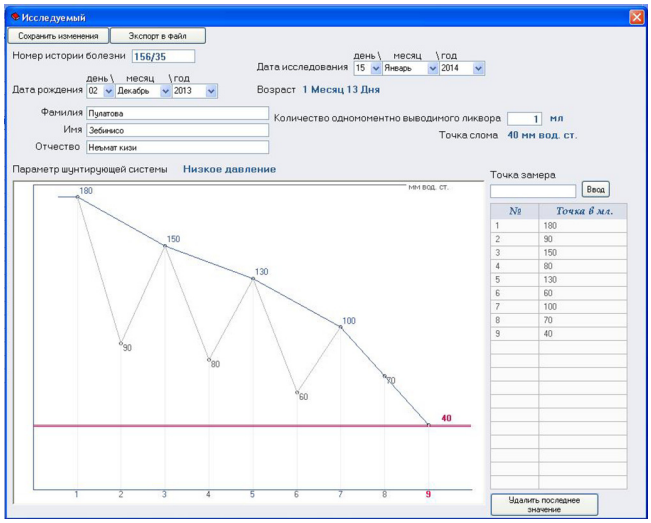

Fig. 6. The interface of the program “Choose right shunt”.

ventricular system is set, which corresponds to the pressure of flow in implantable drainage system (Fig. 6).

The essence of the method is as follows: puncture of the MMC sac with a needle from the infusion system, measurement of the initial pressure of CSF, then through the rubber connector of the infusion system, withdrawal of 1 ml of spinal fluid with the measurement of CSF pressure prior to arrest the level of the CSF. Parameters are entered in the computer program “Choose right shunt”, where the critical point of deformation of the ventricular system is calculated. Based on which, the parameter of the implantable shunting system is chosen – high, medium or low pressure. The method allows reducing the risk of postoperative complications in the form of under or overdrainage states.

As a result of a comprehensive assessment of clinical-neuroimaging and clinical-CSF studies, the evaluation of the peculiarities of the manifestation of hydrocephalic syndrome was carried out. Based on the method “Choose right shunt” the determination of the point of critical deformation of the ventricular system was established, 52 (64.2%) patients for the 1st stage underwent VP shunt surgery with a low-

pressure valve, 23 (28.4%) patients with medium pressure and 6 (7.4%) with high pressure valve. Under or overdrainage complications were not observed in postoperative period. The 2nd stage of surgery for MMC repair was planned in 1–3 months with the follow-up head US and EMG study.

Some case example:

Patient N., 3 months, No 169/41, the diagnosis: "Congenital anomaly of the central nervous system. Chiari malformation type 2. Spina bifida aperta with hydrocephalus. Meningomyelocele VTh7-S1 with the threat of rupture of the hernial sac". On MRI of the brain, there is severe degree of hydrocephalus, width of the third ventricle 15 mm, lateral ventricles: left 22 mm, right 27 mm. A puncture of the hernial sac was carried out, the initial pressure of the CSF was measured that is equal to 100, then an withdrawal of 1 ml of CSF was conducted through the rubber connector of the infusion system, measuring the CSF pressure prior to arrest the level of the CSF, which was 80. These parameters were entered into the computer program "Choose right shunt", where the point of critical deformation of the ventricular system is equal to 60, which corresponds to the parameter of the shunting system for low pressure. It was decided to implant the selected shunting system. After surgery, the child's condition was compensated, hypertensive symptomatology was not observed, the threat of rupture of the MMC sac was solved. After 2 months on the follow uphead US, the positive changes of hydrocephalus is determined: a decrease in ventriculomegaly, an increase in the thickness of the brain tissue. In a comfortable condition, the second stage of the surgery back closure is performed.

4. Conclusion

The implantable shunt valve parameters chosen before surgery in the surgical management of hydrocephalus in children with MMC are important in the causes of under or overdrainage states, CSF leakage from the MMC sac, to prevent further reoperations and anesthesia. Management of hydrocephalus should be performed by considering MMC affecting craniospinal status. The presence of the CSF infection, high cell count and protein, presence of acute associated diseases are considered as contraindications for VP shunt placement.

Conflict of Interest

On behalf of all authors, the corresponding author states that there is no conflict of interest.

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CRediT authorship contribution statement

M.M. Akhmediev: Visualization, Investigation, Supervision, Conceptualization. **G.M. Kariev:** Data curation, Methodology, Validation. **T.M. Akhmediev:** Software, Investigation, Writing - original draft, Writing - review & editing.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.inat.2019.100644.

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