

NEUROSURGICAL TECHNIQUES FOR MENINGOMYELOCELE REPAIR: A LITERATURE REVIEW

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Abstract: Introduction: Myelomeningocele (MMC), resulting from alterations in neural tube closure during embryonic life, is the most severe form of spina bifida. MMC is diagnosed through morphological ultrasound between the 18th and 22nd gestational week through direct visualization of the dorsal malformation. With the emergence of fetal medicine, early treatment of MMC became possible and its prognosis more satisfactory. **Goal:** To understand the feasibility of neurosurgical techniques for meningomyelocele repair and post-surgical effects. **Method:** Bibliographic review carried out in April 2021 at the Virtual Health Library. The descriptors used, defined by DeCS/MeSH, were “*meningomyelocele*” AND “*surgery*”. Articles published between 2016 and 2021, in all languages, were considered. By reading the titles, abstracts, eliminating duplicates and including articles manually selected for the relevance of the theme, original articles were chosen to compose this review article. **Results:** Early separation of the spinal cord with amniotic fluid can prevent major neurological damage to the patient. Prenatal surgery lessens the occurrence of tonsillar herniation and hydrocephalus. In cases of deformities secondary to MMC, fusion surgery instrumented by constructions of pedicle nails and screws promotes improvement in the neurological and functional status and correction of spinal angulation. The island flap technique based on perforating arteries (KDPIF), performed after birth, enables adequate vascularization of the tissue, preventing future deformities and necrosis of the skin flap. Fetal surgery proved to be more effective than postnatal surgery because it represents a possibility of fetal development of the nervous system closer to what is considered normal, reducing the need for a shunt and promoting a better urological prognosis. However, it has limitations

because it poses risks to the mother and fetus. **Conclusion:** Post-birth surgery entails prolonged contact with amniotic fluid. Thus, when comparing pre and postnatal surgery, it is clear that intrauterine surgery has greater advantages by providing repair in a timely manner so that there is a better quality of life for the patient with MMC.

Keywords: Myelomeningocele. Spinal Dysraphism. Neurosurgery.

INTRODUCTION

Myelomeningocele (MMC) is a pathology resulting from improper closure of the neural tube during embryonic life, being the most severe form of spina bifida, in which the spinal cord and meninges protrude out of the spine, being exposed to amniotic fluid and mechanical damage during pregnancy.^{1,2} The prenatal diagnosis can be based on morphological ultrasound, between the 18th and 22nd week of pregnancy. Examination findings include direct visualization of dorsal malformation.³

The incidence of MMC in Brazil is 2.28:1000 births. The disease then entails major socioeconomic impacts, since those with this anomaly have limitations in their development, in addition to representing increased costs for the family and loss of productivity for their caregivers.¹ One way to prevent spina bifida in fetuses is the use of folic acid by pregnant women. However, encouraging its consumption during the prenatal period, despite reducing the number of cases of MMC, was not able to eradicate it.⁴

With the development of Fetal Medicine, MMC repairs that could only be done after childbirth, could be performed even with the pregnancy in progress, allowing corrections to be made throughout the embryonic development. The expectation is that fetal surgery will promote a reduction in the exposure time of the medulla and meninges

of the fetus with MMC to mechanical injuries and those developed by contact with the amniotic fluid, improving its prognosis.⁴

Thus, it is necessary to gather scientific evidence capable of subsidizing the importance of intrauterine MMC repair, compared to postnatal surgery.

Therefore, the purpose of this literature review is to understand the feasibility of neurosurgical techniques for meningocele repair and its postoperative effects.



Figure 1 - Full MMC (non-rotate)

Source: author's own production (2015).

GOAL

To understand the feasibility of neurosurgical techniques for meningocele repair and post-surgical effects.

METHOD

The bibliographical research was carried out in April 2021, at the Virtual Health Library (BVS). The descriptors used were “myelomeningocele” and “surgery”, defined by the Medical Subject Headings (MeSH) and Health Sciences Descriptors (DeCS), with the

combination used “myelomeningocele” AND “surgery”. MEDLINE filtering was used. We considered all articles published from 2016 to 2021, in all languages, freely available in full and addressing neurosurgical procedures as the main subject. Systematic review articles and case reports were excluded, as well as those that did not correspond to the objective of this study, by reading the title, abstract and, subsequently, reading the entire article. With the inclusion of six articles selected manually by the relevance of the theme, thirteen original articles were selected to compose this review article.

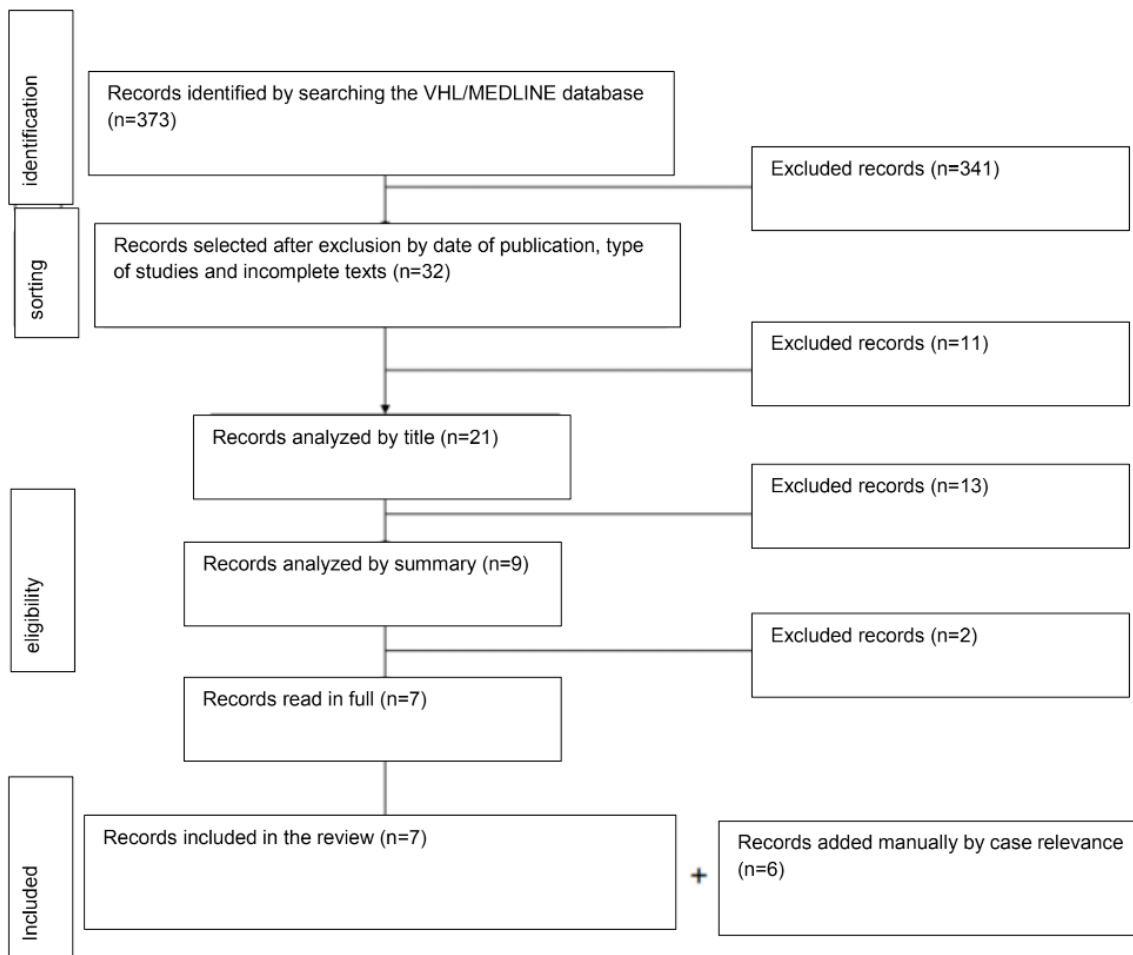


Figure 2 - PRISMA flowchart
 Source: Author's own production (2021).

RESULTS AND DISCUSSION

After reading the articles selected to compose this review in full, it was noted that amniotic fluid toxicity, as well as mechanical injuries to the spinal cord and spinal nerves at the time of delivery, lead to irreversible changes in the innervation of the individual affected by MMC. Different neurosurgical techniques are available to correct this pathology, with post-surgical impacts to be considered.

In a study carried out with 72 patients with MMC, divided into prenatal group (36 patients) and postnatal group (36 patients), where pre and post natal surgeries were performed by the same medical team, urodynamic tests and urinary incontinence

assessments. In the prenatal group, open fetal surgery was performed between the 24th and 26th gestational weeks, and the pregnancy in this group lasted between 35 and 38 weeks. In both groups, the same therapy was applied immediately after birth, with the use of cyclobenzaprine, anticholinergic drugs and urine sterilizing drugs. The study demonstrated that children operated on prenatally have an 81% chance of achieving urinary retention after the third year of life. Thus, it is possible to accept that, by early separating the spinal cord from contact with the amniotic fluid, greater neurological damage to the individual can be avoided, making the urinary system function better.⁵

The percutaneous fetoscopic technique

consists of placing a biocellulose adhesive on the exposed placode and can be adapted with the use of a bilaminar cutaneous substitute, formed by a dermal matrix and a silicone sheet, for better management of complex cases of spina bifida. In this case, the bilaminar skin substitute is placed on the biocellulose adhesive, with its silicone blade facing the amniotic fluid, and then they are sutured to the edge of the fetal skin, preventing it from sliding. A study carried out in 45 patients diagnosed with spina bifida with hindbrain herniation reported that fetal surgical intervention did not cause complications during childbirth, but premature birth proved to be a problem in 80% of cases. Fetal surgery is still quite challenging and, in addition to having a prolonged surgical time, premature delivery is a challenge. In this context, the importance of new studies capable of quantifying the risk/benefit of the percutaneous fetoscopic technique for repairing the MMC is highlighted.⁶

The Management Of Myelomeningocele Study (MOMS), performed with women, between 19-25 weeks of gestation, who had the fetus diagnosed with myelomeningocele were randomized and underwent surgery for prenatal or postnatal repair, after delivery at 37 weeks. Compared with postnatal repair, prenatal MMC repair prior to 26 weeks of gestation led to improved mental and motor function, increasing the likelihood of being able to walk independently. Despite the greater chance of preterm delivery, neurodevelopmental outcomes were no less favorable for children who underwent prenatal surgery. Therefore, the MOMS study makes it clear that intrauterine surgical repair is effective and more promising, and must therefore be the parameter of comparison adopted with regard to MMC repair surgeries.⁷

The MOMS and the Endoscopic Surgery for Antenatal Meningocele Correction

(CECAM) reported that in patients with spina bifida, fetal surgery brings many benefits when compared to postnatal repair, since prenatal surgery leads to a reduction in the placement of shunts, better neuromotor performance and improvement of hindbrain herniation. Thus, it is important to emphasize that, despite the great improvements in the neuropsychomotor prognosis after birth, fetal surgery has some limitations, among which the need for specialized centers and multidisciplinary teams of specialists stand out.^{7, 8, 9, 10}

The development of Arnold-Chiari II malformation is common in babies with MMC, this malformation is characterized by displacement of the brainstem through the foramen magnum. Patients with Chiari type II may develop abnormalities in the brain and hydrocephalus, with effects on nervous and cognitive motor function associated with this pathology, which is responsible for the highest number of deaths in children with MMC aged up to 2 years. The rhombencephalon herniation obstructs the outflow of liquor in the fourth ventricle causing hydrocephalus, which if not properly treated can cause an increase in intracranial pressure. This pressure results in caudal herniation of the brainstem and cerebellum.¹¹

An average of 80% of patients with Chiari type II have hydrocephalus, requiring the use of a shunt in the first months of life, which can cause morbidity and mortality. During the study that took place in neonatal centers in the USA, two children died who had undergone postnatal MMC surgery due to symptoms related to Chiari II malformation, both of whom had already undergone shunt placement. One of the treatments to avoid the consequences of hydrocephalus is the placement of a shunt. The occurrence of herniation and hydrocephalus are minimized when prenatal surgery is performed.¹¹

A study carried out in maternal-fetal surgery centers in the United States showed an improvement in the development of patients who undergo prenatal surgery for MMC, in relation to patients who undergo postnatal surgery. There was an average 50% reduction in shunt placement in infants aged 12 months when they had had intrauterine MMC surgery. In this group, there was also a decrease in the demonstration of hindbrain herniation in babies, and when they developed a hernia, the number of severe cases was lower compared to patients participating in the postnatal surgery group. At 30 months, the score measured by the Bayley Development Index and the anatomical and functional difference of the lesion showed better results for children who underwent prenatal surgery. Therefore, prenatal surgery proved to be more beneficial in most cases, despite the risks of prematurity and maternal and fetal morbidity, showing a better development of the child affected by MMC. One of the pieces of evidence that explains these results is the opportunity for the fetal development of the nervous system to be closer to normal when the surgery is performed early.¹¹

A study was carried out with 20 patients, with a mean age of 6.3 years, who presented spinal cord deformity related to myelomeningocele, between 2010 and 2017, compared the status of these patients before and after fusion surgery instrumented by constructions of rod and pedicle screws. Before surgery, the mean neurological status of the subjects, as given by the modified Japanese Orthopedic Association score (mJOA score), was 7.3 points, while the mean Motor Function Measure (MFM) was 41 points. Five subjects had focal and intermittent back pain, eleven patients had kyphoscoliosis, four had lordoscoliosis, four had lumbar kyphosis, and one had an inferiorly “flat” spine. The mean angle of kyphosis was 83.7 degrees Cobb, that

of scoliosis was 36.7 degrees Cobb, and that of lordosis was 67 degrees Cobb.

After surgery, the mean correction angle for kyphosis was 57 degrees Cobb, for scoliosis it was 25 degrees Cobb, and for lordosis it was 25 degrees Cobb. Neurological status improved by an average of 0.6 points, while functional status improved by 6.6 points. In addition, there was an improvement in back pain in all patients. However, ten patients experienced long-term complications associated with the implant, including screw misplacement seen in six patients. Thus, it is evident that, despite the low improvement in the neurological status, this surgical technique allowed the significant progress of the functional status and considerable correction of the spinal angle, providing better quality of life for these patients.¹²

The keystone design perforator island flap (KDPIF) is a plastic surgery technique that assists in the postnatal repair of complex congenital malformations such as MMC. The technique is projected onto the dermatomes, making cutaneous incisions parallel to the longitudinal axis of the injured area, in the outline of pre-established flaps, based on the muscular and fasciocutaneous perforating vessels of the back. The incision reaches the skin, the subcutaneous tissue up to the fascia, preserving the muscle layer. Thus, in addition to the geometric versatility and distribution of wound tension across the flap, the technique promotes adequate vascularization of the tissue and preserves a large part of the musculature and fascia, preventing late deformities.⁹

KDPIF promotes good healing, reducing the chances of dehiscence or necrosis of the skin flap, as well as minimizing the chances of infectious processes and cerebrospinal fluid leakage. The study carried out with 7 newborns with MMC observed, however, that many patients still needed ventriculoperitoneal shunting for the treatment of hydrocephalus

after repair. Thus, it is evident that KDPIF offers patients good aesthetic results, being feasible and effective for postnatal surgical correction of MMC, but it does not rule out the need for specialized follow-up and treatment of cases with concomitant hydrocephalus.⁹

Postnatal surgery seeks to reconstruct the dural sac, the superficial muscles and the dermal layers superimposed on the neural placode. Dural reconstruction is performed by duraplasty, using dural substitutes that fill in and reinforce the affected area or muscle fascia flaps. A study carried out with 5 patients with spinal dysraphism points to the cryopreserved amniotic membrane (AM) as a good option for a dural substitute, since it has high resistance and elasticity and promotes the reduction of inflammatory processes and adhesions to the neural tissue, in addition to benefiting re-epithelialization and preventing scars. With neurophysiological monitoring, the placode and meninges are then closed and covered by the homologous amniotic membrane layer. The use of AM as a dural substitute is shown to be quite effective and efficient for the surgical correction of MMC, both for its mechanical and insulating efficiency and for its anti-adhesive characteristic.¹⁰

In a concept survey where 446 neurosurgeons or neurosurgery residents from around the world were interviewed, members of the Congress of Neurological Surgeons (CNS) and the International Society of Pediatric Neurosurgery (ISPN) showed a diversity in the management of patients with MMC. Open fetal surgery and foetoscopic surgery are procedures offered in few centers and 83.1% of respondents do not agree that open fetal surgery must be the standard procedure to treat MMC. This is due to the restricted number of cases available to make the technique efficient, in addition to the risks involving fetal and maternal complications. This survey showed that 62.6% of respondents

agree that open fetal surgery causes a greater risk of neonatal death. Thus, even though open fetal surgery presents positive clinical results, it remains an issue due to risks to the fetus and also to maternal risks.¹³

CONCLUSION

The KDPIF technique presents good aesthetic results, reduces the chance of infection and extravasation of cerebrospinal fluid. In postnatal surgeries, it is also worth noting the good performance of using cryopreserved amniotic membrane in dural reconstruction. However, bearing in mind that in surgery after birth there is prolonged contact between the medulla and the amniotic fluid, there are disadvantages, such as the permanence of concomitant hydrocephalus. Prenatal surgical techniques, such as fetoscopy and open fetal surgery, also reduce the occurrence of hindbrain herniation and the need for a shunt. In the long term, patients undergoing these surgeries have better neurological performance and better urological outcomes. In this context, prenatal techniques, when feasible, offer greater benefit to the patient, despite the risk of premature delivery and the complexity of the surgery, as they offer a better quality of life for patients with MMC. Fusion surgery instrumented by constructing a pedicle screw rod is another tool that seeks to improve the patient's quality of life and, despite having reported complications, it was beneficial for the majority, improving back pain and functional status.

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