Prenatal myelomeningocele repair: report of an inaugural case and considerations about the implementation of a new fetal neurosurgery program

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Myelomeningocele (MMC) or open spina bifida is a neural tube defect which consists of a cleft in the vertebral column and a correspondent defect in the skin, causing the meninges and the spinal cord to be exposed. Despite some recommendations for nutrient supplementation during pregnancy, MMC remains very prevalent, especially in poor regions. MMC causes paralysis, urinary and gastrointestinal dysfunction, motor and neurological deficits, hydrocephalus and hindbrain herniation. Until 1997, the only way to repair this defect was performing surgery right after birth. In 2011, MOMS trial revealed a positive impact of prenatal surgery compared to postnatal. This paper reports the first case of one of the first myelomeningocele prenatal surgery programs in the North and Northeast regions of Brazil and discusses some issues about the implementation of new programs. The patient was a 34 years old woman in her second pregnancy, who was diagnosed at 22 weeks. She was submitted to an open fetal surgery applying MOMS protocol at 26 weeks of pregnancy and underwent cesarean at 36 weeks and five days of pregnancy. After two-years follow up, the baby girl did not need shunt for hydrocephalus, but has motor deficit in the lower limbs. Although fetal surgery is a high-tech procedure that demands a well-trained team, many families come from low-income regions. This way, the implementation of new services has an important social impact.

Keywords: Meningomyelocele, Fetal Therapies, Intra utero, open Spina Bifida

INTRODUCTION

Myelomeningocele (MMC), or open spina bifida, is a neural tube defect which consists of a cleft in the vertebral column and a correspondent defect in the skin, causing the meninges and the spinal cord to be exposed. It is the most common congenital anomaly of the nervous system compatible with life and can cause severe long-term disabilities [1, 2]. Although the recommendations of folic acid supplementation for couples planning a pregnancy, incidence in Brazil remains at approximately 1 per 1000 live births [3]. MMC causes paralysis, urinary and gastrointestinal
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dysfunction, motor and neurological deficits, hydrocephalus and hindbrain herniation [2,4].

As the additional methods have advanced, the diagnosis can be made early in the first trimester of pregnancy. Suspicion begins after tests such as ultrasound and serum alpha-fetoprotein levels are altered and can be confirmed with magnetic resonance imaging (MRI) and genetic testing if the ultrasound findings are nonspecific [2,5].

Until then, postnatal surgical repair of MMC has been the gold pattern and data has shown that it should be done as soon as possible. However, intrauterine repair of MMC began in 1997 through hysterotomy and early results suggested it could be beneficial in lowering hydrocephalus rates and improving motor function. On the other hand, despite the decrease in hindbrain herniation rates, there was an increase in preterm births, uterine dehiscence and fetal suffering. [5]

The two-hit theory explains that MMC neural damage can be the result of the primary congenital defect, a secondary event, as amniotic fluid exposure, trauma or hydrodynamic pressure, or both. Therefore, a primary congenital defect is susceptible to get secondarily damaged. This hypothesis shows why early fetal repair may have better outcomes [2].

In 2011, a randomized controlled trial named as Management of Myelomeningocele Study (MOMS) compared prenatal versus postnatal repair. As a result, the intra uterine repair was shown to decrease the need for ventriculoperitoneal shunt, as well as to improve the possibility of unassisted ambulation. Nevertheless, it was also more associated with oligohydramnios, chorioamniotic membrane separation, placental abruption, premature rupture of membranes, preterm delivery and uterine scar dehiscence [1,4].

A Brazilian group demonstrated that MOMS protocol could be replicated successfully outside the United States in 2012 [6]. In 2019, a group based in Fortaleza performed the first prenatal myelomeningocele repair in the state of Ceará and started one of the first myelomeningocele prenatal surgery programs on the North and Northeast regions of the country.

The aim of this paper is to report this inaugural case and discuss some issues about the implementation of a new fetal neurosurgery service.

CASE REPORT

A 34-year-old woman in her second pregnancy underwent morphological ultrasonography at 22 weeks of pregnancy. The exam revealed a defect of spinal closure with upper anatomical level of the lesion at L2, measuring 32 x 22 x 22 mm. The image also evidenced ventriculomegaly with a 13mm maximum ventricular width, cerebellum herniation through the foramen magnum and unilateral clubfoot. The first child of the woman had a myelomeningocele.

A Fetal magnetic resonance image (MRI) confirmed the sonographic findings (Figure 1). At 26 weeks of pregnancy, she was submitted to open fetal surgery according to the technique described in the Management of Myelomeningocele Study (MOMS), with exposure of the uterus and a small hysterotomy performed by the obstetric team, sufficient for good visualization of the myelomeningocele. The defect was then closed in three layers: dissection and repositioning of the placode, dissection and suture of the dural-fascial layer and skin suture (Figure 2). Although it was the first case operated in the region, the local team was assisted by a very experienced team of two obstetricians and two neurosurgeons from an experimented group from São Paulo.

Afterwards, the mother continued with uneventful prenatal care and underwent a cesarean section at 36 weeks and 5 days after having gone into labor. A female child was born weighing 2195 grams, with APGAR 9/10 and was sent to a rooming-in unit. She was then discharged uneventfully after 72 hours following a neurosurgical assessment. The lumbar scar was then in a very good state, the fontanel was not tense and there were no signs of active hydrocephalus. After one year and ten months follow-up, there was no need for hydrocephalus treatment. The girl requires daily bladder...
catheterization each 4 hours and presents distal motor deficit on the lower limbs.

Although the girl keeps presenting a relative macrocephaly and dilated ventricles at the follow up neuroimages, no objective signs of intracranial hypertension were noted.

On clinical evaluation, the patient has preserved hip flexor and adductor strength, good lower limb sensitivity, greater on the left. At one year and ten months, the child moves from dorsal and ventral decubitus to sitting, remaining seated for long periods for playing. She carries out locomotion through dragging but does not assume a posture of four supports or crawls.

Regarding communication, she speaks a few words, interacts and understands very well. The mother reports that despite not having sphincters control and being dependent on bladder catheterization the child reports intention and sensitivity of bladder functions.

DISCUSSION

Fetal surgery for MMC has changed the prognosis of the disease around the world, bringing lower rates of hydrocephalus and improving the motor status of children victims of this malformation. However, performing the surgery under adequate conditions requires qualified personnel and appropriate facilities, so that the benefits are achieved with the least possible risk. The prenatal repair of MMC requires a multidisciplinary team of obstetricians, neurosurgeons, neonatologists, anesthesiologists and nurses among other professionals [7]. The local team responsible for this first case in our state had experience in experimental models and visited several times a very experienced team in another region of the country. This same team assisted the local professionals performing this first case surgery.

Our case was in accordance with MOMS study inclusion and exclusion criteria. The fetus must present lesions from T1 to S1, with gestational age between 19 to 25 weeks and 6 days, and the mother must be older than 18 years. Exclusion criteria were ventriculomegaly over 16 mm, non-MMC-related abnormalities, prior risk of preterm delivery, severe kyphosis and abrupted placenta. Positive outcome is directly
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related to an interval of 19 to 27 weeks and 6 days of pregnancy, with a minimum maternal age of 18 years [1,6].

In several systematic reviews published in 2019, data that were congruent to this case report were obtained. It was demonstrated that intrauterine fetal surgery decreases the need for ventriculoperitoneal shunt. In the present case, after nearly two years of follow up, the patient did not require treatment for hydrocephalus. Although she presents a head circumference (HC) close to the 98 percentile, this variable growth speed decreased consistently after the first year and the child had no clinical findings suggestive of intracranial hypertension. This is in accordance with the literature that demonstrates a great chute on the need for hydrocephalus treatment in children operated before birth.

Fetal surgery for MMC can also improve the patients’ ambulation status. Our patient is not able to walk and presents an important distal lower limbs paralysis, probably due to the severity of the malformation in this particular case.

As a curiosity, there was a higher rate of tethered cord syndrome in prenatal surgeries in these series, as well as an earlier onset of this syndrome. Broader and more accurate observations could confirm this phenomenon [7-9]. When long-term outcomes were evaluated in a cohort study, a better motor development was evidenced, as well as a decrease in the rates of Chiari II malformations and hydrocephalus. In addition, quality of life was improved after intrauterine surgeries in the same study. [10]

Brock et al (2015) studied the urinary function of patients undergoing prenatal and postnatal myelomeningocele surgeries through randomized analysis. There was no significant difference in the urological status of patients [11]. In a study that evaluated the impact of gestational age on urinary pattern, it was evidenced that the influence is minimal [12]. The case described in the present report is in accord with the finding that the intrauterine intervention does not significatively changes the prognosis in terms of control of sphincters. Maybe technical improvements could change this situation in the future.

Preterm birth is one of the possible complications of oligodramnios after prenatal repair of myelomeningocele. In addition, early maternal age at surgery, as well as separation of the chorioamniotic membrane are important risk factors for a poor follow up described in the study of Johnson et al (2016) [13]. The first case of our service had a not severe preterm labor and the child presented no complications attributed to prematurity.

Despite the recent implementation of prenatal MMC repair, other even less invasive techniques are already being studied. The fetoscope technique, although promising, has not shown superior results when compared to the open technique until this moment. There are concerns about the cerebro-spinal fluid leak control due to difficulties to perform a watertight dural closure. The two-port endoscopic approach with uterus exposure is also a promising technique, waiting for larger series to gain terrain and to be proven advantageous [4,5]. We chose to start our program with the traditional technique of uterus exposure and hysterotomy in order to properly accomplish the learning curve, although we consider incorporating other techniques in the future. [14]

In conclusion, fetal surgery for myelomeningocele is a paradox in the sense it is a high-tech procedure with sophisticated personal and material resources for a malformation that is very prevalent in poor and underdeveloped regions. Furthermore, long trips during pregnancy to perform the surgery in specialized centers can represent discomfort for the mother and family. Thus, we believe that the creation of fetal surgery programs for myelomeningocele in centers like Fortaleza can provide access to treatment for more families and make the city a regional hub for this type of procedure, which results in a very positive social impact.

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the local Ethics Committee

Consent to participate

The patient gave consent to use his information and images for research proposes.

Consent for publication

The patient gave consent to use his information and images for publication.

Conflict of interest

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

Funding

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DOI: 10.46900/apn.v4i1(January-April).107
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This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors

ACKNOWLEDGMENTS

The authors are deeply grateful to the staff at Escola Paulista de Medicina (UNIFESP) and Hospital Santa Joana for their welcome in technical visits and for their in-person assistance during the inaugural procedure in our fetal surgery program for myelomeningocele. We are especially grateful to Drs. Antônio Moron, Maurício Barbosa (Obstetrics and Fetal Medicine), Sérgio Cavalheiro, Ítalo Suriano, Patrícia Dâstoli (Pediatric Neurosurgery) and Cid Ura (anesthesiology).

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