

# Functional performance of children with Myelomeningocele submitted to intrauterine repair and conventional surgery

## Abstract

**Introduction:** Myelomeningocele (MMC) is an embryonic malformation that occurs in the first four weeks of gestation due to a failure in the closure of the neural tube. They have multifactorial etiology such as genetic and environmental factors that affect folic acid metabolism. Several anomalies may be associated with MMC, with the occurrence of hydrocephalus and Arnold Chiari type II malformation being most present. Surgical repair should ideally occur within the first 24 hours after birth to prevent or decrease the risk of infection, preservation of all viable nerve tissue, anatomical reconstitution. However, surgical repair of MMC intrauterine has shown substantial benefit in all babies submitted to this intervention.

**Objective:** To compare the functional performance of children with MMC who underwent intrauterine surgical repair and those who did traditional surgery at birth.

**Methods:** Cross-sectional study carried out at the UCB School-Clinic, where children treated with MMC were evaluated, analyzing their motor status and functional performance using the Pediatric Evaluation of Disability Inventory (PEDI) for a quantitative record of functional capacity and autonomy in daily activities, in the areas of self-care, mobility and social function.

**Results:** There was a statistically significant relevance on the mobility function ( $p=0.01$ ) for children who underwent intrauterine repair when compared to children who underwent traditional surgery.

**Conclusion:** There is a strong relationship of better functionality of children who underwent intrauterine surgery when compared to children who underwent traditional surgery.

**Keywords:** myelomeningocele, children, intrauterine repair, traditional surgery, PEDI

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## Introduction

Myelomeningocele, also known as open spina bifida, is a congenital malformation of the child's spine in which the meninges, medulla and nerve roots are exposed. Myelomeningocele is the most common and the most severe type of spina bifida.<sup>1</sup> There is no specific causal factor; this condition appears to result from a combination of genetic and environmental factors, such as a family history of spinal malformations and folic acid deficiency. This pathology is the second leading cause of chronic deficiencies of the locomotor system in children.<sup>2</sup>

Normally, during the first month of pregnancy, the two sides of the spinal cord close over the spinal cord and all the accompanying nerves and meninges. The spine protects the spinal cord, preventing it from suffering any type of damage. When any type of congenital malformation occurs in which complete closure of the spine does not occur, it is called spina bifida.<sup>3</sup> Three different forms have been described: spina bifida occulta, which is asymptomatic, meningocele, in which only the meninges of the spine are exposed at birth, and myelomeningocele, in which, in addition to the meninges, there is avulsion of the spinal cord and innervation.<sup>4</sup> It is more common among whites and Hispanics and is more common in females.<sup>5</sup> Symptoms depend on the location and degree of spinal cord exposure. This malformation manifests itself through motor, sensory and trophic alterations, bladder and bowel incontinence and sexual dysfunction such as decreased muscle strength, flaccid paralysis, hydrocephalus, and sphincter incontinence.<sup>6</sup>

The prognosis is linked to the level of injury and worsens with the presence of hydrocephalus, spinal deformities, or additional injuries. The diagnosis can still be made intrauterine, which increases the chances of treatment. This is possible and should happen in the second trimester, during the morphological ultrasonography, in which characteristics in the shape of the skull can already be observed, and the inverted cerebellum, resulting from the traction of the brainstem, in addition to the presence of a defect in the thoracic, lumbar, or sacral spinal column. If diagnosed before the 24th week of pregnancy, there is the possibility of performing intrauterine surgery or fetal surgery, which is performed by exteriorizing the uterus. Laparotomy, hysterotomy are performed, the fetus is exposed, and the neurosurgeon addresses the myelomeningocele. A hysterorrhaphy is then performed and the pregnancy continues for about two months.<sup>7</sup> Recent studies prove that the benefits of the intervention during pregnancy are superior to those of the postpartum technique.<sup>7</sup>

Brazil was the first country in Latin America to perform intrauterine open surgery for the correction of myelomeningocele, the practice began in 2011 at Hospital Maternidade Santa Joana and at UNIFESP.<sup>8</sup> Until then, the only possible surgical option was conventional postnatal surgery, where the birth defect is corrected within 72 hours after the baby's birth.<sup>8</sup> When the child has associated hydrocephalus, it is necessary to implant a ventriculoperitoneal shunt to drain the cerebrospinal fluid, where it will be reabsorbed. Hydrocephalus is present in 80% of children with myelomeningocele and is one of the worst prognostic factors.<sup>8</sup>

Studies report that intrauterine intervention is an extremely important advance, as the technique corrects the defect in the baby’s spine, avoiding hydrocephalus in most cases.<sup>9</sup> Therefore, the importance of research to evaluate children submitted to intrauterine surgery and conventional surgery comparing both treatments to reinforce the benefits in the fetal approach. Studies prove that the benefits of the intervention during pregnancy are superior to those of the postpartum technique and that, over the years, it has been proving even more positive.

Thus, the present study seeks to evaluate the children assisted at the school-clinic of Universidade Católica de Brasília (UCB) who were submitted to both surgical techniques (intrauterine and traditional), verifying their functional acquisitions as well as quantifying their functional performance.

## Materials and methods

Cross-sectional study carried out in November 2018 with the evaluation of all children with Myelomeningocele treated at this Center, in the Neuropediatric sector, during this period. Thirteen children whose family members agreed to participate in the study by signing the informed consent form met the inclusion criteria. However, 3 children were excluded, two of which were hospitalized and one who did not attend at the time of collection. Thus, 10 children were included. An interview was scheduled with the families where

data on the gestational history and the child history were collected and subsequently the *Pediatric Evaluation of Disability Inventory* (PEDI) was applied.

The PEDI reports three important aspects of functional performance: Part I) the skills in the child’s repertoire; Part II) independence in daily activities; Part III) the modifications of the environment used to facilitate functional performance. The scale was created to measure, in each one and its parts, functional changes in self-care activities, mobility and social function.<sup>10</sup> Statistical analysis was performed using the *Statistical software Package for Social Science (SPSS)* version 20.0 in which means, variances and frequencies were calculated. Associations were verified using the Chi-square test and correlations using Pearson’s coefficient, adopting 5% as the significance level.

## Results

Ten children participated in the present study, 3 females and 7 males, with a mean and standard deviation of age of 32.2±15.5 months. Table 1 shows the data found regarding the profile of the studied children. It is observed that there was a higher prevalence in boys (70%) than in girls (30%) with a mean age equivalent to 2.6 years (32.20 months) SD: 15.5; and that 50% of the children have lumbar sacral injuries, while 40% have lumbar injuries and 10% have high thoracic injuries.

**Table 1** Profile of the studied children

Patients initials	Age (Months)	Sex	Level of injury	Hydrocephalus	Arnold chiari II	Surgical repair	Ambulation
HRO	13	male	Low back	Yes	Yes	Postnatal	Dont walk
CSM	48	male	Low back	Yes	Yes	Postnatal	Dont walk
MAM	29	male	Low back	No	No	Intrauterine	Walk with a walker
TFA	11	male	Low back	Yes	Yes	Postnatal	Dont walk
AOC	17	female	Lumbosacral	No	No	Intrauterine	Walk independently
AF	48	female	Thoracic	Yes	Yes	Postnatal	Walk with a walker
LV	40	male	Lumbosacral	Yes	Yea	Postnatal	Walk with a walker
GCH	22	male	Lumbosacral	No	No	Intrauterine	Walk with a walker
AVS	48	female	Lumbosacral	No	No	Intrauterine	Walk independently
LHSM	46	male	Lumbosacral	No	No	Intrauterine	Walk independently

Hydrocephalus was present in 50% of the patients and the Arnold Chiari II malformation was also present. Regarding walking, 30% do not walk, 40% walk with a walker and 30% walk independently. Five of the children participating in the study were submitted to intrauterine surgical treatment and the other five to the traditional postnatal

procedure. Table 2 presents the scores obtained on the PEDI by the children grouped according to intrauterine and postnatal surgical treatment, in the areas of self-care, mobility and social function. The Mann-Whitney U t test verified a statistically significant difference for the mobility function (p=0.01).

**Table 2** Mann-Whitney U test

PEDI	Intrauterine surgery (Mean±SD)	Postnatal surgery (Mean±SD)	p-value
Self care	25.8±13.3	13.6±12.8	0.31
Mobility	24.6±11.6	3.6±3.7	0.01
Social Function	25.0±14.0	16.2±15.7	0.59

## Discussion

Initial studies of intrauterine surgery repair suggested an important decrease in the occurrence of hydrocephalus and Chiari II malformation, but also showed an increase in maternal-fetal risk

including premature labor, uterine dehiscence, prematurity, and fetal death. To assess the effectiveness and risks of such an intervention there was a need for a randomized controlled trial. In 2003, the MOMS (Management of Myelomeningocele Study) began, with the aim of recruiting 200 randomized patients either for intrauterine repair

(prenatal) or for traditional postnatal repair. For ethical reasons, the study was interrupted with 183 patients recruited and published in 2011, showing that for the selected population, intrauterine surgery was significantly beneficial, although there were maternal-fetal complications.<sup>11</sup>

In the present work, it was observed that of all the children assisted, 5 had hydrocephalus and they had a Chiari II malformation at birth. 50% underwent intrauterine surgery and 50% conventional surgery. Of the children who underwent fetal surgery repair, 3 had intrauterine hydrocephalus seen on morphological ultrasound, but were born without any signs of it, showing superiority of the intrauterine technique over conventional surgery. Studies show that injury levels are directly proportional to limitation in functional performance, being greater the higher level of the malformation.<sup>12-18</sup> In the present study, a child had an upper thoracic injury, and his motor performance was not the worst, which disagrees with these studies.

In patients with MMC, therapy has traditionally focused on improving muscle strength, adjusting tone, and preventing contractures, as well as optimizing child development and functionality. In the present study, the PEDI was important to allow a functional analysis of the performance of the child with MMC, since functionality is the primary focus in the multidisciplinary intervention for rehabilitation. Studies have found low PEDI scores, that is, functional limitations in activities related to self-care, mobility, and social function,<sup>13,14</sup> as in the present study.

In the PEDI self-care area, activities that require degrees of mobility and body control are quantified, such as washing, dressing, and combing the hair. In general, MMC generates limitations in these motor degrees, resulting in sensorimotor paralysis, which is directly proportional to the level of malformation.<sup>12</sup> In the present study, this was the highest score found, which could be attributed to the age of the children studied (2.6 years old), the age at which the child really needs more parental care.

Mobility is the individual's ability to move efficiently in the environment. In general, children with MMC have limited mobility, which can influence their participation in different activities.<sup>15</sup> In this study, the score for this area was like those cited in the literature,<sup>11,12</sup> however, if we compare this area of children who underwent to intrauterine surgery and those who underwent conventional surgery, this was the comparison with the greatest statistical evidence showing the superiority of open fetal surgery. It is observed that these children show better mobility daily, where of the 5 children who were operated on while still in the womb, they acquired independent gait.

The area of social function, for some authors, represents the best results.<sup>12</sup> Cognitive impairment is often attributed to hydrocephalus and the percentage of hydrocephalus associated with MMC in the literature is described as present in more than 85% of patients.<sup>16-19</sup> In the present study, this area was affected not by hydrocephalus or cognitive alterations, but probably because of dealing with a very young population of children, which made it difficult to assess this function.

## Conclusion

The present study concluded amongst the children studied, those who underwent intrauterine surgical treatment had better motor and functional development, while those who underwent conventional treatment showed worse scores, especially in mobility. Fetal repair of myelomeningocele has been widely debated and studied, providing important data that point to a reduction in the incidence of

hydrocephalus and improvement in motricity, as well as a better future prognosis. New analysis should be carried out to confirm the findings in the present study, mainly considering the limitation of this sample, with associated symptomatic neurological disorders.

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## Conflicts of interest

The author declares that there is no conflict of interest.

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