Clinical and ultrasonographic criteria for using ventriculoperitoneal shunts in newborns with myelomeningocele

Critérios clínicos e ultrassonográficos para a indicação de derivação ventrículo peritoneal em neonatos portadores de mielomeningocele

Jose Roberto Tude Melo¹, Pollyana Pacheco¹, Emília Nunes de Melo¹, Ângela Vasconcellos¹, Rosane Klein Passos²

Myelomeningocele (MM) is the most common form of spinal dysraphism being the complex congenital malformation of the central nervous system (CNS) compatible with life, of higher incidence²,³,⁴. Despite prophylactic measures, such as use of folic acid before and during pregnancy⁵,⁶, incidence of MM remains high, ranging between 0.1 and 10 per 1,000 live births⁷. Moreover, MM is more common in countries showing low socioeconomic development⁸. According to the World Health Organization, among 41 evaluated countries, Brazil ranks fourth in the incidence of spina bifida, at a rate of 1.1-1.000 live births⁹. Hydrocephalus is present in most children with MM⁵, thus making it essential to establish criteria for indicating ventriculoperitoneal (VP) shunts in these children⁴,¹⁰,¹¹,¹².

Brazil is a large country with socioeconomic imbalance¹³ and poor accessibility to radiological methods of diagnosis, such as computed tomography (CT) or magnetic resonance imaging (MRI), for routine examination of neonates with neurological diseases in public health hospitals, which makes it difficult to apply standardized protocols followed in developed countries⁹,¹¹,¹⁴. This study aimed to describe the epidemiological profile of children with MM by considering the records of a public pediatric reference hospital and identify clinical and transcranial ultrasonographic criteria for the indication of VP shunts in these children.

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METHOD

This research was approved by the Brazilian Research Ethics Committee (registration number 14990213.5.0000.5543).

This retrospective cohort study included the medical records of all newborns who underwent surgical closure of MM at a reference public pediatric hospital in Salvador da Bahia, Brazil, between 2009 and 2013. All the children included in this study underwent the same protocol for hydrocephalus in case of MM, as those described in previous reports9,11,12. Transcranial ultrasonography (TUS) was chosen as the radiological method for diagnosing hydrocephalus in all neonates, as that described in previous studies12,16,17,18, because of the difficulty in performing CT scan or MRI in these children. TUS was performed with a 1.9- to 6 MHz curvilinear transducer (Toshiba Aplio 100® with color Doppler), with a classical trans-fontanelar window approach by a trained operator (senior radiologist with > 10 years experience in TUS).

Based on previously established measurements, a neonate was considered as having mild hydrocephalus when the transversal lateral ventricular atrium (LVA) width, measured slightly above the level of the thalami at the level of the choroid plexus, was < 15 mm (10-14mm)19. When this value was ≥ 15 mm (15-19mm), the neonate was considered as having moderate hydrocephalus18. Thus, the study population was divided into two groups:

- Group A: Children with LVA width of < 15 mm (n = 12)
- Group B: Children with LVA width of ≥ 15 mm (n = 31)

Because hydrocephalus mostly develops in the first 43 days after the closure of MM9, we recommended a biweekly follow-up during the first two months after the closure of MM to measure head circumference (HC) and LVA width (by TUS). We consider the measure of HC and evaluation of the fontanels as the most important clinical signs to be observed in these children, according to previously published studies11,15,16,17,19. Infant girls with HC of 37.7 cm and infant boys with HC of 38.5 cm who showed an increase in the ventricular system were considered as having hydrocephalus since birth17,18,20. In this group, the proposal for treatment was with closure of MM and VP shunt at the same surgical time. VP shunt was chosen as the method of choice for treating hydrocephalus in these children after considering the benefits and drawbacks of endoscopic third ventriculostomy (ETV) in neonates with MM12,17,21.

Statistical analyses

Based on previous study on the incidence of hydrocephalus in children with MM9, our study included 43 children, considering a sampling error of 5% and a confidence interval of 95%. Some results were exposed in descriptive manner, dispensing statistical analyses. Measures of central tendency (mean, mode, and median) were calculated and presented where relevant. We compare groups (exposed and control groups) and verified the odds ratio. The differences were considered statistically significant when p < 0.05 (Z statistic test). The sensibility, specificity, predictive values and efficiency (the proportion of correct predictions; sum of true positives and true negatives) were calculated, considering the LVA width, 15 mm cutoff point.

RESULTS

Forty-seven newborns with MM were admitted for treatment during the proposed period. Six newborns (14%) were considered premature (< 37 weeks’ gestation). Follow-up was irregular in 4 babies; therefore, they were excluded from the final analysis. Forty-three newborns were consecutively included in the study. Nineteen mothers (44%) received regular prenatal care (≥ 6 medical appointments) that involved regular folic acid replacement during pregnancy and 70% came from the countryside area. Eleven mothers (25%) underwent a transvaginal delivery. Our hospital did not have a maternity unit (obstetric department), and all the children were transferred from other hospitals usually 24 h after birth. Among the newborns included in this study, 51% were boys and 95% had lumbar and lumbosacral lesions. MM was present as a ruptured lesion in 50% of our sample. Intrauterine corrections were not performed for the closure of MM in our hospital until the end of this research. Table 1 shows the main epidemiological aspects of children with MM that were evaluated in this study.

The overall incidence of hydrocephalus and the need of VP shunts were observed in 32 (74%) children. LVA width was measured by performing TUS at 48-72h after the closure of MM. VP shunt were required in 50% (6/12) children with an LVA width of < 15 mm (group A) and in 84% (26/31) children with an LVA width of ≥ 15 mm (group B; p = 0.02; OR = 5.2). Evaluating the sensitivity and specificity of LVA width as a measure to guide the indication of VP-shunt, we found sensitivity of 81%, a positive predictive value of 84%, and efficiency of 75% considering 15 mm cutoff value (Table 2).

Progressive increase in HC and bulging fontanels were the earliest signs of intracranial hypertension (ICH) (observed in 94% children; 30/32); these signs were predominant in children with LVA width of ≥ 15 mm. In the remaining two children (6%; 2/32), VP shunts were indicated based on other signs of ICH such as respiratory distress, hypotonia, and drowsiness; these were consistent with the increases in the ventricular system identified using TUS before hospital discharge. The average duration for the development of hydrocephalus was 35 days (range, 0-180 days) after the closure of MM. Figure shows the protocol for VP shunt in children with MM after considering TUS observations and clinical criteria. Three children simultaneously underwent closure of MM and ventricular drainage because at hospital admission, they had
Table 1. Epidemiological aspects of 43 children with myelomeningocele (MM) who were treated in a referral pediatric hospital in the state of Bahia, Brazil (2009-2013).

<table>
<thead>
<tr>
<th>General Characteristics</th>
<th>n (%)</th>
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<tbody>
<tr>
<td>Regular prenatal care (≥ 6 medical appointments)</td>
<td>19 (44)</td>
</tr>
<tr>
<td>Regular use of folic acid</td>
<td>19 (44)</td>
</tr>
<tr>
<td>Prenatal diagnosis of MM</td>
<td>10 (23)</td>
</tr>
<tr>
<td>Region of origin</td>
<td></td>
</tr>
<tr>
<td>Capital (city of Salvador da Bahia)</td>
<td>13 (30)</td>
</tr>
<tr>
<td>Rural zone (countryside)</td>
<td>30 (70)</td>
</tr>
<tr>
<td>Males</td>
<td>22 (51)</td>
</tr>
<tr>
<td>Association with hydrocephalus*</td>
<td>32 (74)</td>
</tr>
<tr>
<td>*Hydrocephalus requiring a VP shunt.</td>
<td></td>
</tr>
</tbody>
</table>

Table 2. Comparison among the 43 newborns undergoing evaluation by transcranial ultrasonography (TUS) and lateral ventricular atrium (LVA) width, measured slightly above the level of the thalami at the level of the choroid plexus (15 mm cutoff), and the need of VP-shunt.

<table>
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<tr>
<th>TUS</th>
<th>VP shunt</th>
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<tbody>
<tr>
<td></td>
<td>Yes</td>
</tr>
<tr>
<td>LVA ≥ 15 mm</td>
<td>26</td>
</tr>
<tr>
<td>LVA &lt; 15 mm</td>
<td>6</td>
</tr>
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We verified a sensitivity of 81%; specificity of 55%; positive predictive value of 84%; a negative predictive value of 50%, and efficiency of 75%, considering the TUS and LVA width measure (15 mm cutoff point).

DISCUSSION

A previous study identified the irregularity and deficiency of prenatal monitoring in pregnant women, assisted by a poor public health system, in the state of Bahia (Brazil) and indicated that these may be associated with the high incidence of spinal dysraphism. We recommend, as suggested by other authors, that at least 400 μg of a folic acid supplement should be consumed daily; patients with an intermediate to high risk of neural tube defects (such as patients with a previous well-known history of neural tube defects) should consume 4-5 mg folic acid. Investigation of proposed type of delivery revealed high rates (25%) of vaginal delivery. The study suggested that women undergoing transvaginal delivery may receive irregular prenatal care that would consequently lead to the failure of diagnosing MM in the fetus, which in turn may lead to misguided management based on the type of delivery: Vaginal delivery may increase the risk of rupture and infection of MM during the passage of the fetus from the birth canal; however, indication of cesarean delivery in these pregnant women and its benefits in newborn are still controversial.

In our study, all the children were transferred from other hospitals in the city of Salvador da Bahia or from a countryside, which is justified by the fact that our hospital is a referral center for treating children with MM in this region. Because of the lack of an obstetric department in our hospital, these children usually arrive at our hospital 24 h after birth. This delays the immediate closure of the lesions, which should in fact be corrected immediately, and hinders the training of a multidisciplinary team involved in intrauterine closure and correction of MM. The same situation is observed in other Brazilian states where many maternity hospitals do not have a unit for pediatric neurosurgery, thus making it necessary to transfer these children to other hospitals.

The relationship of gender distribution of children with MM can be verified in other studies as well as the predilection of lumbar and lumbosacral region involvement. Probably, the fact that all children with MM who were evaluated in this study have awaited transfer for treatment, besides the relatively high number of vaginal delivery, justify the large number of ruptured MM (50%). MM, particularly when ruptured, must be considered as a neurosurgical emergency because of the high risk of CNS infections, which is a large public health concern in our state. All children with MM undergo surgical closure of the MM as soon as they are transferred to our hospital to prevent the risk of infection and the risk associated with keeping the placode exposed.

Data indicate that the incidence of hydrocephalus and the requirement of VP shunts in these children vary from 52% to 90%; however, some of these data do not provide a clear definition of the optimal timing and criteria for CSF diversion in children with MM. We verified that 74% children with MM who underwent an institutionalized protocol based on the clinical and TUS criteria required VP shunts, which is the same as the percentage of children who underwent CT scan-based protocols. The mean duration of 35 days between the closure of MM and development of hydrocephalus observed in our study was similar to that observed in other studies, which emphasizes the importance of regular follow-up, especially in the first 60 days after the closure of MM. However, it should be noted that in some children, hydrocephalus may develop later (within 180 days after the closure of MM).

VP shunt should be considered after a careful clinical and radiological evaluation. According to the results reported here, LVA width of 15 mm, as per the TUS, could be considered as a threshold to classify children with a lower or higher
Measurement of LVA width by using TUS was performed 48-72 h after the closure of MM in our study. It was observed that VP shunt were required in 84% children with LVA width ≥ 15 mm, which was statistically different (p = 0.02) from that required by children with lesser LVA width, with a sensitivity of 81% and a positive predictive value of 84%. Because of the high incidence of hydrocephalus in these children, it is essential to closely monitor them for signs of ICH after the closure of MM9,11,12,22,26. Besides the LVA width measured slightly above the level of the thalami at the level of the choroid plexus, other criteria for hydrocephalus such as bifrontal diameter, bicaudate diameter, diameter of the body of the lateral ventricle, ventricular index, frontooccipital horn ratio, and thalamooccipital distance had already been studied in patients with MM and hydrocephalus11,29. Was not the aim of our study comparison between these methods and measures.

In children who still have opened fontanels and sutures, increased HC and bulging fontanels are the main clinical manifestations11,15,17,19 that require imaging methods to confirm the diagnosis and to implement an appropriate treatment11,12,16,17,29. In children with greater LVA width (≥ 15 mm), signs such as progressive increase in HC or bulging fontanels occur more frequently, which lead to the indication of VP shunts18. We emphasize that 6% of the children who required a VP shunt showed progressive increase in the ventricular system (identified using TUS) without an increase in HC or bulging fontanels, however showed signs suggestive of neurological impairment due to Chiari II malformation. Other studies also highlight children with signs of ICH but without bulging fontanels11,15.

The best surgical time for VP shunt in these children remains unknown (i.e., at the same time of the closure of MM or not). Results of other studies verify that closure of MM and VP shunt sometimes should not be performed in the same surgical time. It is emphasized that concomitant surgeries (closure of MM and VP shunt) should be avoided when MM is corrected 24 h after birth and that there is a possibility of infection at the site of MM and higher risk of complications associated with the shunt9,12. Complications rates were similar to those reported by other authors, considering CSF diversion12,17.

We recommend that these children should be monitored regularly by a multidisciplinary team in a systematic manner in the long-term mainly due to orthopedic and urinary malformations. Protocols should be followed correctly, according to the possibilities of each region (city, state, or country), to avoid overindication of VP shunt in these children and to avoid systematic realization of CT scan and unnecessary irradiation in these neonates. It should be noted that the results presented here are from a single referral center treating children with MM and may not reflect the reality in other regions. No single criteria can be considered completely safe, and TUS parameters used in the present study seeks to integrate a possible measure to be used, and under any circumstances overrides other previously published ventricular and ultrasound measurements. We plan to extend the protocol used in this study along with the
inclusion of transcranial Doppler to figure out the resistive values of the cerebral vascular vessel and in postoperative follow-up study of these children to better identify the functioning of the VP shunts.

In conclusion, definition of criteria that identify children with MM who need a VP shunt is crucial to eliminate unnecessary CSF diversion. By following our institutionalized protocol, we observed that 74% children who had MM required a VP shunt, which was closely the same observed with other protocols based on CT scan. In our sample VP shunts are usually indicated in children with an IVA width of ≥ 15 mm, increased HC (2 standard deviations for age), or bulging fontanels. We all know that no method or measure used in isolated form can be considered completely safe. We believe that the measures presented here considering the IVA width can be added to ventricular measurements and parameters previously described in the literature, assisting in the monitoring of children with MM.

References


