



# Clinical and Radiological Predictors of Ventriculoperitoneal Shunt Insertion in Myelomeningocele Patients

Ahmed M. F. Ghoul\*, Ahmed Hamdy Ashry, Mohamed Hamdy El-Sissy, Ibrahim Mohamed Ibrahim Lotfy

Department of Neurosurgery, Kasr Alainy Faculty of Medicine, Cairo University, Cairo, Egypt

## Abstract

**Edited by:** Ksenija Bogoeva-Kostovska  
**Citation:** Ghoul AMF, Ashry AH, El-Sissy MH, Lotfy IM. Clinical and Radiological Predictors of Ventriculoperitoneal Shunt Insertion in Myelomeningocele Patients. Open Access Maced J Med Sci. 2021 Jul 02; 9(B):522-532. <https://doi.org/10.3889/oamjms.2021.6265>  
**Keywords:** Myelomeningocele; Hydrocephalus; Ventriculoperitoneal shunt; Chiari malformation type II; Cerebrospinal fluid; Intracranial pressure; Intra uterine myelomeningocele repair  
**\*Correspondence:** Ahmed M. F. Ghoul, Department of Neurosurgery, Faculty of Medicine, Cairo University, Egypt. E-mail: [ahmedghoul9@gmail.com](mailto:ahmedghoul9@gmail.com)  
**Received:** 24-Apr-2021  
**Revised:** 19-May-2021  
**Accepted:** 20-May-2021  
**Copyright:** © 2021 Ahmed M. F. Ghoul, Ahmed Hamdy Ashry, Mohamed Hamdy El-Sissy, Ibrahim Mohamed Ibrahim Lotfy  
**Funding:** This research did not receive any financial support  
**Competing Interests:** The authors have declared that no competing interests exist  
**Open Access:** This is an open-access article distributed under the terms of the Creative Commons Attribution-NonCommercial 4.0 International License (CC BY-NC 4.0)

**BACKGROUND:** Myelomeningocele (MMC) is one of the most common developmental anomalies of the CNS. Many of these patients develop hydrocephalus (HCP). The rate of cerebrospinal fluid diversion in these patients varies significantly in literature, from 52% to 92%. MMC repair conventionally occurs in the post-natal period. With the technological advances in surgical practice and fetal surgeries, intra uterine MMC repair IUMR is adopted in some centers. Cerebrospinal fluid shunting has numerous complications, most notably shunt failure and shunt infection. Studies have suggested that patients with greater numbers of shunt revisions have poorer performance on neuropsychological testing. There is also good evidence to suggest that the IQs of patients with MMC who do not undergo shunt placement are higher than that of their shunt treated counterparts.

**AIM:** In this study, we are trying to identify strong clinical and radiological predictors for the need of ventriculoperitoneal (VP) shunt insertion in patients with MMC who underwent surgical repair and closure of the defect initially. This will decrease the overall rate of shunt placement in this group of patients through applying a strict policy adopting only shunt insertion for the desperately needing patient.

**METHODS:** Prospective clinical study conducted on 96 patients with MMC presented to Aboul Reish Pediatric Specialized Hospital, Cairo University. After confirming the diagnosis through clinical and radiological aids, patients are carefully examined, if HCP is evident clinically and radiologically a shunt is inserted together with MMC repair at the same session after excluding sepsis or cerebrospinal fluid (CSF) infection, (GROUP A). If there are no signs of increased ICP, MMC repair shall be done alone (GROUP B). Those patients shall be monitored carefully postoperatively and after discharge and shall be followed up regularly to early detect and promptly manage latent HCP. Multiple clinical and radiological indices were used throughout the follow-up period and statistical significance of each was measured.

**RESULTS:** Shunt placement was required in 45 (46.88%) of the 96 patients. Eighteen patients (18.75%) needed the shunt as soon as they presented to us (GROUP A), because they were having clinically active HCP. Twenty-seven (28.13%) patients were operated on by MMC repair initially without shunt placement because they did not have signs of increased ICP at the time of presentation. Yet, they developed latent HCP requiring shunt placement during the follow-up period (GROUP B2). Fifty-one patients of the study population (53.13%) underwent surgical repair of the MMC without the need of further VP insertion and they were followed up for 6 months period after the repair without developing latent HCP (GROUP B1). Patients of GROUP B were the study population susceptible for the development of latent HCP. Out of 78 patients in GROUP B, only 27 patients (34.62%) needed a VP shunt.

**CONCLUSION:** In our study, we found that the rate of shunt insertion in patients with MMC is lower than the previously reported rate in the literature. A more thorough evaluation of the patient's post-operative need for a shunt is mandatory. We suggest that we could accept postoperative (after MMC repair) ventriculomegaly provided it does not mean any deterioration in the patient's clinical or developmental state. We assume that reduction of shunt insertion rate will eventually reduce what has previously been an enormous burden for a significant proportion of children with MMC.

## Introduction

Myelomeningocele (MMC) is one of the most common developmental anomalies of the CNS. Many of these patients also develop hydrocephalus. The rate of CSF shunt placement in these patients varies significantly in published studies, from 52% to 92% [1]. Myelomeningocele repair conventionally occurs in the post-natal period. With the technological advances in surgical practice and fetal surgeries, intra uterine myelomeningocele repair IUMR is adopted in some centers in the developed countries. IUMR appears to substantially reduce the incidence of shunt-dependent

hydrocephalus when compared to conventional treatment [2]. Cerebrospinal fluids hunting has numerous complications, most notably shunt failure and shunt infection. Studies have suggested that patients with greater numbers of shunt revisions have poorer performance on neuropsychological testing [3]. There is also good evidence to suggest that the IQs of patients with myelomeningocele who do not undergo shunt placement is higher than that of their shunt treated counterparts. Furthermore, it has been suggested that it is the infective complications of shunt-treated hydrocephalus rather than the hydrocephalus per se that has the greater impact on intelligence in this population [4].

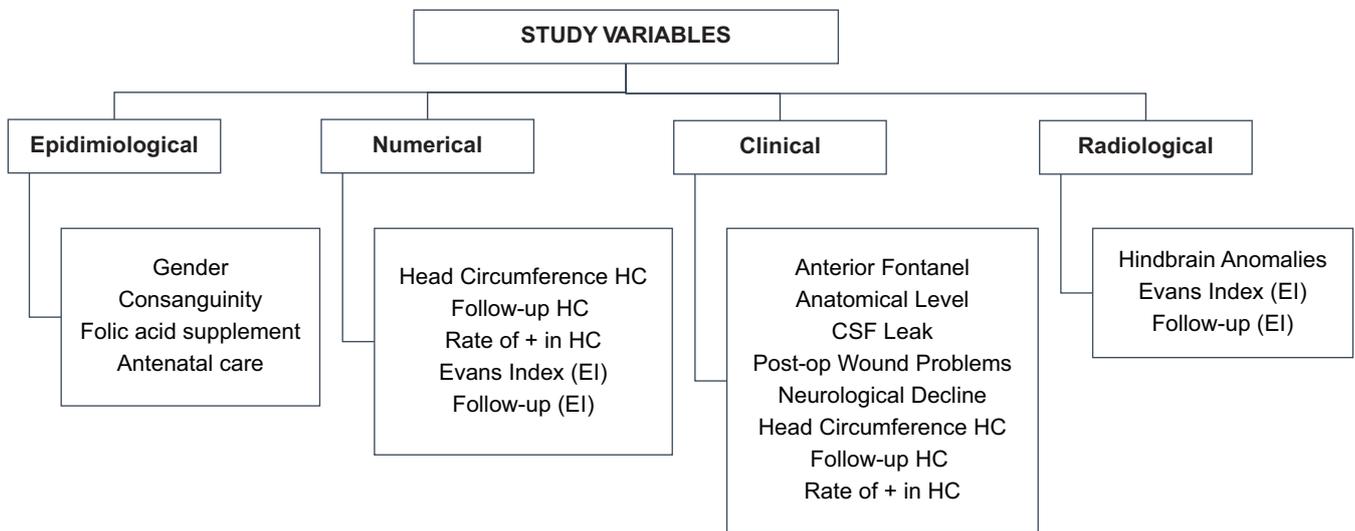


Figure 1: Study variables used in the study

## Methods

This prospective clinical study was conducted on infants with myelomeningocele (MMC) fulfilling the inclusion criteria presented to Aboul Reish Specialized Pediatric Hospital, Cairo University Hospitals, either in the outpatient clinic, or at the emergency department. Diagnosis will be confirmed through history (swelling, apnea, dysphagia, and enlarged head), thorough clinical examination (presence of MMC, and motor weakness), and radiological evidence of cranial and spinal anomalies (hydrocephalus [HCP], hindbrain herniation, and syrinx).

Strict protocol will be applied to manage those patients regarding shunt placement. The decision of shunt placement will be taken by two senior neurosurgeons in the pediatric neurosurgery unit at Aboul Reish Hospital and it will be strictly exclusive for patients with early or latent active HCP confirmed by clinical signs and radiological tools.

All patients will undergo the routine preoperative workup such as full routine labs and echocardiography to rule out the presence of other anomalies requiring special attention. They will also undergo non-contrast CT scan of the brain, MRI brain, MRI Spine (according to the level of MMC), and MRI cranio-cervical junction.

Ninety-six patients were included in the study. The duration of follow-up was 6 months. All patients presented to us postnatally and they were examined meticulously and included into the study according to our inclusion criteria mentioned in the methodology section.

We did not meet any technical difficulty in closure and repair in patients with large MMC. We did not have any patient with very large problematic MMC, requiring surgical insertion of tissue expansion devices as a primary procedure; followed by delayed closure of the skin defect and closure of the MMC.

The mean age at which repair was completed was 16.84 days (median 6.50 days). The elevated mean and median age for intervention are attributed to the fact that some of the patients included in the study were operated on in a delayed age due to various medical and non-medical causes such as [1] anesthetic consideration regarding fitness for surgery, [2] the need to exclude or prove the presence of multiple congenital anomalies to promptly execute the perioperative planning which might take some time until finished, and [3]. unequal distribution of the tertiary health-care facilities in which patient can find highly trained medical personnel that could deal with such complicated cases in a multi-disciplinary manner. Hence, parents who cannot reach a nearby hospital for their child to be treated tend to travel hundreds of kilometers to reach a specialized institute in the capital.

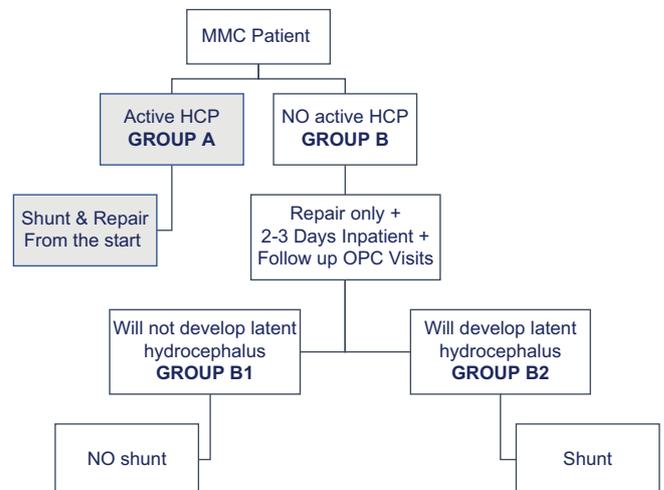


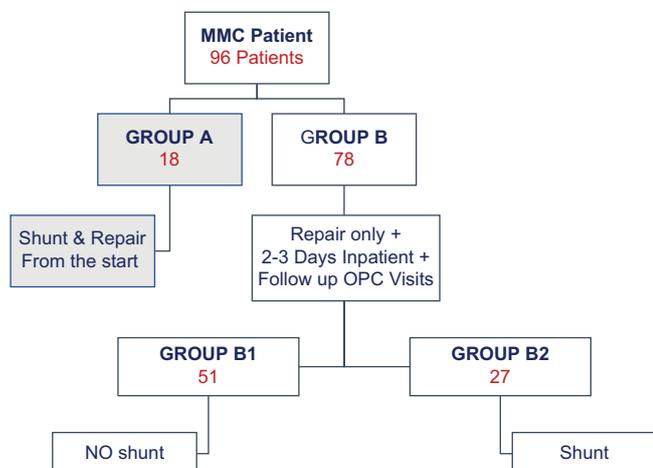
Figure 2: Study population groups

The group of patients (Figures 2 and 3) who presented with MMC with signs of increased intracranial pressure ICP (active HCP) was assigned to have a ventriculoperitoneal (VP) shunt insertion and MMC repair at the same session (starting with the

shunt placement) after exclusion of CSF infection or septicemia on clinical and laboratory basis. This group of patients was named (GROUP A).

The group of the patients who presented with MMC without signs of increased intracranial pressure (active HCP) was assigned to do MMC repair only without VP shunt insertion. This group was named (GROUP B).

The latter group of patients was followed up in the early post-operative period in the pediatric neurosurgery inpatient ward at Aboul Reish University Hospital for 3 days. Clinical and radiological signs of latent HCP were meticulously monitored. If latent HCP develops, the patient is scheduled for a VP shunt. Patient is discharged if there are no signs of active HCP, and his/her parents receive a thorough detailed program for post-operative follow-up in the outpatient clinic together with certain instructions to be aware of warning signs of latent HCP at home. If any of these signs is evident, they were instructed to return back to our institute as soon as possible. Patients of Group B were categorized into two subgroups according to the fact that they developed latent HCP and needed a shunt or not:



**Figure 3:** Patients distribution among the groups of study population

- GROUP B1 MMC repair only WITHOUT VP shunt insertion
- GROUP B2 MMC repair WITH VP shunt insertion (Afterwards/2 sessions)

All patients without CSF diversion will be monitored in the outpatient clinic for clinical signs of active HCP. Clinical evaluation will be associated with radiological investigations through non-contrast CT scans of the brain and/or MRI brain at the stage the patient will likely need a VP shunt.

Several clinical parameters shall be used to predict active HCP hence the need for VP shunt as: Increased head circumference (HC), increased rate of increase of HC per day, tense anterior fontanel, wound leak, developmental milestones decline, repeated vomiting, or seizures.

This will be monitored during the post-operative stay in the hospital and during the follow-up visits. The follow-up outpatient clinic visits will be conducted within the first 6 months after MMC repair. They should be weekly in the 1<sup>st</sup> month, twice a month for the next 2 months and monthly during the following 3 months.

Several pre-operative (before MMC repair and before shunt surgery) radiological linear indices and ratios have been previously used in several studies to follow-up the resolution of HCP after VP shunt insertion in cases of obstructive HCP and it has been proved it was significantly important to predict the group of patients who will need shunt revision due to shunt malfunction.

From those parameters and ratios, we used Evan's index which is the ratio of maximum width of frontal horns of the lateral ventricles and the maximal internal diameter of the skull at the same level employed in axial CT and MRI images. It is measured at presentation and at follow-up either just before shunt insertion (if needed) or at a 6-month follow-up visit. Their statistical significance in predicting the need for VP shunt in those patients will be evaluated.

### Inclusion criteria

The following criteria were included in the study:

1. Patients newly diagnosed with MMC without limitations regarding the age.
2. Patients fit for surgical intervention.

### Exclusion criteria

The following criteria were excluded from the study:

1. Patients presented with ruptured MMC.
2. Patients with MMC diagnosed with meningitis and/or sepsis prior any surgical intervention.

### Outcomes/objectives

1. Identification of clinical and radiological predictors of shunt insertion HCP in MMC patients.
2. Reducing the incidence of shunt placement in MMC hydrocephalic patients as much as possible except for those who desperately need it to avoid its socio-economic burden and related complications.
3. Identification of a follow-up and surveillance protocol for those who will undergo MMC repair without CSF diversion to avoid non-treated HCP related complications such as psychomotor delay and decreased IQ.

**Primary outcomes (most important outcomes to be assessed)**

1. Incidence of shunt placement in cases with MMC.
2. Clinical predictors of shunt placement in MMC-related HCP and their significance.
3. Radiological predictors of shunt placement in MMC-related HCP and their significance.

**Secondary outcome (other outcomes to be assessed)**

1. Percentage of patients not requiring CSF diversion.
2. Time of shunt insertion in the follow-up period
3. Correlation between HCP and the presence of hindbrain anomalies.
4. Correlation between the level of MMC and the development of HCP.
5. Possible complications that could be encountered during various management plans.

**Possible risk**

No additional risks could be addressed other than the ordinary known risks of surgical intervention (e.g., wound infection, CSF leakage, wound dehiscence, and shunt malfunction).

**Statistical methods**

Data were coded and entered using the Statistical Package for the Social Sciences version 26 (IBM Corp., Armonk, NY, USA). Data were summarized using mean, standard deviation, median, minimum and maximum in quantitative data and using frequency (count) and relative frequency (percentage) for categorical data. Comparisons between quantitative variables were done using the non-parametric Mann-Whitney test. For comparing categorical data, Chi-square ( $\chi^2$ ) test was performed. Exact test was used instead when the expected frequency is  $<5$ .

Receiver operating characteristic (ROC) curve was constructed with area under curve analysis performed to detect best cutoff value of significant parameters for detection of failure.  $P < 0.05$  was considered as statistically significant.

**Results**

It is to be noted that the group of concern in our study is Group B because its patients are susceptible for development of latent hydrocephalus and the need

of VP shunt. Thus, a variety of epidemiological, clinical and radiological and numerical variables (predictors) are studied and their significance in predicting the occurrence of latent hydrocephalus and the need for VP shunt is identified and proved statistically. These variables are summarized in Figure 1.

**Epidemiological variables**

Epidemiological variables include gender, consanguinity, antenatal care, folic acid supplementations, and time of discovery of MMC. The statistical comparison between these variables in Group B1 and Group B2 is illustrated in Table 1. All of these variables are statistically insignificant in predicting the occurrence of latent HCP in MMC patients after surgical repair.

**Clinical and radiological variables**

Clinical variables in Group B1 and Group B2 are illustrated and compared in Table 2 and in Figures 4-9. State of the anterior fontanel, CSF leakage, neurological deterioration, and severity of hindbrain anomalies are the variables of statistical significance in predicting the occurrence of latent HCP in MMC patients after surgical repair.

**Table 1: Comparison between Group B1 and Group B2 regarding the epidemiological variables**

Epidemiological variables	Group B1 Only repair (n=51)		Group B2 Repair then shunt (n=27)		p value
	Count	%	Count	%	
Gender					
M	22	43.1	11	40.7	0.838
F	29	56.9	16	59.3	
Consanguinity					0.077
Yes	16	31.4	14	51.9	
No	35	68.6	13	48.1	
Antenatal care					0.287
Yes	38	74.5	17	63.0	
No	13	25.5	10	37.0	
Folic acid supplements					0.201
Yes	34	66.7	14	51.9	
No	17	33.3	13	48.1	
Time of discovery					0.121
Delivery	19	37.3	16	59.3	
At 2nd T	17	33.3	4	14.8	
At 3rd T	15	29.4	7	25.9	

Ghoul Statistically significant, Ghoul Statistically insignificant,

The statistical analysis of the data collected in this study group failed to say which of these significant variables is stronger in predicting the occurrence of latent HCP. The statistician stated that it might be applicable if we had a larger sample size of the study population and if the study was conducted over a more prolonged period of time.

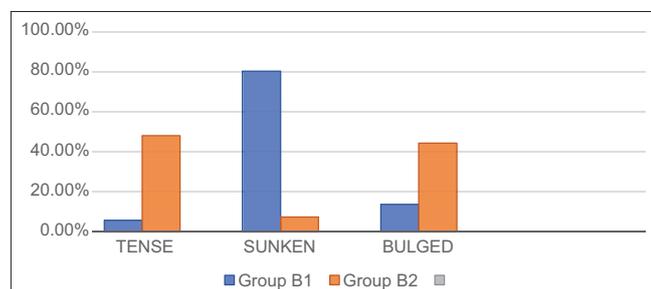
**Numerical variables**

There are multiple variables used to predict the occurrence of latent HCP in this group of patients. Table 3 illustrates the comparison between Group B1 and Group B2 regarding those variables.

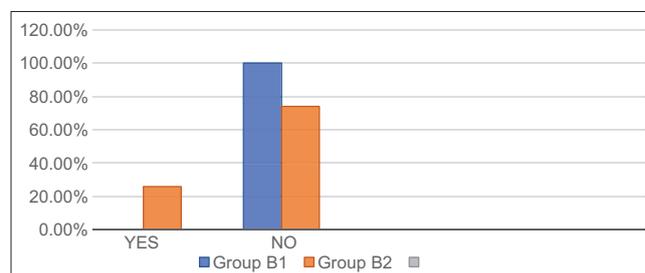
**Table 3: Comparison between Group B1 and Group B2 regarding the different numerical variables used in the study**

	GROUP B1 (repair only)					GROUP B2 (repair followed by shunt)					p value
	Mean	SD	Med	Min	Max	Mean	SD	Med	Min	Max	
1 <sup>st</sup> HC <sup>a</sup>	37.86	1.48	37.00	36.00	42.00	39.70	1.92	40.00	37.00	42.00	<0.001
Follow-up HC <sup>b</sup>	44.25	0.80	44.00	43.00	46.00	43.15	1.90	43.00	40.00	46.00	0.004
RATE +HC/d <sup>c</sup>	0.035	0.005	0.039	0.022	0.039	0.400	0.277	0.330	0.050	1.000	<0.001
1 <sup>st</sup> Evans Index EI <sup>d</sup>	0.30	0.02	0.31	0.28	0.34	0.33	0.03	0.32	0.28	0.41	<0.001
Follow-up EI <sup>e</sup>	0.31	0.01	0.31	0.30	0.34	0.36	0.05	0.33	0.29	0.45	<0.001

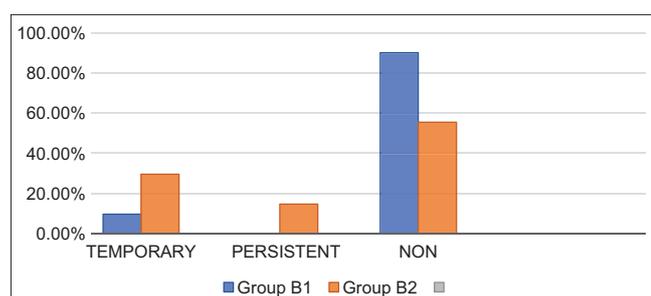
HC: Head circumference, <sup>a</sup>1<sup>st</sup> HC: Head circumference measured at first presentation, <sup>b</sup>Follow-up HC: Head circumference measured either after 6 months of presentation (Group B1) or just before shunt placement (Group B2), <sup>c</sup>Rate + HC/d: Rate of increase in head circumference per day either for non-shunted patients (Group B1) or during the duration prior to shunt insertion for shunted patients (group B2), <sup>d</sup>1<sup>st</sup> EI: Evan's index measured at first presentation, <sup>e</sup>Follow-up EI: Evan's index measured either after 6 months of presentation (Group B1) or just before shunt placement (group B2).



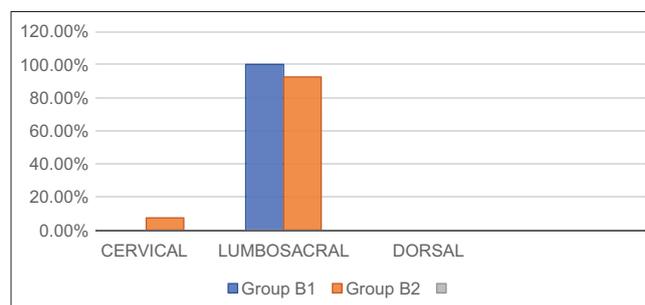
**Figure 4: Comparison between Group B1 and Group B2 patients regarding the state of anterior fontanel (n=78)**



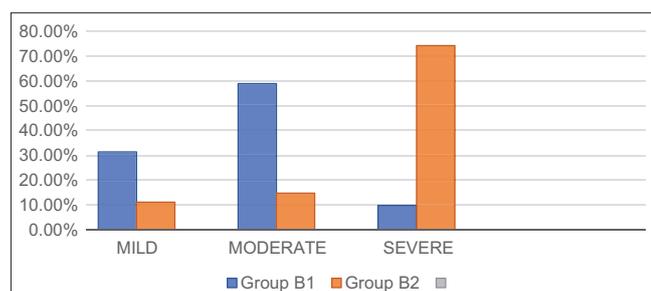
**Figure 8: Comparison between Group B1 and Group B2 patients regarding neurological deterioration (n=78)**



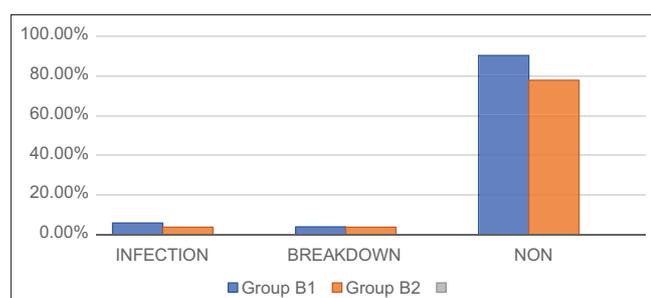
**Figure 5: Comparison between Group B1 and Group B2 patients regarding the CSF leakage (n=78)**



**Figure 9: Comparison between Group B1 and Group B2 patients regarding the anatomical level of MMC (n=78)**



**Figure 6: Comparison between Group B1 and Group B2 patients regarding the severity of hindbrain anomalies (n=78)**



**Figure 7: Comparison between Group B1 and Group B2 patients regarding the complications of surgery (n=78)**

**Primary outcome assessment**

*Incidence of HCP*

Shunt placement was required in 45 (46.88%) of the 96 patients. Eighteen patients (18.75%) needed the shunt as soon as they presented to us (GROUP A), because they were having clinically active HCP. Twenty-seven (28.13%) patients were operated on by MMC repair initially without shunt placement because they did not have signs of increased intracranial pressure at the time of presentation. Yet, they developed latent HCP requiring shunt placement during the follow-up period and the diagnosis was supported by clinical and radiological evidence (GROUP B2). Fifty-one patients of the study populations (53.13%) underwent surgical repair of the MMC without the need of further VP insertion and they were followed up for 6 months period after the repair without developing latent HCP (GROUP B1).

Patients of Group B were the study population susceptible for the development of latent HCP. Out of 78 patients in Group B, only 27 patients (34.62%) needed a VP shunt.

Compared to the study published in 2008 by Dr. Aabir Chakraborty and his fellows where the

incidence of shunt insertion was 51.9%, the incidence of shunt insertion in our study is lower, Tables 4.

**Table 4: Comparison between our study and Chakraborty, et al. 2008 and Tulipan, et al. 2015 studies regarding the development of MMC-related hydrocephalus requiring a VP shunt**

	Our study	Chakraborty, et al. [1]	Tulipan, et al. [11]
Year	2020	2008	2015
Type of study	Prospective	Retrospective	Randomized controlled
Time of repair	Postnatal	Postnatal	Prenatal
Number of patients	96	54	91
Shunted patients	46.88%	51.9%	44%
Predictors	<ul style="list-style-type: none"> <li>· HC</li> <li>· Rate + HC/d</li> <li>· Fontanel</li> <li>· CSF Leak</li> <li>· Evan's Index</li> <li>· Follow Up EI</li> <li>· Hindbrain Anomalies</li> </ul>	<ul style="list-style-type: none"> <li>· HC</li> <li>· Ventriculomegaly</li> <li>· Pseudomeningocele</li> <li>· CSF Leak</li> <li>· IQ Tests</li> </ul>	<ul style="list-style-type: none"> <li>· Bulging fontanel</li> <li>· OFC<sup>a</sup></li> <li>· Split sutures</li> <li>· Syringomyelia</li> <li>· Severe CM-II signs</li> <li>· Persistent leak</li> </ul>
Latent HCP	27/78 (34.6%)	-	-
Mortality	One (unrelated RTA)	1 (sepsis)	2 (extreme prematurity)

<sup>a</sup>OFC: Occipito-frontal circumference, MMC: Myelomeningocele, VP: Ventriculo-peritoneal, HCP: Hydrocephalus, HC: Head circumference, CSF: Cerebrospinal fluid.

### Clinical predictors

Throughout the early post-operative period and the follow-up in outpatient clinic visits in which patients of Group B were monitored for development of latent HCP, several clinical variables were used to assess patient's status. These variables included initial HC at the first presentation just prior to repair surgery, follow-up HC (either after 6 months of follow-up in Group B1 patients or just before shunt placement in Group B2 patients), anterior fontanel state, CSF leakage from the repair wound (Group B2 patients), and neurological deterioration attributed to the severity of the CM-II.

State of the anterior fontanel was a strong indicator of the need of VP shunt ( $p < 0.001$ ). About 48.1% of the patients that had a shunt later on, initially had a tense anterior fontanel and 44.4% of them bulged fontanel. Whereas, the non-shunted patients exhibited 80.4% of patients with flat anterior fontanels at presentation and 5.9%, 13.7% of patient's tense and bulged fontanels, respectively.

CSF leakage from the repair wound was also a strong clinical predictor for the need of shunt due to latent HCP. Almost more than 90% of the non-shunted patients did not experience CSF leak postoperatively neither early nor late. On the other hand, more than 44% of the shunted patients did experience CSF leak in the post-operative period either temporarily (stopped by local measures as repeated dressing and suturing) or persistently (only stopped after shunting).

It has to be noted that from the shunted patients in this group (27 patients), seven patients (25.9%) have experienced clinical manifestations of neurological decline due to severe CM-II and HCP as respiratory problems and bradycardia. That was statistically significant denoting it is a powerful predictor

of the need of VP shunt in those patients. Non-shunted patients have never experienced such clinical signs throughout the post-operative period.

HC is a valuable clinical tool in assessing progressive head enlargement in pediatric population generally especially in certain instances like management of MMC-related latent HCP. It is measured several times throughout the management journey of each patient. The most important measurement of HC is the initial one at presentation just before surgical repair of the MMC and either after 6 months of follow-up in Group B1 patients or just before shunt placement in Group B2 patients. In our study, we abbreviated for those parameters as follows (1<sup>st</sup> HC, FU-HC, and Rate + HC/d). All three of them were strong predictors for the need of VP shunt.

### Radiological predictors

Almost all of MMC patients have their condition as a part of CM-II. Several hindbrain anomalies could be encountered in these patients. Some of the hindbrain anomalies in cases of MMC include; (1) tonsillar and vermian herniation, (2) stretched and low-lying, (3) peaking of the quadrigeminal plate, (4) kinked cervico-medullary junction, (5) elongated aqueduct and fourth ventricle, (6) concave clivus and widened pre-pontine space, and (7) abnormally lying torcular herophili.

We noticed that not all of the previously mentioned anomalies are present in all patients. They do exist in variable numbers and frequencies. We have used the terms (mild, moderate, and severe) to denote the presence of these anomalies in each patient individually and to describe the degree of severity.

- Mild cases include < 2 hindbrain anomalies
- Moderate cases include those who have three or four anomalies
- Severe cases have 5 or more anomalies.

Severity of hind brain anomalies was a strong radiological predictor for the need of shunt in these patients ( $p < 0.001$ ). It has been noted that in the shunted patients (Group B2) almost 74.1% of patients have severe hindbrain anomalies in comparison with the percentage in the non-shunted patients (Group B1) which was 9.8%. This means more severe the malformation, the more likely to develop latent HCP the more likely the need for shunt placement.

Evan's index is a radiological parameter used to describe the extent of ventriculomegaly in patients with HCP. With normal individuals, Evan's index should be <0.3. If it is more than this it indicates ventriculomegaly. We used this radiological variable to denote the extent of ventriculomegaly in MMC patients. Most of those patients have large ventricles, but on the contrary not all of them will need a VP shunt eventually to treat ventriculomegaly. Hence, this raised attention to the fact that ventriculomegaly associated with MMC cases

might not be true HCP. Hence, Evan's index could be used to describe in a numerical manner the cutoff limit after which we could deal with the ventriculomegaly as true HCP.

Evans's index is measured initially at presentation just before surgical repair of the MMC. Another measurement of Evans index (called follow up Evans's index) either after 6 months of repair surgery (Group B1) or just before shunt placement (group B2).

In our study, we abbreviated for those parameters as follows (1<sup>st</sup> EI, FU-EI). Both of them were strong predictors for the need of VP shunt ( $p < 0.001$ ).

### Secondary outcome assessment

Out of 96 patients included in the study, 51 patients (53.13%) did not need a VP shunt after MMC repair without developing latent HCP. Those patients were monitored after the surgical repair for a 6-month follow-up period for clinical and radiological findings of HCP. They developed throughout this period of time in a normal manner without significant motor delay (that can be attributed to HCP) with a range of ventriculomegaly on their serial imaging accepted for age.

### Time of shunt insertion

For the group of patients (27 patients) who developed latent HCP (**Group B2**), 13 patients have developed HCP in the 1<sup>st</sup> week following MMC repair. Seven patients required the shunt in the 2<sup>nd</sup> week after MMC repair. Only 4 days required the shunt in the 3<sup>rd</sup> and 4<sup>th</sup> week after repair. There were three patients who required shunt insertion in the 3<sup>rd</sup> month after MMC repair. Hence, it is valuable to conclude that it is more likely for the patient who does the surgical repair with higher susceptibility to develop latent HCP to develop it with subsequent need of a shunt during the 1<sup>st</sup> week after surgery.

### Anatomical level of MMC

From the 96 patients included in the study, only two patients had cervical MMC and both patients (100%) developed latent HCP and required a VP shunt. Although this result is statistically insignificant to say that the higher anatomical level of MMC is a strong predictor for the need of VP shunt, yet, it is believed in some genuine publications that the higher the level of MMC, the more the need for VP shunt due to latent HCP [1].

### Associations

Fifty-seven of the 96 patients (59.4%) had associated orthopedic congenital deformities

at birth. Five patients (5.2%) showed evidence of urinary tract malformations. Fourteen patients (14.6%) had associated cardiac defects. Six patients (6.3%) had associated congenital infantile inguinal hernia.

### Complications

MMC-related wound complications occurred in eight patients (14.8%) as follows and included infections (five patients), localized wound breakdown (three patients), All of these cases were treated with a combination of antibiotics and and/or wound revision.

### The ROC curve

ROC curve (Figure 10) was constructed with area under curve analysis performed to detect best cutoff value of significant parameters for detection of failure. Failure here means that the patient, who was previously operated on by MMC repair with subsequent close follow-up for the detection of development of latent HCP, has eventually been in need for a ventriculoperitoneal shunt to treat HCP.

Detection to certain numerical values for the previously mentioned clinical and radiological parameters (variables) is essential to know the cutoff point after which further increase in such values carries significant additional risk for the development of true latent HCP thus the need for VP shunt, Table 5.

**Table 5: Interpretation of the ROC**

	Area	p value	95% Confidence interval		Cut-off	Sensitivity %	Specificity %
			Lower bound	Upper bound			
HC	0.663	0.018	0.532	0.794	39.5	55.6	62.7
Rate +HC/d	1.000	<0.001	1.000	1.000	0.0445	100	100
1 <sup>st</sup> Evans Index	0.747	<0.001	0.629	0.866	0.315	55.6	72.5
Follow up EI	0.771	<0.001	0.650	0.892	0.3250	59.3	82.4

ROC: Receiver operating characteristic, HC: Head circumference.

### Risk stratification for MMC-related latent HCP

As mentioned before in the statistical analysis of the results, patients who underwent initially MMC repair (Group B) in their follow-up period into the following two main groups; (a) high risk post-MMC repair group and (b) standard risk post MMC repair group. Criteria of each group are shown in Table 6.

**Table 6: Risk stratification of post MMC repair patients into high and standard risk groups**

High-risk post MMC repair group	Standard-risk post MMC repair group
<ul style="list-style-type: none"> <li>• Initial HC<math>\geq</math>39.5 cm</li> <li>• Rate of increase in HC per day is <math>\geq</math>0.0445 cm/day</li> <li>• Initial EI measured at presentation <math>\geq</math>0.315</li> <li>• Follow-up EI<math>\geq</math>0.3250               <ul style="list-style-type: none"> <li>· Fontanel: Tense</li> <li>· Pseudomeningocele: Moderate to Significant</li> <li>· Leak: Persistent</li> <li>· Neurological Decline: Present</li> <li>· Hindbrain anomalies: Moderate/Severe</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li>• Initial HC<math>&lt;</math>39.5 cm</li> <li>• Rate of increase in HC per day is <math>&lt;</math>0.0445 cm/day</li> <li>• Initial EI measured at presentation <math>&lt;</math>0.315</li> <li>• Follow up EI<math>&lt;</math>0.3250               <ul style="list-style-type: none"> <li>· Fontanel: Sunken, Bulged</li> <li>· Pseudomeningocele: Minimal</li> <li>· Leak: Temporary, or non-existing</li> <li>· Neurological Decline: Absent</li> <li>· Hindbrain anomalies: Mild</li> </ul> </li> </ul>

MMC: Myelomeningocele.

Extra measures shall be taken and implemented to ensure that the high-risk group patients receive the appropriate care and follow-up at the exact time they need it. This also shall be aiming at not missing a patient with latent HCP without a VP shunt he is badly needing.

## Discussion

There is a cliché in neurosurgical practice that “the ideal shunt is no shunt.” The shunt placement rate in patients with MMC has historically been quite high [1].

Several reasons have raised the attention to why we should try to decrease the incidence of VP shunt placement in MMC patients and keep this only for the patients with active latent HCP after initial surgical repair.

CSF shunting is fraught with many complications, whatever the cause of HCP; however, the rate of shunt obstruction is considered higher in MMC-related HCP than in other causes of HCP. Tuli calculated that the hazard-ratio of shunt revision for MMC-related HCP was higher in comparison with other causes of congenital HCP [12].

Patients with MMC who have had CSF diversion have reduced longevity compared with patients who have not required shunt placement [5], [6].

It is hard to evaluate the impact of HCP on the intellectual development and autonomy of patients with MMC because of the presence of multiple anomalies affecting the CNS in these patients. It is believed that there is greater severity of affection of these intellectual functions when HCP is present [7].

The mean IQ has been reported to be lower in shunted patients with MMC when compared to the non-shunted patients. Studies have shown that the functional outcome in non-shunted MMC patients is better than the shunted patients. Early and efficient treatment is the most important factor in achieving a better functional outcome [4], [7].

The majority of complications CSF shunting occurs early. The rate of complications during the year

following shunting for MMC-related HCP is generally higher than the next years. Among these 1<sup>st</sup>-year complications, Caldarelli *et al.* concluded that 73% were mechanical and 27% were infectious. The 1-year revision rate was higher in MMC-related HCP than in non- MMC-related HCP. Long-term adverse effects of shunt-related complications in this group of shunted patients [8], [12], [13], [14], [18].

It is not uncommon that there might be some increase in ventricular size following MMC closure. This initial increase is not necessarily progressive, however, and ventricular size will commonly stabilize without intervention [9].

Increasing experience with endoscopic third ventriculostomy in the treatment of HCP has shown us that excellent clinical outcomes can occur despite only minimal improvement in ventricular configuration. This leads neurosurgeons to tolerate moderate ventriculomegaly. It has been suggested that most infants who have undergone fetal MMC closure have persistent ventriculomegaly but do not have overt clinical signs or symptoms of increased ICP. Hence, previous criteria for shunt placement based on ventricular indices or cortical mantle thickness alone may be less relevant than the clinical context in which they occur [10].

It has to be taken into consideration that shunt placement in patients with MMC-related HCP is sometimes a lifesaving procedure and therefore at certain circumstances it is mandatory. Caution must be taken while dealing with these patients and the concept of decreasing shunt insertion shall never be at the expense of endangering patient's neurological and developmental state [15], [16], [17].

It would, therefore, seem reasonable to ensure that shunts are only placed in patients who truly need them and can benefit from them. It is obviously essential that we continue to closely monitor this group of children to ensure that this reduction in shunt placement rate does not affect them with respect to neurodevelopmental outcome.

In our study, out of the 78 patients in Group B who underwent surgical repair of the MMC initially on presentation, 27 patients (34.6%) required a VP shunt later on due to development of latent HCP. Decision-making regarding these patients was concluded based on the relevant clinical and radiological data obtained about these patients in the serial outpatient clinic visits in the post-operative period. This was very important to construct a strict protocol that shall ensure the insertion of a VP shunt for the desperately needing patient with true latent HCP rather than having mild to moderate ventriculomegaly without any clinical consequences.

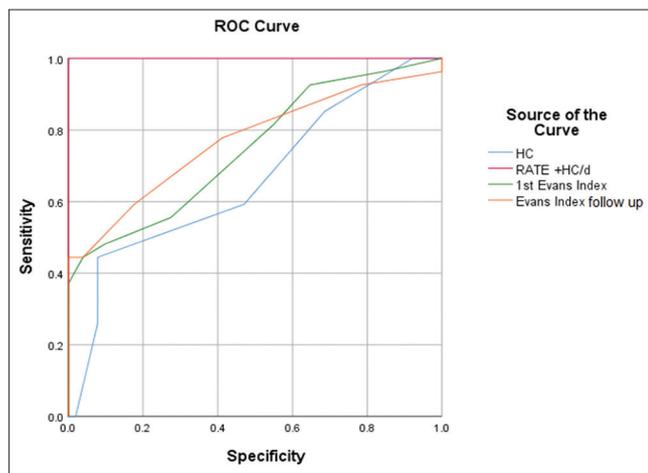


Figure 10: The receiver operating characteristic curve

### Management protocol

The following protocol (Figure 11) was applied in the Neurosurgery Unit in Aboul Reish University Hospital from January 2019 to September 2020.

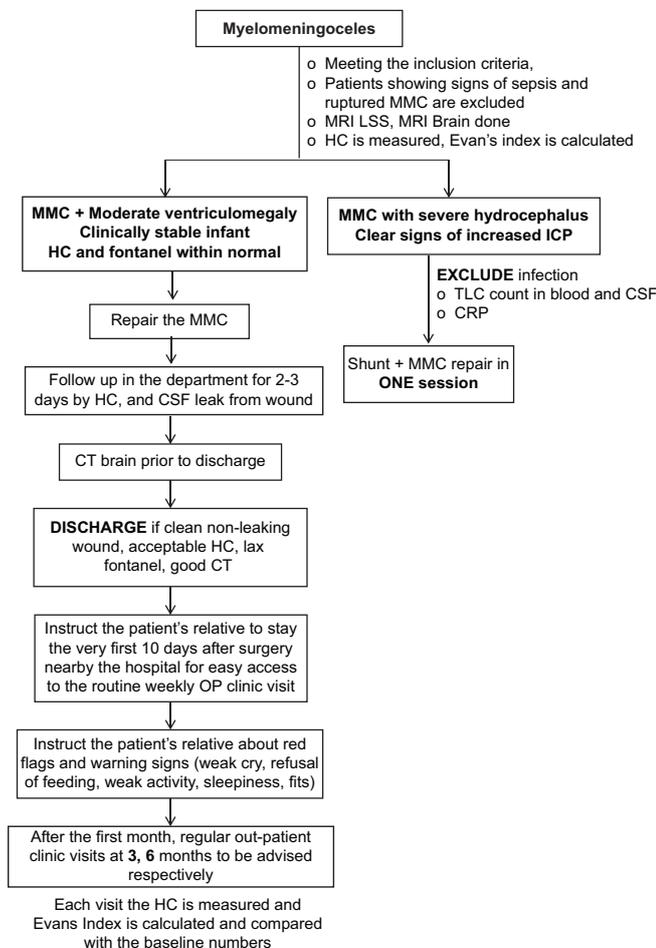


Figure 11: Diagram illustrating the algorithm used in management of patients with MMC

The aim was to conduct proper treatment of MMC patients taking in consideration that a certain group of these patients could develop latent HCP with the need for VP shunt placement. Shunt placement was exclusively

spared for those who desperately need it and this led to decreased overall shunt insertion rate in comparison with the last years.

The following considerations were adopted and followed strictly in management of all MMC patients:

- All patients presented postnatally shall undergo detailed neurological exam and investigations with a special focus to exclude other associated congenital anomalies that might need certain anesthetic considerations.
- MR lumbosacral spine is requested for all patients together with CT brain and MRI brain +/- craniocervical junction.
- All patients shall have laboratory work up to exclude CSF infection or sepsis. This is achieved by withdrawing a blood sample and requesting a complete blood count with total and differential leukocytic count, ESR, and CRP. When indicated, a trans-fontanel tapping of CSF is done and CSF parameters are studied to exclude CSF infection.
- Meticulous surgical technique of MMC repair with water-tight dural closure is mandatory.
- Post-operative follow-up is essential in the neurosurgery unit inpatient ward for 3 days.
- In the early post-operative hospital stay, follow-up shall focus on the cardinal manifestations of increased ICP and on CSF leak from the wound.
- If there is leakage, we did not rush to shunt placement, we closely monitor the patient, local wound measures may be sufficient for the leak to stop.
- These measures include; application of additional stitches, repeated dressing, frequent aspiration of pseudomeningocele, and wound revision in the operating theatre. Care must be taken not to introduce infection and all procedures must be done under complete aseptic conditions.
- If leak persists, assess the whole status of the patient with other clinical signs including increased HC or clinical decline and radiologically evident ventriculomegaly. If all variables are favoring the presence of latent HCP, shunt insertion shall be adopted. Decision of shunt insertion shall be approved of two senior consultants of pediatric neurosurgery.
- If the patient did not develop latent HCP in the early post-operative period, discharge the patient home and give his/her parents the following instructions:
  - It is better to stay nearby the hospital to facilitate reaching the medical facility in case of emergency. This is applied especially for the families living in distant areas. If it is not practical for the family, talk to hospital administration to facilitate and provide

logistics to avoid such a problem.

- Outpatient clinic visits will be weekly in the 1<sup>st</sup> month of follow-up, every 2 weeks in the 2<sup>nd</sup> and 3<sup>rd</sup> months, and every month starting from the fourth, till the 6<sup>th</sup> month.
- In OP clinic visits; examine the patient, HC to be measured, assess the fontanel, neurodevelopmental assessment to be done, history taken from the parent(s) regarding any warning signs (irritability, poor feeding, delayed milestones, vomiting, or seizures). If you are suspicious about the infant condition, assess ventriculomegaly through asking for a non-contrast CT brain and compare it with the previous studies.
- Decision-making of shunt placement in the follow-up period shall be based on clinical and radiological parameters and it is to be taken by two senior consultants of pediatric neurosurgery.
- It is said that Group B patients are the ones susceptible to develop latent HCP, so they are the target group of follow-up. Stratification of patients in this group according to being at high risk or at a standard risk to develop latent HCP is advisable (Table 6). It is more logic to exert more efforts to follow-up patients with high risk for developing latent HCP by certain extra-measures.
- These extra-measures might include:
  - More frequent follow-up visits (weekly in the first 2 months, and half-monthly afterward for the remaining of 6 months).
  - Reach out team to visit patient at home to do a quick check (done by house officers and for nearby families).
  - Surveillance call center to check for patient's quality of life after the repair procedure and to make sure that the infant is doing well without any complications.

## Conclusion

This study has demonstrated that the rate of shunt insertion in patients with MMC is lower than the previously reported rate in the literature and comparable, but still higher than, to the incidence reported following in utero repair [1], [11]. A more thorough evaluation of the patient's post-operative need for a shunt is mandatory with the aid of clinical and radiological particular signs. This has led us to implementing a strict policy for shunt insertion in those MMC patients, taking in consideration their ongoing neurological development. This policy has been achieved by the genuine attitude of the neurosurgeons to accept ventriculomegaly provided it does not mean any deterioration in the patient's clinical or developmental state.

We assume that reduction of shunt insertion rate will eventually reduce what has previously been an enormous burden for a significant proportion of children with MMC. Studying such emerging attitude shall be continued and measuring its impact on the clinical and economic basis could be an interesting target for the upcoming future studies.

## References

1. Chakraborty A, Crimmins D, Hayward R, Thompson D. Toward reducing shunt placement rates in patients with myelomeningocele. *J Neurosurg Pediatr.* 2008;1(5):361-5. <https://doi.org/10.3171/ped/2008/1/5/361> PMID:18447669
2. Tulipan N, Sutton LN, Bruner JP, Cohen BM, Johnson M, Adzick NS. The effect of intrauterine myelomeningocele repair on the incidence of shunt-dependent hydrocephalus. *Pediatr Neurosurg.* 2003;38(1):27-33. <https://doi.org/10.1159/000067560> PMID:12476024
3. Dennis M, Landry SH, Barnes M, Fletcher JM. A model of neurocognitive function in spina bifida over the life span. *J Int Neuropsychol Soc.* 2006;12:285-96. <https://doi.org/10.1017/s1355617706060371> PMID:16573862
4. Mapstone TB, Rekate HL, Nulsen FE, Dixon MS Jr., Glaser N, Jaffe J. Relationship of CSF shunting and IQ in children with myelomeningocele: A retrospective analysis. *Pediatr Neurosurg.* 1984;11(2):112-8. <https://doi.org/10.1159/000120166> PMID:6723425
5. Davis BE, Daley CM, Shurtleff DB, Duguay S, Seidel K, Loeser JD, et al. Long-term survival of individuals with myelomeningocele. *Pediatr Neurosurg.* 2005;41(4):186-91. <https://doi.org/10.1159/000086559> PMID:16088253
6. Tuli S, Tuli J, Drake J, Spears J. Predictors of death in pediatric patients requiring cerebrospinal fluid shunts. *J Neurosurg Pediatr.* 2004;100(5):442-6. <https://doi.org/10.3171/ped.2004.100.5.0442> PMID:15287452
7. Hunt GM, Oakeshott P, Kerry S. Link between the CSF shunt and achievement in adults with spina bifida. *J Neurol Neurosurg Psychiatry.* 1999;67(5):591-5. <https://doi.org/10.1136/jnnp.67.5.591> PMID:10519863
8. Iskandar BJ, Tubbs S, Mapstone TB, Grabb PA, Bartolucci AA, Jerry Oakes W. Death in shunted hydrocephalic children in the 1990s. *Pediatr Neurosurg.* 1998;28(4):173-6. <https://doi.org/10.1159/000028644> PMID:9732242
9. Sankhla S, Khan GM. Reducing CSF shunt placement in patients with spinal myelomeningocele. *J Pediatr Neurosci.* 2009;4(1):2-9. <https://doi.org/10.4103/1817-1745.49098> PMID:21887167
10. Sutton LN. Fetal surgery for neural tube defects. *Best Pract Res Clin Obstetr Gynaecol.* 2008;22(1):175-88. <https://doi.org/10.1016/j.bpobgyn.2007.07.004> PMID:17714997
11. Tulipan N, Wellons JC III, Thom EA, Gupta N, Sutton LN, Burrows PK, et al. Prenatal surgery for myelomeningocele and the

- need for cerebrospinal fluid shunt placement. *J Neurosurg Pediatr.* 2015;16(6):613-20. <https://doi.org/10.3171/2015.7.peds15336>  
PMid:26369371
12. Steinbok P, Irvine B, Douglas Cochrane D, Irwin BJ. Long-term outcome and complications of children born with meningomyelocele. *Childs Nerv Syst.* 1992;8(2):92-6. <https://doi.org/10.1007/bf00298448>  
PMid:1591753
13. Bowman RM, McLone DG, Grant JA, Tomita T, Ito JA. Spina bifida outcome: A 25-year prospective. *Pediatr Neurosurg.* 2001;34(3):114-20. <https://doi.org/10.1159/000056005>  
PMid:11359098
14. Caldarelli M, Di Rocco C, La Marca F. Shunt complications in the first postoperative year in children with meningomyelocele. *Childs Nerv Syst.* 1996;12(12):748-54. <https://doi.org/10.1007/bf00261592>  
PMid:9118142
15. Tamburrini G, Frassanito P, Iakovaki K, Pignotti F, Rendeli C, Murolo D, *et al.* Myelomeningocele: The management of the associated hydrocephalus. *Childs Nerv Syst.* 2013;29(9):1569-79. <https://doi.org/10.1007/s00381-013-2179-4>  
PMid:24013327
16. Phillips BC, Gelsomino M, Pownall AL, Ocal E, Spencer HJ, O'Brien MS, *et al.* Predictors of the need for cerebrospinal fluid diversion in patients with myelomeningocele. *J Neurosurg Pediatr.* 2014;14(2):167-72. <https://doi.org/10.3171/2014.4.peds13470>  
PMid:24877604
17. Norkett W, McLone DG, Bowman R. Current management strategies of hydrocephalus in the child with open spina bifida. *Top Spinal Cord Inj Rehabil.* 2016;22(4):241-6. <https://doi.org/10.1310/sci2204-241>  
PMid:29339864
18. Khan B, Hamayun S, Haqqani U, Khanzada K, Ullah S, Khattak R, *et al.* Early complications of ventriculoperitoneal shunt in pediatric patients with hydrocephalus. *Cureus.* 2021;13(2):e13506. <https://doi.org/10.7759/cureus.13506>  
PMid:33786215