



بسم الله الرحمن الرحيم

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A Systematic Review for Simultaneous versus Delayed Ventriculo-Peritoneal Shunting in Surgical Repair of Meningomyelocele Sac in Patients with Chiari Type II Malformation

A Systematic Review

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بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

قالوا

لَسْبَدَانِكَ لَا نَعْلَمُ لَنَا
إِلَّا مَا عَلَّمْتَنَا إِنَّكَ أَنْتَ
الْعَلِيمُ الْعَظِيمُ

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List of Contents

Title	Page No.
List of Tables.....	i
List of Figures.....	ii
List of Abbreviations	v
Introduction	1
Aim of the Work.....	5
Review of Literature	
Chapter 1: Neuroanatomy.....	6
Chapter 2: Neuro-embryology.....	15
Chapter 3: Pathophysiology	25
Chapter 4: Clinical Presentation	33
Chapter 5: Investigations.....	40
Chapter 6: Treatment.....	55
Patients and Methods.....	69
Results.....	78
Discussion	84
Summary and Conclusion	90
References	91
Arabic Summary	

List of Tables

Table No.	Title	Page No.
Table 1:	Summary of inclusion and exclusion criteria used in this review.....	70
Table 2:	Study Characteristics:.....	76
Table 3:	Clinical findings (Shunt infection in Simultaneous shunting vs Delayed shunting) of the study population post operative follow up	78
Table 4:	Clinical findings (Shunt Revision in Simultaneous vs Delayed Shunting) of the study population post operative follow up	79
Table 5:	Clinical findings (Wound infection (meningeal sac) in Simultaneous shunting vs Delayed shunting) of the study population post operative follow up	80
Table 6:	Clinical findings (Time of VP shunt insertion related to age of patient and associated percentage of shunt infection).....	81

List of Figures

Fig. No.	Title	Page No.
Fig. 1:	Coronal posterior view showing brain stem and upper cervical segments and their relations in posterior fossa.....	7
Fig. 2:	Inferior view showing dissected brainstem and its topography.	8
Fig. 3:	Sagittal view of dissected brain stem specimen and its connections.	8
Fig. 4:	Axial, sagittal, & coronal sections of ventricular system.	14
Fig. 5:	Multiple stages of primary neurulation by electron microscope.....	16
Fig. 6:	Steps of 2ry neurulation.....	17
Fig. 7:	Illustration for steps of formation of neural tube and neural crest.....	19
Fig. 8:	The role of morphogenic proteins in development of spinal cord.....	21
Fig. 9:	Development of Mesencephalon & Prosencephalon.....	23
Fig. 10:	Development of early ventricular system.....	24
Fig. 11:	Different types of neural tube defects.....	26
Fig. 12:	A. typical myelomeningocele with placode B. typical myeloschisis with no underlying CSF sac.	29
Fig. 13:	Epidemiological distribution of cases of MMC in middle east.....	33
Fig. 14:	Ultrasound sagittal view shows a large massa intermediate in long arrow, tectal beaking in short arrow.	42

List of Figures cont...

Fig. No.	Title	Page No.
Fig. 15:	Ultrasound sagittal view of the thoracic spinal cord (arrowheads) and myelomeningocele (arrows). Note the tethering of the placode at the site of the dysraphic defect.	44
Fig. 16:	Sagittal T1 cuts reveling extension of vermis till level of C4.....	45
Fig. 17:	MRI Axial T2 showing culpocephaly resulted from corpus callosum agenesis, the white arrows show heterotopias.	47
Fig. 18:	Sagittal T1 MRI show low insertion of the tentorium & polygyria.	48
Fig. 19:	Sagittal T1 MRI showing medullary kink at level of C3-4.	48
Fig. 20:	Axial T2 MRI showing enlarged massa intermedia.....	49
Fig. 21:	Sagittal MRI T1 showing large syrinx.....	51
Fig. 22:	Axial CT 3D reconstruction of inferior surface of skull showing the abnormal rotation of C1 (long arrow) related to C2 (short arrow) associated with Chiari II Malfunction.	53
Fig. 23:	Coronal CT 3D reconstruction of base of skull showing post operative decompression at foramen magnum, with removal of posterior arches of atlas.	54
Fig. 24:	Different steps of MMC closure. A) separation of placode from pathological membranes, B) separation of pial layer and recanalization of the placode C) approximation and closure by lumbar fascia.	59
Fig. 25:	Huge defect closed primarily (A) and with mesh (B).	61

List of Figures cont...

Fig. No.	Title	Page No.
Fig. 26:	The proposed positioning for simultaneous shunting and MMC repair.	64
Fig. 27:	Methodology of Systematic review.....	72
Fig. 28:	Clinical findings (Shunt infection in Simultaneous shunting vs Delayed shunting) of the study population post operative follow up	78
Fig. 29:	Clinical findings (Shunt Revision in Simultaneous vs Delayed Shunting) of the study population post operative follow up	79
Fig. 30:	Clinical findings (Wound infection meningeal sac in Simultaneous shunting vs Delayed shunting) of the study population post operative follow up	80
Fig. 31:	Clinical findings (Time of VP shunt insertion related to age of patient and associated percentage of shunt infection).....	81

List of Abbreviations

Abb.	Full term
AFP.....	Alfa feto protein
BMP.....	Bone morphogenic protien
CM II	Chiari malformation type 2
CNS	Central Nervous System
CSF.....	Cerebrospinal fluid
CT.....	Computed tomography
DT.....	Delayed shunting
ETV	Endoscopic Third Ventriculostomy
EVD	External ventricular drain
HCP	Hydrocephalus
JLS	Jarcho–Levin syndrome
LL	Lower limb
MGS.....	Meckel-Gruber syndrome
MMC.....	Myelomeningocele
MRI.....	Magnetic resonance imaging
NIS	Nationwide inpatient sample
NTD	Neural Tube Defect
SB.....	Spina bifida
SCM.....	Split cord malformation
SD.....	Standard deviation
SHH.....	Sonic Hedgehog Gene
ST	Simultaneous shunting
TCUS.....	Transcranial Ultrasound
UL.....	Upper lower
VPS	Ventriculo-peritoneal shunt

INTRODUCTION

Most of the features which characterize the hydrocephalus associated to myelomeningocele (MMC) were already pointed out in late 1970s of the last century, for example, its high incidence and its adverse prognostic significance in terms of intellectual development and survival as well as its multifactorial and complex pathophysiology.^[74]

It was noticed in fact that only one out of six infants born with MMC presented signs of increased intracranial pressure at birth and that only one out of eight of them had a head circumference (HC) above the 98th percentile.

It was also observed how the hydrocephalus became obvious clinically, eventually in some cases after the spinal defect repair, in a further 65 % of the affected children in early postnatal life with a peak in its recognition at 2–3 weeks of age and how irregular its progression could be subsequently. Consequently, it was emphasized that the HC at birth—in most cases inferior to the 50th percentile—did not have any predictive value for the occurrence of the hydrocephalus as well as for its successive evolution.

Despite the numerous studies aimed at understanding the pathogenesis of the ventricular dilation accompanying MMC, this peculiar type of hydrocephalus remains still relatively obscure. Most of its pathogenetic interpretations appear to have been influenced by the mere consideration of the associated anatomical

abnormalities which could impact on the CSF dynamics rather than be based on objective scientific demonstrations. However, the changed attitude of the neurosurgeon who has become reluctant to insert a CSF shunt apparatus in this particular condition because of the related high number of complications.^[6] In the past time, most researchers suggest that repair of MMC sac in first 24-48 hours decreases risk of infection. The repair after 48 hours of MMC sac causes a significant increase in mortality and morbidity rate.^[75]

In MMC, however, it is not rare that the hydrocephalus may slow down its progression after a transient phase of increased intracranial pressure and reach a spontaneous arrest in a significant percentage of the cases. On the other hand, those surgeons in favor of the simultaneous approach emphasized the relatively common occurrence of CSF leak from the site of the spinal malformation repair.^[76]

There is a general agreement that CSF leak represents a major risk of infection, further advantages were also discussed, namely avoiding a second operation and reducing the duration of the hospitalization.

It is likely that the optimal time for the placement of a CSF shunt device is still far to be established, there are no widely applied criteria for CSF shunting in other patients with MMC with less profound hydrocephalus. Previously reported indicators of the need for a shunt include level of the lesion, clinical signs of

elevated intracranial pressure such as a tense or bulging fontanelle, bradycardia, sunseting eyes, increasing head circumference, and increasing ventricular size. ^[77-79]

In a retrospective study, the incidence of infective complications was particular high in newborns receiving the shunt in the first 2 weeks of age. Furthermore the 1-year revision rate was higher in MMC-related HCP than in non- MMC-related HCP, as well as in infants which underwent a delayed shunt insertion after an excessively long period of watchful waiting in the hope to avoid the shunt operation and the related risk to develop shunt dependency, the comparison between both procedures has been addressed in the literature. ^[79]

Historical Background

The first mention in literature to what's will be called CM II appeared in 1891.^[40] The first published series on unknown syndrome of hindbrain herniations based on autopsy findings by Hans Chiari, Professor of Pathology at the German University of Prague, Chiari then identified four distinct types of these hindbrain herniations including the Type II malformation he found that this type exclusively appears in patients with myelomeningoceles. It was first defined as caudal migration of vermis fourth ventricle & brain stem, later CM II was discovered to affect all the CNS. Two contemporaries of Chiari earlier shared in description of CM II and thus deserve mention. In 1883, Cleland wrote about an infant with myelomeningocele associated

with hind brain abnormalities.^[41] Although Eight years before Chiari's contributions his publishes passed unnoticed. Three years after the initial report by Chiari, Arnold then published on a case again with hindbrain herniation & myelomeningocele.^[42] Although great effort helped to establish a connection, it is obvious that Chiari's contribution was the greatest. Chiari held detailed study understanding the pathology in order, his classification system remains basically unchanged for more than 100 years, thus deserved his name to be associated with the disease. And so, its preferred to use the term of Chiari type II malformation over Arnold-Chiari malformation. The first "Chiari decompression" occurred in 1932 reported by Ben-Sira, et al.^[44]