بسم الله الرحمن الرحيم

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بقسم التوثيق الإلكتروني بمركز الشبكات وتكنولوجيا المعلومات دون أدنى مسئولية عن محتوى هذه الرسالة.

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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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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, sunsetting 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]