Multiple modalities in the management of Meningomyelocele

Thesis

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By

Hesham Atef Abdelkader Elshitany
(M.B.B.Ch.)
Faculty of Medicine, Cairo University

Supervised by

Prof. Dr. Fahim Ali Elbassiouny
Professor of General Surgery
Faculty of Medicine
Cairo University

Prof. Dr. Amr Mahmoud Safwat
Professor of Neurosurgery
Faculty of Medicine
Cairo University

Prof. Dr. Ibrahim Galal Khalifa
Professor of General Surgery
Faculty of Medicine
Cairo University

Faculty of Medicine
Cairo University
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ABBREVIATIONS

VP	Ventriculo-peritoneal
MMC	Meningomyelocele
CSF	Cerebrospinal Fluid
ICP	Intracranial Pressure
CT	Computed Tomography
MRI	Magnetic Resonance Imaging
US	Ultrasonography
POD	Post-ovulatory day
NTD	Neural tube defect
TCS	Tethered cord syndrome
IQ	Intelligence quotient
AFP	Alpha foeto-protein
CNS	Central Nervous System
LD	Latissimuss dorsi

Abstract

The aim of this study is to review the literature and recent publications regarding the management of myelomeningocele.

myelomeningocele. is the most common and complex presentation of neural tube closure defects with wide range of management modalities as regarding the association with Chiari malformation type II and hydrocephalus ,the need for shunt placement and its timing and the ideal method for closure of the defect.

In this study we tried to clarify the best methods and timing to deal with myelomeningocele. and the associated hydrocephalus as well as to reduce the risk of post operative wound breakdown with associated CSF leakage and subsequent serious infection other possible complications and the final outcome.

The study was done prospectively on 20 cases of myelomeningocele ,of whom 6 cases were operated upon by v-p shunt before presenting to us,10 cases were presented with associated hydrocephalus and were managed accordingly and 4 cases were presented without evident hydrocephalus of whom 1 case required shunt placement and the other 3 cases were followed -up and did not require CSF diversion.

Despite the size of the defect of the myelomeningocele ,primary skin closure was efficient in restoration of the skin contour sometimes helped by more lateral dissection or lateral releasing incision with limited role for skin flaps and grafts.

Key words: spina bifida, myelomeningocele, hydrocephalus, pediatric neurosurgery.

INTRODUCTION

INTRODUCTION

Meningomyelocele is the most common and complex presentation of neural tube closure defects **(Gaskill, 2004)**, with an incidence of 1.6 per 1000 live births **(Kazmi et al., 2006)**.

It affects the central nervous system at different and multiple levels, and a number of other collateral conditions accompany the spinal cord malformation including: hydrocephalus, Chiari malformation type II, syringomyelia, cerebral midline anomalies, abnormal corticocerebral development **(Giuseppe Talamonti et al., 2007)**.

It results from a failure of primary neurulation during Days 18 through 27 of human embryogenesis **(Cohen & Robinson, 2004)**. A portion of the neural plate fails to invaginate, and lies on the dorsal surface in continuity with the remainder of the spinal cord.

Meningomyeloceles have been reported at all spinal levels: lumbosacral in 30% of cases, thoracolumbar in 26%, lumbar in 26%, sacral in 10%, thoracic in 5%, and cervical in 3% of cases **(Akar, 1995)**.

Meningomyelocele is so diverse that no single theory can be cited to explain all forms of spinal dysraphism, although a deficiency in maternal dietary folate has been postulated as a cause **(Beeker et al., 2006).**

Hydrocephalus is commonly associated with meningomyelocele and develops in 80 to 90% of patients with this anomaly. Its incidence varies according to lesion location. Other abnormalities associated with meningomyelocele include paresis of the lower extremities with associated gait disturbance or paraplegia, Chiari malformation, vertebral anomalies, genitourinary dysfunction, epilepsy and ophthalmological complications **(Akar, 1995)**.

The goal of surgery in meningomyelocele closure is to preserve neurological function and prevent infection.

Although early surgery has not been shown to improve neurological status, it can significantly decreases the risk of death or disability related to infections **(Brocklehurst et al., 1997)**.

The operation includes repair of the defect in the back microscopically to avoid increasing the neurological deficit, hydrocephalus will be dealt with whether simultaneously, before or after closure of the defect.

Cases with large defects necessitates collaboration of plastic surgery with different modalities regarding the design of skin incision, the technique of repair and closure of the defect.

In this work we are aiming at clarifying the best methods and timing to deal with meningomyelocele and the associated hydrocephalus as well as to reduce the risk of post operative wound breakdown with associated CSF leakage and subsequent serious infection other possible complications and the final outcome. The study will include also a detailed survey of the multiple techniques to deal with this problem. All our results will be compared to that of the literature.

REVIEW OF LITERATURE

NORMAL EARLY HUMAN DEVELOPMENT

During the first four days after fertilization post-ovulatory day (POD) 1 to 4, stages 1 to 3, the human embryo undergoes about five cell divisions to form a mass of approximately 32 cells (the blastocyst), which surrounds a central cavity (the blastocystic cavity). The blastocyst contains an eccentrically located inner cell mass, the embryonic cell proper, and a thinner surrounding ring of cells, the trophoblast. By stage 3 (POD 4), the inner cell mass develops two distinct layers: cells of the dorsal surface, adjacent to the trophoblast, form the epiblast while cells on the ventral surface, adjacent to blastocystic cavity, form the hypoblast **(O'Rahilly and Muller, 1987)**.

By stage 5 (POD 7 to 12), two additional cavities develop. The amniotic cavity appears between the epiblast and the overlying trophoblast cells, and the umbilical vesicle (or yolk sac) appears below the hypoblast. The epiblast is therefore adjacent to the umbilical vesicle. By stage 6 (POD 13), the hypoblast thickens cranially; this portion of the hypoblast is the prochordal plate and is the first morphologic feature of craniocaudal orientation. The prochordal plate eventually will give rise to the cephalic mesenchyme and to portions of the foregut **(O'Rahilly and Muller, 1987)**.

The primitive streak first develops at the caudal end of the blastocyst at stage 6 (POD 13) and elongates cranially over the next three days. It reaches its full length by stage 7 (POD 16), at which time it occupies the midline in the caudal half of the human embryo;

beyond this time, the primitive streak begins to regress - that is, it becomes shorter and moves back toward the caudal pole of the embryo. The primitive streak is contiguous cranially with the primitive knot, or Hensen's node; in the midline of Hensen's node is a small indentation, the primitive pit. Along the length of the primitive streak is located a midline trough, the primitive groove, which is contiguous with a primitive pit. Hensen's node is regarded as the cranial extension of the primitive groove **(O'Rahilly and Muller, 1987)**. During both primitive streak elongation and regression, cells of the epiblast migrate toward the primitive streak and invaginate through the primitive groove.The first cells to ingress are the prospective endodermal cells, which intercalate with the hypoblast and displace the hypoblast cells laterally. The prospective endoderm cells will form the definitive endoderm while the displaced hypoblast cells ultimately will form extraembryonic tissues **(Fontaine and Le Douarin, 1977)**. A short time later, prospective mesodermal cells that have ingressed through the primitive groove; these remaining epiblast cells will form the ectoderm (both neuroectoderm and surface ectoderm). This process, referred to as "gastrulation" transforms the embryo from a two-layered structure containing an epiblast and a hypoblast into a three-layered structure containing ectoderm, mesoderm, and endoderm **(Nicolet, 1971)**.

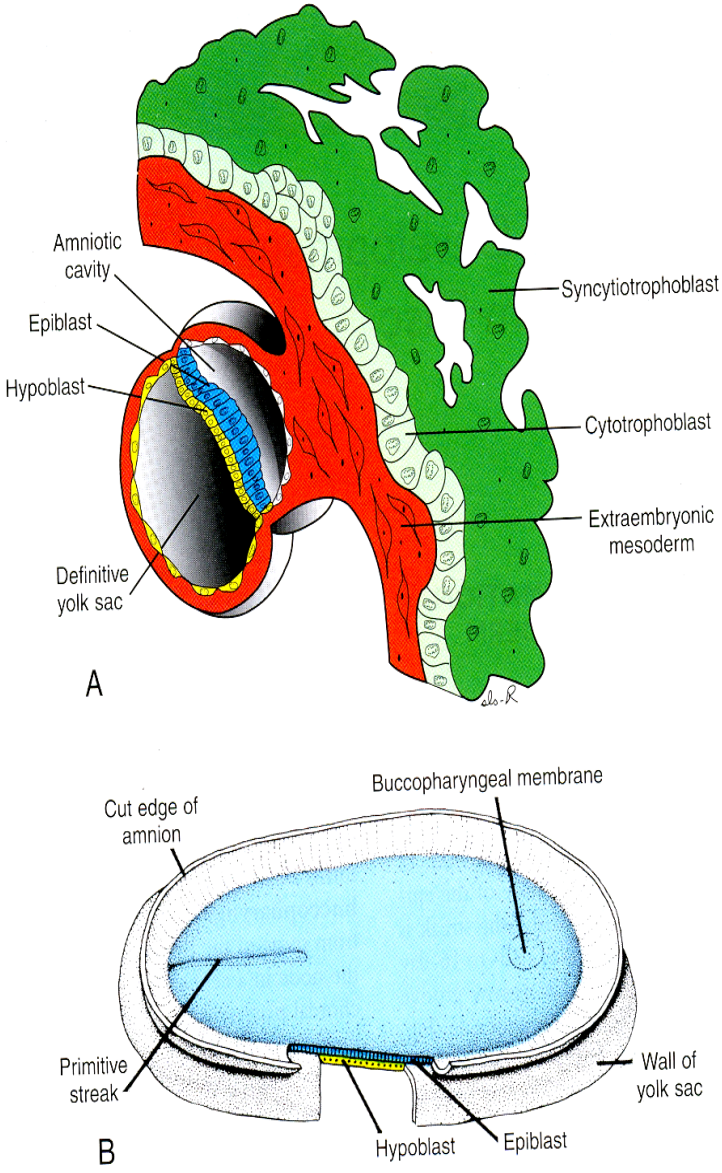

Fig. (1): A. Implantation site at the end of the second week. **B.** Representative view of the germ disc at the end of the second week of development. The amniotic cavity has been opened to permit a view of the dorsal side of the epiblast. The hypoblast and epiblast are in contact with each other, and the primitive streak forms a shallow groove in the caudal region of the embryo (Adapted from Sadler, 2006).