Multidisciplinary Approach in the management of Myelomeningocele

Eassy

Submitted in Fulfillment for the Requirements of Master Degree (M.Sc.) in General Surgery

By

Beshoy Wadie Abd El-Sayed Iskandar

(M.B.B.Ch.)

Faculty of Medicine, Minia University

Supervised by

Prof. Dr. Mohammed Ashraf Ghobashy

Professor of Neurosurgery

Faculty of Medicine

Ain-Shams University

Prof. Dr. Sameh Abdallah Maaty

Professor of General Surgery

Faculty of Medicine

Ain-Shams University

Dr.Sherif Morad Girgis

Lecturer of General Surgery

Faculty of Medicine

Ain- Shams University

Faculty of Medicine

Ain-Shams University

2015

بسم الله الرحمن الرحيم

ACKNOWLEDGEMENT

Above all, my deepest thanks go to God, for giving me the patience, power, and health to finish this work.

I am greatly honored to express my deepest thanks, gratitude and respect to my mentor ***Prof. Dr. Ashraf Ghobashy***, Professor of Neurosurgery, Faculty of Medicine, Ain Shams University, for his guidance, supervision, and continuous advice. I am greatly honored to learn from his experience and wise counsel, and thankful for him for giving me some of his precious time.

I am greatly honored to express my thanks and gratitude to ***Prof. Dr. Sameh Maaty***, Professor of General Surgery, Faculty of Medicine, Ain Shams University, for guidance, great help encouragement and his creative support throughout the whole work up.

I am deeply thankful to ***Dr. Sherif Morad***, Lecturer of General surgery, Faculty of Medicine, Ain Shams University.I am greatly honored to learn from his experience and wise counsel, and thankful for him for giving me some of his precious time**.**

النهج المتعدد التخصصات في علاج القيلة النخاعية السحائية

رسالة
الجراحة العامة توطئة للحصول على درجة الماجستير في

مقدمة من

طبيب/ بيشوى وديع عبد السيد اسكندر
بكالوريوس الطب والجراحة

تحت إشراف

الأستاذ الدكتور/ محمد أشرف غباشى
أستـــاذ جراحة المخ والاعصاب
كلية الطب – جامعة عين شمس

الأستاذ الدكتور/ سامح عبد الله معاطى
أستاذ الجراحة العامة
كلية الطب – جامعة عين شمس

الدكتور/ شريف مراد جرجس
الجراحة العامة مدرس
كلية الطب – جامعة عين شمس

كلية الطب
جامعة عين شمس
2015

CONTENTS

LIST OF FIGURES

LIST OF TABLES

ABBREVIATIONS

Posterior longitudinal ligament	PLL
Anterior root exit zone	AREZ
Deep venous thrombosis	DVT
ALL-Terrain Vehcles	ATV
Naso Gastric Tube	NGT
Computed Tomography	CT
Magnetic Resonance Imaging	MRI
Myelomeningocele	MMc

INTRODUCTION

Spinal dysraphism (Spina bifida) is classified into spina bifida occulta with no exposure of meninges or neural tissue and spina bifida aperta which include meningocele & my elomeningocele.

Myelomeningocele is the most common, serious and complex presentation of neural tube closure defects compatable with life ***(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, 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)***, 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).***

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).***

Neural tube defects can usually be detected during pregnancy by testing the mother's blood (AFP screening) or a detailed fetal ultrasound, also the management can be done intra-utero either open or foetoscopic approach.

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

Multidisciplinary management includes dealing with the associated conditions either congenital, psychological or as a result of myelo-meningocele ***(Kemal et al., 2003).***

AIM OF THE WORK

In this study we will clarify the need for a team of doctors (Neonatologist, Urologist, Psychologist and Orthopaedic surgeon) plus the neurosurgeon in this approach to manage lower limb spasticity or flaccidity, urine retention, psychosocial, ophthalmological problems or other associated congenital problems. Also dealing with myelomeningocele either intra-uterine or post-natal. Associated hydrocephalus or Chiari malformation type II and the need for shunt placement and its timing.

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 continuous 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 definitiveendoderm 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)***.

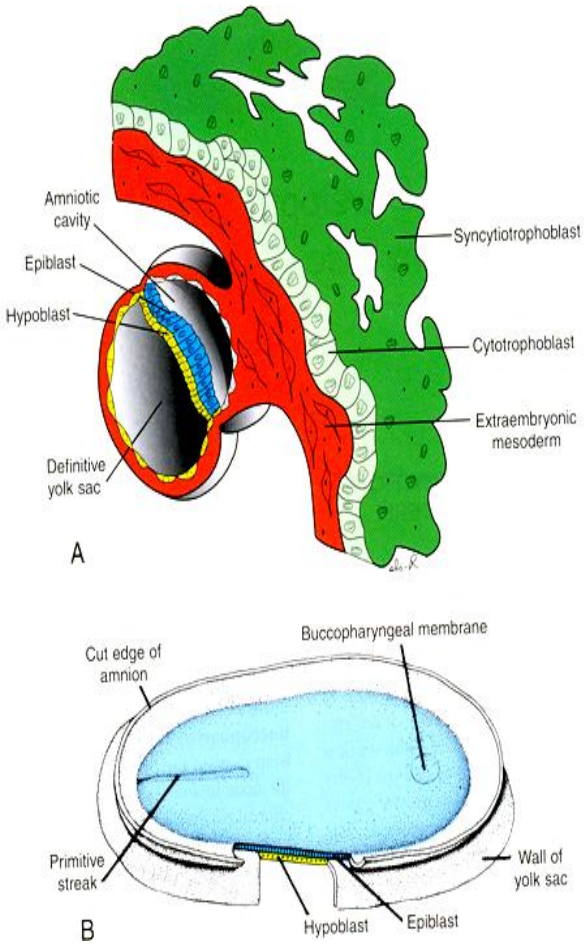

Fig. (1): **A.** Implantation site at the end of the second week. **B.** Representative view of the germ disc at the end of the second week of development. The amniotic cavity has been opened to permit a view of the dorsal side of the epiblast. The hypoblast and epiblast are in contact with each other,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

Gastrulation:

The most characteristic event occurring during the third week of gestation is gastrulation, the process that establishes all three germ layers (ectoderm, mesoderm and endoderm) in the embryo. Gastrulation begins with the formation of the primitive streak on the surface of the epiblast. Initially, the streak is vaguely defined, but in a 15- to 16 day embryo, it is clearly visible as a narrow groove with slightly bulging regions on either side. The cephalic end of the streak, the primitive node, consists of a slightly elevated area surrounding the small primitive pit. Cells of the epiblast migrate toward the primitive streak. Upon arrival in the region of the streak, they become flask-shaped, detach from the epiblast and slip beneath it. This inward movement is known as invagination. Once the cells have invaginated, some displace the hypoblast, creating the embryonic endoderm, and the others come to lie between the epiblast and newly created endoderm to form mesoderm. Cells remaining in the epiblast then form ectoderm. Thus, the epiblast through the process of gastrulation, is the source of all of the germ layers, and cells in these layers will give rise to all of the tissues and organs in the embryo.