MANAGEMENT OF SPINA BIFIDA CYSTICA

THESIS

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of M.D. in « Neurosurgery »

BY

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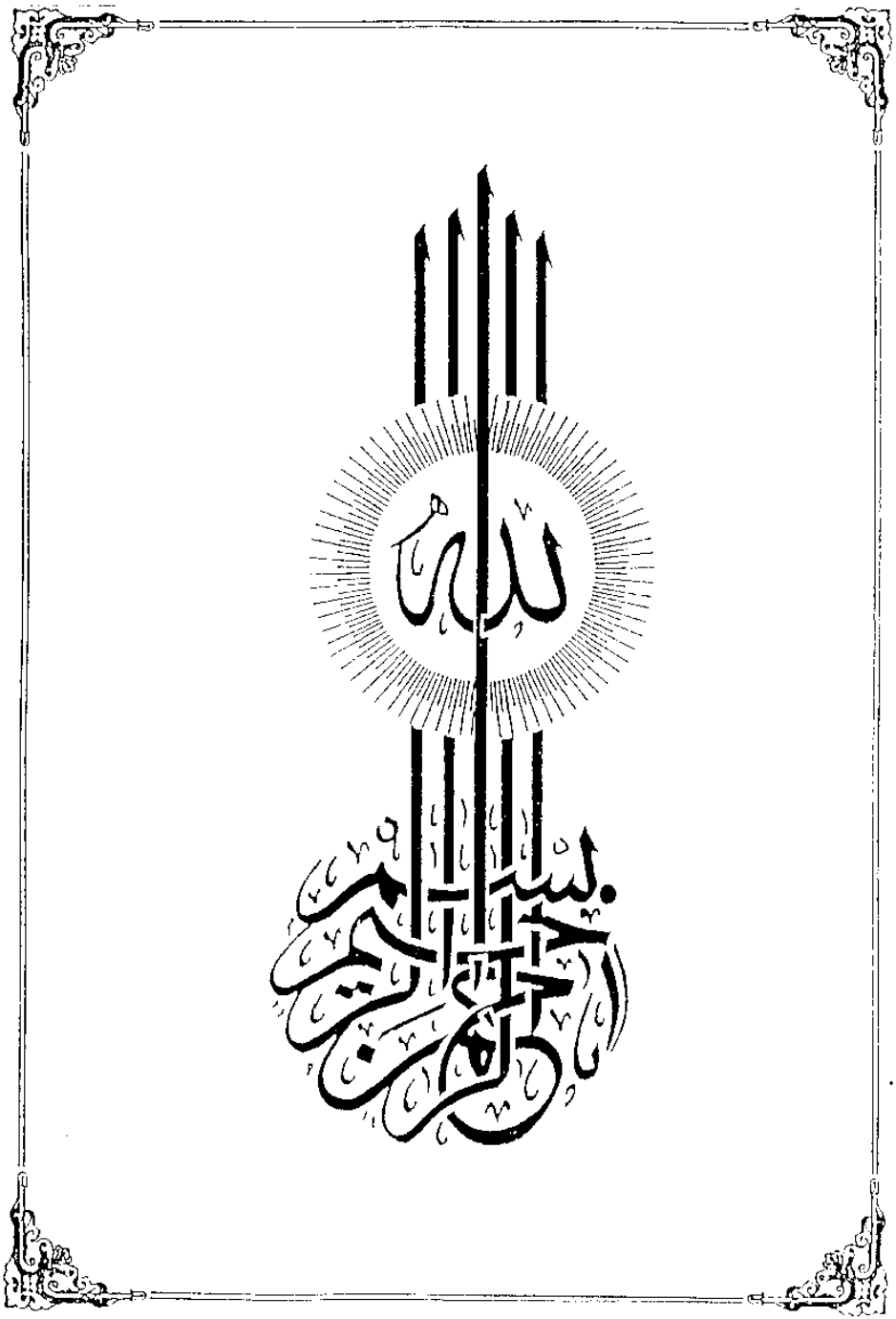
The usefulness of histological data rests largely upon the quality of its illustrations and that was properly directly by **Prof. S. MUSA.**



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C O N T E N T S

PAGE

PAGE

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INTRODUCTION AND AIM OF WORK

INTRODUCTION

The term spina bifida is applied to a dysraphic state of the osseous elements that enclose the vertebral canal. Dysraphism defines the failure of closure of two structures that are normally fused. **Willis in 1958** divided spina bifida into two groups, spina bifida cystica and spina bifida occulta.

The lack of bony closure in spina bifida may be anterior involving the body of the vertebra or posterior involving the neural arches of the vertebra (pedicles, laminae, articular processes and spinous process).

Menkes [81] classified spina bifida cystica added the lipomyelomeningocele to the Willis classification.

Our research on spina bifida cystica includes 35 patients, most of them at newborn or infancy period, 25 myelomeningocele patients were managed in Ain Shams Hospital 'Demardash' in the department of neurosurgery during the last two years 1987-1988 with review of five years back in the same hospital.

Spina bifida is one of the commonest anomalies of the nervous syster, with an incidence that ranges from 0.1 per 1000 live births in Japan to 3.4 per 1000 live births in Ireland [24].

A recurrence rate of 10 percent is seen in affected families [24]. Myelomeningocele is one of the most frequent congenital abnormalities encountered in paediatric and neonatal practice. No satisfactory explanation for the geographical variation has been produced but social factors may be partly involved in an explanation.

The aetiology of the malformation is not known and is considered to be multifactorial [20].

Excessive tea drinking on the one hand, and the eating or touching of blighted potatoes on the other hand, alcohol ingestion [19] has been incriminated, but neither has been

substantiated as a significant factor. There must be an inherited factor, as the random chance of any couple having a child with a major CNS abnormality is about 3 per 1000 live births. If the couple have had a previous myelomeningocele, their chance of having another such affected child rises ten folds to about 1 in 30 and following two similarly affected children, the chance for the third one having a similar lesion becomes about 1 in 6. The inheritance does not appear to be x-linked. [23].

The diagnosis of spina bifida cystica is patently obvious midline defect in the cranio spinal axis. Myelomeningocele occurs most often in the lumbosacral area and in clinical practice it is relatively uncommon in the cervical spine. All cervical myelomeningoceles do not live for more than a few minutes after birth, as they usually have paralysis of the respiratory muscles and die of anoxia. [17].

The number of meningoceles in my work was small (4 patients) and the lesion was covered by normal looking skin, and presented simply as a midline swelling on the back. The management of those patients was simple and the result was excellent. That is simply due to the fact that there was no associated neurological deficit and most of the patients had had no hydrocephalus pre or postoperatively.

The initial surgical management of myelomeningocele was not difficult because we used the paravertebral fascia in repairing the dural defect and in creating artificial dura, the skin of the sac was used in closing the skin defect.

Four patients needed musculocutaneous flaps, that was in the beginning of the work when my experience was limited, I can say now after completion of the work that only one patient actually needed a musculocutaneous flap. We did histological study of the sac components in the department of histology, Ain Shams Faculty of Medicine under supervision

of Professor Dr. Said Musa, the head of the department of histology and with the help of Dr. Mahmoud Hassan, we could proove that there is no dura in the specimen the bulging sac may be a membrane formed by arachnoid or the subcutaneous fascia. We also proved that the skin of the sac can be used in repairing the defect because it is normal thin skin and the atrophic part of it can be removed.

Some newborns after birth tend to have a flat lesion but if untreated it becomes elevated in a posterior direction, by the accumulation of cerebrospinal fluid. Presumably, the intra amniotic pressure prevented such fluid accumulation before birth.(34)

The size of the lesion can vary greatly from 1 to 2 cm to one extending from the mid thoracic to the sacral spine, this could be up to 5 cm or more across.

The aim of this work is to show the initial surgical management of spina bifida cystica by early spinal surgery to close the defect without sac, with sac or musculocutaneous flaps and also to show the management of other anomalies spinal or cranial like hydrocephalus and Arnold Chiari malformation (no patient give early symptom inspite of that this lesion occurs in more than 80% of cases). (58).

Another aim is to follow up the patients for the neurosurgical problem and to refer other to the departments of orthopaedics, urology or physiotherapy for more agressive management.

The myelomeningocele patient is a real problem not only for doctors but also for himself, his family and the society.

REVIEW OF LITERATURE

Basic Science

Neurosurgery

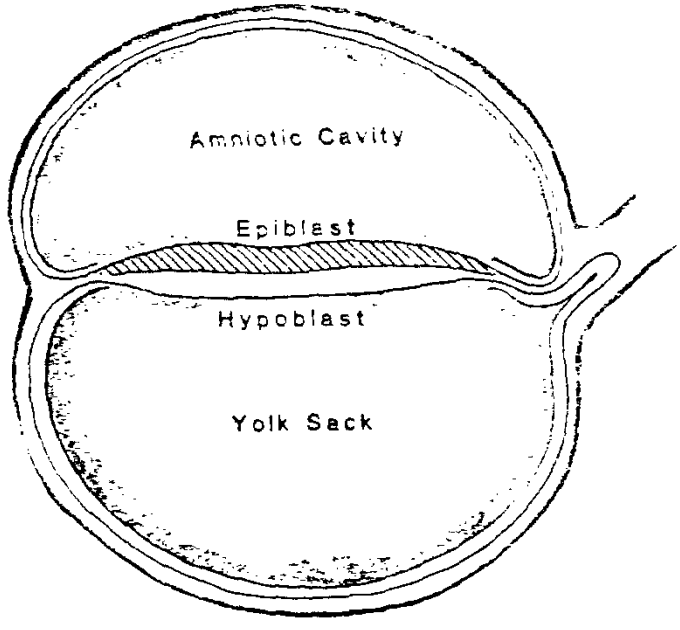
Orthopaedics

Urology

BASIC SCIENCE

Development of the spinal cord and column :

The first two weeks of development after fertilization (embryonic stage) consists of implantation of the conceptus into the uterine wall and development of bilaminar (two layered) embryo as in fig.(1-1) [7]



Diagramatic longitudinal section through the bilaminar embryonic disc within the blastocyst. The epiblast is adjacent to the amniotic cavity and the hypoblast to the yolk sac. The two cavities are seperated by the embryonic disc. [8]

A mesoblastic layer is then formed between the epiblast and the hypoblast converting the bilaminar embryo into the trilaminar embryo consisting of :.

outer ectoderm.
middle mesoderm.
inner entoderm.

The embryo then takes a longitudinal orientation, a linear thickening called the primitive streak in the middle of the caudal aspect of the embryonic disc appear Later,

cells from the epiblast migrate towards the primitive streak and groove and form the mesoblast, thus a trilaminar embryo is formed.

The epiblast (ectoderm) is destined to form the CNS (neurectoderm). The ectoderm which overlies the notochard begins to thicken to form the neural plate, alongside which a longitudinal depression called the neural groove develop, as the edges of the plate heap up forming the neural folds.

At days 22-23, the neural groove deepens, the neural folds meet in the dorsal mid line to form the neural tube as in fig. (27)

Pig.(1-2)