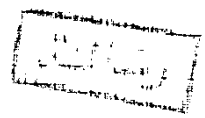
MANAGEMENT OF SKULL DEFORMITIES IN CRANIOSYNOSTOSIS



Thesis Submitted for
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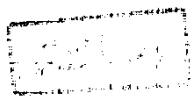
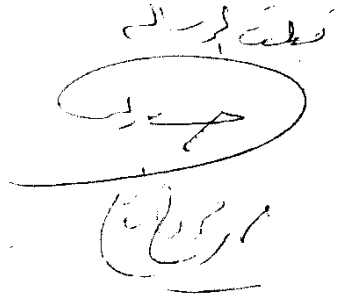
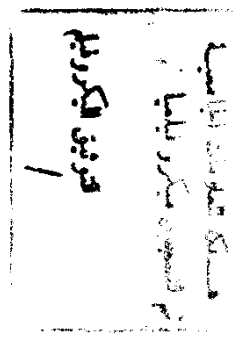




By
Hazem Maher Mohamed Ali,
M.B., B.Ch., M.Sc.
Ass. Lct. of Plastic Surgery



SUPERVISED
BY
Prof. Dr. Hassan Adel Badran
Prof. of Plastic & Reconstructive Surgery
Faculty of Medicine
Ain Shams University

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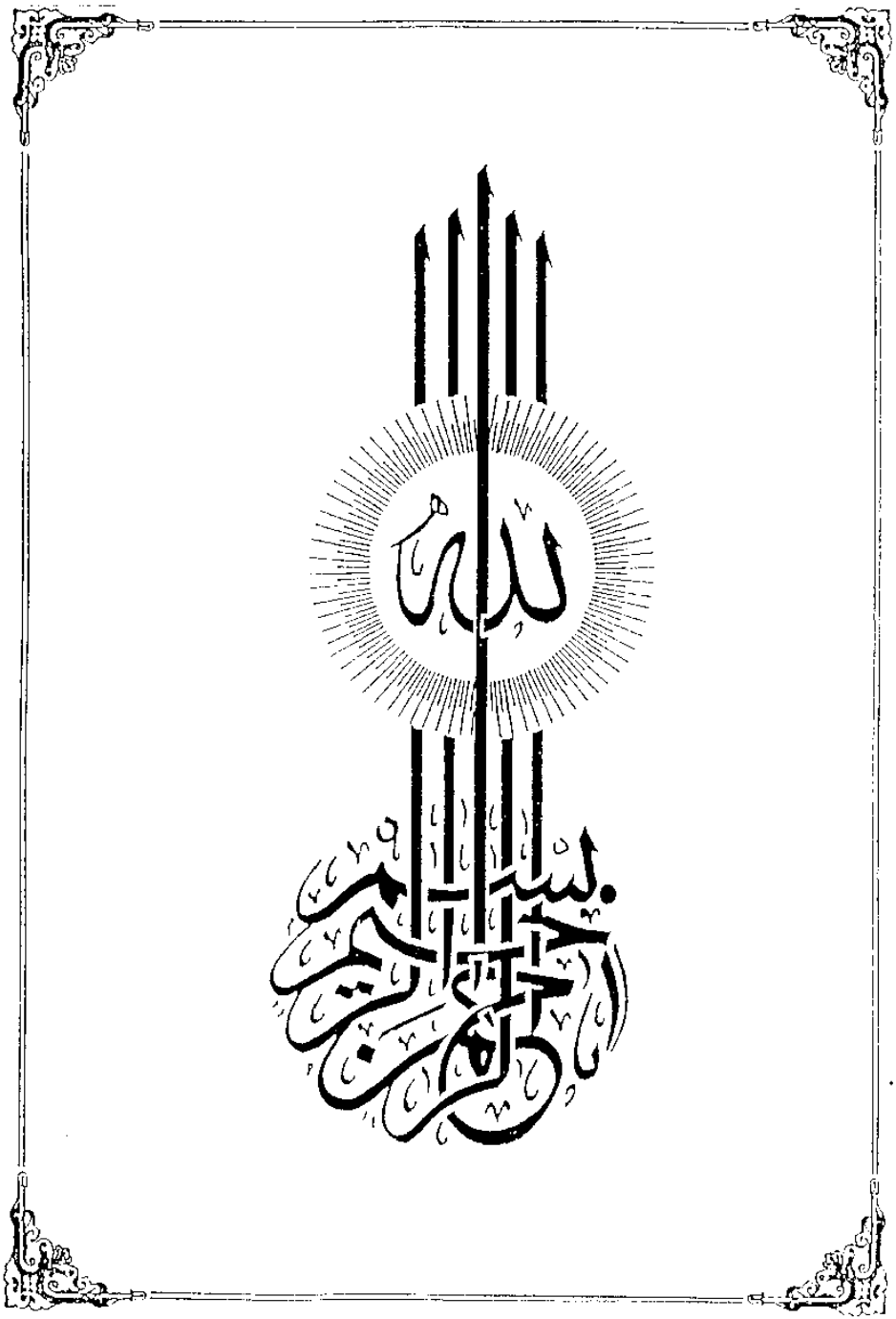

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Introduction

and

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MANAGEMENT OF SKULL DEFORMITIES IN CRANIOSYNOSTOSIS

Introduction and Historical Review:

Craniosynostosis is defined as "the premature fusion of one or more cranial sutures,, with changes in cranial shape and/or dimensions". Sutures are the strips of connective tissue interposed between the adjacent skull bones, usually they refer to the sutures at the vault, however, they are also present at the skull base. The fusion of these sutures means the partial or complete transformation of these sutures into bone which may be focal or total along the whole length of the suture. Ossification of sutures is a physiologic behavior if occurred at the proper time. The normal timing of suture ossification differs from one suture to another, as well as from one race to another. (Cohen, 1986).

Virchow's classical study was published in 1851 about what he called at that time craniostenosis, the term craniosynostosis is now preferred as it describes the pathology and its effects rather than just describing cranial cavity constraint as the craniostenosis implicate. The importance of Virchow's study comes from his description of the deformities of that disease and his explanation that there is an arrest of growth at right angles to the closed suture and compensatory growth at the other sutures. This study, however, was based on pathologic skulls at anatomy museums so, it didn't include clinical data. The deformities described in this study included Macrocephaly, Microcephaly, Dolicocephaly, Brachycephaly (unilateral or bilateral anterior or posterior), and Oxycephaly.

Welker in 1862, described trigonocephaly, and mentioned that it resulted from coalescence of two ossification centers of the frontal bones. He also noted that there was smaller optic nerve in two of the patients with that deformity, however, it wasn't until 1866 when Von Graefe described papilledema with "strikingly high, long and narrow cranium".

Friedenwald in 1893, is the first author to describe increased intracranial pressure as the cause of blindness in patients with craniofacial deformities.

Apert's in 1906, described acrocephalosyndactyly or Apert's syndrome from a case of his own and nine other similar reported cases, in this original description the disease is characterized by a cranial deformity similar to oxycephaly and symmetrical syndactly of both hands and feet. In the few years that followed the clinical picture has been further described by Gadelius (1915) and Wigert (1927). Park and Powers (1920) tried to study the etiology of the disease. (Cohen, 1986).

Crouzon (1912) first described the syndrome that named after him by dysostosis craniofacialis hereditaria. He noted a diseae in a mother and her son, characterized by exophthalmos, cranial deformity, facial deformity specially around the nose.

Lannelongue in 1890 was the first to perform surgery trying to treat craniosynostosis, he originally tried coronal linear craniectomy as a treatment for microcephaly. It was eleven years later (1901) when Bull discovered that microcephaly is the result

of a small brain not the opposite. As a result, linear craniectomy was abandoned as a treatment of microcephaly.

Faber and Towne in 1927 re-employed Lannelongue operation but for a patient with oxycephaly and they stressed on that the outcome depends on performing the procedure as early as possible to avoid the effects of increased intracranial pressure. In a trial to improve this procedure Simmons and Peyon in 1947 and Ingraham et al in 1948 added a non absorbable material in the created gap. Several materials were used including silastic sheets but due to their high incidence of complications and the inadequacy of the results they have been abandoned.(Tessier, 1982).

Total vault craniectomy has been advocated by Powiertowski in 1965, this technique radically released the brain but the reossification that followed was usually incomplete and its shape was defective.

Tessier after introducing the principals and technqiues of craniofacial surgery in 1967 was the first to publish the use of these technqiues in the management of craniosynostosis. In 1971 he described forehead advancement with the supraorbital bar using tenon in groove technique for Crouzon's syndrome teenagers.

Rougerie et al in 1972 proposed an early treatment of craniosynostosis by mobilization of free bony segments decompressing the cranium and remodeling it. They did not move the supraorbital bar but used a bone graft to augment it. They had

good results in cases of sagittal synostosis but not as good results in cases of coronal suture synestosis due to their failure to address the anterior skull base. (Marchac, 1987).

Striker and Montaut in 1972 described moving the supraorbital bar in children with oxycephaly keeping the temporalis attached to the bar which did not allow them to freely adjust the bar position.

Marchac et al in 1973 described a Z osteotomy at the lateral end of the supraorbital bar which allowed slight advancement of the supraorbital bar and forehead repositioning. In 1979 Marchac and colleagues introduced the floating forehead technique with frontocranial remodeling for treating trigonocephaly, plagiocephaly, and brachycephaly.

Jackson in 1978 described different transposition techniques for cranial vault remodeling and in 1988 he described orbital expansion in plagiocephaly.

McCarthy and his associates in 1979 advocated the extension of linear craniectomy to the skull base in cases of coronal synostosis.

In the 1980's different methods of bone fixation have been developed and different osteotomies have been modified which influenced the results as well as a few outcome studies became available to critically evaluate the older techniques. This will be discussed in more details in the next chapters.

The data about the outcome of surgery for craniosynostosis is lacking. Most of the reports on the surgical outcome (Marchac, 1983, Shillito, 1968) do not show a quantitative method for assessing the result, instead they indicate the authors opinion "good or bad" result. In this study we will try to give an alternative solution to answer this question in a more objective way.

AIM OF THE WORK

This work was conducted as a trial to improve and if possible optimize the results of craniofacial surgery for patients with craniosynostosis. Evaluation of the present surgical procedures will be done. Using three dimensional computerized tomography as a method for estimating the skull deformity in an objective and quantitative way will be tried. Using this technique, comparison of the deformed skulls of patients with craniosynostosis and a control group will be carried out. A comparative study of pre and post operative skull measurements will be done in relation to age matched normal. If this technique proves successful it will be used to help in preoperative planning for surgery on patients with craniosynostosis and in the estimation of the success of the present surgical techniques. It also can be used to evaluate the effect of the continued growth on the result.

Review of literature

REVIEW OF LITERATURE

Embryological aspects related to craniosynostosis:

The cranial vault bones originate from the mesenchymal mass that surrounds the brain with development in three overlapping stages (Kier, 1976).

(1) Membranous stage:

At the start of the second month of fetal life a thick mesenchymal mass appears in the occipital region where it surrounds the notochord, extending forward towards the hypophysis to outline the clivus and the sellae. Then the mesenchymal mass extends into the region of the nasal septum, the ethmoid and greater sphenoid wings, closing the optic and autidory centers, suroudning the nerves forming the cranial foramina. Subsequently, chondrification takes place in the mesenchyma of the base of the skull, while the bones of the cranial vault form directly in membrane from the mesenchyma surrounding the brain.

(2) Cartilagenous stage:

Cranial chondrification begins during the second month of fetal life and is limited to the base. It begins in three regions, posteriorly arround the notochord and the auditory centers, medially around the hypophysis, and anteriorly in the region of the nasal septum. The chondrocranium is continuous with the vault and diminishes in size as clacification proceeds, so that at birth its residues are in the nasal, sphenoid and occipital bones and in the foramen lacerum and in the spheno-occipital and petro-occipital

synchondroses.

(3) Ossification stage:

* Frontal bone: The bone originates in the external layer of the membranous coverings of the brain from two centers that appear at the 8th week in the superciliary arch region and centers extend upward and backward forming the oribital plane, and downward forming the nasal segment. At birth the frontal bone is made up of two halves separated by the metopic suture.

* Parietal bone: begins ossification by the 7th week of intrauterine life from two centers that join each other then spread in a radial manner.

* Occipital bone: This is formed by four segments that surround the foramen magnum (a) the basioccipital anteriorly (b) the squamous posterior and inferior (c,d) the two exoccipitals at lateral sides of the foramen magnum.

At birth the four segments are separated by strips of cartilage. They start to ossify from the ossification center that appears between the 6th - 12th week of intrauterine life.

* Temporal bone: At birth temporal bone is made up of three parts (a) the squamous (b) the tympanic, both from a membranous origin and (c) the petrous bone from a cartilagenous origin. The squamous bone starts ossifi from a center that appears at the 8th week I.U life which extends then to the zygomatic process. The tympanic center of ossification appears between the