DIAGNOSTIC ASPECTS OF ULTRASONOGRAPHY IN INTRA - UTERINE FOETAL NEUROLOGIC ANOMALIES.

THESIS

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Of Master Degree In Radiodiagnosis

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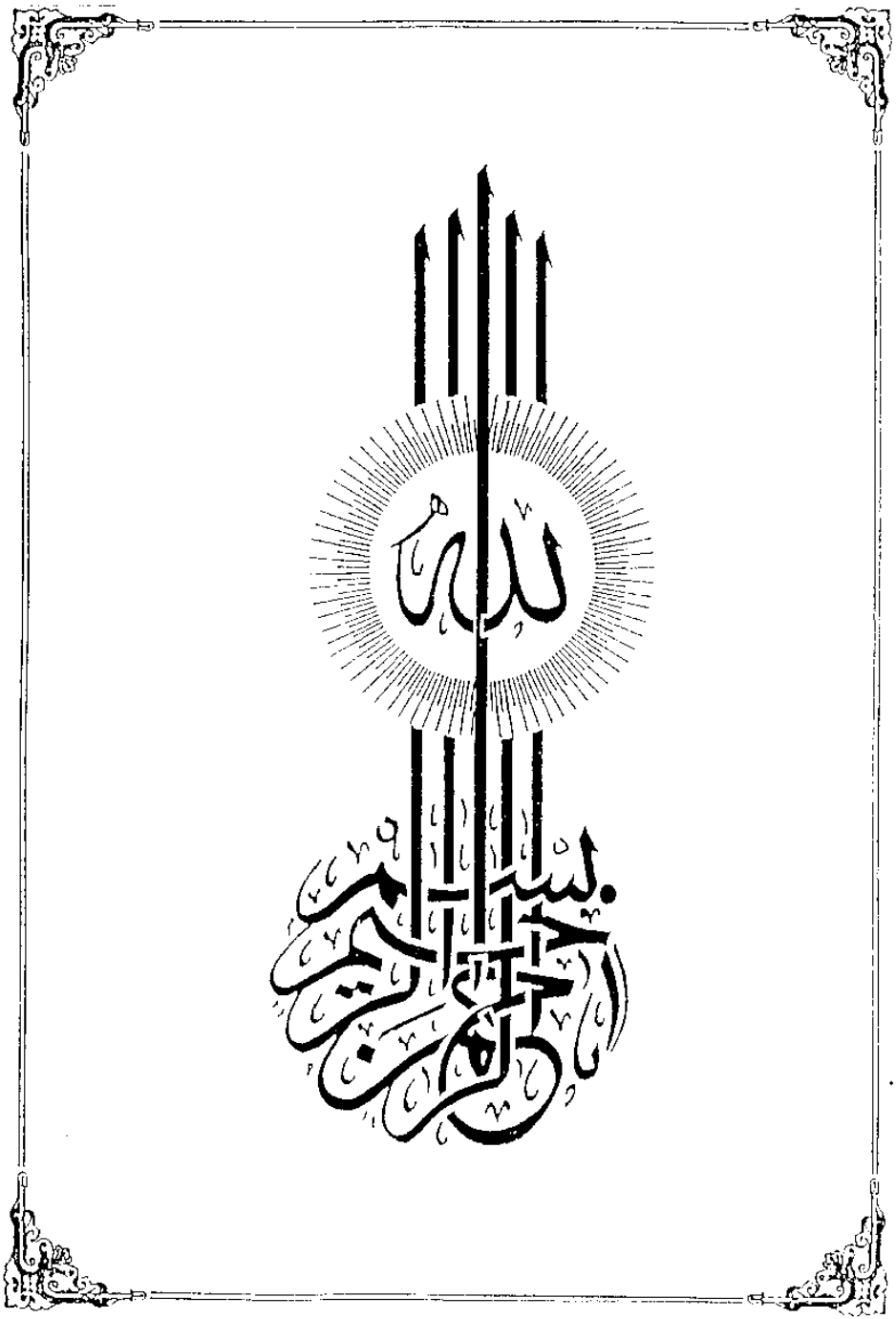

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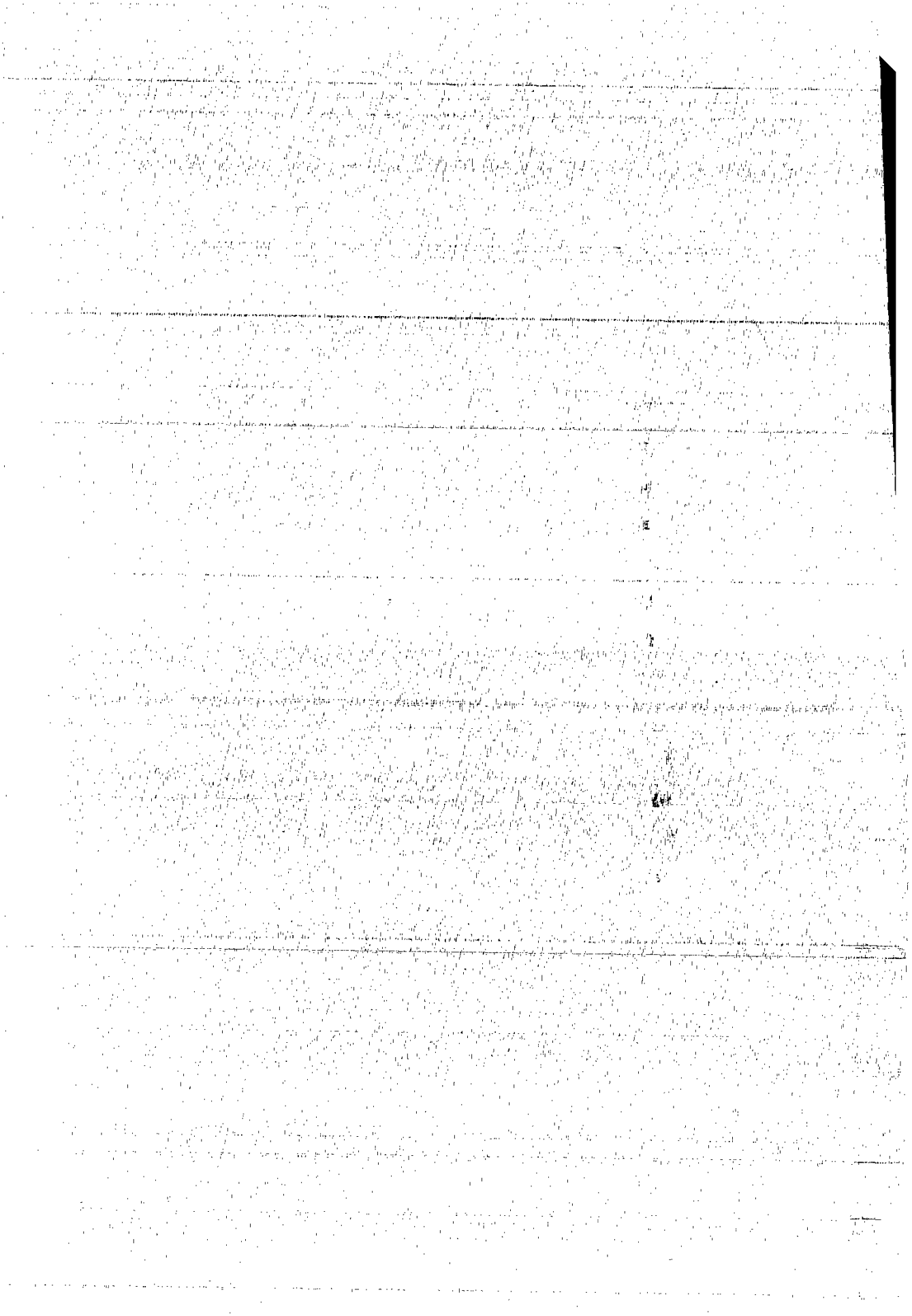
INTRODUCTION AND AIM OF THE WORK



INTRODUCTION AND AIM OF THE WORK

Ultrasonography is assuming an important and increasing role in the Prenatal diagnosis of foetal anomalies.

Ultrasonography, as it is safe and non-invasive, has become the Principal means of obtaining morphological information about the contents of the uterus which can be evaluated at any time during Pregnancy.

Since the early 1970, ultrasound has been used to examine the foetal spine and head for diagnosis of neural tube defects (Macri and Weiss, 1982).

Normal and some abnormal conditions of the brain especially hydrocephalus can be detected using the anterior fontanel as a sonographic window (Babcock and Han, 1981 & Sauerbrei and Cooperberg, 1981).

PATHOLOGY

PATHOLOGY

Malformations result from an error in the complex development of the nervous system. The etiology of such malformations has excited interest in recent years and although knowledge is regrettably incomplete, it is known that a variety of influences operating in the early stages of fetal development may result in errors of development (Ford, 1960 & Potter, 1961). Among the known causes are radiation, anoxia, drugs and viral illnesses, especially rubella in the early months of pregnancy, (Foley, 1974).

According to Thomson & Cotton, 1983, these can be classified as follows:

I- Congenital abnormalities of the brain :

- Anencephaly.
- Microcephaly.
- Spastic diplegia.
- Tuberose sclerosis.
- Microgyria.
- Porencephaly.
- Mongolism.

II- Congenital abnormalities of spinal or cranial fusion:

- Spina bifida occulta.
- Spina bifida vera.

- Myelocoele.
- Syringomyelocoele.
- Meningomyelocoele.
- Meningocoele.
- Encephalocoele.
- Arnold-Chiaria malformation.
- Spondylolisthesis.
- Platybasia.
- Klippel - Feil deformity.

III- Hydrocephalus

According to Nelson (1975), these can be classified as follows:

I- Failures of fusion or dysraphic states:

- Anencephaly.
- Cephalocele.
- Spina bifida.
- Myelodysplasia.

II- Failures of cleavage :

- Cyclopia.
- Arhinencephaly.

III- Disorders of migration :

- Heterotopias.
- Ectopias.

IV- Disorders of sulcation :

- Lissencephaly.
- Pachygyria.
- Microgyria.

V- Miscellaneous lesions :

- Agenesis of the corpus callosum.
- Cerebellar agenesis.
- Hydranencephaly.
- Porencephaly.
- Microcephaly.

The number of possible malformations is very great and malformations of the nervous system are frequently associated with abnormalities in other parts of the body, (Foley, 1974).

These lesions will be dealt with according to the first classification :

A- <u>Skull & Brain Lesions :</u>

1- Anencephaly :

Is a condition in which there is a deficiency of the cranial vault with absence of the brain. This condition, which is incompatible with life, is sometimes associated with non-closure of the spinal canal or rachischisis (Anderson, 1985).

According to Nelson (1975), anencephaly is one of the dysraphic states or failure of fusion because of complete or partial discontinuity of the neural tube or its boney coverings.

Anencephaly is the most common human congenital anomaly incompatible with sustained extrauterine life.

It is more frequent in female infants than in male, and its incidence ranges from 0.5 to 3.7 per 1,000 births. It is especially frequent in Scotland and Ireland. The lesion is more likely to occur in infants born to women over 40 years of age or to parents who have had an anencephalic infants previously.

Anencephalic infants may survive a number of days after birth. Motor responses, including head rotation, limb flexion, partial Moro reflex, reflex sucking, and changes in facial expression, can be elicited with appropriate stimuli. No therapy is available.

Many anatomic variants of anencephaly have been described. The basic defects are absence of the cranial vault (acrania), the cerebral hemispheres, and the diencephalon.

In place of the neural structures is an exposed mass of tissue, the area cerebrovasculosa. It is composed

of thin-walled vascular channels, masses of neural tissue, and structures resembling choroid plexus. The optic nerves are absent, although the eyes may be well formed. Some of the cranial nerves are identifiable.

The caudal extent of the lesion varies. Occasionally, the midbrain, or the pons, or rarely, the cerebellum is present. Commonly, the malformation involves the cervical cord.

The entire neural axis may be affected-**Craniorachischisis totalis.*** The spinal nerves and sympathetic structures are well formed (Nelson, 1975).

* **Craniorachischisis totalis** is the most extreme degree of failure of closure of the neural tube. Here, the convexity of the skull is absent and the spine in represented only by its bodies with no posterior covering.

The nervous system lying in its underdeveloped container is equally underdeveloped, consisting of blood vessels, glia and scattered unorganized nests of nerve cells and processes. Such a defect is not compatible with extrauterine existence (Foley, 1974).

Bony defects include absence of the frontal bones above the supraorbital ridge, absence of the parietal bones, and usually absence of squamous portions of the

occipital bones and the foramen magnum. The base of the skull is well developed, but deformities of the petrous temporal bone, the sphenoid wings, or abnormally fused bones are often present.

When the malformation extends into the spine, the number of cervical vertebrae is reduced. The bodies of the vertebrae are present, but the vertebral arches are not formed and the vertebral canal is exposed.

The adrenal glands and pituitary gland are small in anencephalic infants. Other anomalies commonly occuring in anencephalic infants include malformation of the main cerebral arteries and club foot. Palatal clefts, defects of the abdominal wall or the diaphragm, abnormalities in the shape and number of bones in the upper extremity, and visceral anomalies have been described.

Maternal hydramnios often accompanies anencephaly (Nelson, 1975).

2- Microcephaly :

Congenital smallness of the brain which is otherwise anatomically and proportionately normal. This results in varying grades of mental deficiency (Thomson & Cotton, 1983).

It is thought to occur in an incidence of 1:6,200 to 1:8,500, however, studies from which these figures were derived did not include stillborns or children who had died early in childhood (Kurtz et al., 1980).

There is deficiency in the convolutions, and the sulci are imperfectly formed, it is associated with severe mental retardation. The whole brain may be abnormal, but usually the cerebellum and brain stem are less affected than the cerebrum. In other cases, there is a local defect in growth, often associated with a small size of the convolutions or microgyria. Some virus infections, e.g. cytomegalovirus can cause microgyria, but the cause of most cases of micrencephaly is not known (Anderson, 1985).

The causes of microcephaly include:

1- Craniosynostosis.

2- Inherited factors, in association with the Meckel-Gruber syndrome (Polydactyly, encephalocele, polycystic kidney disease, and microcephaly).

3- Chromosomal abnormalities, including trisomies.

4- Exposure to environmental teratogens including radiation and viruses (Fiske & Filly, 1982 and Smith, 1982).