CONGENITAL ANOMALIES OF THE TESTES.

Thesis Submitted In Partial FullFilment

For The Master Degree In Urology.

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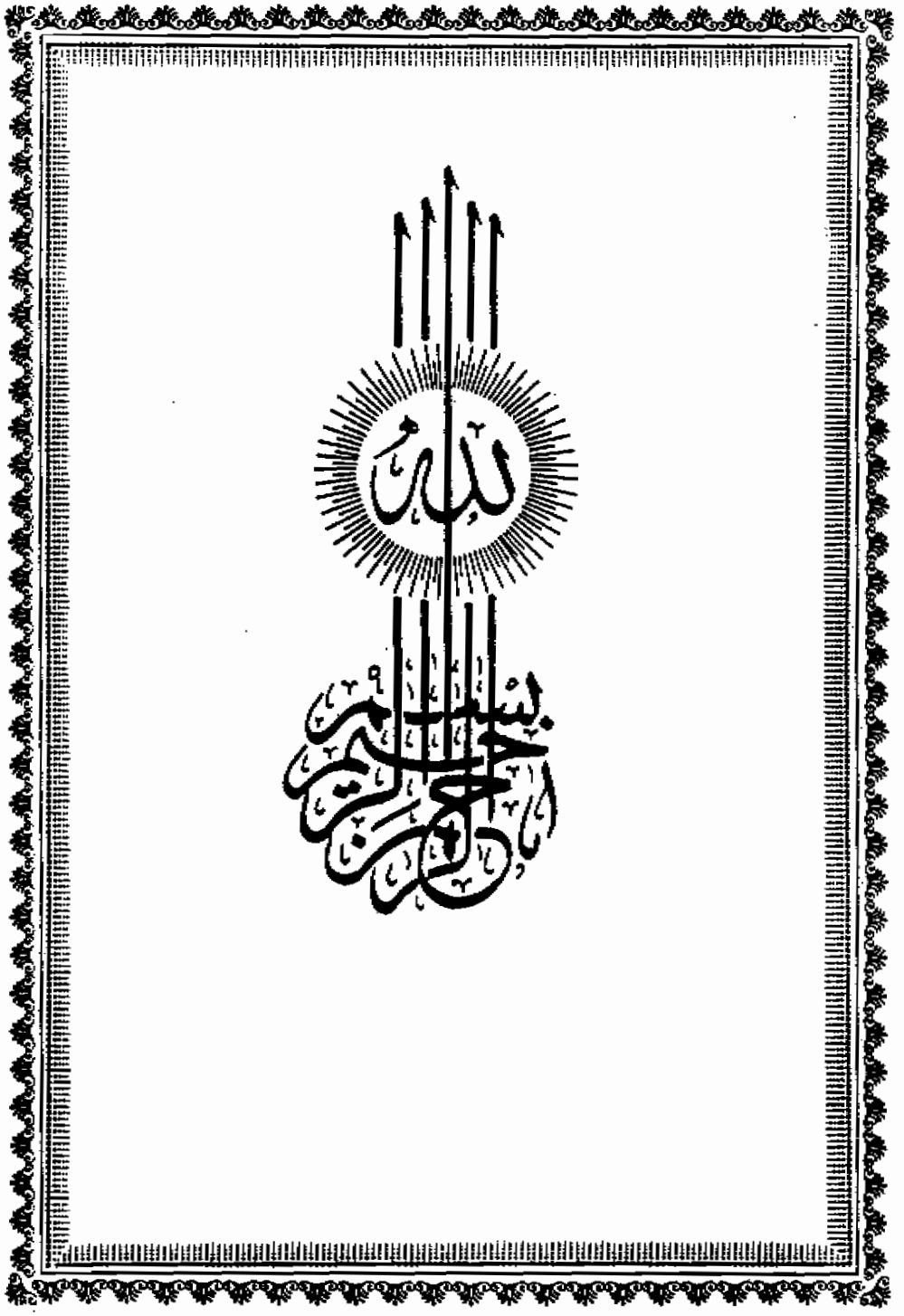

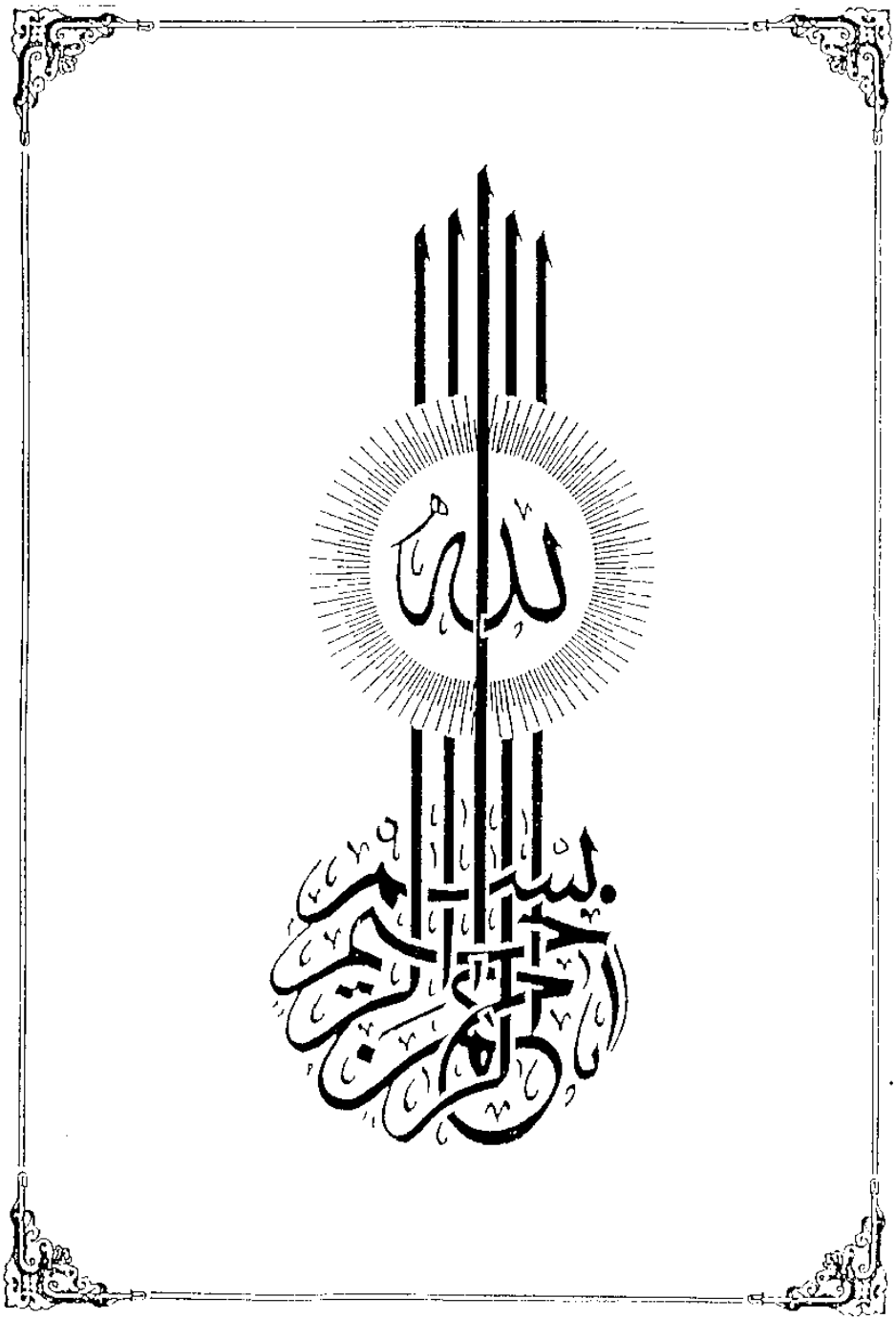

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I dedicate this volume to
my mother who made this
endeavor possible.

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INTRODUCTION

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The embryos of both sexes develop in an identical fashion until approximately 40 days of gestation (about 15 to 20 mm crown-rump (C-R) length), and only thereafter does anatomic and physiologic development diverge to result in the formation of the complete male and female phenotypes.

The gross anatomic aspects of this critical developmental process are largely completed by the end of the second trimester, but certain functional and structural aspects of sexual development, including maturation of the genital and gonads, are not completed until postnatal life

There is probably no aspect of embryonic development for which the forces that regulate the process of differentiation are so well understood as sexual differentiation. Moreover, there is probably no other developmental process in which abnormalities result in such a large variety of human disease states. This is thought to be the consequence of the fact that normal sexual development is essential only to the survival of the species, not to the life of the individual .

On the other hand , many developmental defects in organ systems that are vital to life cause lethal abnormalities that result in abortions.

Congenital anomalies of the tests include cryptorchidism, testicular torsion and inguinal hernia and hydrocele.

The testis is hidden, that is the meaning of the word cryptorchidism. When absent from the scrotum, the testis is hidden from sight. Palpation may show that it is concealed in the upper part of the scrotum but can be drawn down, or it may be found in the groin, relatively fixed in position. Rarely,it may have diverted from the path of descent to a totally abnormal situation(true ectopia). At times, repeated searching fails to locate the organ in any site, it is also hidden from the examining fingers. This common developmental abnormality of childhood-cryptorchidism-needs to be more widely understood, accurately diagnosed, and effectively treated. If it is

overlooked until adult years, it is then too late to give useful help .

Torsion of the testis is possible at any age, although it is most common in the adolescent, from 12 to 18 years of age. A few cases present in neonates, and there are occational examples in infancy and childhood. Above the age of 18 years the incidence slowly decreases, although cases are still seen in patients, third and fourth decades of life .

Congenital inguinal hernias and hydroceles are frequent reminders of the past history of the descending testis. In a normal neonate, the peritoneal sleeve of the processus vaginalis, which has been drawn down through the inguinal canal during descent, is in the process of closure. It generally becomes obliterated between the eighth month of fetal life and the end of the first month of brith.

This closure may be delayed for weeks or months, or it may never occur. Failure of the processus vaginalis to disappear may be complete or partial .

Although an open processus provides the defect that allows herniation of the abdominal viscera to occur, it is only when the herniation happens that a clinical problem is discernible by the parent or physician. if closure is incomplet , yet is sufficient to prevent the entery of abdaminal viscera, the patent channel may still allow the accumalation of peritoneal fluid within the tunica vaginalis, resulting in a vaginal hydrocele. If the distal portion of the processus should close but the proximal part remain open, a hydrocele of the spermatic cord may result .

EMBRYOLOGY

EMBRYOLOGY OF THE TESTIS

Formation of the Gonads:-

The primordial germ cells are discernible at about day 21 of embryonic life and are seen among the entodermal cells in the wall of the yolk sac near the origin of allantois. With the formation of body folds, this part of the yolk sac is taken into the embryo and forms the hindgut. Later these cells migrate via the dorsal mesentery and, by the sixth week, they come to lie beneath the coelomic epithelium on the medial side of the mesonephros. The presence of the primordial sex cells beneath the coelomic epithelium stimulates the epithelial cells to multiply, and cords of epithelial cells now grow down into the underlying mesenchyme. The cords of cells are called sex cords, and they come to surround the primordial sex cells, maintaining their connection with the surface epithelium(Snell, 1975).

The thickening rapidly extends in a longitudinal direction until it covers nearly the whole of the medial surface of the mesonephric ridge. The thickened epithelium continues to proliferate, displacing the renal corpuscles of the mesonephros in a dorse lateral direction, and forming a projection into the coelomic cavity, the gonadal ridge.

Surface depressions form along the limits of the ridge which is thus connected to the mesonephros by an originally broad mesentery " the mesogenitale." In this way the mesonephric ridge becomes subdivided into a lateral part containing the mesonephric "Wolfian" duct and paramesonephric duct, whi may termed the tubal fold and a medial part, termed the gonadal fold.

The tubal fold contains the nephric tubules and glomeruli at its base.

Up to 7 week the gonad possesses no differentiating features. The gonadal cords, remain at the periphery of the primordium to form a cortex, more centrally a proliferation

of the mesenchyme of the mesonephros constitutes a madella. In the male all the progenitors of the definitive gonocytes become incorporated in the cords, but in female a large nu number remain behind under the surface epithelium. At this stage in the male, an extension of the mesechyme cuts off the gonadal cords from the surface and rapidly thickens to form the tunica albugina. This also develops in the female but to a lesser extent and at a later stage.

In embryo with an XX chromosome complex, the cortex forms an ovary, and the medulla regress, in one with an XY chromosome complex, the medulla differentiates into a testis and the cortex regresses.

Development of the testis:- (Fig. 1)

The male gonad develops into the testis toward week 7 of of development. Until then, it is undifferentiated. The differentiation is determined by XY genetic constitution and proceeds as follows,the primary sex cords condense and extend into the medulla of the gonad where the cords branch, their deep ends anastomose, and they form the rete testis.

The prominent sex cords become the seminiferous or testesticular cords which soon lose their connections with the germinal epithelium because of the development of a thick fibrous capsule, the tunica albugina which is a layer of connective tissue interposed early, between the coelomic epithelium(parietal peritoneum) and the rest of the gland. It produces partitions which compartmentalize the gland, closing off the seminiferous ducts, about day 50, into testes cords.

The development of the tunica albugina is a characteristic and diagnostic feature of testicular development.Then the seminiferous or testicular cords develop into the seminiferous tubules, whose deep portions narrow to form the tubuli reeti, which converge on the rete testis.

The seminiferous tubles become separated by mesenchyme which gives rise to the interstitial cells of leyding where the androgenic hormones are secreted which help in the differentation of the genital tract and the external genital organs.

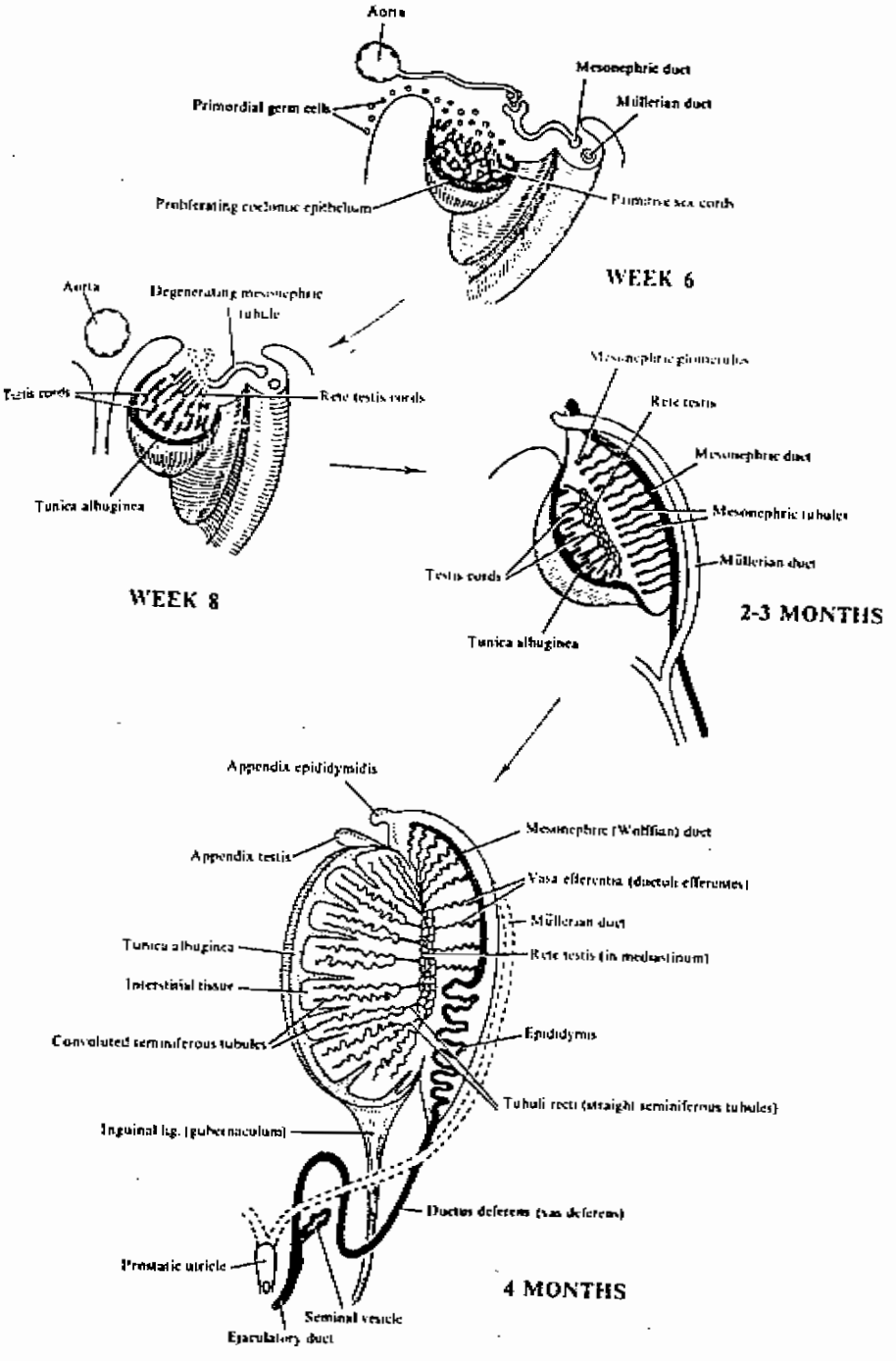

Figure 1.

The interstitial cells of Leydig reach their maximum development between 3½ -4 months.

The walls of the seminiferous tubules, as a result of their cellular duality of origin, are composed of 2 types of cells: supporting or sustentacular cells of Sertoli, derived from the germinal epithelium, make up most of the seminiferous epithelium in the fetal testis and the spermatogonia, derived from the primordial germ cell(unlimited in number, in contrast to the oogonia).

Gradually, the enlarging testis separates from the regressing mesonephros and is suspended by its own mesentery, the mesorchium. Later the germinal epithelium flattens to form the mesothelium on the surface of the testis and the rete testis becomes continuous with the 15-20 adjacent persistant mesonephric tubules, forming vasa efferentia or efferent ductules which open into the adjacent mesonephric duct which becomes the ductus epididymis(epididymis).

Primitive Genital Tracts and sex determination:-

The genital tracts have the same appearance in both male and female embryos until week 7 of development, consisting of the 2 paramesonephric or Mullerian ducts and the 2 mesonephric or Wolffian ducts. The latter drain the mesonephric kidneys, but persist as the male genital ducts when the mesonephric system undergoes degeneration.

The paramesonephric ducts develop bilaterally from invaginations of the coelomic epithelium on the lateral aspects of the mesonephroi. In the 10-mm embryo the Müllerian ducts induce an invagination of the coelomic epithelium opposite the cranial end of each mesonephric duct which reates an epithelial bud that penetrates the mesenchyme and progresses caudally along the mesonephric duct. The bud hollows out at the same time it grows and thus becomes an open paramesonephric duct in the coelomic or peritoneal cavity. The paramesonephric ducts cross in front of the Wolffian ducts at the lower pole of the mesonephric body and then run along side it. The terminal parts of the paramesonephric ducts fuse to form a

small,single median duct (uterovaginal primordium or canal) which ends blindly at the posterior surface of the urogenital sinus to create an elevation, the sinus or Müllerian tubercle which is located between the opening of the mesonephric ducts into the urogenital sinus.

The mesonephric and paramesonephric ducts are located in the urogenital cord, which is attached to the anterior external edge of the mesonephric (Wolffian) body by the urogenital mesentery. The latter attaches the urogenital cord to the body wall, below the mesonephric body. Furthermore, the 2 urogenital mesenteries (righ and left) join below the median line. The mesonephric body is attached to the abdominal wall by the mesonephric mesentery through its entire length. Above the body the urogenital mesentery and mesonephric mesentery extend upward and form the diaphragmatic ligament. The lower pole of the mesonephric body is attached at the inguinal region by the inguinal ligament.

Histogenesis of Gonads:-

It is now pertinent to give attention to the late histological changes by which such indifferent gonads gradually become differented into testes or ovaries.

If an individual is destined to become a male, the cords of potential sex cells which were but vaguely defined in the gonads of the indifferent stage become much more sharply delimited from the intervening embryonic connective tissue, they may now properly be called testis cords. At the same time there is developed directly beneath the germinal epithelium a conspicuous zone of young connective tissue. This is the start of a layer which in the adult is called the tunica albugina, it is composed of such densely interwoven white fibrous connective tissue. By the fourth month of development the testis has lost the original elongated, more or less spindle-shaped, configuration characteristic of the early gonad and become more rounded and compact.

At the same time its original broad attachment to the mesonephros is reduced to a mesentery like attachment known as the mesorchium.