MANAGEMENT OF INTERSEXUALITY

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By

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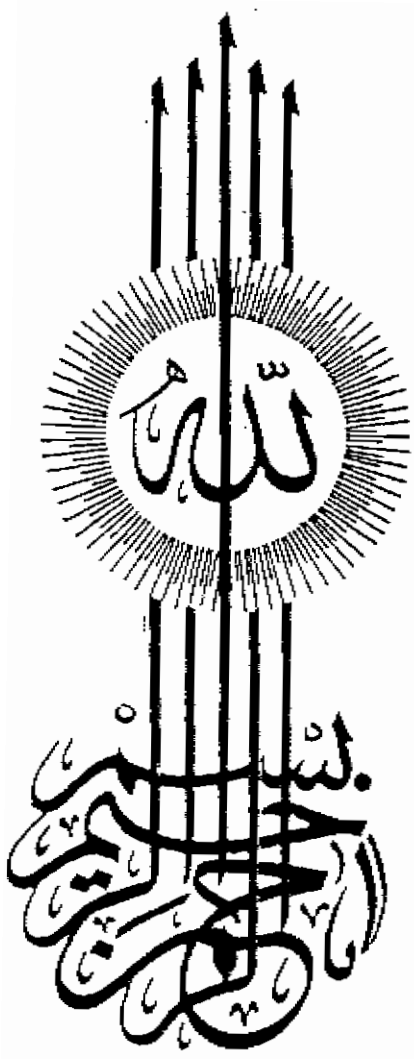

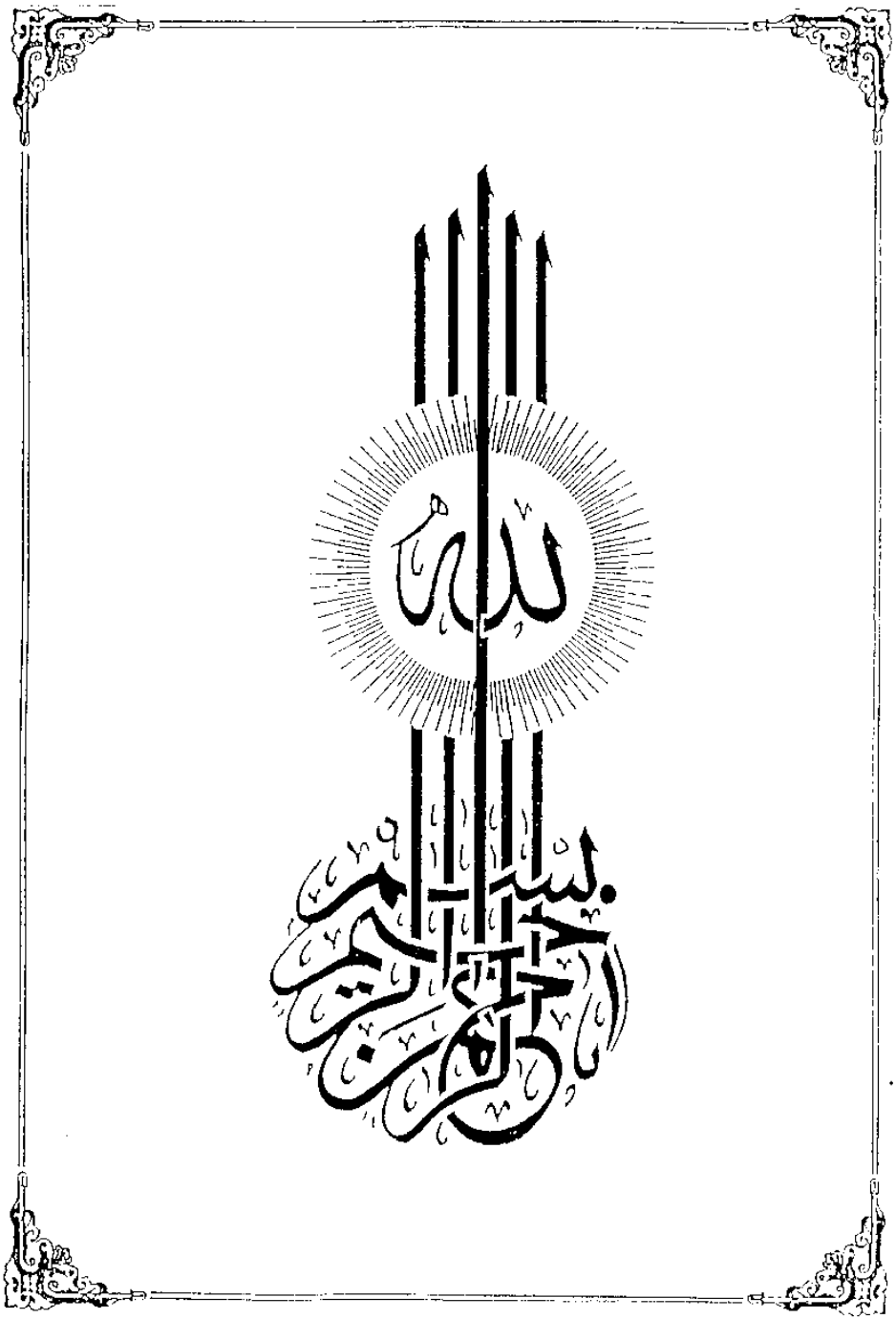

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C O N T E N T S

INTRODUCTION
AND
HISTORICAL REVIEW

I N T R O D U C T I O N

A N D

HISTORICAL REVIEW

Intersex may be defined as a disorder of sexual differentiation or development at the chromosomal, gonadal, or genital level. Some intersex disorders have a fixed phenotype like Klinefelter's and Turner's dyndromes; they mainly present with infertility, and will be discussed briefly. Other disorders cause ambiguity of the genitalia of varying degrees, and this type will be the main interest of this thesis.

A true hermaphrodite is a person who possesses both ovarian and testicular tissue, while a male pseudo-hermaphrodite is one whose gonads are exclusively testes, but his genital ducts, or external genitalia,or both exhibit incomplete masculinization in one or more aspects. In female pseudohermaphrodites, the gonads are purely ovaries, but the genital development exhibits a masculine appearance.

Human embryos of both sexes develop in an identical fashion until about 40 days of gestation, and only thereafter does anatomic and physiologic development diverge to result in the formation of the complete male and female phenotypes.

The main part of this critical developmental process is largely completed by the end of the second trimester, although certain functional and structural aspects of sexual development,

including maturation of the gonads and the genital tracts are not completed until postnatal life.

An aberration at any stage of sexual development has profound clinical consequences that range from the common defects involving the terminal stages of male development (hypospadias, cryptorchidism, and microphallus) to the more fundamental abnormalities that result in a variety of problems of intersex.

There is no other aspect of embryonic development for which the forces that regulate the process of differentiation are so well understood as sexual differentiation and that is why I discussed them in a separate chapter. Moreover, there is no other developmental process in which abnormalities result in such a large variety of human disease states. This is thought to be the result 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 abortion .

A complete historical review of the subject may be difficult because of the much controversy about terminology and classification, as a representative, the following disorders may be considered: Turner (1938) described gonadal dysgenesis, for the first time, as a syndrome of short stature, sexual infantilism, congenital

webbed neck, and cubitus valgus. In 1942, Klinefelter, Reifenstein, and Albright delineated an entity in men characterized by small firm testes, varying degrees of eunuchoidism, azoospermia, gynecomastia, and elevated levels of urinary gonadotropins.

Sohval (1963) was the first to use the term (mixed gonadal dysgenesis) to describe an entity in which phenotypic female or male patients were found to have a testis on one side and a streak gonad on the other. More than 300 cases of true hermaphroditism have been reported in the literature, Lee (1971) reported an incidence of 1.08%, it is the commonest cause of genital ambiguity in the black populations.

Morris (1953) was the first to employ the term (testicular feminization) in those "hairless women" recognized in medicine.

The goal of the physician who has a patient with ambiguous genitalia is to diagnose the problem and to assign a sex for rearing which is most compatible with a well-adjusted life and sexual adequacy. In recommending male sex assignment, the adequacy of the size of phallus should be the most important consideration.

Once the sex for rearing is assigned, the gender role is reinforced by the appropriate employment of whatever surgical, hormonal, or psychologic measures that are indicated.

ANATOMY

OF

THE EXTERNAL GENITALIA

A N A T O M Y

O F

THE EXTERNAL GENITALIA

External Genitalia in the Male

The male external genital organs comprise the scrotum and the penis. The scrotum is situated below and in front of the urogenital triangle, part of the penis is located in the urogenital triangle and the remainder is located in front of the scrotum (Gray D.J. 1960).

The urogenital triangle :

It is contained between the ischio-pubic rami and the line passing between the anterior parts of the ischial tuberosities. It is divided into deep and superficial triangles by the perineal membrane (urogenital diaphragm).

The urogenital diaphragm is an unyielding sheet of fibrous tissue which forms the basis upon which the penis and penile musculature are fixed; below it the scrotum depends, above it the membranous urethra lies, surrounded by the sphincter urethrae, below the apex of the prostate. It is pierced by the urethra, and by the foramina for nerves and vessels. It is attached to the ischio-pubic rami from the subpubic angle back to the level of the anterior part of the ischial tuberosities, along a ridge

which lies on the inner aspect of the medial surface of each ramus. Its antero-posterior extent is almost 1½ inches (3.5 cm). The fascia of Colles is attached to its posterior margin. The superficial perineal pouch is a name given to the space enclosed between the perineal membrane and the fascia of Colles. It contains the testes and their spermatic cords as well as the penis and the muscles of the corpora .. in a word it constitutes the external genitalia. (Last R.J. 1981).

The Scrotum :

The scrotum is a cutaneous and fibromuscular sac containing the testes and the lower parts of the spermatic cords, and dependent below the pubic symphysis in front of the upper parts of the thighs.

It is divided on its surface into right and left halves by a cutaneous ridge, or raphe, which is continued anteriorly to the inferior surface of the penis and dorsally along the middle line of the perineum to the anus; the left usually descends a little more than the right, in correspondance with the greater length of the left spermatic cord. The raphe indicates the bilateral origin of the scrotum from the genital swellings (Fig. 1).

The external appearance varies in different circumstances : thus under the influence of warmth, and in old and debilitated

persons, the scrotum is smooth, elongated and flaccid; but under the influence of cold, and in the young and robust, it is short, corrugated, and closely applied to the testes (Warwick, R. and Williams, P.L. 1973).

It consists of the skin and dartos muscle, together with the external spermatic, cremasteric, and internal spermatic fasciae. The inner surface of the internal spermatic fascia is loosley attached to the parietal layer of the tunica vaginalis. The scrotal skin is very thin, of brownish colour, and often thrown into folds or rugae (Warwick, R. et al. 1973) that run at right angles to the raphe and correspond to the dermal muscle fibres lying parallel to the surface of the skin (Robert Lich, J.R.; Howerton, L.W & Amin, M. 1979). It is beset with thinly scattered hairs, the roots of which are visible through the skin; they are provided with sebaceous glands, the secretion of which has a peculiar odour.

It also contains numerous sweat glands, pigment cells, and nerve endings responding to mechanical stimulation of the hairs and skin, and to variation in the circumambient temperature. Subcutaneous adipose tissue is lacking. (Warwick, R. et al. 1973).

The dartos, consisting of elastic fibres, connective tissue and smooth muscle fibres which are directed at right angles to the derma, lies immediately beneath the skin from which it is

virtually inseparable (Robert Lich, J.R. et al. 1979). It is supplied by sympathetic fibres that are carried in the perineal brancn of S_4. (Last, R.J. 1981).

It is composed of superficial and deep fibres, the superficial part is continuous around the scrotum, while its deeper part passes inward at the raphe to form the septum of the scrotum which divides the scrotum incompletely into two compartments (Gray, D.J. 1960).

The scrotal septum is composed of all the layers of the scrotal wall except the skin (Warwick, R. et al. 1973). Deep to the dartos muscle is the deep layer of the superficial fascia (Colles' fascia) attached behind to the posterior edge of the perineal membrane, at the sides to the ischio-pubic rami and bodies of the pubic bones, and in front continuous with the deep membranous layer of the superficial fascia of the anterior abdominal wall or Scarpa's fascia (Last R.J. 1981).

The scrotal size varies greatly because of the action of the dartos; however its action must not be confused with that of the cremasteric muscle. The cremasteric reflex affects the testicle and not the scrotum, it is occasioned by irritating the skin of the inner surface of the thigh, since the genitofemoral nerve (L_3) supplies both the skin of this area and the cremasteric muscle. (Robert Lich J.R. et al. 1979).

Blood Supply :

The front of the scrotum is supplied by the superficial and deep external pundendal arteries from the femoral artery, whereas the back is supplied by the scrotal branches of the internal pudendal artery. Branches of the testicular and cremasteric arteries which run in the spermatic cord help to supply the scrotum, the veins accompany the arteries (Gray D.J. 1960).

The subcutaneous plexuses of the scrotal vessels are dense and carry a substantial blood flow to effect loss of heat, arteriovenous anastomoses of a simple but large calibre type are prominent (Warwick, R. et al. 1973).

Lymph Drainage :

The lymphatic vessels of the scrotal halves freely anastomose with one-another and with those of the penis; they drain into the inguinal and femoral nodes. There are no connections between the lymphatics of the scrotum and the testes or their tunics. The scrotal lymphatics do not accompany the scrotal vessels. (Robert Lich J.R. et al. 1979).

Nerve Supply :

The anterior part of the scrotum is supplied by the ilioinguinal nerve and the genital branch of the genitofemoral nerve