SURGICAL MANAGEMENT OF INTER SEX

Submitted for Partial Fulfilment of the
Master Degree
In
[GENERAL SURGERY]

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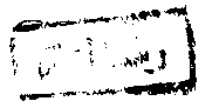

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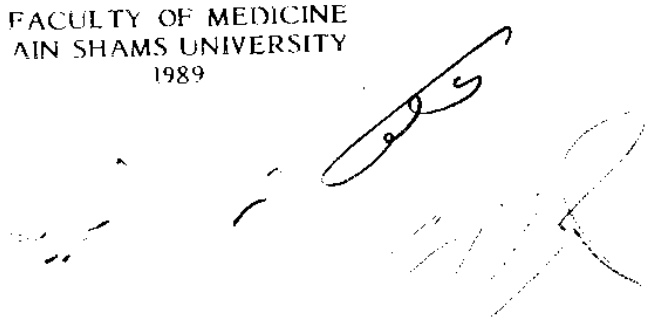

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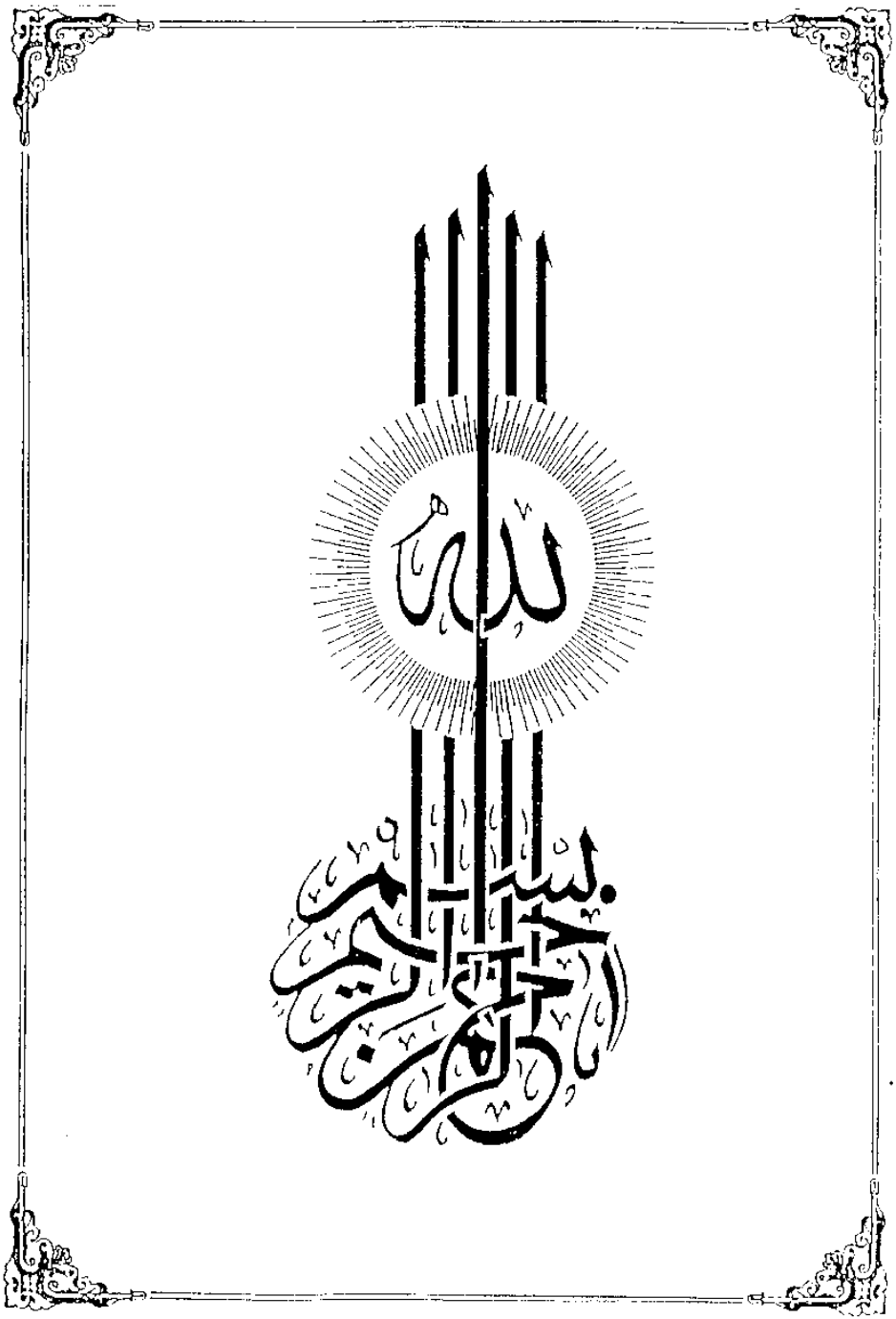

ACKNOWLEDGEMENT

I am greatly indebted to Prof. AHMED FAWZY BAHNASY. Professor of General and Pediatric Surgery, Faculty of medicine, Ain Shams University to him to express my thanks and deep gratitude.

I would like to express my deepest gratitude and cardinal thanks to Prof. **IBRAHIM BASSIONY** Assist. Professor of General Pediatric Surgery , Faculty of Medicine Ain Shams University for his valuable advice and great effort.

I am really thankful and greatful to all members of Pediatric Surgery Department for their sincer cooperation with me.

Finally I would like to express my love and obedynce to my father and my mother for their great effort and constant support.

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INTRODUCTION

Disorders of sexual development is one of the most challenging and intersting problems in pediatric urology.

In our culture, the differentiation between male and female is expected to be absolute and these terms are used to optimize opposites. Thus the patient and family who encounter this problem are often confused, afraid, unreasonably ashamed, guilty, and angry.

Surgeons and physicians dealing with those problems must offer understanding knowledge, precise and prompt diagnosis, optimal therapy and long term follow up and support.

Two other general problems must be considered:-

1. Intersex remains one of the last hidden diseases in our culture with minimal assurance.

2. Physicians, pediatricians, urologists and residents are often not optimaly informed because of lack of training, confusing and overwhelming literature or the relatively rare incidence of these various conditions.

There are ten major characteristics that determine sexual development and eventually sexual identity:-

1. Chromosomal composition.
2. Presence or absence of H.Y. antigen.
3. Gonadal structures and function.
4. Hormonal production, environment and response.
5. Internal genital ducts.
6. Development and differentiation of urogenital sinus.
7. External genitalia.
8. Secondary sexual characteristics and malignancy potential.
9. Sex of rearing.
10. Psychological cerebral and individual behavior factors.

These ten major factors are used as a frame work of analysis and understanding of normal as abnormal sexual development, and to diagnose clinical problems and implement therapy.

There is rapid advance in evaluation and treatment of the infants born with ambigious genitalia in the recent years.

Patho-physiology and appropriate medical and Surgical treatment is much advanced over just a few years ago.

Today it is generally possible to make an acurate diagnosis and plan appropriate gender assignment and Surgery promptly.

The focus of this Eassay will be on newborn evaluation and surgery that is carried out shortly after that.

An understanding of these concepts is based on an adequate understanding of normal as well as abnormal sexual differentiation.

Definition of intersex:

It is the clinical condition in which there is one or more inconsistencies, incongruities, ambiguities or significant, imperfections in one or more of the ten major factors.

Normally, the ten factors, of course, are concordant usually in intersex the inconsistency involves factors 1-8 inconsistencies or ambiguities involving only psychological, behavioral or perceptual factors are not normally classified as intersex.

* NORMAL SEXUAL DIFFERENTIATION.

NORMALSEXUAL DIFFERENTIATION

An understanding of the normal process of sexual differentiation is basic to any consideration of the abnormal.

Figure (I-1) shows in the schematic form the sequential but continuous events that take place prenatally and that when complemented by the events of puberty, define the sex of the person.

These processes can be considered in four stages:

1. The differentiation of a testis or an ovary from a common structure. (the indifferent gonad).

2. The development of male or female accessory sex organs from separate primordia. The Wolffian and Müllerian ducts, which coexist transiently in embryos of either sex.

3. The conversion of external genital analagen common to both sexes, the urogenital sinus and genital tubercle to the male or female form.

4. At puberty the appearance of secondary sex characteristics and gametogenesis, which together complete the preparation of the person for his procreative role.

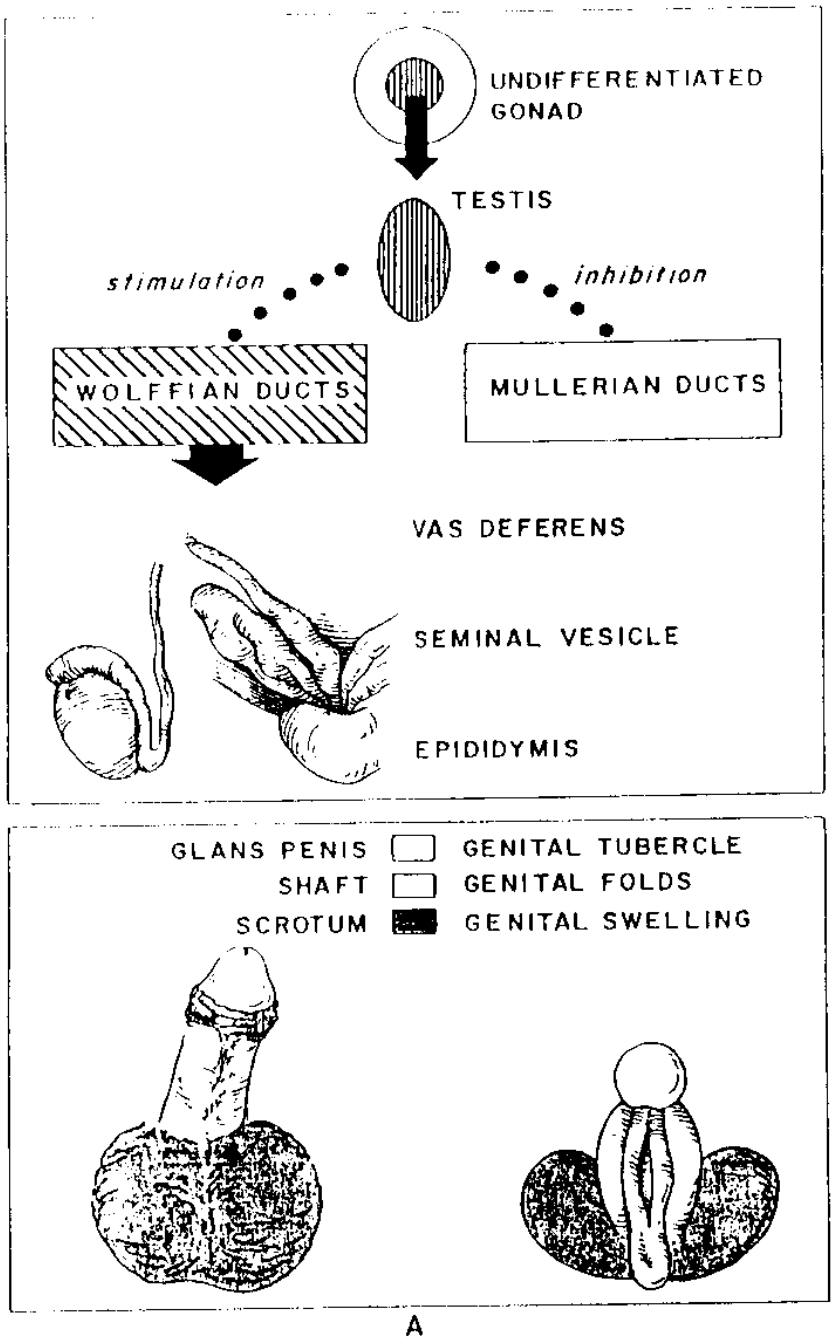

FIGURE 1–1. Schematic outline of normal male (*A*) and female (*B*) differentiation. Note that distinct internal duct primordia coexist in both sexes; that normal development involves growth of one system plus regression of the other; and that a testicular substance (s) plays the major role in this process. In contrast, the external genitalia develop in a continuous transformation from an anlage common to embryos of both sexes.

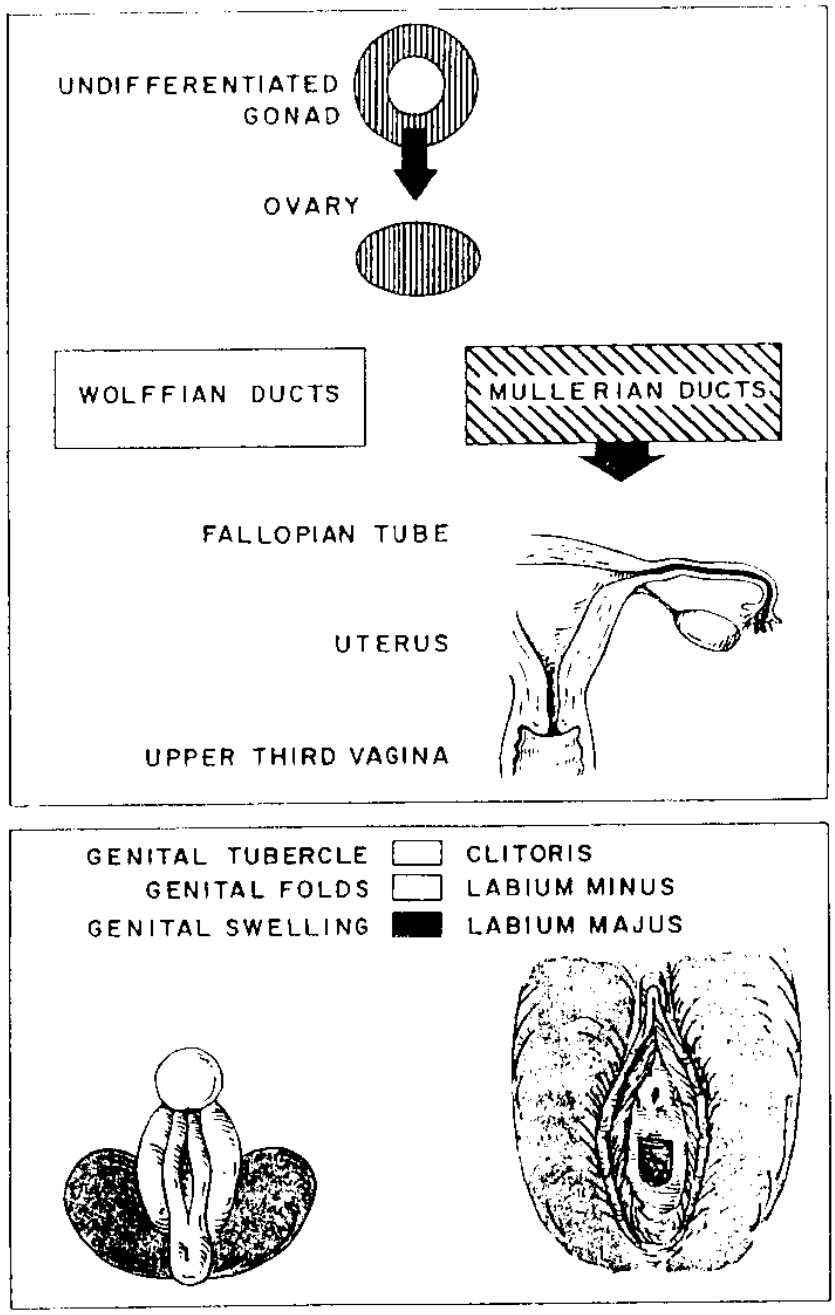

FIGURE I-1.

In the first stage a recognizable gonad appears as a thickening of the coelomic epithelium by the fourth week of embryonic life.

The primary sex cords grow down from coelomic epithelium into the mesenchymal cell mass of the gonad. While retaining attachment to their epithelial origins these cords anastomose distally to form the rete complex in which the germ cells will bedispersed.

Up to this point the process is the same in both sexes and the resulting structure , barely differentiated into a cortex and medulla is called "indifferent gonad."

The germ cells do not arise in the gonad but migrate to it from their origin in the entoderm of the yolk sac caudal to the embryonic disc. They can be histochemically identified by a unique alkaline phosphatase reaction that has made it possible to trace their course [Mckay, et al., 1953].

The arrival of these cells is requisite for further development of the gonad, Without them, there is gonadal agenesis.

The germ cells themselves are probably biopotential, or able to develop into sperm or eggs. Which course they take appears to depend on whether they settle in the part of the