# INTRODUCTION

The finding of ambiguous genetalia at birth is considered a neonatal emergency requiring the same attention as any other neonatal emergency**.** Intersex disorders are rare and complex, with the prevalence of 1 in 5500 individuals ***(Sax, 2002).***

Before the 8th week of gestation, the external genetalia of both male and female embryos are indistinguishable. External genetalia musculinization of the male embryo begins between the 9th and 12th week of gestation, which results from the systemic delivery of testosterone**.** By the 10th week, the genital tubercle grows to become the phallus and the labioscrotal folds fuse to form the scrotum ***(Walsh et al, 2004).***

Differentiation of the female external genetalia occurs between the 12th to the 16th week of gestation in absence of fetal androgen ***(Low and Husten, 2003).***

Error at any of the sexual developmental processes would lead to malformed external genetalia ***(Money, 1985).***

From the physiologic standpoint, ambiguous genetalia can result from;

-Gonadal defect that result in inadequate effects on the internal sexual ducts & the external phenotype.

-Extra gonadal problem such as virilization of the female fetus secondary to congenital adrenal hyperplasia.

-End organ Failure as in 5 alpha reductase deficiency and androgen insensitivity ***(Snyder et al., 2000).***

The most popular classification is one that is based on the gonadal sex;

1-True hermaphroditism; with ovarian and testicular gonadal tissue.

2-Male pseudohermaphroditism; an infant with an XY karyotype, but deficient musculinization of the external genetalia.

3-Female pseudohermaphroditism (46 XX); these infants have ovarian tissue and commonly have congenital adrenal hyperplasia.

4-Mixed gonadal dysgenesis; usually imperfectly formed gonads and retained mullarian structures, the typical karyotype is mosaic "45XO, 46XY".

5- Pure gonadal dysgenesis; 46XX or 46XY ***(Brunicardi et al., 2010).***

A baby born with ambiguous genetalia must have thoughtful evaluation to determine whether gender assignment can be made. It is important to minimize the emotional trauma to the family and later to the child ***(Grosfeld et al., 2006).***

Sex identification may be described in terms of at least 7 characteristics, the last two are psychological:

1-Sex chromosome.

2-Gonadal structure.

3-Morphology of the external genetalia.

4-Morphology of the internal genetalia.

5-Hormonal status.

6-Sex of rearing.

7-Gender role: by which is meant the sex that the individual considers himself or herself to be. ***(Wentz, 2002).***

Surgical reconstruction should be performed after a full work up and with the involvement of pediatric endocrinologists, pediatric plastic surgeons & ethicists with the family ***(Brunicardi et al., 2010).*** Current practice is to have genital reconstruction when the child still very young to attempt ensuring normal sexual development. It also aims to avoid embarrassment and anxiety for both the parents and the child .The operative procedures currently employed give reasonably good esthetic and functional results ***(Minto and Liao, 2003).***

Virilizing genital reconstruction include orchiopexy, hypospadius repair, gonadectomy, repair of the scrotum, removal of contradictory structures, insertion of artificial testis, phalloplasty and mastectomy ***(Kroovand and Glenn, 2000)***.The requisites for penile reconstruction include erection (or stiffness), sensibility, male fashion micturation and aesthetic appearance. Pre-operative testosterone is helpful in the repair of micropenis and indicates whether response to testosterone is possible ***(Grosfeld et al., 2006).***

For feminizing genital surgery, it is important to preserve an innervated but non-erectile clitoris, to separate vagina from the urogenital sinus and to create a normal vagina. In order to achieve these goals various genital reconstructive surgical techniques have evolved including clitroplasty either resection or reduction, labial surgery, vaginoplasty and gonadectomy in some instances ***(Alizai et al, 1999).*** However, it is easier to transform a sexually ambiguous individual into female than into male ***(Lobe and Woodall, 1995).***

Intersex abnormalities are caused by several pathophysiological entities, each of which must be systemically considered by pediatric surgeon working in callobration with pediatric endocrinologist with the goal of giving the child the best quality of life possible ***(Grosfeld et al., 2006).***

Many ethical and moral issues regarding intersex management have been mentioned in the media ***(Minto and Liao, 2003)***. The aim of informed consent requires that parents of the infant be given all relevant information about the diagnosis, treatment options and expected outcomes ***(Grosfeld et al., 2006).***