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# GENDER ASSIGNMENT FOR CHILDREN WITH INTERSEX PROBLEMS: AN EGYPTIAN PERSPECTIVE

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***Gender identity is a complex process of differentiation that is affected by numerous variables. Three-hundred and fourteen cases of intersex were diagnosed and surgically managed between 1986 and 2000. Cases of male pseudohermaphroditism (MPH) formed the major group (59 %) ; 27 % had female pseudohermaphroditism (FPH) . Mixed gonadal dysgenesis (MGD) and true hermaphroditism (TH) were the most common variants of the abnormal gonadal differentiation group ( 13 1and 18 cases respectively ) . Congenital adrenal hyperplasia (CAH) due to 21-hydroxylase deficiency constituted the major part of FPH (n =76 ) . Fifty-three per cent of patients were raised as males at the time of presentation. Forty-three patients (13 % ) reassigned their sex after diagnosis of their disease : 31 cases (10 % ) changed their sex from females to males, the majority was due to either 5 a-reductase deficiency or partial androgen insensitivity syndrome(PAIS), (16 and 14 cases respectively). Twelve cases (4 % ) reversed their sex from males to females, CAH was the cause in 10 patients . Ninety-eight cases (31 % ) were considered as " sex misassignment" , while 41 cases (13% ) retained a "discordant sex" even after their sex-counseling. Only 16 cases (5% ) were diagnosed in the first three months of life which reflected the major defect in early diagnosis of such conditions in the community; a problem which was also proved by the large number of cases diagnosed at adolescence (n = 9). Familial occurrence was reported in 36 patients. Gonadoblastoma was detected in 2 cases of PAIS and TH. Several social, cultural, and religious factors related to the this local area influencing the gender assignment of such patients were elicited. Several reconstructive surgical procedures were performed after reassignment with excision of all discordant tissues e.g. gonadectomy (n=131) and hysterectomy (n=33).This included a wide variety of either feminizing genitoplasty e.g. clitoroplasty (n=104), vaginoplasty (n =110) and labioplasty (n =84) or male genital reconstruction such as urethroplasty (n=160), scrotoplasty (n=78) ,orchiopexy (n=40), etc…The different factors influencing the decision of rearing in this community should be properly comprehended so as to avoid the problems of mismanagement of intersex conditions with early screening of such cases.***

## INTRODUCTION

Gender assignment,the process by which the sex of rearing in a newborn with ambiguous genitalia is decided, must be considered a psychological emergency. It is usually a multidisciplinary team effort carried out against time in terms of days, even hours [1]. Comprehensive and rapid evaluation is necessary to assign the appropriate gender to organize any necessary medical, surgical and psychological intervention. Diagnostic uncertainty can delay or lead to inappropriate gender identity, with a need for reassignment at a later stage [2].

The goal cannot be simply regarded as restoring the normal anatomy or identifying a common link between genetic complement, gonadal tissue and appearance of the external genitalia, which in many situations is impossible. Instead the main objective remains the rehabilitation of one of the most important aspects of the human life: its sexuality. Rearing the child in a sex discordant with the anatomic and functional possibilities to achieve a normal erotic and coital performance may render life an ordeal [1].

The objective of the physician in the management of patients with ambiguous genitalia is to establish a diagnosis and to assign a sex of rearing that is most

compatible with a well–adjusted life and sex adequacy. Once the sex of rearing is assigned, the gender role is reinforced by the use of appropriate surgical, hormonal or psychologic measures [3].

The purpose of this study is to review the Egyptian experience with intersex patients in Cairo university: their incidence , clinical variants , different patterns of presentations , methods of diagnosis and surgical management with special emphasis to the different ethical, social, cultural and religious aspects influencing the gender assignment of these cases in our local community.

## PATIENTS AND METHODS

Three-hundred and fourteen cases presenting with intersex problems were studied in the Pediatric Surgical Division , Cairo University over a period of 14 years , between 1986 and 2000 . Their ages at presentation ranged between 10 days and 20 years with a mean of 5.5 years .

History taking included the chronological age, sex of rearing , consanguinity and any similar conditions in the family . Genital ambiguity was evaluated and described as regards the degree of differentiation of the labioscrotal folds , the size of the phallus , the site of external urethral meatus and the gonads on both sides . Puberty was assessed using Tanner and Whitehouse classification [4]. Prader staging was used to evaluate the degree of virilization of the external genitalia in female pseudohermaphroditism [5].

Karyotype examination was performed using the G banding technique. Chromosomal study for sex-determining region Y gene (SRY) was performed in some cases of true hermaphroditism (TH). Hormonal assessment included determination of the serum gonadotropins (FSH and LH), testosterone (T) and its precursors, dihydrotestosterone (DHT) and T / DHT ratio, progesterone, 17 hydroxyprogesterone, and 11-deoxycortisol. Dynamic tests were done for cases of MPH including the short and prolonged HCG stimulation tests to assess androgen biosynthesis and the ACTH stimulation test to exclude 3B-hydroxysteroid defects (3B-OHSD) .

Radiologic assessment of the internal genital organs was essentially achieved by genitography and ultrasonography, computed tomographic scan was uncommonly needed.

Surgical diagnostic procedures were performed in certain cases: e.g. exploratory laparotomy, laparoscopy, gonadal biopsy etc.. . Fibroblast culture in skin biopsy from the gonad was performed in some cases of MPH to assess the DHT receptors and the 5 α-reductase activity.

After being properly diagnosed, the patients were then grouped according to the Grumbach and Conte criteria [6].Unclassified local genital pathologies e.g. isolated hypospadias , cloacal extrophy, etc… were excluded from the study. The parents were then counseled about the nature of their child's abnormality and guided about the appropriate sex of rearing. Care has been taken to reassure them that their child will be definitely either a boy or a girl and not something in 'between". All other members of the staff were asked not to express an opinion about the child's sex until a clear decision has been taken. The expected child's future sexual function and fertility were clarified to the parents. Cultural, traditional and religious factors related to our community were considered with great care when counseling parents about the recommended sex of rearing.

Patients who presented with discordance between the genotype and sex of rearing were labeled as "sex misassignment". Those who retained their sex of rearing inappropriate with his expected future anatomical and functional possibilities to achieve a normal sexual life were defined as having "discordant sex". Once this has been decided, treatment was then organized including surgical correction of the genitalia to concord with the assigned sex either by female genitoplasty e.g. clitoroplasty, vaginoplasty, labioplasty etc… or through male genital reconstructive procedures e.g. urethroplasty, scrotoplasty, orchiopexy etc... Hormonal supplementation if needed and psychological support for adolescents were then completed.

Follow-up of the patients which ranged between 6 months and 8 years was performed to evaluate the results of their corrective surgery and the general behavior, acceptance and adaptation of the patients and/or their parents to the assigned sex. Traditional, social, cultural and religious factors related to the Egyptian community that have strong influences on the decision of sex rearing and further management of intersex cases were then carefully analyzed and discussed.

## RESULTS

The records of 314 patients with intersex problems were studied and analyzed . Cases of MPH formed the major group (n=186), (Table 1). Among the patients with abnormal gonadal differentiation (n=45), those with mixed gonadal dysgenesis (n=13) and true hermaphroditism (n=18) were the most common pathologic types, (Fig.1,2) . Almost all of the cases of FPH (n=83) were due to congenital adrenal hyperplasia resulting from 21-hydroxylase enzymatic deficiency (n=76) .

Early diagnosis – within the first three months of life- was only achieved in 16 cases, the majority of them were due to salt losing 21- hydroxylase deficiency (n=10) . A

relatively large number of cases were diagnosed in the post pubertal and adolescent periods (n=49), (Fig. 3). Most of these cases were either misdiagnosed males with CAH (n=8), (Fig. 4); or females with pubertal virilization due to 5 α-reductase or PAIS (9and 11 respectively), (Fig.5). Adolescents with abnormal gonadal differentiation were mainly either due to phenotypic females presenting with primary amenorrhea due to Turner's, syndrome or pure gonadal dysgenesis (3 and 4 respectively) or due to phenotypic males with true hermaphroditism having mild degrees of hypospadias and unilateral impalpable gonad (n=5), (Fig. 6) .

In general, at the time of presentation, the number of patients with intersex problems reared as males was slightly larger than those reared as females, 166 and 148 respectively. The misassigned patients- i.e. their sex of rearing was discordant with their genotype – were 98 cases (31 %) .

After completion of all investigations and the accurate diagnosis has been established and clarified to the parents, and after all measures were tried and directed toward restoring the concordance of the phenotype with the appearance of the genitalia and the sex of rearing, 43 patients have reassigned their sex so as to achieve a better social and sexual lives in their future. Those who have changed their sex to males (n=31) were exceeding those reassigned to females (n=12) . Forty-one patients remained with a "discordant sex" – i.e. their accepted sex of rearing was not the most appropriate and suitable to the expected anatomical and functional possibilities of their future sexual life . Most of these patients were reared as males (n=30).

Almost all of the cases of true hermaphroditism– had masculinized external genitalia (grade III , IV , and V). Only one case of unilateral type (ovary on one side) was grade II , but the clitoris was greatly enlarged and the child retained his sex of rearing as a male . All the cases of true hermaphroditism were reared as males except one case of unilateral type: it was a sib to a normal non-identical male twin that reassigned his sex as a female at the age of 1.5 years , (Table 2) .

On the other hand , cases of gonadal dysgenesis had their external genitalia more toward the feminine appearance (Grade II), although their clitoris was enlarged in most of the cases. More than 60 % (n=8/13) were assigned as males .

In all cases of Turner's syndrome (n=3) and pure gonadal dysgenesis, (n=4), the external genitalia were totally feminine. All these cases presented late with primary amenorrhea. Bilateral gonadectomy with excision of the streak gonads was performed in only one case of pure gonadal dysgenesis with a 46 XY chromosomal pattern. Estrogen replacement therapy was given for all these cases .

Orchiopexy, correction of the hypospadias and hysterectomy were performed in cases of dysgenetic male pseudohermaphroditism (n=6) as all of them retained their sex of rearing as males.

Among all the studied cases, familial occurrence was reported in 36 patients ,the highest incidence occurred in cases of PAIS (n=17) and 5 α- reductase deficiency (n=10), (Fig. 7). Other uncommon familial cases resulted with 21-hydrxylase deficiency (n=4), leydig cell hypoplasia or deficiency (n =3) , and 3-B hydroxylase deficiency (n = 2). Although one sib of a non identical twin was reported to have a unilateral true hermaphroditism , yet the other sib was normal .

Several techniques have been performed for reconstruction of the male external genitalia such as urethroplasty repairs, orchiopexy , scrotoplasty (either for bifid scrotum or associated penoscrotal transposition) , subcutaneous excision of abnormal breasts and insertion of testicular prosthesis which was available only in two cases, (Table 3) . In two adolescents of PAIS and a case of true hermaphroditism aged 16 and 17 years respectively, gonadoblastoma was detected in the excised gonads.

Mild degrees of hypospadias were uncommon. These were corrected by either urethral advancement, Snodgrass or Mathieu's techniques. The severe forms were reconstructed as a one-stage repair using an onlay graft, a transverse preputial or double-faced island flaps. Long urethral defects were repaired either with a bladder or buccal mucosal graft or by using a combined Duplay's and transverse preputial island flaps .

Female genitoplasty included clitoroplasty, vaginoplasty and labioplasty . Early in the study large-sized clitoris was repaired by either total clitrorectomy (n=6) or by resection of its shaft with wedge-excision glanuloplasty (n=28) . Later on, clitoroplasty was performed by a subtunical resection of its hypertrophied shaft with preservation of the dorsal neurovascular bundle to the glans with subtotal de-epithelialization and partial concealment of the glans (n=70), (Fig. 8) .

In the majority of the cases , U-shaped perineal flap vaginoplasty was sufficient to widen the introitus and exteriorize the vaginal orifice (n=70) . Mild forms of labial fusion were treated by simple cut-back where as in severely virilized forms , vaginoplasty was performed mostly via a vaginal pull-through (n=18).Only in two cases , a colonic patch was used to repair the high vaginal defect . Recently, urogenital sinus mobilization was performed in 6 cases. Labioplasty (n=86) was then performed using the skin covering the clitoris after

reconstructing the mons pubis. This completed the feminine appearance of the external genitalia. Excision of all contradictory internal genitalia to the assigned sex e.g. gonadectomy and / or a hysterectomy were performed to complete the surgical reconstruction of the sexual ambiguity to be concordant with the assigned sex .

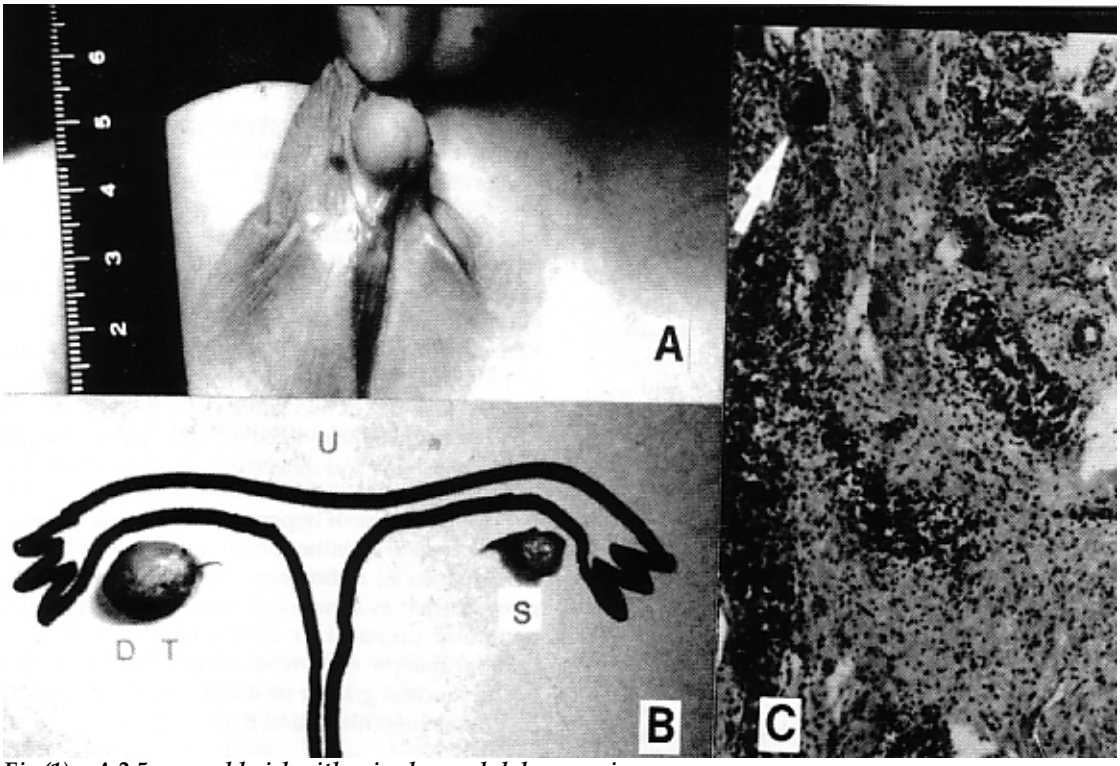


*Fig (1) : A 2.5 year-old girl with mixed gonadal dysgenesis :*

- *A- Appearance of the external genitalia showing enlarged ,penis-like clitoris and non-fused labioscrotal folds.*
- *B- Bilateral gonadectomy with a left streak gonad (S), and a dysgenetic testis (DT).*
- *C- Histopathologic picture of the dysgenetic testis showing wavy ovarian-like stroma with ghosts of seminiferous tubules and " intratubular eggs"(arrow), (H &E x40).*

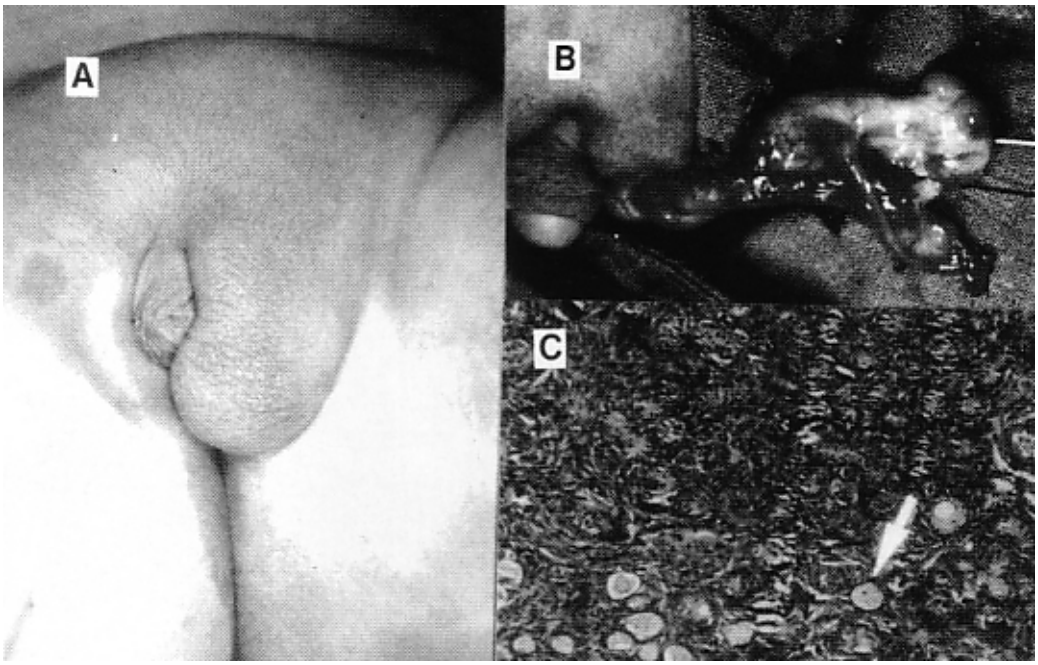


*Fig (2) : A 2 year-old male child with true hermaphroditism :*

- *A- Appearance of the external genitalia showing small penis, left scrotal gonad and undescended right gonad.*
- *B- Exploration of left gonad showing adjacent fallopian tube having short fimbrial ends.*
- *C- Histopathology showing both ovarian tissue with follicles ( arrow) and wavy stroma; and testicular tissue with seminiferous tubules in the same histological section ( H&E x100).*

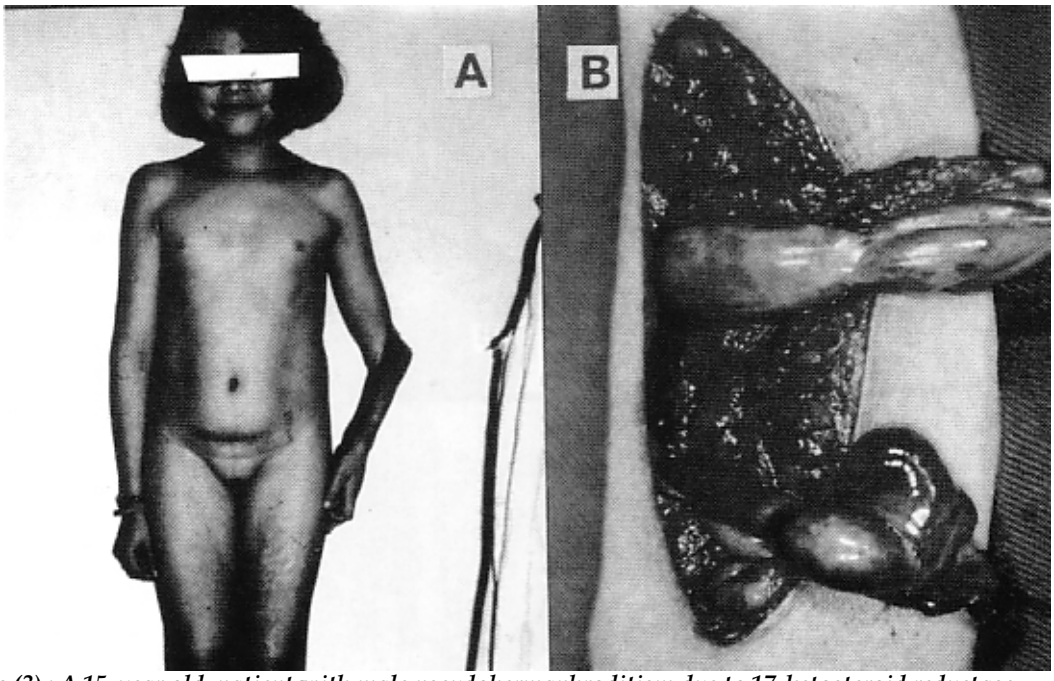


*Fig (3) : A 15-year old patient with male pseudohermaphroditism due to 17-ketosteroid reductase , reared as a female.*

- *A- Female phenotype.*
- *B- Exploration of both inguinal canals showing bilateral testes.*

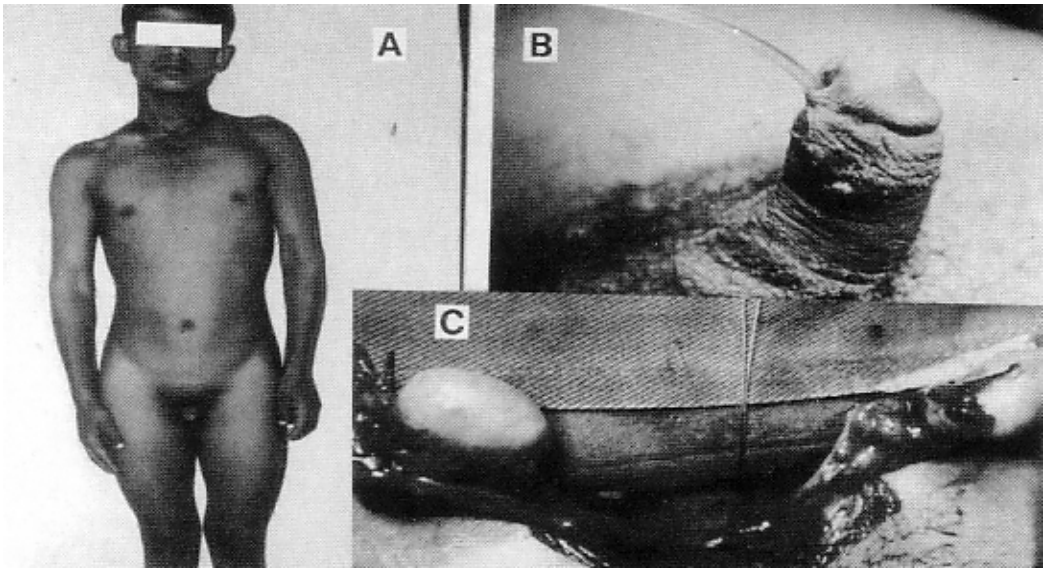


*Fig (4) : A 14-year old patient with female pseudohermaphroditism due to congenital adrenal hyperplasia – ( 21-hydroxylase deficiency ) reared as a male:*

- *A- Male phenotype.*
- *B- External genitalia showing mild degree of hypospadias (coronal) and bilateral undescended gonads.*
- *C- Exploration revealed bilateral normal ovaries and a midline uterus.*

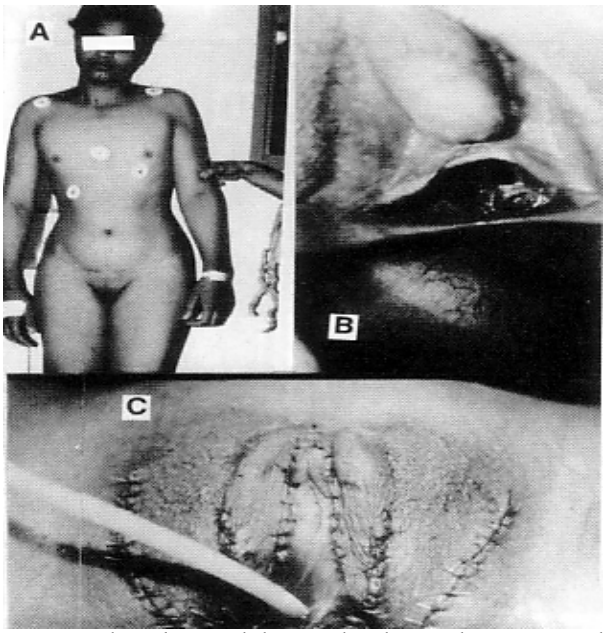


*Fig (5) : A 16 year-old patient with male pseudohermaphroditism due to 5 a-reductase deficiency :*

- *A- Female phenotype.*
- *B- Appearance of external genitalia showing enlarged clitoris and both separate urethral and vaginal openings.*
- *C- External genitalia after female genitoplasty : clitoroplasty, labioplasty and introitoplasty.*

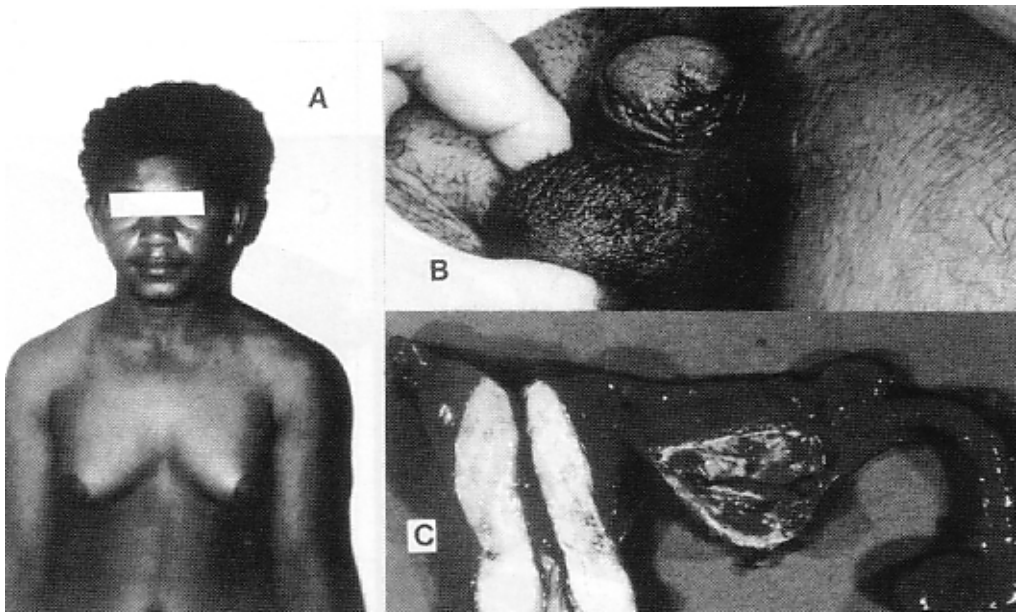


*Fig (6) : A 17 year-old male with true hermaphroditism :*

- *A- Phenotypic male with gynecomastia.*
- *B- The external genitalia showing a good phallus, mild degree of hypospadias (coronal), scrotal right gonad and undescended left gonad.*
- *C- Internal genitalia after excision of uterus and left ovary(bisected), with the adjacent fallopian tube.*

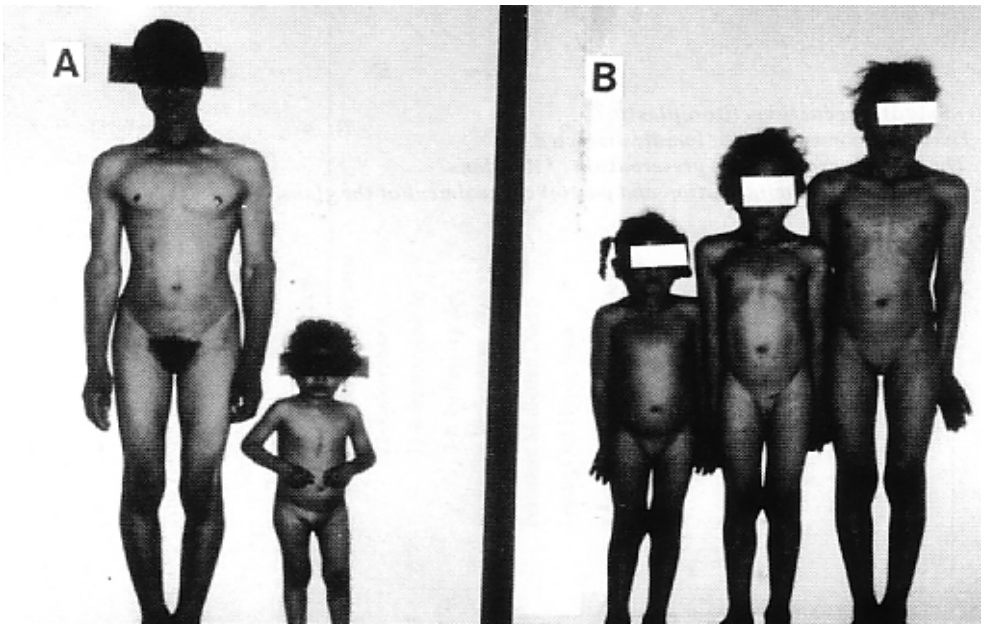


*Fig ( 7 ) : Familial cases of intersex :*

- *A- Two cousins with 5 a- reductase deficiency syndrome : a virilized male aged 16 year-old and a female child aged 1.5 years.*
- *B- Three sisters with male pseudohermaphroditism due to partial androgen insensitivity syndrome.*

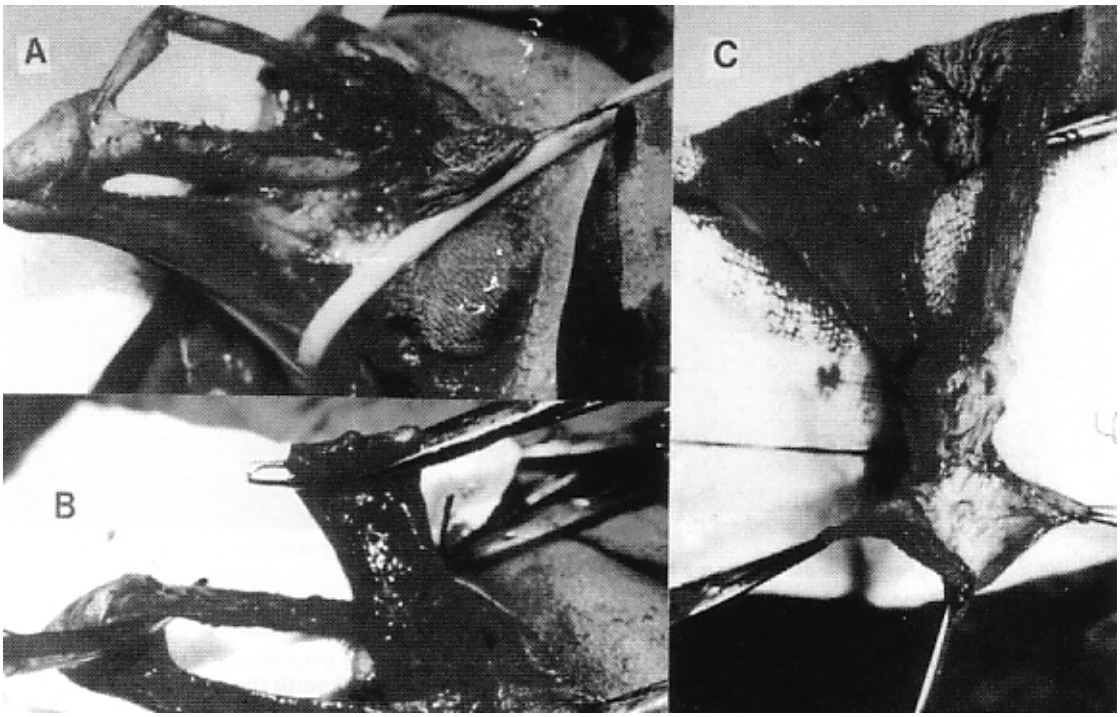


*Fig (8) : Subtotal reduction clitoroplasty:*
- *A- The dorsal neurovascular bundle dissected.*
- *B- The shaft is excised with preservation of the glans.*
- *C- Subtotal de-epithelialization and partial concealment of the glans.*

| CLASSIFICATION | TOTAL | | Type | AGE AT PRESENTATION | | | | | | GENDER AT PRESENT. | | RE-ASSIGN. | | MISASSIGNMENT | DISCORDANT SEX | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | N | % | | N | 0-3 m | 3m - 2y | 2 - 7y | 7 - 12 y | 12 -20 y | M | F | M to F | F to M | | M | F |
| ABNORMAL GONADAL DIFFERENTIATION | 45 | 14 | Turner's S. | 3 | | | | | 3 | | 3 | | | | | |
| | | | Pure Gonadal D. | 4 | | | | | 4 | | 4 | | | | | |
| | | | Mixed Gonadal D. | 13 | | 6 | 6 | 1 | | 8 | 5 | 1 | | 5 | | 1 |
| | | | Chimerism | 1 | | 1 | | | | 1 | | | | | | |
| | | | True Hermaphroditism | 18 | | | | | | | | | | 12 | 10 | |
| | | | *Bilateral 2* | | | 1 | 1 | | | 2 | | | | | | |
| | | | *Unilateral 1 1* | | | 3 | 2 | 2 | 4 | 11 | | 1 | | | | |
| | | | *Lateral 5* | | 1 | 1 | 2 | | 1 | 4 | 1 | | | | | |
| | | | Dysgenetic Male Pseudoherm. | 6 | | 1 | 3 | 1 | 1 | 6 | | | | | | |
| FEMALE PSEUDO-HERMAPHRODITISM | 83 | 27 | Congenital Adrenal Hyperplasia | 78 | | | | | | | | | | | | |
| | | | *21- hydroxylase def. 76* | | 10 | 14 | 29 | 15 | 8 | 18 | 58 | 10 | | 18 | 8` | |
| | | | *11-B hydroxylase def. 1* | | | | | 1 | | | 1 | | | | | |
| | | | *3-B hydroxylase def.* 1 | | | | 1 | | | | 1 | | | | | |
| | | | Idiopathic Virilization | 5 | 1 | 2 | 2 | | | 1 | 4 | | | 1 | 1 | |
| MALE PSEUDO-HERMAPHRODITISM | 186 | 59 | Persistent Mullerian Duct S | 2 | | 1 | 1 | | | 2 | | | | | | |
| | | | Leydig Cell Hypoplasia | 9 | | 1 | 1 | 5 | 2 | | 9 | | | | | |
| | | | Testosterone Biosynthesis Defect | 5 | | | | | | | | | | | | |
| | | | *3-B Hydroxysteroid D.def* 4 | | | 1 | 2 | 1 | | 3 | 1 | | 1 | 1 | | |
| | | | *17ketosteroid R. def* 1 | | | | | | 1 | | 1 | | | 1 | | |
| | | | Steroid 5 α-reductase def. | 82 | | 14 | 20 | 39 | 9 | 56 | 26 | | 16 | 26 | | 10 |
| | | | Androgen Insensitivity S. | 74 | | | | | | | | | | | | |
| | | | *- complete 13* | | | | 1 | 7 | 5 | | 13 | | | 13 | | |
| | | | *- partial 61* | | 4 | 10 | 22 | 14 | 11 | 40 | 21 | | 14 | 21 | 10 | |
| | | | Emb.Testicular Regression S | 14 | | 5 | 8 | 1 | | 14 | | | | | 1 | |
| TOTAL | 314 | | | 314 | 16 | 61 | 101 | 87 | 49 | 166 | 148 | 12 | 31 | 98 | 30 | 11 |
| % | | 100 | | 100 | 5 | 19 | 32 | 28 | 16 | 53 | 47 | 4 | 10 | 31 | 10 | 4 |

**Table (1) :** ***Distribution of the intersex disorders, their age at presentation and their gender before and after diagnosis and management.***

| TYPE | N | GENDER AT PRESENT. | AGE | EXTERNAL GENITALIA | | | | | LUCK'S CLASS. | KARYOTYPE | TESTOSTERONE ng/ml | INTERNAL GENITALIA | | | SEX ASSIGNMENT |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | L-S FOLDS | PHALLUS (cm) | URETHRA | GONAD | | | | | GONAD | | UTERUS | |
| | | | | | | | RT | LT | | | | RT | LT | | |
| MIXED GONADAL DYSGENESIS | 1 | F | 1.5 y | NF | 2.2 | P | Impl | Impl | II | 45X/46XY | 0.9 | ST | DT | + | F |
| | 2 | F | 1.5 y | Fu | 3 | P | Impl | Impl | IV | 45X/46XY | 1.8 | ST | DT | + | F |
| | 3 | M | 1 y | Fu | 2.5 | PS | Impl | Ing | IV | 45X/46XY | 2.2 | ST | DT | + | M |
| | 4 | M | 5 y | PF | 2.5 | P | Impl | Impl | III | 45X/46XY | 1.8 | DT | ST | + | M |
| | 5 | M | 1.5 y | NF | 1.8 | P | Impl | Impl | II | 45X/46XY | 0.8 | ST | DT | + | M |
| | 6 | M | 1 y | NF | 2.2 | P | Impl | Impl | II | 45X/46XY | 2..3 | ST | DT | + | M |
| | 7 | M | 8 y | S | 2.8 | PS | Ing | Impl | V | 46XY | 2.8 | DT | ST | + | M |
| | 8 | F | 3 y | PF | 2 | P | Impl | Impl | III | 45X/46XY | 1.1 | ST | DT | + | F |
| | 9 | M | 1.5 y | NF | 2 | P | Impl | Impl | II | 45X/46XY | O.8 | DT | ST | + | F |
| | 10 | M | 2 y | Fu | 2.2 | PS | Impl | Impl | IV | 45X/46XY | 1.8 | DT | ST | + | M |
| | 11 | F | 3 y | NF | 1.8 | P | Impl | Impl | II | 45X/46XY | 0.8 | ST | DT | + | F |
| | 12 | F | 3.5 y | NF | 3 | P | Impl | Impl | II | 45X/46XY | 1.8 | ST | DT | Bicor | F |
| | 13 | M | 5 y | PF | 2.8 | P | Impl | Impl | II | 46XY | 1.5 | ST | DT | + | M |
| TRUE HERMAPHRODITISM | 1 | M | 7 m | PF | 2.3 | S | Impl | S | III | 46XX | O.1 | OT | T | - | M |
| | 2 | M | 8.5 y | PF | 3..5 | PS | S | Impl | III | 46XX | O.1 | T | OT | HU | M |
| | 3 | M | 14.5 y | S | 6 | S | Impl | S | V | 46XX | O.26 | OT | T | + | M |
| | 4 | M | 2 y | Fu | 4 | S | Impl | Impl | IV | 46XX/46XY | 0.1 | T | O | + | M |
| | 5 | M | 17.5 y | S | 6 | C | S | Impl | VI | 46XY | 1.8 | T | OT | + | M |
| | 6 | M | 8 y | Fu | 5 | S | Impl | Impl | IV | 46XX | O.17 | OT | O | + | M |
| | 7 | M | 3 y | PF | 5 | S | Impl | Impl | III | 46XX | 0.1 | OT | T | - | M |
| | 8 | M | 1 y | S | 4 | P | Impl | Impl | V | 46XX | 0.02 | OT | OT | + | M |
| | 9 | M | 3 y | PF | 4.5 | S | Impl | S | III | 46XX | 1.3 | O | T | + | M |
| | 10 | M | 17 y | PF | 7.5 | P | Impl | Impl | III | 46XX | 77.5 | O | DYS | + | M |
| | 11 | M | 3 y | S | 4 | PS | Impl | Impl | V | 46XX | 0.9 | OT | OT | + | M |
| | 12 | M | 1 y | PF | 2.4 | PS | Impl | S | III | 46XX | 1.8 | O | OT | + | F |
| | 13 | M | 5.5 y | PF | 3.8 | S | S | Impl | III | 46XX | 1.4 | OT | O | + | M |
| | 14 | M | 16 y | S | 8 | PS | S | S | IV | 46XY | 9.6 | OT | T | - | M |
| | 15 | M | 1 m | PF | 2.8 | S | Impl | S | III | 46XX | 1.2 | O | T | + | M |
| | 16 | M | 17 y | S | 6.5 | C | Impl | S | VI | 46XY | 8.5 | O | T | + | M |
| | 17 | M | 1.5 y | PF | 2.5 | P | Impl | S | III | 46XX/46XY | 1.8 | O | T | + | M |
| | 18 | M | 1 y | NF | 1.5 | P | Impl | S | II | 46XX | 0.9 | O | OT | HU | M |

**Table (2) :** ***Clinical presentations, diagnosis and sex assignment of cases of mixed gonadal dysgenesis and true hermaphroditism.***
***(M = male, F = female,L-S = labioscrotal, NF = non-fused, Fu = fused , PF = partially fused, S = scrotal, P = perineal, PS = penoscrotal, C = coronal, ST = streak, DT = dysgenetic testis, OT= ovotestis, T = testis, O = ovary, Dys = dysgerminoma, Bicor = bicornuate, HU = hemiuterus).***

| *SURGICAL PROCEDURE* | | *NUMBER OF CASES* |
|---|---|---|
| **GONADECTOMY** | | 131 |
| - Streak gonad | **21** | |
| - Ovotestis | **14** | |
| - Oophorectomy | **20** | |
| - Orchiectomy | **76** | |
| HYSTERECTOMY | | 33 |
| **URETHROPLASTY** | | 160 |
| - Urethral advancement | **3** | |
| - Tubularized-incised urethral plate | **6** | |
| - Mathieu's technique | **15** | |
| -Transverse preputial island flap | **20** | |
| - Onlay preputial flap | **14** | |
| - Double-faced preputial flap | **24** | |
| - Simple Duplay's technique | **12** | |
| - Combined Duplay's / T. preputial flap | **48** | |
| - Bladder mucosal graft | **13** | |
| - Buccal mucosal graft | **5** | |
| **SCROTOPLASTY** | | 78 |
| **ORCHIOPEXY** | | 40 |
| **SUBCUTANEOUS EXCISION OF ABNORMAL BREAST** | | 4 |
| **EXCISION OF GONADAL TUMOR** | | 2 |
| **INSERTION OF TESTICULAR PROSTHESIS** | | 2 |
| **CLITORAL SURGERY** | | 104 |
| - Total clitoridectomy | **6** | |
| - Clitoroplasty | **98** | |
| **VAGINOPLASTY** | | 110 |
| - Simple cut-back | **12** | |
| - Flap vaginoplasty | **70** | |
| - Vaginal pull-through | **18** | |
| - Colo-vaginoplasty | **2** | |
| - Urogenital-sinus mobilization | **6** | |
| **LABIOPLASTY** | | 84 |

**Table (3)***: Surgical procedures performed for genital reconstruction of the intersex cases*

## DISCUSSION

Ambiguity of the genitalia in the newborns and children still remains a poorly understood subject and even during the postgraduate teaching programs, very little attention has been paid to make this subject popular. There is also no chapter "without tears " on the subject for the proper management and to prevent the agony of the parents resulting from the ignorance and the social stigmata [7]. The purpose of this study was to project the practical management of children with intersex disorders and to discuss the different physical, surgical, social, cultural and religious factors influencing the gender assignment based on an Egyptian experience from the Pediatric Surgical Division at Cairo University.

In the present study, 314 patients were diagnosed and managed over a period of 14 years. The MPH constituted the majority of the cases(59%). Those reared as males at the time of presentation were 53 % (n=166). These results were similar to those of Krstic et al from Belgrade who reviewed 84 children with ambiguous genitalia between 1986 and 1993. MPH formed 58 % and those raised as males were 57%[8] .On the contrary, Newman et al reported 79 patients assigned as females among 91 children studied in a 25-year period [9]. Coran and Polly reviewed 69 children between 1974 and 1989, only 10 patients were male pseudohermaphroditism, 32 had congenital adrenal hyperplasia , 10 cases of mixed gonadal dysgenesis and 3 had true hermaphroditism [10] .

In the present study, classification of the age distribution of the cases was selected so as to reflect the concept of managing the intersex conditions in our community. Only 16 cases (5 %) were diagnosed within the first 3 months of age which denotes that there is a severe defect in early diagnosis and dealing of such cases.This could be attributed to lack of awareness of the primary care physicians and nurses . Forty-nine patients (16%) were diagnosed at the age of adolescence. Most of the cases (n=20) were due to the appearance of virilization in misassigned females due to either 5 α-reductase deficiency or PAIS . Less commonly this was attributed to either a primary amenorrhea in assigned females (n=12) due to gonadal dysgenesis or complete androgen insensitivity syndrome or during investigating severely virilized males with CAH for impalpable testes (n=8) .

The American Academy of Pediatrics recommended the repair of the external genitalia and hypospadias before the age of 30 months for the following reasons: (1) awareness of the different sexes as well as the presence of a physical deformity occur at that time , (2) socialization of boys of that age creates situations in which comparison of genitalia occurs e.g. in nursery schools; (3) from the age of 30 months to at least 5.5 years , the boys' fears of physical harm are significant and may be exacerbated by surgery. It is also stated that a child who has attained the capacity for operational thought (7 years of age or older) will be able to understand the casualty in a more adult-like fashion but still may unconsciously associate surgery with punishment [13].

In the present study, more than 75% (n=237) were diagnosed and managed later than the optimal time for their surgical reconstruction (30 months) which may be due to poverty or the wrong thoughts of some pediatricians that the surgery is better to be performed at the ages older than 6 years. This might have exposed the children to some psychological trauma.

The limited number of cases of Turner's syndrome and pure gonadal dysgenesis (n=7) may be due to their late presentation by primary amenorrhea, without genital ambiguity. Most of the other cases may have been referred to adult gynecologists. All these cases retained their sex of rearing as females as they were phenotypically completely females.

As it is proved that the presence of even a small part of a Y chromosome greatly increases the risk of gonadal malignancy in a person with dysgenetic gonads [14,15], bilateral gonadectomy was performed in one case of 46 XY complete gonadal dysgenesis and excision of the streak gonads in all cases of partial gonadal dysgenesis.

It is stated that chimerism involving the Y chromosome is quite rare [16]. Only one case was recorded in this study (0.31%) being raised as male with reconstruction of the urethra and fixation of the testes.

In the current study; 13 cases (4 %) of MGD were recorded. Although all of them had a well-sized phallus, yet 5 cases were raised as females, the others were reared as males due to the presence of unilateral palpable scrotal gonad. As has been stated in literature [1] , most of the cases had a karyotype of 45X/46XY, only two cases were 46XY. Fertility was not a decisive issue in gender assignment of these cases as it is reported that post pubertal testicles uniformly have absence of spermatogenesis.Izquierdo and Glassberg stated that in the presence of intra-abdominal testicle and poorly virilized genitalia, the decision is straightforward in the female direction. In this case, bilateral gonadectomy is advisable in the newborn. Neonates with adequate phallic size and a scrotal testicle can be considered for male assignment [1].

True hermaphroditism is the least common and least understood variant of intersex disorders. Luks et al stated that out of 528 cases reported in the literature since 1899, only 114 have provided sufficient information correlating the appearance of the external genitalia, age at diagnosis, karyotype, findings at laparotomy and sex of rearing [17].

Krstic et al treated 97 patients with intersex disorders over a period of 10 years, there were 4 with true hermaphroditism [18]. The current study added 18 new cases to the literature with all data concerning their clinical and surgical managements.

Most of the authors agreed that the decision of sex assignment in infants with TH should be based solely upon the functional capability of the external genitalia: If the infant has an inadequate phallus, the child should be reared as a male regardless of the gonadal sex .In the patient with both a vagina and well developed phallus, the sex of rearing should be based on findings at exploratory laparotomy. If there is a normal testis that can be placed in the scrotum, the patient should be reared as a male. However, if the patient has normal Mullerian duct structures, a normal ovary on one side with a testis or ovotestis on the other side, the patient should be reared as a female. These decisions will provide the greatest prospects for future fertility [17, 19]. On the contrary, Relly and Woodhouse stated that a small penis does not preclude normal male role and a micropenis or microphallus alone should not dictate a female gender reassignment in infancy [20].

In the present study , almost all the cases of TH, 94% (n=17) showed male phenotypic expression, a result similar to that of Izquierdo and Glassberg [1]. All of them – except one case who reversed the sex to a female – retained their sex of rearing as males although in ten cases, the patients could have been a fertile female due to the presence of an ovary on one side with the presence of the Mullerian duct organs. This was partially due to the old age of the patients at presentation or to the preference of their parents to the reared sex as males being supported by the virilized appearance of their genitalia; in spite of their knowledge that if a testis is spared, it is lacking the spermatogenesis with nearly no possibility for fertility as male in the future.

In females with CAH, female sex assignment must be the rule as these individuals have entirely normal female internal sexual organs and a reasonably good chance of fertility [21]. In the present study, among the 78 reported cases, 18 (23%) were misassigned as males due to their severe virilization. In 10 cases, the parents were convinced to reassign their children as females for better fertility and sexual life. The age of these patients ranged between one month and 3.5 years.

In eight cases, the patients and/or their parents refused completely sex reassignment because of their late diagnosis, severe virilization and other social, cultural and psychological factors. They preferred to retain their sex and become infertile males after performing the hystero-salpingo-oophorectomy than to be declared as females in their community and family after this long social life, which they considered to be a great "shame". They were refusing to pass in a long series of problems to change the sex of their child e.g. birth-certificate, change of school, friends and neighbors, etc.. They were satisfied with their degree of virilization so that they can be able to marry in the future with no expectation of much problems, in spite of having no children: a condition that they considered commonly to happen for a normal male. They considered a male is more strong to live independently in the community than a female who will remain always dependable on their parents for life.

These results reflects the severe defect in neonatal screening for CAH as this misery could have been prevented – as recommended by all endocrinologists- by the early diagnosis and proper dexamethazone treatment of the pregnant mother sparing the newborn female consequences of genital ambiguity, sex misassignment and gender confusion [22].

The results are nearly similar to those of Kandermin and Yordam from Turkey who reported 91 girls reared as males among 273 cases of CAH, 31 of them (34%) refused sex reassignment [23]. Also, Jarayyan from Saudi Arabia reported a higher incidence (58%) of misassignment in similar cases [24].

Patients with MPH included in the study were assigned according to its pathological cause. As it is reported in literature [25,26], patients with persistent Mullerian duct Syndrome were reared as males as they are phenotypically normal males with excision of the corpus of the uterus and fallopian tubes enabling fixation of the testes in the scrotum.

Patients with leydig cell hypoplasia (n=9) and complete androgen insensitivity (n=13) have completely feminine phenotype and external genitalia, thus all of them retained their sex of rearing.

Among the cases of testosterone biosynthesis defects, only one case of 17-ketosteroid reductase (17B-Hydroxysteroid Dehydrogenase) deficiency with raised androstenedione levels was reported, being reared as a female. The patient had severely feminized genitalia that became virilized at the age of 15 years. This was contradictory to what has been stated by Rosler that a little more than 100 cases are reported in literature and a major aggregation of 68 cases has been identified among a highly inbred Arabic population that originated in the mountainous areas of Lebanon and Syria and now lives in Gaza strip [27].

Patients with MPH due to either 5 α-reductase deficiency or PAIS showed a great variation in the age of presentation due to the variability of the phenotype in these disorders: the more ambiguous, the earlier the presentation. This may be delayed until puberty which is marked in cases of 5 α-reductase deficiency when significant virilization without gynecomastia occurs with acquisition of male

gender identity [28]. These findings have suggested to some investigators that the prenatal influence of testosterone on the central nervous system overrules the socially induced process of gender identity and role acquisition [29.30].

In the present study, 31 patients have changed their sex from females to males, the majority was due to either 5 α-reductase (n=16) or PAIS (n=14), 13 of these were adolescents. These outcomes are similar to those in a Turkish family in which 4 of 5 individuals and the results in the Dominican Republic family in which17 of 18 individuals with 5 α-reductase deficiency had changed from female to male social sex [28].

Mendonca et al stated that it is of interest that most , but not all, subjects who have changed their social sex from female to male come from developing countries and they reported ten other similar cases. They proposed that if androgens have a role on the sexual behavior in humans, that critical period might be either during embryogenesis or in the neonatal period and not at time of sexual maturation[31]. This proposal is still opposed by the results of the present study which showed that still 10 peri-pubertal patients with 5 α-reductase have elected to maintain their female social sex with no apparent changes on their gender identity. Ten other patients with PAIS preferred to retain their sex as males although the size of their phallus might remain smaller and inadequate compared to a normal male with expected to face some problems in their future sexual life. It was apparent that their decision was nearly totally influenced by their social and cultural environment with their consideration for the "male" to be stronger to live in this community with such kind of problems than a "female".

Sex misassignment was defined as the presence of discordance between the genotype and the sex of rearing [32]. This may not be an accurate factor for evaluating the degree of proper management of patients with intersex problems in a certain community. In the present study the terminology of "discordant sex" was suggested as it demonstrates the magnitude of resistance of this community to assign these cases - after being counseled by specialized physicians- in a sex that is appropriate to their anatomical and functional possibilities to achieve a normal future sexual and social lives.

During the present study, it was noticed that there are several non-medical factors e.g. religious, social and cultural that were governing and influencing the view and the decision of sex assignment in this local community :

(1) With a person's death, the treasure - according to the local laws and the Islamic rules - is distributed in a way that the male inherits double the amount of a female and the presence of a son in the family preclude distribution of the inheritance to some other members in the family. This concept is based on the fact that the male is the responsible person of the family as regards protection, financial support, guidance etc… In the absence of the son, the family heritage will be distributed to the far relatives in certain proportions. Some families consider this as loss of the "family treasure" to other non-relative families.

(2) Males are considered to be free to circulate in outdoors and to travel alone which is restricted for females according to their cultural level.

(3) Males are obliged to do their military services if they have a son in the family.

(4) The sons transmit the name of their father's family to their offsprings which is considered by many individuals an important issue of continuity to "the name of their family" in their community. On the other hand, offsprings of daughters acquire the name of their father ,who is considered to be of a different family.

(5) Males are obliged according to the Islamic rules to perform their essential daily prayers at Mosque, for females it is preferable to do it at home.

(6) According to the Islamic rules, although not applied by law but sometimes adopted in certain very closed communities, the male witness is considered the double as that of a female.

(7) Fertility if compared to male assignment was viewed and accepted with little importance as they considered a male with no children is not going to face much problems in his future life, even if he remained unmarried with no excuses to be clarified to others. This is not acceptable for females.

(8) The religious view to the sex assignment in the Islamic communities has been previously discussed. All juristic religious opinions (Fatwa) considered the change of sex in a totally feminine or masculine human being with no physical abnormalities in his body (only due to refusal of the person to his native sex born-with i.e. transexuality) is a religious doctorinal crime as it changes " what God has created".

Only those patients who have an organic physical abnormalities in his body and genitalia making his sex characters as a male or female being confused are allowed to change their sex and perform the appropriate surgery to achieve this after being examined by a competent trustable doctor who approve the new sex assignment . It is essential that these details should be fully explained to the parents.

These "Fatwa" stated if both masculine and feminine characters are detected in a person, the doctors should detect what is predominant and to remove what can cause any suspicion in his sex in an attempt to achieve the best benefit and avoid any future corruption [33,34,35,36].

The issues that still demand the decision of religious leaders is about the criteria for definition of the sex characters as "male" or "female": is it the chromosomal sex, the gonadal sex, the phenotype, the appearance and function capability of the external genitalia? Also a decision is needed about the legality of excision of the gonads or the uterus in cases of females with PAIS and misassigned males with CAH specially if they are diagnosed in old age.

(9) The attachment of the majority of general population to their religion is strong. The patients and/or their parents are sharp and precise in their decision for sex assignment either male or female , nothing in "between". Transexuality is a rejected problem in this community and is not considered to be a commonly and freely discussed issue.

After sex assignment, most of the patients included in the current study were subjected to reconstructive surgical procedures with excision of all discordant tissues. Patients reared as males underwent simultaneous repair of their urethral defect and simultaneous correction of the scrotal anomalies and removal of contradictory uterus and gonads as a one-stage reconstruction. Orchiopexy and excision of abnormal breast were performed separately at a later schedule.

Urethroplasty included several techniques according to the degree of urethral defect .Mild degrees were repaired using urethral advancement , Mathieu's repairs, onlay or the transverse island flaps. Recently the tubularized incised plate urethroplasty -Snodgrass technique, [37]- was adopted with acceptable results. In severe forms, it was much preferred to use the combined techniques of Duplay's and the preputial flaps over the use of either the bladder or the buccal free mucosal grafts as the formers are considered as vasculalirized flaps and for the higher reported incidence of complications with the later techniques [38, 39].

The best time for female genital reconstruction is not clearly established. Some authors have favored treating only the clitoral enlargement early, postponing the vaginoplasty specially in severe degrees until puberty [40].Others prefer to perform the entire reconstruction in the first weeks of life thus ensuring a better relationship and proportion of the perineal structures[41].

In the present study, it was preferred to perform the entire reconstruction as a one-stage repair . As most of the patients presented late, they were operated upon once fully diagnosed with settlement of their proper sex assignment. Generally, it was preferred to perform surgery between the age of 6 - 18 months. Very early in the study , cases with severe virilization and penis-like clitoris were treated with total clitrorectomy and simple introitoplasty , postponing the vaginoplasty for older ages. Later on, preservation of the neurovascular supply to the glans was then adopted as recommended by most of the authors for better sexual function [41,42,43]. Subtotal de-epithelialization and partial concealment of the glans as described by Bellinger [44] was applied in a large number of cases. Very recently, the urogenital sinus mobilization technique [45,46] was applied on a small number of patients (n=6). Although the results of such technique were satisfactory, yet it needs a longer period of follow-up for its better evaluation.

With respect to evaluating and reporting results of this study, several variables are at play. First is the poor understanding and knowledge about the subject matter. Doctors and nurses dealing with young infants should be aware of the intersexual anomalies and should expect to encounter a wide spectrum of anatomic and physiologic derangements. Early screening of such conditions is simple but of utmost importance specially those of congenital adrenal hyperplasia. Cosmetic appearance alone is an inadequate measure of success because endocrinologic, social, psychological, religious and sexual factors must be blended into comprehensive evaluation of these patients. The management plan must be individualized incorporating long-term follow-up to adulthood.

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