Original Article

Managing disorder of sexual development surgically: A single center experience

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ABSTRACT

Introduction: Ambiguous genitalia are a major cause of parental anxiety and create psychological and social problems to patient, if not managed properly. Here we present our experience in managing patients with ambiguous genitalia.
Material and Methods: We retrospectively reviewed clinical records of all patients with ambiguous genitalia managed surgically at out institute between December 1989 and January 2011. Relevant history, clinical examination , investigations and surgical procedures performed were analyzed and results were evaluated in terms of anatomical, functional and psychosexual outcomes.
Results: Female pseudohermaphroditism was the most common cause of genital ambiguity in our patients. Male and female genitoplasty was done according to gender of rearing, genital anatomy and parental choice. Twenty six patients (86.6%) reported satisfactory cosmetic outcome and 22 (73.3%) satisfactory functional outcome on long term follow-up. Among the 24 patients diagnosed as male pseudohermaphroditism 14 (82.3%) patient have reported satisfactory cosmetic outcome and 13 (76.4%) have reported satisfactory functional outcome. In patients with mixed gonadal dysgenesis and true hermaphroditism satisfactory cosmetic and functional outcome was seen in 70% patient.
Conclusion: Managing patients of genital ambiguity according to gender of rearing, genital anatomy and parental choice carries good prognosis in terms of anatomical, functional and psychosexual outcome.

Key words: Disorder of sexual differentiation, genital ambiguity, intersex, sexual ambiguity

INTRODUCTION

Individuals with a congenital discrepancy between external genitalia, gonadal, and chromosomal sex are classified as having a disorder of sexual development (DSD).[1] Whenever there is difficulty in attributing gender to child based on appearance of external genitalia, they are termed "ambiguous genitalia." The appearance of external genitalia is a result of complex interaction between genetic and endocrine processes during fetal development.[1,2] Abnormalities of external genitalia sufficient to warrant genetic and endocrine studies are said to occur in 1 in 4,500–10,000 births.[3,4] Diagnosis and management of this condition can be challenging requiring a multidisciplinary approach.[5] Ambiguous genitalia are a major cause of parental anxiety and can create psychological and social problems to the patient, if not managed properly.

Data on disorder of sex development from Indian subcontinent are limited. The purpose of this article is to highlight the various surgical options available and surgical outcome on long-term basis. Herein, we present our experience in surgical management of patients with DSD.

MATERIALS AND METHODS

The clinical records of all patients with ambiguous genitalia managed surgically at out institute between December 1989 and January 2011 (N=88) were retrospectively reviewed. After thorough history and clinical examination, patient had undergone chromosomal analysis, relevant blood and

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urinary steroid estimation, abdominal and pelvic ultrasound, as well as retrograde genitogram as indicated. The genitogram was performed by low pressure injection of a water-soluble contrast agent through Foley catheter, inserted into the very distal part of the common perineal opening with care and repeated fluoroscopy in order to identify the perineal internal anatomy. Cystoscopy was done just before the surgery to confirm the findings. The frequency of various disorders according to the classification system utilized by Grumbach and Conte (1998) and the surgical procedures done were retrospectively reviewed.

No specific statistical tests were applied, but only the calculation of mean, frequency, and percentages using SPSS 17 software has been done.

RESULTS

Between December 1989 and January 2011, we managed 88 patients with ambiguous genitalia. The age at diagnosis ranged from new born to 23 yrs. The final diagnosis of causes of genital ambiguity in these patients were: female pseudohermaphroditism (N=42) that included congenital adrenal hyperplasia (CAH)-35 and non-adrenal cause (maternal androgens) – 1 [Figure 1a]; Male pseudohermaphroditism (N=24) and disorder of gonadal differentiation (N=22) [syndrome of gonadal dysgenesis {mixed gonadal dysgenesis (MGD) – 11, testicular vanishing syndrome – 4, 46 XY pure gonadal dysgenesis-1 } and true hermaphroditism-6].

Female hermaphroditism

This was the most common diagnosis of the cause of genital ambiguity in our study. As obviously, all the patients were genetically females (46 XX non-mosaics). Forty-one patients were diagnosed to have adrenogenital syndrome due to congenital adrenal hyperplasia with raised 17 alpha hydroxy progesterone; however, the exact enzyme deficiency assessment was not done due to non-availability of the facility. One patient was diagnosed to have female pseudohermaphroditism due to maternal administration of synthetic progestational agents.

The age at diagnosis ranged from new born to 14 yrs (mean=17 months) and the age at operation ranged from 3 months to 18 yrs (mean=3.5 years). The grade of ambiguity ranged from grade III to IV according to Prader classification.[6] Due to extreme masculinization of external genitalia and social reasons, seven patients underwent male genitoplasty, and laparoscopic excision of mullerian structures with bilateral gonadectomy, staged urethroplasty for hypospadias and testicular implantation [Figures 1b and 1c]. In this subgroup of patients, the mean age was 5.5 years (range1.5 years to 8.0 years). Follow up is available for only four such patients, among these only one has reached age of sexual activity and is fully sexually active and satisfied. The gender orientation in all four patients is that of a male.

In one patient, blind ending vagina was present; therefore, fulguration of vagina with staged urethroplasty with chordee correction was done. Thirty-three patients underwent female genitoplasty. In all 33 such patients, clitoral recession was done with perineal vaginoplasty, vaginal pull through and sigmoid vaginoplasty in 22, 5, 6 patient, respectively. One patient did not need a vaginoplasty as she had a normal looking vagina and was 18 years of age. The mean age for vaginal pull through was 2.5 years and for perineal vaginoplasty was 2.1 years, while the mean age for sigmoid vaginoplasty was 15.3 years. The patency in these patients was maintained by St'Mark's dilator use by parents in small patients and themselves in grown patients. In patients in whom there was single perineal opening of vagina and urethra (urogenital sinus) with confluence below urogenital diaphragm, the perineal vaginoplasty was done. While, in patients with high urethra-vaginal confluence, the vaginal pull through was done. Sigmoid vaginoplasty was done in patients with almost absent and rudimentary vaginal stump. Patency rates on long term were 100% for sigmoid vaginoplasty on regular dilatation. It is 60% for vaginal pull through (i.e. introital stenosis occurred in two out of five patients). For perineal vaginoplasty, patency rates is 91% (i.e. 2 patients out of 22 developed introital stenosis) and the cause was poor compliance to the use of dilator.

Follow up ranged from 3 months to 21. 3 years. Post-operative complications were seen in six patients in the form of clitoral sloughing in two, clitoral atrophy in one, and bleeding in three patients.

Male pseudohermaphroditism

Twenty-four patients were diagnosed as male pseudohermaphroditism [Figure 2a]. The age at diagnosis ranged from 3 months to 23 years (mean= 4.4 years), by definition the karyotype in each case was 46XY. Fourteen of the patients were raised as female and 10 were raised as males. In 12 patients who were raised as female underwent bilateral gonadectomy and clitoral recession. Eight of these patients underwent perineal vaginoplasty. Two patient had a high confluence of vaginal pouch and urogenital sinus at the area of urinary sphincter so sigmoid vaginoplasty was done [Figures 2b and 2c]. For the remaining two patients, parents insisted on delayed vaginoplasty but did not turn for follow up. Among these two, one had adequate vaginal length. In 3 of these 12 patients, simultaneous introitoplasty was also performed. In two patients, diagnostic laparoscopy was done, gonadal biopsy taken, and are awaited for follow up.

Ten patients were raised as males as they had somewhat better length of phallus with 3 even having palpable testis in labioscrotal folds. Six of these patients underwent bilateral orchiopexy with penile reconstruction and staged

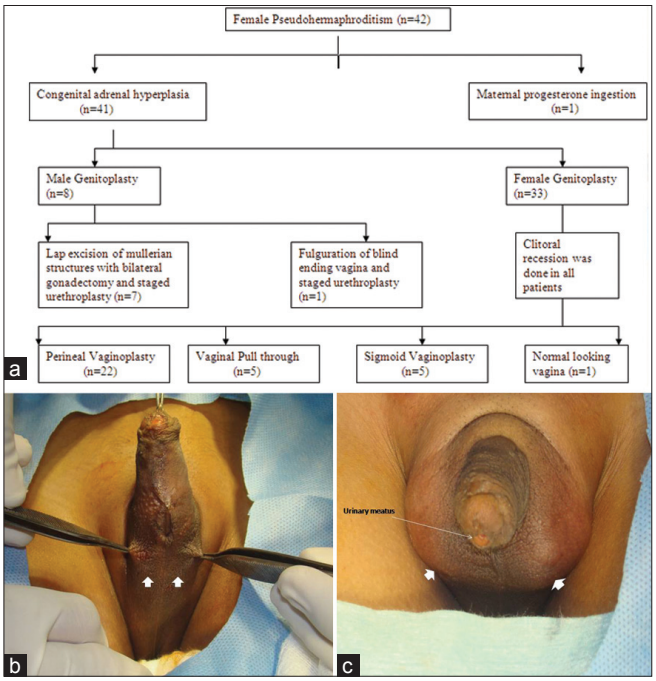

Figure 1: (a) Management of female pseudohermaphrodite (46XX DSD). (b) A case of CAH with well formed phallus, perineally located urinary meatus and empty labioscrotal folds (arrows). (c) The same case after male Genitoplasty and bilateral testicular implants (arrows).

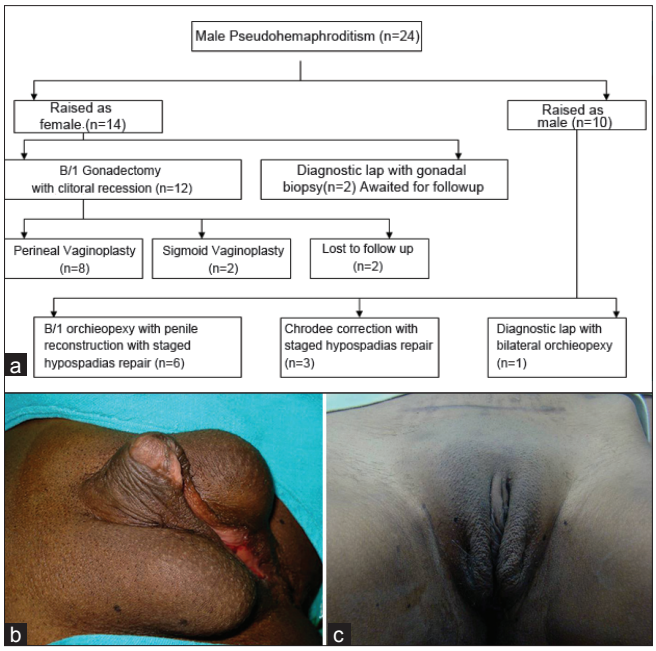

Figure 2: (a) Management of male pseudohermaphrodite (46XY DSD). (b) A child with 46 XY DSD raised as female with reasonable phallus and bilateral palpable gonads. (c) Same child after female genitoplasty.

hypospadias repair. One patient needed only diagnostic laparoscopy with bilateral orchiopexy, while three underwent chordee correction with staged hypospadias repair. The above-mentioned six patients had perineoscrotal hypospadias and bilateral testis in the inguinal region. Diagnostic laparoscopy revealed no mullerian structures. The patient underwent bilateral orchiopexy and penile reconstruction stage 1 initially, and subsequently Johansons's urethroplasty and glanuloplasty. One patient only had stage 1 repair and is awaited to come for follow up. Follow up ranged from 10 months to 18 yrs. Post-operative complications as urethrocutaneous fistula were seen in three patients. Long-term follow up is available for 17 patients, out of them 10 are married. 14 (82.3%) patient have reported satisfactory cosmetic outcome, and 13 (76.4%) have reported satisfactory functional outcome.

Disorders of Gonadal differentiation

These patients included 6 patients having true hermaphroditism and 16 having syndrome of gonadal dysgenesis (11 patients with mixed gonadal dysgenesis (MGD), 4 with vanishing testes syndrome and one with 46 XY pure gonadal dysgenesis). The age at diagnosis ranged from 1 month to 15 yrs.

True hermaphroditism

Chromosomal analysis in all cases was 46 XX/XY mosaic. Four out of six patients were raised as female. All the four patients underwent bilateral gonadectomy and perineal vaginoplasty and recession clitoroplasty. In one patient, a female gender was recommended; however, the parents refused it. This patient underwent laparascopic excision of mullerian structures (uterus, unilateral gonad as ovary, fallopian tubes, and vagina). Reconstruction of penoscrotal hypospadias was done using dartos-based tubularized prepucial flap. The sixth patient in this group was diagnosed at the age of 15 years. This patient has been raised as male due to fairly masculinized external genitalia and presented to us with complaints of breast enlargement and suprapubic pain. The patient also complained of having a short phallus. Clinical examination revealed the presence of tender suprapubic lump. Contrast enhanced computed tomography revealed the presence of hematocolpos and hematometra. The patient underwent exploratory laparotomy, hysterectomy, and bilateral sapingo-oopherectomy.

Mixed gonadal dysgenesis

Eleven patients were diagnosed as having a mixed gonadal dysgenesis with chromosomal analysis revealing 46 XX/XY. The age of presentation varied from 4 weeks to 12 years (mean=3.8 years). Four of these patients underwent bilateral gonadectomy with clitoral recession and perineal vaginoplasty, three underwent bilateral gonadectomy with clitoral reduction and vaginal pull through. The remaining four patients were assigned male sex of rearing. In all these patients, bilateral gonedectomy, laparoscopic mullerian structures excision with penile reconstruction, and hypospadias repair were done.

Follow up of above two categories of patients ranged from 39 months to 17.8 years (mean=10.3 years). Long-term follow up is available for 10 patients only out of these 5 are married, with satisfactory cosmetic and functional outcome in 7 (70%) patient. Post-operative complication in the

form of vaginal stenosis was seen in three patients and one patients in the MGD category needed buccal mucosal staged urethroplasty for failed hypospadias repair.

Vanishing testes syndrome

These four patients presented late at the age of 22-29 years. Three had partially developed sexual character with small size penis with no evidence of mullerian structures and testicular tissue. One had normal penile length. All the patients underwent bilateral testicular implantation

One patient was diagnosed as 46 XY pure gonadal dysgenesis. The physical examination showed clitoris normal looking female genitalia, normal looking vagina and cervix, high arched palate, low set ears, and breasts development. The Karyotype revealed 46 XY diagnostic laparoscopy and mullerianectomy was done.

DISCUSSION

Genitalia are ambiguous whenever there is difficulty in attributing gender to a child based on the appearance of the external genitalia.[1,2] Children born with the intersex problem comprise about 1.7% of all live births; these are recognized immediate in the new born nursery.[7] Proper gender assignment to a neonate born with ambiguous genitalia is a social emergency to minimize the psychosocial impact of a parent rending an equivocal or evasive answer to the age-old question from loved ones about the sex of new born child.

For the past 50 years practice has been informed by the so-called optimal gender policy, which proposed that in ambiguous genitalia, gender assignment should be that which allows optimal psychosexual and psychosocial functioning later in life.[8] This approach assumes the socialization is the decisive factor in gender identity development. In practice to support the assigned gender, early feminizing or masculinizing surgery of external genitalia was recommended, as socially acquired sexual identity is established by the age of 2 years.[5] The optimal gender policy has been challenged; in particular, the underlying assumption that gender identity is socially constructed. Parents are now routinely advised that children have the right to full details about their diagnosis and the medical and surgical management of this. Evidence suggests that parents have significant fears about the impact of ambiguous genitalia ad disclosure of diagnosis on their child's development. They are often uncertain about what to say and fear child will blame them or develop psychological difficulties.

An accurate and appropriate sex assignment should ideally be a multidisciplinary approach sex assignment and can be issued with the help of diagnostic tools which permit assessment of biochemical derangements, determination of genotype pattern, and definition of internal genital anatomy. Once an appropriate sex assignment has been made, the next critical step is treatment which depends on the type of the disorder, but will usually include reconstructive surgery in a timely fashion to remove or create reproductive organs appropriate for the gender of the child. Treatment may also include hormone replacement therapy.

There had been inconsistencies in various classification of abnormal sexual differentiation. We used the classification system utilized by Grumbach and conte (1998); however, recently the American Academy of Pediatrics proposed a new classification and obviated the term like "pseudohermaphroditism" and recommended the term disorder of sexual development (DSD).[9]

The four major pathological groups of patients with ambiguous genitalia are: female pseudohermaphroditism now called as 46 XX DSD, male pseudohermaphroditism now called as 46 XY DSD, mixed gonadal dysgenesis, and true hermaphroditism. Overall, CAH is the most frequent cause of ambiguous genitalia in the new born constituting approximately 60% of all intersex cases. Excessive androstenedione production results in a gonadal female with a virilized phenotype. The basic biochemical defect is an enzymatic block that prevents sufficient cortisol production (enzymatic deficiencies namely 21-hydroxylase 11-hydroxylase and 3-beta-hydroxysteroid dehydrogenase, etc.) or when the mother receives progesterone to prevent a miscarriage or has a hormone producing tumor, biofeedback via the pituitary gland causes the precursor to accumulated above the block. Clinical manifestation of CAH depends on which enzymatic defect is present. CAH presents a spectrum of abnormalities including the degree of phallic enlargement, the extent of urethral fold fusion, and the size and level of entry of the vagina into the urogenital sinus. Although the degree of virilization seen in CAH can be extreme, internal mullerian structure are consistently present. In these children, endocrine stabilization must be individualized, a process that usually takes several weeks. These patients require lifetime replacement with glucocorticoids and mineralocorticoids if they represent the salt loosing form of CAH. The psychosexual evidence to date supports maintaining female gender in masculinized CAH patients diagnosed in infancy.[7,10] In our series of CAH cases, 33 patients underwent female genitoplasty, whereas 7 patients underwent male genitoplasty (laparoscopic excision of mullerian structures with bilateral gonadectomy and staged urethroplasty of hypospadias) due to extreme masculinization of external genitalia and social reasons as they were reared as male. Since ours is a male dominant society and due to extreme masculinization these patients were assigned as male sex. It was in regard to parental request and child psyche, to avoid any psychosocial complication which may occur when grown up. It is in spite of the fact that female genitoplasty is more technically simple as compared to male genitoplasty.

Long-term follow up is available for 30 patients. Out of them, 18 are married. Twenty-six (86.6%) reported satisfactory cosmetic outcome and 22 (73.3%) satisfactory functional outcome. In our series, postoperative complications like clitoral sloughing and atrophy occurred, but these complications occurred in the initial phase of learning curve and can be avoided by meticulous use of sharp instruments, and avoiding the use of electrocautery and harmonics in and around the region of neurovascular bundle.

Bocciardi *et al.* reported 66 cases of CAH with satisfactory outcome in 65% and reintervention rate in 35%, follow up is of 5 months to 8 years.[11] In another study, Hoepffner *et al.* reported 41 cases of CAH with satisfactory functional outcome in 84.5% and reintervention rate in 12%, no follow up is available.[12]

The term undermasculinized male (male pseudohermaphroditism better defined as 46 XX DSD) refers to 46 XY individuals with differentiated testes who exhibit varying degrees of feminization phenotypically. Impaired male differentiation in these patients is secondary to inadequate secretion of testosterone by the testis at the necessary period in development or inability of target tissue to respond to androgen appropriately. The most common form of this entity is known as the testicular feminizing syndrome or androgen insensitivity syndrome. Sex assignment should be extremely individualized in these cases. Most of these patients are usually raised as females as the phallus is inadequate for the male gender role.[1] If female gender is chosen, gonadectomy is delayed until puberty unless the testes are palpable in the hernial sac, groin, or in the labia. Estrogen supplementation after gonadectomy is indicated. Most of these patients can be managed with perineal vaginoplasty; however, a few require vaginal replacement.[13] In our series out of 20, such patient 11 were raised as females and underwent female genitoplasty; 8 had perineal, and 1 sigmoid vaginoplasty. In those cases, who are raised as male, an early surgical reconstruction of the external genitalia is recommended.

True hermaphroditism (now called ovotesticular disorder of sexual development) represents an uncommon cause of genital ambiguity, accounting for fewer than 10% of DSD cases. The appearance of the genitalia varies widely in this condition. While ambiguity is the rule, the tendency is towards masculinization. Gonadal findings may be any combination of ovary, testis, or ovotestis. An ovotestis is most common and is found in approximately two thirds of patients. When an ovotestis is present, one third of the patients exhibit bilateral ovotestes. An ovary, when found is situated most commonly in the normal anatomic intra-abdominal position although Van Niekerk reported an ovary in the right hemiscrotum.[14] The least common gonad to ovotesticular DSD is the testis; when present the testis is found in approximately two thirds of the time lying in the scrotum, emphasizing that normal testicular tissue is not likely to descend fully and 80% of these patients have a 46XX karyotype although mosaicism is common. Gender assignment is again based on size of the phallus, extent of the labioscrotal fusion. Most of the children have an inadequate phallus and raised as females. In these cases, the testis should be removed and the testicular portion of the ovotestis should be removed leaving the ovarian portion in place.[3] In the present series of true hermaphroditism, all the patient were 46 XX/XY mosaics. Two patients had to be assigned male sex, one due to parental desire and other due to fairly masculinized external genitalia and social reasons. In a study by Hadjathanasiou CG *et al.*, 22 patients were described, no functional and cosmetic outcome were studied.[15] In another study by R. Wiersma, 71 patients were described and gonadectomy was done in all, no functional outcome is available.[16]

The syndrome of mixed gonadal dysgenesis (MGD) is associated with dysgenetic gonads and retained mullerian strictures. Most patients with MGD have a mosaic karyotype 45 XX/46 XY, in MGD, 25% of the gonads, including streak gonads can be expected to undergo malignant change, most commonly to gonadoblastoma. Early gonadectomy appears wise because tumors have been reported to arise in the first decade. Gender assignment of patients with MGD remains under debate. For example, Glassberg, citing that no case has been reported of a tumor developing in a fully descended testis in a patient with MGD, argues for assigning male gender to patients who are sufficiently virilized.[17] However, Rajfer and Walsh prefer an elective feminine gender assignment for patient with MGD because as uterus and vagina always are present and one half of patient have high incidence of inadequate external virilization.[18] Depending on the size of the phallus and extent of the labiaoscrotal fusion, gender assignment can be done. In our patients of MGD, all were mosaics, and bilateral gonadectomy was done in all. Seven of these patients underwent female genitoplasty while in four patients, bilateral gonadectomy, laparoscopic mullerian structures excision with penile reconstruction and hypospadias repair was done.

The timing of surgery is judged keeping in view a balance between the psychological advantages of early surgery and the technical limitations imposed by the small size of the structures. It is generally felt that surgery that is performed for cosmetic reasons in the first year of life relieves parental distress and improves attachment between the child and the parent; however, the systematic evidence for this belief is lacking.[9] Canty reporting his series of 18 patient with several forms of ambiguous genitalia suggested 2.5 to 3 yrs of age as the optimal time for reconstruction.[19] In contrast, Sharp *et al.*,[20] in their series presented nine infants with various forms of ambiguous genitalia all of whom were operated at an age of less than 6 weeks and Donohoe and Hendren[21] also favor early reconstruction, suggesting that

3-6 months of age may be the optimal time. However, they feel that children with high insertion of the vagina into the urinary tract should be operated at an older age (around 2 years). The rationale for early reconstruction is based on guideline on the timing of genital surgery from the American Academy of pediatrics.[9] It is anticipated that surgical reconstruction in infancy will need to be refined at the time of puberty, vaginal dilatation should not be undertaken before puberty. Emphasis should be on functional outcome rather than at strictly cosmetic appearance. Nihoul *et al*. and Randolph *et al*.[22] emphasized the importance of clitoris for orgasm and normal sexual function in their series and they recommend preservation of all or part of the clitoris with its nerve supply.[23] In our series clitoris was recessed underneath the pubic symphysis.

Lastly, it is necessary to discuss the social issues attached to these disorders in a country like India where male genitoplasty is insisted upon by parents of such children irrespective of the genotype and the surgical feasibility. This is because of the social stigma attached with the female child in the lower socioeconomic status. Feminizing genitoplasty as opposed to masculinizing genitoplasty requires less complex, surgical reconstruction to achieve an acceptable outcome and results in fewer urologic difficulties.

Our is a male-dominant society, and definitely there is a male gender bias but due to parental request and child psyche. This is not at the level of clinicians. Single stage or staged procedure for hypospadias repair is individual surgeons preference and according to their past experience in such procedures. Author prefers to do it in two stage as an institutional policy.

CONCLUSIONS

Managing patients of genital ambiguity according to gender of rearing, genital anatomy, and parental choice carries good prognosis in terms of anatomical, functional, and psychosexual outcome. The gender assignment policy, as described by Mayer and Bahlberg, does not always govern the management of such patients. Clinician has to regard the parent's will many a times, which may be contradictory to the anatomical facts and optimally feasible reconstruction.

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