Surgical Management of Ambiguous Genitalia in the Infant and Child

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● **Proper gender assignment to a neonate born with ambiguous genitalia is a social emergency of the newborn period. Once an appropriate sex assignment has been made, the next critical step is performance, if needed, of a reconstructive procedure in a timely fashion. In an attempt to evaluate our experience with this unique group of patients, we have retrospectively reviewed the course of 69 children with ambiguous genitalia managed surgically at this institution between 1974 and 1989. This series consists of 32 genotypic females with congenital adrenal hyperplasia (CAH), 10 children with mixed gonadal dysgenesis (MGD), 10 male pseudohermaphrodites, 3 true hermaphrodites, 8 genotypic females with urogenital sinus anomalies (UGS), and 6 genotypic males with bilateral undescended testes and penoscrotal hypospadius. All newborns initially seen at this institution received proper sex assignment within the first week of life. Prior to 1980, 17 of the children with CAH underwent clitorectomy and vaginoplasty and three underwent clitoral recession and vaginoplasty. After 1980, 10 patients with CAH were managed with clitoral recession and vaginoplasty and 2 with vaginoplasty alone. Eight of 10 cases of MGD were given a female sex assignment and all 8 underwent gonadectomy due to the high risk of gonadoblastoma; the other 2 children were raised as males. There were 3 true hermaphrodites of which 2 were assigned female gender roles and were managed with a clitoral recession and vaginoplasty. All 10 male pseudohermaphrodites were raised as females and all underwent bilateral orchidectomy. The 8 children with UGS were raised as females and underwent vaginal reconstruction. Six genotypic males with penoscrotal hypospadius and bilateral undescended testes were raised as males and underwent hypospadius repair and bilateral orchidopexy. This retrospective review underscores the complexities of assessment and management of ambiguous genitalia in infants and children. It also reconfirms the data from other series showing that the vast majority of infants born with the four most common forms of ambiguous genitalia (CAH, MGD, male pseudohermaphroditism, and true hermaphroditism) must be raised as females because of inadequacy of the phallus.**

INDEX WORDS: Ambiguous genitalia.

WE HAVE HAD the opportunity to gain extensive experience with the diagnosis and management of ambiguous genitalia, which may require several different approaches for definitive management. During the past 15 years we have cared for 69 such children who have required surgery. These patients represent a subgroup of infants and children with abnormalities of sexual differentiation seen at this institution, many of whom did not undergo an operation. This group of 69 patients forms the basis for this report.

MATERIALS AND METHODS

A retrospective review of all patients with ambiguous genitalia managed surgically at this institution between July 1, 1974 and July 1, 1989 was carried out. The cases were categorized into the following groups: congenital adrenal hyperplasia (CAH; 32), mixed gonadal dysgenesis (MGD; 10), male pseudohermaphrodites (10), true hermaphrodites (3), urogenital sinus anomalies (UGS; 8), and males with bilateral undescended testes and penoscrotal hypospadius (6). The patients primarily evaluated in the newborn period at this institution were jointly seen by the pediatric endocrine service, the neonatology service, and the pediatric surgical service. This group of children is part of a larger series of children (200) with ambiguous genitalia seen during this same period of time, most of whom were managed nonsurgically. Diagnostic workup included chromosomal analysis, blood and urine steroid measurements, abdominal x-rays and ultrasound, as well as a retrograde genitogram. The genitogram was performed in the x-ray suite with insertion of a Foley catheter into the very distal part of the common perineal opening. Following gentle inflation of the balloon outside the perineal opening, low-pressure injection of a water-soluble contrast agent is performed with careful and repeated fluroscopy in order to identify the perineal internal anatomy (Fig 1). Each of the groups was analyzed according to karyotype, age at diagnosis, age at operation, operative procedure, sex of rearing, and follow-up.

RESULTS

There were 32 patients with CAH. The age at diagnosis ranged from newborn to 16 years and the age at operation ranged from 2 weeks to 16 years. Fifteen infants were diagnosed in the newborn period; however, in 13 the diagnosis was delayed until after 1 year of age. All of the patients were genetic females (Table 1). Twenty-nine underwent a perineal vaginoplasty; the remaining 3 required a vaginal pull-through. Of those 29 patients who had a perineal vaginoplasty, 16 underwent clitoral resection and 13 were managed with clitoral recession. Two of the 3 children who underwent a vaginal pull-through were treated with clitoral recession; the other patient managed early in the series had a clitorectomy (Table 2). Prior to 1977, all patients underwent clitoral resection (10). Between 1977 and 1980, 7 children underwent clitoral resection and 3 clitoral recession.

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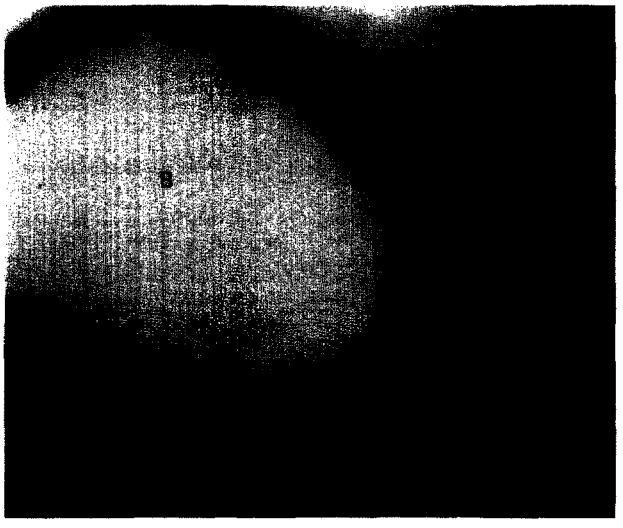


Fig 1. Retrograde genitogram. This procedure is performed with a Foley catheter inserted just inside the perineal opening with the balloon inflated. Note the low insertion of the vagina into the urogenital sinus. V, vagina; B, bladder.

Since 1980, ten patients have undergone clitoral recession; no clitoral resections have been performed during this latter period.Two children did not require clitoral surgery (Table 3).

Ten patients received the diagnosis of MGD. The age at diagnosis ranged from newborn to 20 years. Chromosomal analysis showed most commonly a 45, XO/XY composition with several patients having unusual forms of mosaicism, including 46,XY/49,XXXXY; 45,XO/46,XY/47,XXY; and 45,XO/XY + Y. Eight of the 10 children were raised as females and 7 have undergone bilateral gonadectomy. One child underwent bilateral gonadal biopsy in 1976 and was lost to follow-up. There were 2 children raised as males in this group. In the first patient, the female gender was recommended but the parents refused because the child had been designated a boy by the local physician. He was operated on at 3 months of age (bilateral inguinal herniorrhaphy, hysterectomy, right gonadectomy, and left hemigonadectomy). The second male underwent resection of a midline pelvic mass, probably of Mullerian origin, at 13 years of age. Among the remaining 7 females, 7 underwent perineal vaginoplasty, 4 had a clitoral recession, and 1 underwent a clitorectomy. Five children also underwent bilateral inguinal herniorrhaphies, including one of the male patients (Table 4).

Table 1. Congenital Adrenal Hyperplasia: Age at Diagnosis

Age At Diagnosis	No. of Patients
Newborn	15
1-6 mo	1
6-12 mo	3
1 yr	13

Table 2. Congenital Adrenal Hyperplasia: Surgical Reconstruction

	No. of Patients
Perineal vaginoplasty	29
Clitoral resection	16
Clitoral recession	11
No clitoral surgery	2
Vaginal pull-through	3
Clitoral resection	1
Clitoral recession	2
Total	32

There were 10 male pseudohermaphrodites in this series. The most common form of this entity is called the testicular feminization syndrome (TFS). The age at diagnosis ranged from 2 weeks to 17.5 years. All were raised as females and, by definition, the karyotype in each was 46,XY. All patients underwent bilateral gonadectomy and, in 5 children, bilateral herniorrhaphy was done at the same operation. One patient underwent bilateral inguinal herniorrhaphy as an infant, followed years later by bilateral gonadectomy. Five of these 10 children required vaginoplasties; 2 of these were performed via the perineal approach, one child had a colovaginoplasty using a vascularized loop of sigmoid colon, one patient underwent a pull-through of an ileal segment, and the fifth child underwent a cystovaginoplasty (use of portion of bladder to create a neovagina). The patients with the colovaginoplasty and the ileovaginoplasty are old enough for evaluation of sexual function. In both cases, the neovaginal depth is normal and sexual intercourse (by history) is normal. One female with the incomplete form of the TFS underwent a clitoral recession. Of the 5 patients who required a vaginoplasty, 3 underwent the procedure after the original gonadectomy and inguinal herniorrhaphy; the other two had the vaginoplasty performed at the same time as the gonadectomy and inguinal herniorrhaphy. Follow-up has ranged from 1 to 15 years and there have been no postoperative complications (Table 5).

True hermaphrodites represent the least common form of ambiguous genitalia; there were 3 children in this series (Table 6). The age at diagnosis ranged from newborn to 3 months. Two of these children were raised as females and were operated on at 2 and

Table 3. Congenital Adrenal Hyperplasia: Clitoral Resection Versus Clitoral Recession by Time Period

Operation	1974 to 1980	1981 to 1989
Clitoral resection	17	0
Clitoral recession	3	10
No clitoral surgery	0	2

Table 4. Mixed Gonadal Dysgensis

Case No.	Age at Diagnosis	Age at Operation	Operation	Length of Follow-Up	Chromosomal Analysis	Gender of Rearing
1	Newborn	5 mo	BIH, bilateral gonadectomy, clitoral recession, vaginoplasty	16 mo	45,XO/XY	Female
2	2 wk	2 wk	Bilateral gonadectomy, clitorectomy, vaginoplasty	9 yr	45,XO/XY	Female
3	4 yr	4 yr	Bilateral ovarian biopsy	Lost to follow-up	45,XO/XY	Female
4	13 yr	13 yr	Hysterectomy, bilateral gonadectomy, hypospadius repair	4.5 yr	46,XY/49,XXXXY	Male
5	Newborn	Newborn, 11 yr	Bilateral gonadectomy, vaginoplasty	10 yr	45,XO/XY	Female
6	1 mo	4 mo	BIH, bilateal gonadectomy, clitoral recession, vaginoplasty	2 yr	45,XO/46,XY/47,XXY	Female
7	1 mo	3 mo	BIH, hysterectomy, right gonadectomy, left hemigonadectomy, hypospadius repair	5 yr	45,XO/46,XY+Y	Male
8	Newborn	3 mo	BIH, bilateral gonadectomy, clitoral recession, vaginoplasty	4 yr	45,XO/XY	Female
9	Newborn	2 wk	BIH, bilateral gonadectomy, clitoral recession, vaginoplasty	4 yr	45,XO/XY	Female
10	10 yr	10 yr	Bilateral gonadectomy, vaginoplasty	1 yr	45,XO/XY	Female

Abbreviation: BIH, bilateral inguinal herniorrhaphy.

4 months of age, respectively. They both underwent bilateral inguinal herniorrhaphy, bilateral gonadectomy, and vaginoplasty. Clitoral recession was performed in both these females. Chromosomal analysis in these cases was 46,XY. The third child in this group was raised as a male and was 3 months old at time of diagnosis and operation. His chromosomal analysis showed a 46,XX/XY mosaic. This child was referred to us at the age of 3 months with a large but inadequate phallus. Careful and repeated suggestions were made to the family to raise this child as a female; however, the parents refused this approach. There have been no postoperative complications and follow-up has ranged from 3.5 to 8 years (Table 6).

Eight children with UGS anomalies presented with ambiguous genitalia. These patients represent a small

Table 5. Male Pseudohermaphrodites

Case No.	Age at Diagnosis	Age at Operation	Operation	Length of Follow-Up	Gender of Rearing
1	2.5 yr	2.5 yr	BIH, bilateral gonadectomy	2 yr	Female
2	4 yr	5 yr	BIH, bilateral gonadectomy	15 yr	Female
3	1 wk	1 wk	Bilateral gonadectomy	5.5 yr	Female
4	17.5 yr	17.5 yr	Bilateral gonadectomy, vaginoplasty with ileal pull-through	5 yr	Female
5	17.5 yr	17.5 yr	BIH, bilateral gonadectomy	2 yr	Female
6	2 yr	2 yr; 3 yr	BIH, bilateral gonadectomy; perineal vaginoplasty	2.5 yr	Female
7	17 yr	18 yr	Bilateral gonadectomy, colovaginoplasty	11 yr	Female
8	17.5 yr	17.5 yr	BIH as infant, bilateral gonadectomy	3 yr	Female
9	17.5 yr	17.5 yr	Bilateral gonadectomy, perineal vaginoplasty	1.5 yr	Female
10*	11 yr	11 yr	BIH, bilateral gonadectomy, clitoral recession, cystovaginoplasty	1 yr	Female

Abbreviation: BIH, bilateral inguinal herniorrhaphy.
*Incomplete form of testicular feminization syndrome.

Table 6. True Hermaphrodites

Case No.	Age at Diagnosis	Age at Operation	Operation	Length of Follow-Up	Chromosomal Analysis	Gender of Rearing
1	Newborn	2 mo	BIH, right gonadectomy, clitoral recession, perineal vaginoplasty	3.5 yr	46,XY	Female
2	3 mo	3 mo	Hysterectomy, right salpingoophorectomy	8 yr	46,XX/XY	Male
3	1 mo	4 mo	Bilateral gonadectomy, clitoral recession, perineal vaginoplasty	8 yr	46,XY	Female

Abbreviation: BIH, bilateral inguinal herniorrhaphy.

Table 7. Urogenital Sinus Anomalies

Case No.	Age at Diagnosis	Age at Operation	Associated Anomalies	Operation	Length of Follow-Up	Chromosomal Analysis	Gender of Rearing
1	Newborn	6 mo	Vaginal atresia	Perineal vaginoplasty	1 yr	46,XX	Female
2	Newborn	Newborn	Vaginal atresia	Perineal vaginoplasty	5 yr	46,XX	Female
3	5 mo	5 mo	Vaginal atresia	Transabdominal drainage of mydrometrocolpos, perineal vaginoplasty	3 yr	46,XX	Female
4	Newborn	10 yr	Imperforate anus, bicornuate uterus	Perineal vaginoplasty, colostomy, sacroperineal pull-through	1 yr	46,XX	Female
5	Newborn	Newborn, 14 mo 19 mo	Imperforate anus	Ileostomy, cecal augmentation cystoplasty, vaginal reconstruction via thermal ileal pull-through, bilateral gonadectomy, clitoral recession	3.5 yr	46,XX	Female
6	2 yr	2 yr	Bilateral renal dysplasia	Perineal vaginoplasty	3.5 yr	46,XX	Female
7	Newborn	Newborn, 16 mo	Imperforate anus	Loop sigmoid colostomy, perineal vaginoplasty, posterior sagittal anoplasty	4 yr	46,XX	Female
8	Newborn	Newborn, 2 yr	Imperforate anus	Colostomy, perineal vaginoplasty, colostomy closure	12 yr	46,XX	Female

percentage of all the females with a urogenital sinus seen at this institution during the same time period. The age at diagnosis ranged from newborn to 2 years with 6 of the 8 patients being diagnosed during the neonatal period. All patients were genotypically female and were raised as such. The age at operation ranged from newborn to 20 years (Table 7). Three of the 8 patients required perineal vaginoplasty (opening of the urogenital sinus) only. One patient underwent transabdominal drainage of hydrometrocolpos at birth secondary to vaginal atresia and a subsequent perineal vaginoplasty. Four of the children were also born with imperforate anus, treated with a sacroperineal pull-through in 2 children, a perineal anoplasty in 1, and a permanent ileostomy (because of inadequate colon) in the fourth. One of these 4 also underwent cecal augmentation cystoplasty, vaginal reconstruction with pull-through of the terminal ileum, bilateral gonadectomy, and clitoral recession. The follow-up has ranged from 1 to 12 years. This group will not be discussed further in this report.

All 6 patients born with severe penoscrotal hypospadius and bilateral undescended testes, who were referred because of ambiguity of the external genitalia, were genetic males and were reared as boys. They all subsequently underwent staged repair of their hypospadius and bilateral orchidopexies. This group will not be discussed further in this report.

DISCUSSION

Essential in the satisfactory management of children with ambiguous genitalia is early diagnosis with prompt gender assignment. This has obvious long-term effects on the well-being of both the child and family. The psychological impact of delayed proper sex assignment is illustrated by patient 9 in Table 5, who was not diagnosed until adolescence and, as a result, suffered severe emotional disability and eventual sexual abuse. In general, female gender assignment is made much more commonly because surgical reconstruction is far more successful. The only excep-

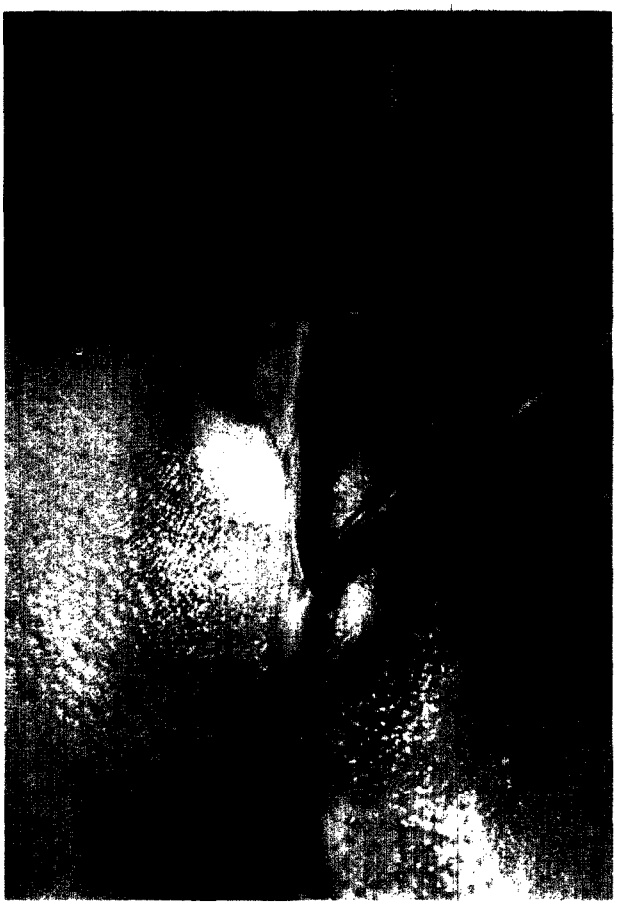


Fig 2. - CAH with moderate clitoral hypertrophy and UGS.

Fig 3. CAH with severe clitoral hypertrophy, UGS, and fusion of the labia to form a scrotum. There was a high insertion of the vagina into the urethra seen on genitogram and at surgery.

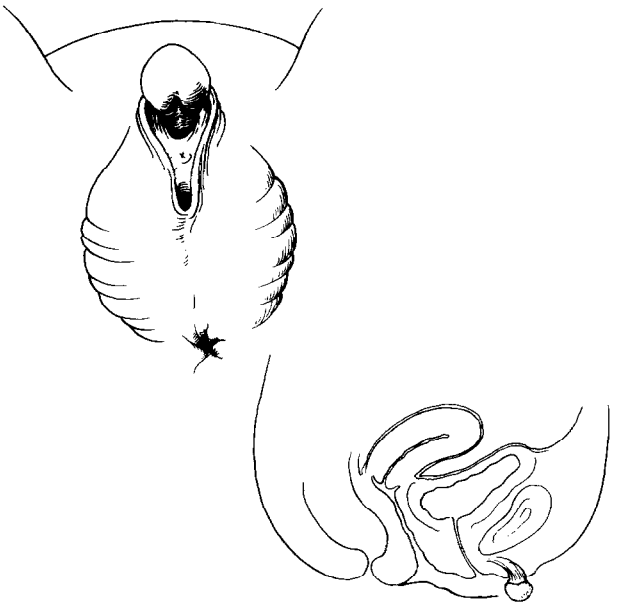


Fig 5. Upper diagram shows clitoral hypertrophy and UGS. Lower diagram shows low insertion of the vagina into the UGS.

tion to this is the genetic male with severe penoscrotal hypospadius and bilateral undescended testes, who is always reared as a male. It is uniformily agreed that an inadequate phallus cannot be surgically corrected and that the patient will fare much better in the female gender role.

The four major pathological groups of patients with ambiguous genitalia are CAH, MGD, male pseudohermaphroditism, and true hermaphroditism. Children born with these syndromes are generally raised as females because the phallus is usually inadequate for assumption of the male gender role. Infants born with congenital adrenal hyperplasia are genetic females who have been exposed to excessive levels of exogenous androgens. There are three enzymatic deficiencies associated with this syndrome, namely 21-hydroxylase, 11-hydroxylase, and 3-B-hydroxysteroid dehydrogenase. All these enzyme deficiencies result in overproduction of androgenic intermediary metabolites that masculinize the external genitalia of the female. The phenotypic picture varies from mild clitoral enlargement alone to complete masculinization of the urethra with a normal appearing male penis and the urethral meatus at the tip of the penis (Figs 2, 3, and 4). This later clinical picture is usually associated with complete fusion of the labia. These patients require lifetime replacement with glucocorticoids (cortisol) and mineralocorticoids (florinef) if

Fig 4. (A, B) CAH. No clitoral hypertrophy, UGS present. (C) Following cutback vaginoplasty.

they represent the salt-losing form of CAH. All these children are raised as females and have normal fertility. The ideal surgical management is a cut-back or flap vaginoplasty together with a clitoral recession at 3 to 6 months of age (Figs 5 and 6). If the child has the rare form of CAH in which the vagina has inserted into the urethra proximal to the external sphincter, then the vaginoplasty is delayed until at least 2 years of age (Fig 7).

The syndrome of MGD is associated with dysgenetic gonads and retained Mullerian structures. A mosaicism of the karyotype, usually 45,XO/46,XY, is always present. There is a very high incidence of malignant tumor development in the dysgenetic gonads (50%). Gonadoblastoma is the most commonly seen tumor; however, seminoma and dysgerminoma can occur, especially in the streak gonads.[1] For these reasons, we recommend bilateral gonadectomy in all patients with MGD. Surgical reconstruction is the same as with CAH in that all these patients should be reared as females and undergo appropriate reconstruction of the external genitalia (Figs 8 and 9).

Male pseudohermaphroditism occurs in genetic males (46,XY) with deficient masculinization of their external genitalia. The most common form of this entity is known as the TFS. Male pseudohermaphroditism can be due to the following: (1) inadequate testosterone production due to deficiencies of the

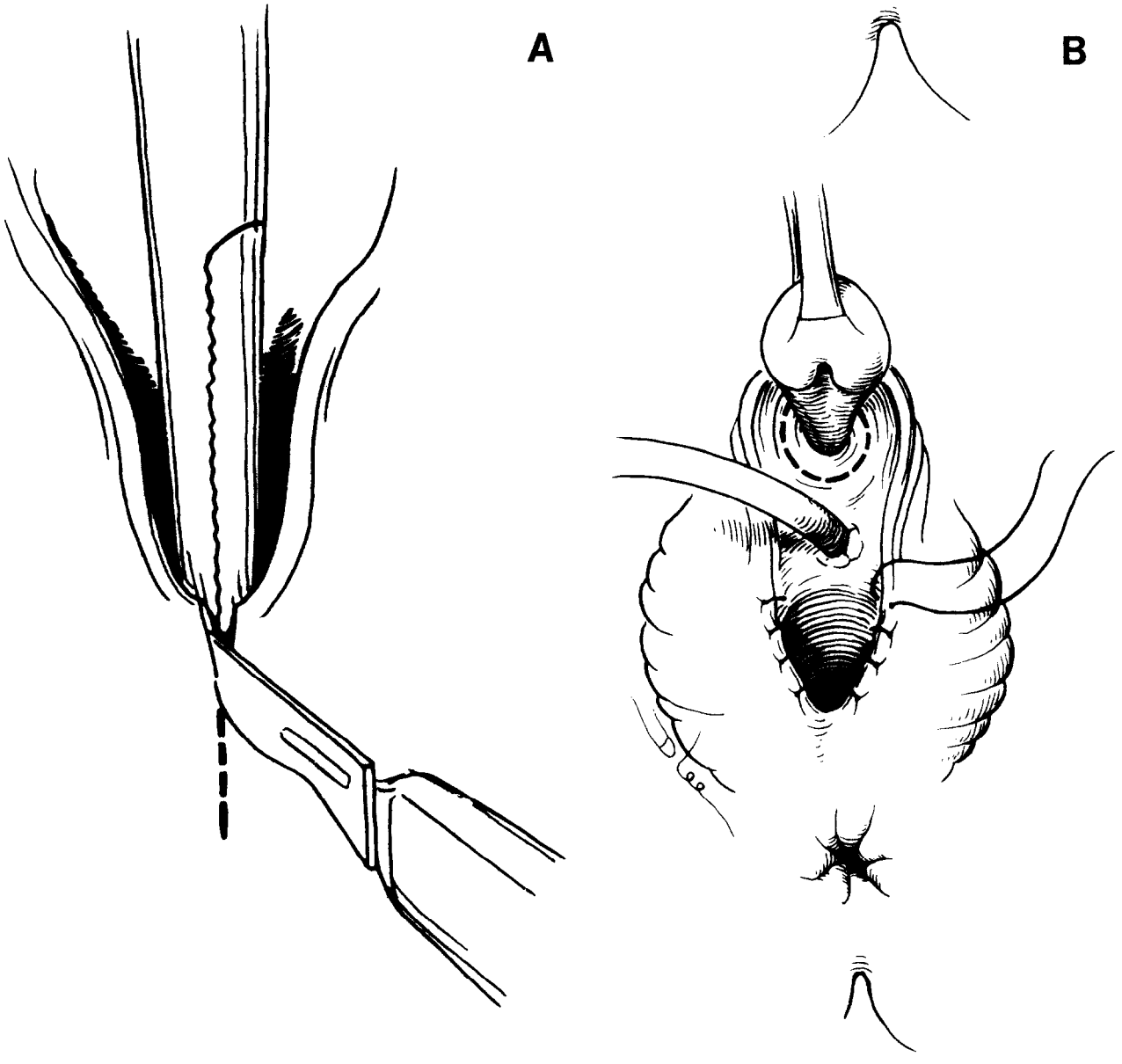
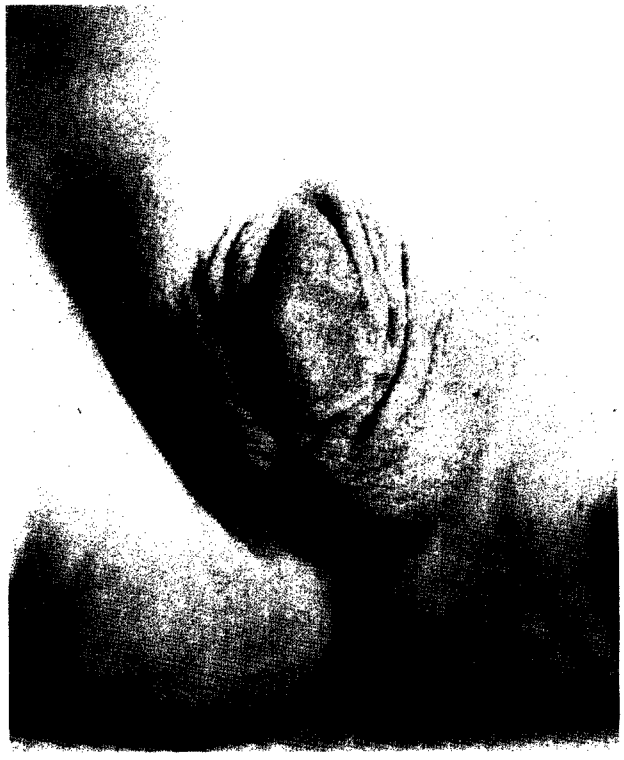


Fig 6. (A) Cutback vaginoplasty done over a hemostat. (B) Completed cutback vaginoplasty with vagina marsupialized to the perineal skin. The clitorus is being mobilized for the clitoral recession.

Fig 7. Child with CAH and high insertion of the vagina into the urethra. Appearance of the perineum immediately following a vaginal pull-through procedure.

enzymes necessary for its biosynthesis; (2) inability of the external genitalia target organs to convert testosterone to dihydrotestosterone due to 5-α-reductase deficiency; and (3) deficiencies in androgen receptors. These patients are all raised as females because in every case the phallus is inadequate for the male gender role.[2] In a few of these children, the diagnosis is made during a routine inguinal herniorrhaphy, at which time testes are found.[3] Our approach is to repair the hernias and remove the testes at that operation rather than remove the testes at the time of puberty. If no other surgery is planned, bilateral orchiectomy should be carried out just before puberty to prevent masculinization during puberty. All these patients have a very short vaginal vault; however, most of them can be treated with vaginal dilatation and will end up with a functionally adequate vaginal cavity. However, a few require a vaginal replacement; we prefer the use of colon for this operation. An occasional child with this abnormality will have significant clitoral hypertrophy and will require a clitoral recession.

True hermaphrodites represent the rarest form of ambiguous genitalia. These patients have both normal male and female gonadal tissue with an ovary on one side and a testis on the other or an ovotestis on one or both sides (Fig 10). Eighty percent of these patients have a 46,XX karyotype. Most of these children have an inadequate phallus and should be 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.[4] Surgical reconstruction is similar to that used for the child with CAH and MGD. If the phallus is adequate for the male gender role, then all ovarian and Mullerian structures are removed and a hypospa-



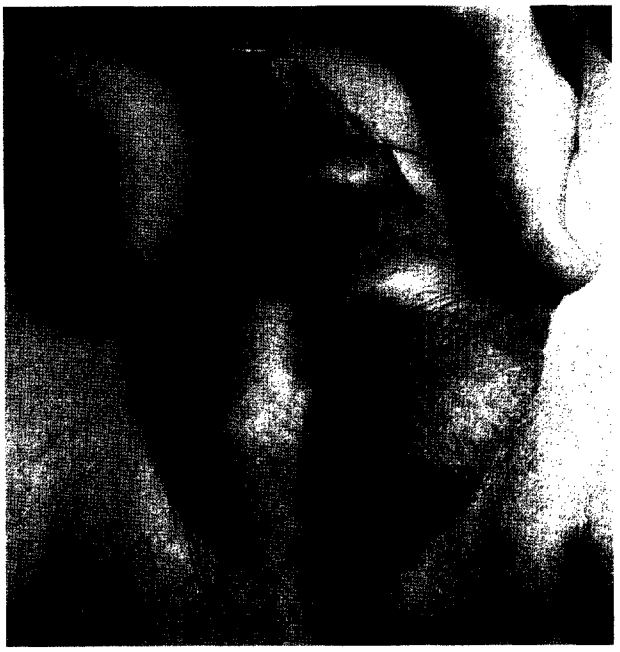


Fig 8. (A, B) Child with MGD (46,XO/XY). Note the significant clitoral hypertrophy, the gonad in the left labioscrotal fold, and the UGS.

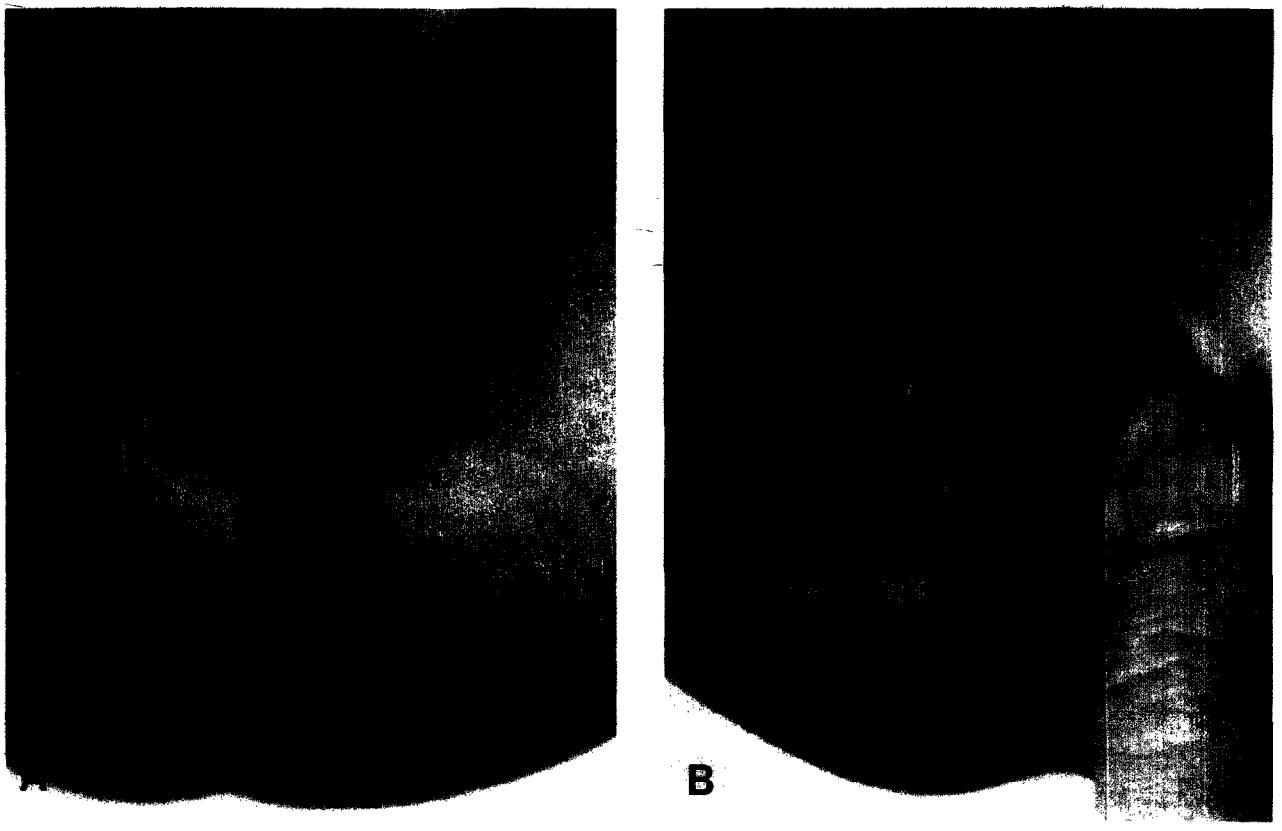


Fig 9. (A) Severe clitoral enlargement in an infant with MGD. Note the absence of an anal opening. (B) The urethral opening and UGS opening are at the base of the phallus.

dius repair is carried out. Just after puberty, testicular prostheses are inserted.

The timing of the surgical procedure represents a balance between the psychological advantages of early surgery and the technical limitations imposed by the small size of the structures. The trend is clearly toward earlier reconstruction when appropriate. Canty[5] reported on 18 patients with several forms of ambiguous genitalia; he suggested that 2.5 to 3 years of age was the optimal time for reconstruction. In contrast, Sharp et al,[6] in 1987, presented 9 infants with various forms of ambiguous genitalia, all of whom were operated on at less than 6 weeks of age. Donahoe and Hendren[7] also favor early reconstruction, when appropriate, suggesting that 3 to 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 on at an older age (around 2 years).

Fig 10. Child with true hermaphroditism. On the right side of the picture (the patient's left) are a Fallopian tube and ovary. On the other side a vas deferens and a testis are present. Note the large phallus.

Determination of the exact location of the insertion of the vagina into the UGS or urethra is best done with a retrograde genitogram as well as endoscopy at the time of initial surgery. Perineal vaginoplasty can be performed in the newborn period if the communication between the vagina and the urethra is distal to the external sphincter of the urethra. If communication is at or proximal to the sphincter, as in some cases of the adrenogenital syndrome, then we feel that a vaginal pull-through is the more appropriate operation. This procedure is generally done later in

childhood when the child is bigger so as to decrease the incidence of postoperative vaginal stenosis.[7]

Clitoral resection, a popular operation in the 1950s, 1960s, and early 1970s, is no longer recommended.[8] The importance of the clitoris for orgasm and normal sexual function has been emphasized by several investigators[9,10] and all attempts to preserve all or part of the clitoris with its nerve supply should be made. We prefer the operation in which all of the clitoris is recessed underneath the pubic symphysis.[10] This procedure gives an excellent cosmetic result; however, the long-term functional results are not yet known because most of the patients who have undergone this procedure have not yet reached full sexual maturity. Randolph et al reported on a small series of patients who had undergone clitoral recession and indicated that the functional results were quite good.[11] An alternate approach to clitoral recession is a procedure in which the glans is preserved with its neurovascular bundle and the corpora are resected. As originally described by Spence and Allen,[12] this then allows the clitoris to be recessed underneath the symphysis without the need to accommodate the bulk of the corpora. Theoretically, the advantage to this type of recession is the elimination of dyspareunia. However, there is no evidence that dyspareunia is a significant complication of clitoral recession without corporal resection. In addition, there is the potential of damage to the neurovascular bundle when the corpora are resected. Therefore, we prefer clitoral recession without corporal resection. However, there are no reports with sufficient long-term follow-up comparing either procedure.

In conclusion, management of the child with ambiguous genitalia continues to be one of the more challenging diagnostic as well as therapeutic problems facing the pediatric surgeon. The treatment of these anomalies underscores the importance of a multidisciplinary approach involving pediatric endocrinology, neonatalogy, and pediatric surgery. In addition, because of the rarity of these abnormalities, a large experience is needed in order to properly manage the large number of variations and complexities seen.

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