INTRODUCTION

In the hermaphrodite syndromes, a wide range of clinical presentations exist that have led to confusion. There is a plethora of diagnostic and therapeutic considerations that require a detailed evaluation of every infant born with some manifestation of these intersexual anomalies.

In the material presented below, only patients with ambiguous genitalia will be considered, for it is in this group of infants that errors in diagnosis, leading to incorrect sex assignment, are most injurious. A classification for patients with ambiguous genitalia is presented; we have found it helpful in considering the various anomalies encountered. Diagnostic principles used at the Children’s Hospital National Medical Center for such patients are also set forth. A few of the male patients require only orchiopexy and/or hypoplasias repair. These are performed in accordance with well-described standard techniques. Most patients with ambiguous genitalia, however, are best adapted to the female role; many of these infants require surgical correction of phallic enlargement. Because these surgical considerations are more controversial and have not become standardized, our experience with 29 patients requiring clitorectomy or clitoroplasty is detailed.
CLASSIFICATION OF THE HERMAPHRODITE SYNDROMES

In any consideration of patients with ambiguous genitalia, terminology is important. From our own clinical material, we have derived the following categories for grouping patients with intersexual abnormalities (Table 1):

1. True Hermaphrodite

   The term “true hermaphrodite” is reserved for those individuals in whom both ovarian and testicular gonadal tissue is present. This is most commonly found as ovotestis, but some patients are seen to have an ovary on one side and a testicle on the other. Often a major portion of the gonad is undeveloped, bearing only ovarian struma, for example; not infrequently, however, both ovarian and testicular tissue of excellent histologic quality is found. With respect to the chromosomal base for true hermaphrodites, there are several possibilities. The patients may be XX, XY, or XXY; less commonly, a mosaic form, such as XY/XO, may be present. For those patients who are true hermaphrodites, the sex assignment should not be made solely on the basis of their genotype. Rather, the sex of rearing should be decided after a full evaluation of the external and internal genitalia and an assessment of their capabilities from both anatomic and physiologic bases. When all factors are considered, most patients who qualify as true hermaphrodites will be found to be more adaptable to a feminine role, and this is the sex assignment usually chosen. However, we have one patient who is being raised as a male, and this seems to be quite successful.

2. Male Pseudohermaphrodite

   Patients who are male pseudohermaphrodites fall into three distinct categories. The first group have a marked penoscrotal hypospadias and one or both testicles in the undescended position. This leads, in the nursery, to an appearance of ambiguous genitalia. However, when

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**TABLE 1**

<table>
<thead>
<tr>
<th>Classification of Patients with Ambiguous Genitalia</th>
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<tbody>
<tr>
<td>1. True hermaphrodite (ovary and testicular tissue)</td>
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<tr>
<td>2. Male pseudohermaphrodite (testicular tissue only)</td>
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<tr>
<td>a. hypospadias plus cryptorchidism (normal-size penis)</td>
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<tr>
<td>b. hypospadias, Cryptorchidism, persistence of müllerian duct remains (adequate-size penis)</td>
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<tr>
<td>c. penile hypoplasia</td>
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<tr>
<td>3. Female pseudohermaphrodite (ovarian tissue)</td>
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<tr>
<td>a. congenital adrenal hyperplasia</td>
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<tr>
<td>b. adrenal cortical adenoma</td>
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<td>c. maternal hormones</td>
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<tr>
<td>d. maternal arrhenoblastoma</td>
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<tr>
<td>e. idiopathic</td>
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<tr>
<td>4. Absent or undeveloped gonad</td>
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the smoke has cleared and all diagnostic maneuvers have been carried out, it will be seen that infants in this category simply have two common anomalies. Hypospadias and cryptorchidism respond readily to surgical correction and carry little or no social stigma. With properly timed surgical correction, there should be no psychologic component to these anomalies.

The second category of patients classified as male pseudohermaphrodites are those with a normal-size phallus and only testicles as gonads. These testicles may or may not be descended. In addition, these patients possess elements of müllerian ducts. With the persistence of some müllerian duct structures, these patients not only appear to have ambiguous genitalia but also may have a fully developed vagina, as well as portions of female internal genitalia. A helpful diagnostic sign sometimes seen in these patients is formation of labioscrotal folds around the base of the phallus, giving the penis a clitoral relationship to the other perineal parts. Further diagnostic investigation may disclose the presence of a vagina, a cervix, a uterus, and even fallopian tubes. Any or all of these müllerian duct structures may be represented. Occasionally, however, the only müllerian remnant is a posterior diverticulum of the urethra; this defect is best demonstrated by retrograde urethrogram and panendoscopy. While the gonads may be in an internal position, patients of this group carry histologically normal testes. Surgical correction includes excision of the contradictory female internal genitalia and repair of the hypospadias. Plastic revision of the labial folds around the penis may also be necessary. Not all patients in this group have testicular feminization, but the possibility exists that some of the male pseudohermaphrodites may show feminization at puberty, even though normal testicular tissue is the only gonadal substance.

The third subgroup in the category of male pseudohermaphrodite represents a very complicated group of patients, not only with respect to embryogenesis but also in the assignment of sex. These patients possess testicular tissue of normal histology and are of normal XY genotype. The major defect in these males is a marked hypoplasia of the penis. The penis usually appears as a small, clitorislike button, with little or no shaft visible or palpable. Fortunately, most of these patients are thought to be female when first born, and a female sex assignment is made by the unwary physician, who may not recognize this condition in the early days of life.

Once the abnormal genitalia are recognized, appropriate diagnostic studies will reveal the true clinical nature of these male infants. In nearly all these patients, a female sex assignment provides the most appropriate way to raise the child, even though that designation is contrary to the genotype. The single overriding feature in the management of this form of male pseudohermaphroditism is the fact that the children in this category, while male in all respects, do not have an adequate phallic tissue for the male role. At the present time, there is no biochemical way to obtain penile growth, and surgical efforts have been inadequate and unrewarding. Because there is now experience with a number of these patients who have been raised as males, the clinical data strongly suggest that a female sex assignment is more appropriate in this group.

3. Female Pseudohermaphrodites
   (Ovarian Tissue Only)

   It is in patients categorized as

   continued
female pseudohermaphrodites that the need for surgical correction of the clitoris most often arises. These are patients who are female, usually with normal female physiology and a capacity for female development, yet have been born with a large, offensive, malelike phallus that is contrary to a female sex assignment. For better definition of this group of patients, the following subdivision is suggested:

(a) Congenital adrenal hyperplasia (adrenogenital syndrome). The principal facets of this clinical syndrome are well understood today. As originally described by Wilkins in 1950, the inability of the adrenal gland to produce cortisol leads to pituitary overaction with excessive production of corticotropin. This, in turn, stimulates the adrenal cortex to overproduce those substances it can manufacture — namely, androgens and growth hormone. The deranged metabolic effects begin in fetal life, and newborns with this abnormality show intense masculinization, with elongation of the phallus, scrotal rugation of the labia, and labial fusion. The labial fusion may be complete, so the urethra actually appears to open in the glans, as in the normal male. More commonly, the meatus is located at the base of the phallus and opens into a urogenital sinus. The diagnosis of congenital adrenal hyperplasia can now be assured by chemical means.

(b) Adrenocortical adenoma. It is a rare but definite clinical possibility that an adrenocortical adenoma present in the fetus or newborn can function exactly as adrenal hyperplasia, producing masculinization of the female fetus with all the changes described above. The seminal feature of the cortical adenoma as opposed to the adrenal hyperplasia is the failure of cortisone therapy to suppress the adrenal hypersecretion. This distinguishing feature obtains because there is actually an increased source of adrenocortical secretion, rather than an overactive corticotropin stimulation as in congenital hyperplasia.

(c) Maternal ingestion of hormones during pregnancy. A number of biochemical preparations have been offered to women in the hope of treating threatened abortions. Some of these products are very close to testosterone in their pharmacologic structure and action. These chemicals have resulted in the masculinization of an otherwise normal female fetus. This phenomenon was particularly prevalent a decade ago, when the compounds were not so refined as they are today. Usually, female neonates in this category have only moderate enlargement of the clitoris.

(d) Arrhenoblastoma. Mothers suffering from a masculinizing tumor of the ovary (arrhenoblastoma) are usually unable to conceive and rarely carry a pregnancy to term. There have, however, been isolated instances in which a woman with a masculinizing tumor has been able to deliver a full-term infant. The arrhenoblastoma exerts a masculinizing effect on both mother and fetus. If the baby is female, there is marked clitoral enlargement and labial fusion. Male infants, as with adrenal hyperplasia, are unusually mature.

(e) Idiopathic clitoromegaly. A number of female children have been born with enlargement of the clitoris and some degree of labial fusion, the cause of which escapes detection. In such patients, all historical and biochemical data are normal and the cause remains obscure.

4. Absent or Undifferentiated Gonad

It is unusual for patients with congenital absence of the gonads to have ambiguous genitalia, but this is occasionally seen. When there is a masculinizing influence that has resulted
in phallic enlargement, it can be assumed that there was at one time functioning testicular tissue that has gone on to failure. In any event, hyperplasia of the clitoris, requiring surgical correction, may occur in rare instances.

**DIAGNOSIS**

All patients with ambiguous genitalia should be considered in light of the foregoing classification, and appropriate steps taken to ensure an accurate diagnosis and proper sex assignment. The elements of the diagnostic pursuit are listed in Table 2. It is worthy of note that, because of the more accurate biochemical means to determine congenital adrenal hyperplasia and cortical adenoma, bone-age films are rarely used today. However, when a child has escaped detection in the neonatal period, bone-age films may be helpful. A laparotomy is done only in patients for whom diagnosis remains obscure. Of our own patients, studied jointly with the endocrinology department, only 15 per cent have come to exploratory operation and gonadal biopsy. However, this is a necessary part of the evaluation in patients ultimately shown to be true hermaphrodites, in some male pseudohermaphrodites, and usually in patients with idiopathic clitoral enlargement. Patients suffering from the testicular feminization syndrome with phallic inadequacy, as well as female pseudohermaphrodites with all but the idiopathic form of clitoral hypertrophy, can usually be diagnosed by other means.

1. **History.** With respect to the history, it is of utmost importance to know the details of the pregnancy, determining all ingested compounds that might in any way have influenced sexual development of the fetus.

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**TABLE 2**

**DIAGNOSTIC MEASURES IN PATIENTS WITH AMBIGUOUS GENITALIA**

1. History (including family and pregnancy information)
2. Physical examination
3. Buccal smear
4. Intravenous pyelography
5. Retrograde urogenital sinogram
6. Biochemical studies:
   a. urinary 17-ketosteroids
   b. urinary pregnanetriol
   c. plasma-OH progesterone
7. Chromosomal analysis
8. Panendoscopy
9. Exploratory laparotomy

*continued*
2. Physical examination. On physical examination, the external genitalia must be very carefully noted. It is of great help to perform a rectal examination and determine whether or not there is a midline structure anteriorly corresponding to the cervix and uterus. The importance of this simple physical finding cannot be overemphasized.

3. X-ray studies. An intravenous pyelogram should always be obtained. The urogenital sinogram is an extremely valuable maneuver. This study is best obtained by retrograde injection of the opening underneath the phallus. With gentle retrograde pressure and with careful fitting of the syringe to the external orifice, a vagina, if present, will usually be filled and be demonstrated as a separate structure on lateral films.

4. Cystoscopy and panendoscopy. Panendoscopy — with careful visual assessment of the urethra, bladder neck, and vaginal entry — is of major importance.

5. Biochemical studies. With respect to the biochemical data, the original definitive test was and is the demonstration of excessive 17-ketosteroid excretion in the urine. Other breakdown products, such as pregnanetriol, are now reported accurately as confirmatory evidence.

6. Chromosome studies. Buccal smear and, when needed, full chromosome studies provide the foundation for correct diagnosis and sex assignment. It is usually unnecessary to obtain formal studies of the chromosomes if a repeat buccal smear fits accurately into the diagnostic data.

7. Exploratory laparotomy. Exploratory laparotomy is reserved for those patients in whom the foregoing studies do not pinpoint the diagnosis. In such patients, laparotomy is usually carried out at five to seven days of age if the other data have been studied and a diagnostic quandary persists. Before laparotomy is undertaken, the salt-losing form of the adrenogenital syndrome must be ruled out. Laparotomy is performed under general anesthesia through a small Pfannenstiel's incision, with careful visual assessment of all internal genitalia and biopsy of both

continued

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**TABLE 3**

**SERIES OF 29 SURGICAL PATIENTS**

1. Clitorectomy ........................................ 6
   a. true hermaphrodite 1
   b. adrenogenital syndrome 4
   c. male pseudohermaphrodite* 1

2. Clitoroplasty ....................................... 23
   a. true hermaphrodite 2
   b. adrenogenital syndrome 18
   c. male pseudohermaphrodite* 1
   d. idiopathic clitoral hypertrophy 2

Total ............................................... 29

* Delayed diagnosis and incorrect sex assignment.
gonads. It has been suggested that the peritoneoscope be used in such patients, but we remain convinced that an open inspection of the tiny structures is more accurate.

CLINICAL MATERIAL

Using the categories established on the surgical service of the Children's Hospital National Medical Center, and with the complete cooperation of the department of endocrinology under Dr. Wellington Hung, 29 patients with significant enlargement of the clitoris have been treated surgically (Table 3). Six patients have been seen with such major enlargement of the clitoris that extirpation was thought to be necessary. In most of these patients, the clitoral enlargement has presented such a prominent phallus that operative revision was considered mandatory. Some workers have been reluctant to advocate excision of even the most grotesquely deformed clitoris, in the belief that the clitoris is necessary for normal sexual function. There are, however, a number of sound studies that demonstrate normal sexual response in patients who have undergone clitoral extirpation. Hampson studied six women with clitoral hypertrophy, none of whom had ever experienced orgasm. Following clitorectomy, five of the six women reported normal sexual gratification. Other interesting evidence that the clitoris is not essential for normal coitus may be gained from certain sociologic data. For instance, it is the custom of a number of African tribes to excise the clitoris and other parts of the external genitals at pubertal ceremonies. Normal sexual function is subsequently observed in these females.

When clitorectomy is indicated in a patient with a large, masculinized phallus, complete rather than partial removal of the clitoris should be the goal of the operation. Amputation of only the exposed portion of the phal- lus should be avoided. This maneuver produces a residual phallic mass beneath the skin that can become erect on sexual stimulation, leading to dyspareunia. In addition, the transected nerves in the body of the phallus can produce painful neuromas. Hence, when clitorectomy is to be performed, complete extirpation of all erectile tissue is essential. The size of an enlarged clitoris requiring clitorectomy cannot be stated in exact measurements. Certainly, only those patients with an inordinately large phallus should be selected for complete clinical extirpation.

Operative technique for clitorectomy (Figures 1-6). The infant is positioned on the operating table on his back in the dorsal lithotomy position. Feet are placed laterally or, in some instances, can be comfortably placed

Figure 1. In the dorsal lithotomy position, the clitoris is exposed and the patient catheterized. The base of the phallus is circumscribed with an elliptical incision. Dissection is carried down to the investing fascia of the corpora.
across the lower chest and upper abdomen and restrained in this position if good respiratory excursion can be assured. The buttocks are elevated with a sacral pad. Older children are placed in a standard lithotomy position with the legs in stirrups. The perineum and vagina are adequately prepared with proper solutions, and the area is suitable draped. The patient is catheterized, and the catheter is left in place during the operation. The prepuse is retracted and the clitoris placed on stretch, with a suture in the glans for stretch. The base of the clitoris is then severed circumferentially, using an elliptical incision. This incision is deepened, and dissection is carried to Buck's fascia, the tough enveloping layer extending up over the shaft of the phallus. Care should be taken to continue the dissection.

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**Indications** Vermox (mebendazole) is indicated for the treatment of *Trichuris trichiura* (whipworm), *Enterobius vermicularis* (pinworm), *Ascaris lumbricoides* (roundworm), *Ancylostoma duodenale* (common hookworm), *Necator americanus* (American hookworm) in single or mixed infections. Efficacy varies in function of such factors as pre-existing diarrhea and gastrointestinal transit time, degree of infection and helminth strains. Efficacy rates derived from various studies are shown in the table below:

<table>
<thead>
<tr>
<th>Trichuris</th>
<th>Ascaris</th>
<th>Hookworm</th>
<th>Pinworm</th>
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</thead>
<tbody>
<tr>
<td>care rates</td>
<td></td>
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<tr>
<td>mean</td>
<td>68%</td>
<td>99%</td>
<td>96%</td>
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<td>(91-100%)</td>
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<td>93%</td>
<td>99.7%</td>
<td>99.9%</td>
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<tr>
<td>(range)</td>
<td>(70-99%)</td>
<td>(99.5-100%)</td>
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**Contraindications** Vermox is contraindicated in pregnant women (see: Pregnancy Precautions) and in persons who have shown hypersensitivity to the drug.

**Precautions** Pregnancy: Vermox has shown embryotoxic and teratogenic activity in pregnant rats at single oral doses as low as 10 mg/kg. Since Vermox may have a risk of producing fetal damage if administered during pregnancy, it is contraindicated in pregnant women.

**Pediatric use:** The drug has not been extensively studied in children under two years; therefore, in the treatment of children under two years the relative benefit/risk should be considered.

**Adverse reactions** Transient symptoms of abdominal pain and diarrhea have occurred in cases of massive infection and expulsion of worms.

**Dosage and administration** The same dosage schedule applies to children and adults.

For control of trichuriasis, ascariasis, and hookworm infection, one tablet of Vermox is administered morning and evening on three consecutive days. For control of enterobiasis, a single tablet of Vermox is given. If the patient is not cured three weeks after treatment, a second course of treatment is advised. No special procedures, such as fasting or purging, are required.

**How supplied** Vermox is available as tablets, each containing 100 mg of mebendazole, and is supplied in boxes of twelve tablets.

*Trademark

†Because Vermox has not been extensively studied in children under 2 years of age, the relative benefit/risk should be considered before treating these children. Vermox is contraindicated in pregnant women (see: Pregnancy Precautions) and in persons who have shown hypersensitivity to the drug.
section in the plane immediately adjacent to this fascia. Superiorly, one transects the suspensory ligament and the dorsal vein. This brings the interpubic ligament into view. Lateral dissection frees the clitoral shaft from the subcutaneous tissue. As the base of the clitoris is delineated, branching of the two corpora cavernosa is seen. Inferiorly, caution is urged to avoid the subjacent urethra, which runs between the limbs of the corpora and disappears beneath the pubis.

At this point in the procedure, the presence of the catheter aids in identifying and preserving the urethra. The corpora cavernosa branch, and each extension is closely applied to the undersurface of the inferior ramus of the pubis, from which it must be separated. When this tissue thins out to strands of ischiocavernosus muscle, each corpus is transected. Before this part is cut free, however, a hemostat is carefully placed across the attenuated tip of the corpus to control the clitoral artery. The other corpus is similarly dissected and the specimen removed. All bleeding is controlled with fine hemostats and ligatures of No. 4-0 silk.

Reconstruction is carried out by mobilizing the lateral subcutaneous fat so that it may be brought to the midline by widely placed sutures. If this subcutaneous tissue is carefully brought together in the midline by several separate layers of interrupted sutures, the lower portion of the mons veneris is created, and a desired contour is achieved. The skin is closed with subcuticular sutures of 4-0 chromic catgut. No drains or dressings are used.

**Postoperative management.** If a vaginoplasty has been necessary, a small vaginal pack is left in place for a day or two. The urethral catheter is left in place for four days. A simple
T-binder is used as the dressing, and soft gauze is placed within this binder. Patients can usually be discharged on the fifth or sixth day. Bathing in a tub is contraindicated for 10 days following the operation. Thereafter sitz baths are recommended, and swimming is permitted two weeks after the operation.

**Complications.** There have been no significant complications. There were no urinary infections and only one minor wound hematoma, which corresponded to separation of the corner of the wound and pressure. There have been no wound infections.

**CLITOROPLASTY**

Operations to reduce or cover the enlarged clitoris without removing it have been suggested. In an effort to preserve the erotic properties of the clitoris, Goodwin et al. has dissected the glans and its neurovascular bundle free of the remaining portion of the clitoris and removed the corporal elements, transplanting the glans to the region of the pubis. Final evaluation of this operation is incomplete with respect to sexual gratification and function. Latimer has suggested burying the clitoris in the mons tissue, but the shaft is drawn downward in a way that could interfere with sexual function when erection is gained. Again, evaluation of this operation in adults is incomplete. Uncertainties about these procedures prompted further search in our clinic for a surgical technique that would preserve the clitoris yet permit reconstruction of the normal female appearance during the important growing years. Experience with the procedure to be described now extends over 10 years, and some of our patients are well into their adolescence (Table 3). None have yet reached an age of regular sexual activity.

*continued*
As previously mentioned, there are no criteria of size by which clitorectomy rather than clitoroplasty can be decided on. Instead, each case must be judged on the size of the parts in the perineum. In this series, there have been three patients for whom it seemed that clitorectomy would be necessary, yet at the time of operation it became obvious that a reduction clitoroplasty would be possible. Conversely, reduction clitoroplasty was planned for two patients, but at operation the shaft of the clitoris was of such length that the plastic operation was abandoned and total clitorectomy was performed. This approach of recession and plastic operation of the clitoris has proved unsatisfactory in only one patient, who later required clitorectomy. In three patients, circumferential excision of a portion of the glans has allowed us to reduce the size and prominence of that portion of the clitoris, making the remainder of the plastic procedure more successful by harmonizing the size of the glans with surrounding parts. In all of these patients, a feminine contour has been preserved. There has been no postoperative difficulty with erection, nor has there been irritation or pain. Clitoroplasty can be performed at any age, but we prefer the patient to have reached at least three months of age before undertaking the necessary dissection in the pubic area around the urethra. If operations on the clitoris are performed before one year of age, many psychologic effects are avoided.

**Operative technique for clitoroplasty (Figures 7-9).** As in clitorectomy, the patient is placed in a lithotomy position. The incision is begun approximately 2 cm. above the union of the shaft of the clitoris with the mons veneris in the midline. The incision is then brought laterally to a point on the labia majora at a level with the middle of the shaft of the clitoris. It is important that this incision be placed outward on the crest of the labia majora rather than in the labiocllitoral fold so that there is not an abnormal sulcus when these labia are brought together at the conclusion of the procedure. The midline incision is deepened down to the fascia over the intersymphyseal ligament, and this skin and subcutaneous tissue are removed by sharp dissection. From its most lateral points, the incision is brought to the subco-
ronal sulcus on each side of the glans. About 5 mm. of mucous membrane is allowed to remain around the dorsum of the glans, as in circumcision; in this manner, the incision on each side of the glans is joined in the midline in the subcoronal sulcus. The entire foreskin of the clitoris, along with the skin and subcutaneous tissue over the central portion of the mons veneris, is taken away sharply, using Buck's fascia of the clitoris as the surgical plane of dissection along the shaft.

With the removal of the foreskin covering the shaft of the clitoris, a length of the shaft is now exposed, as are the intersymphyseal ligament and the suspensory ligaments. Using sharp dissection along Buck's fascia, the suspensory ligament is completely divided until the clitoris is free beneath the pubis. Dissection is carried beneath the pubis until the bifurcation of the corpora cavernosa is demonstrated. This dissection must be complete to allow replacement of the corpora beneath the pubis, but care must be taken to avoid injury to the subjacent urethra. If the dissection plane is kept along Buck's fascia, there is only moderate bleeding in this vascular area. Hemostat and cautery are used for hemostasis. The shaft of the clitoris is placed posteriorly beneath the pubis for about a third of its length. A judgment can now be made as to whether reconstruction of the mons veneris in the midline will satisfactorily cover the clitoris. If it is felt at this point that the clitoris cannot be reduced and covered, then dissection is carried beneath each pubic ramus along the corporal extension and total clitorectomy is carried out. If reduction clitoroplasty is feasible, three individual sutures are taken in the periosteum of the lower border of the presenting surface of the pubis and

Figure 8. The shaft has been completely exposed and the suspensory ligament divided. Sutures placed in the investing fascia of the clitoris and the periosteum of the pubis are drawn together to retract the clitoris, which has been dissected free. The shaft of the clitoris slides behind the pubic ramus. Sutures are then used to bring the subcutaneous fat together, reconstructing the mons veneris.

Figure 9. Reconstruction of the mons veneris. The skin is closed in a manner allowing the labia to be brought together over the glans.
then into the tough fascia of the shaft. As these three sutures are tied, the clitoris is recessed beneath the pubis, reducing the length of the exposed shaft.

Reconstruction of the mons veneris using the available lateral fat is the next step. Interrupted sutures of 2-0 chromic catgut in the deepest layer of fat bring the adipose tissue from the sides towards the midline to cover the exposed pubis and the clitoris. Care must be taken not to carry this row of sutures too far inferiorly, or the clitoris will be doubled on itself in a dorsal direction. Rather, it is important to allow the clitoris to point forward and slightly upward and to bring fat around it to ensure its nearly complete cover. A second layer of sutures is now made in the fat of the mons veneris, further accomplishing closure of this mass of fatty tissue over the pubis and clitoris to obliterate any dead space. The skin is closed with interrupted sutures of 4-0 chromic catgut in the subcuticular position. Several sutures must be used to join the mucous membrane of the subcoronal sulcus to the skin, as in a circumcision. At the conclusion of the procedure, the glans of the clitoris should be only partly visible with the legs still in the dorsal lithotomy position. When the legs are brought together, a normal female contour is seen, and the clitoris should not be visible.

Postoperative management. The catheter is left in place for four days. Antibiotics are administered for five days. Opiates are necessary for pain in the older children. Patients are usually discharged on the fifth or sixth day. They are not permitted to sit in a bathtub until 10 days after the operation, and swimming is discouraged for at least two weeks to avoid irritation from a wet bathing suit.

Complications have been minimal.
Three patients have had hematoma formation with discoloration of the perineum, but only one required drainage. There has been no injury to the urethra and no wound infection.

Repositioning the clitoris as described offers excellent cosmetic results at an early age. Not only is the appearance satisfactory, but the shaft of the clitoris is so closely related to the fatty tissue of the mons veneris that frequent erections and constant irritation are prevented. In patients followed six to 10 years, there has been a normal increase in the size of the labia and vaginal introitus without inordinate growth of the clitoris.

It is entirely possible that some of these patients will have dyspareunia or other sexual malfunction as they reach the age of regular sexual activity. Clearly, they must be re-evaluated from the point of view of sexual and psychologic adjustment. This study has been begun in the older patients. In the meantime, an important factor is the normal appearance, which leads to social and psychologic adjustment free of the stigmas of genital malformation.

**CONCLUSIONS**

Patients with ambiguous genitalia must be recognized, if possible, on the day of birth. Then diagnosis of the abnormalities can proceed in a prompt but orderly fashion, leading to a proper sex assignment in the early days of life. This eliminates a great deal of confusion and apprehension on the part of family members. Thereafter, depending on the extent of deformity and the sex assignment, appropriate plastic surgical procedures can be chosen.

At present, clitorectomy is required for some female infants with major enlargement of the glans and shaft of the clitoris. Most, however, can be treated with an operation de-
signed to reduce the size and prominence of the enlarged phallus. In all of these infants, satisfactory appearance can be obtained by surgical means, so the child can grow up free of any significant physical, social, or psychologic consequences of her deformity. Final evaluation of clitoreoplasty awaits further information on children reaching adolescence.

BIBLIOGRAPHY

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ADDITIONAL REFERENCES


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