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# The gynaecological aspect of developmental anomalies of the female reproductive system

by

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Congenital anomalies of the female Reproductive System are not as rare as might be supposed. Their incidence is 1 in about 1500 obstetric cases and 1 in about 2000 gynaecologic cases. They may cause troublesome complications during the menarche, pregnancy and labour.

Until the 1930's most patients with congenital malformations were considered to be beyond medical relief, about the only exception being hare-lip which had been corrected with varying degrees of success for many years.

Within the past 30 years this situation has changed and malformations which were formerly considered hopeless are now being corrected. Surgical procedures have been simplified and modern safeguards introduced, reducing such complications as infections, haemorrhages and other accidents which used to cancel out the results of treatment.

Basically, progress in the treatment of congenital anomalies has been founded on improvement in our knowledge of embryology and anatomy. An anomaly is no longer a conglomerate mass of malformed organs but can be traced back to certain definite stages in foetal development. Embryological knowledge explains how developmental defects in the Mullerian duct system are often accompanied by anomalies in embryologically associated organs such as the kidneys, bladder and large intestine.

Progress in endocrinology and genetics has enlightened problems pertaining to determination of sex and they have a distinct bearing on the management of hermaphroditism.

The recent progress in chemotherapy and antibiotics has also lessened very much the dangers that used to constitute the chief risk in certain procedures like the repair of intestinal malformations and uretero-intestinal anastomosis.

In considering malformations of the genitalia one must remember their frequent association with defects in the urinary system as well as congenital defects in other systems, especially the lumbo-sacral spine.

I shall try, in the short space of time allotted, to outline the gynaecological problems presented by some of the commoner deformities of the female genitalia and to touch upon the general principles involved in their management.

Anomalies of the female genitalia may be classified into faults due to agenesis and defects due to arrested development.

## Agenesis

In the case of complete agenesis of an organ it is well nigh impossible to replace the missing structure. The transplant of an organ from one individual to another in man and higher animals is immediately fraught with a formidable obstacle in the firm rejection by the host tissues of the donor organ. In lower animals this rejection factor is not so marked. Thus the transplantation of rabbit ovaries into spayed rabbits was successfully accomplished even as rarly as the beginning of this century.

## Ovarian Follicular Agenesis

Anatomically, this condition is characterised by absence of the follicular epithelium of the ovary. The gem cells are not there. The ovary consists of connective tissue stroma with the usual blood vessels and nerves. It is represented by a long pencil-shaped strand of white

fibrous tissue in the usual situation of the ovary. The patient never menstruates. Her breasts, uterus, tubes and vagina are rudimentary or undeveloped. The genetic nature of this condition is attested by its frequent association with other developmental defects—short stature, coarctation of the aorta, mental retardation and other anomalies.

### Faults due to Arrested Development

There is as yet no cure for this condition. Ovarian hormones given in large amounts cause temporary stimulation of the breasts and the genital organs, including occasionally genital bleeding, but such temporary results disappear after cessation of treatment.

These may be classified under 4 headings:—

1. Double uterus and double vagina
2. Aplasia of the uterus and vagina
3. Imperforate hymen
4. Hermaphroditism

Except for the few anomalies that are part of general malformation and for hermaphroditism, women with developmental defects of the Mullerian ducts are quite normal otherwise. Some are unusually attractive. Their secondary sex characteristics—breasts, female habitus (contour), voice and external genitalia are normal. The ovaries are usually normal. The fallopian tubes are present, usually well developed, occasionally rudimentary.

There are innumerable variations of genital duplication, the commoner ones being:—

1. Uterus arcuatus
2. Double uterus with single cervix.
3. Septate uterus with single or septate vagina
4. Double uterus with double cervix and double vagina
5. Uterus with rudimentary horn or absence of one horn.

Absence of the uterus and vagina is probably one of the commonest of all gynaecological malformations. This seems to be due to inhibition in the development of the Mullerian ducts rather than to true agenesis. The structures which should form the uterus and vagina are present because the fallopian tubes are in their usual situation in varying degrees of development. The median ends of the tubes are separated from each other by a nubbin of muscular tissue the size of a cherry or larger. This is the rudimentary uterus which might contain a tiny uterine cavity lined by normal endometrium which menstruates, resulting in cryptomenorrhoea. There is no formed vagina. The external genitalia are normal but above the vesibule and hymen there is no space whatsoever. No menstruation occurs and coitus is impossible, and these are the complaints for which she seeks the gynaecologist's attention. The basic condition which stopped the normal development of the uterus and vagina is unknown. Perhaps it is due to persistence of the uro-rectal septum for unless this septum disappears completely it acts as a wall which separates the lower ends of the Mullerian ducts. In some instances there are associated anomalies such as absence of one kidney, ectopic kidney, double kidney, horse-shoe kidney, vulvo-vaginal anus, extrophy of the bladder and other defects.

In regard to the condition of double uterus and double vagina in its innumerable variations, the history and physical findings are frequently bizarre. Their presence should be suspected when extraordinary pelvic situations are encountered and one should then be constantly aware of the possibility of uterine anomalies. Examination under anaesthesia is warranted in such a situation, including recto-abdominal examination and careful exploration with the uterine sound or sounds. Hysterosalpingography is a major diagnostic aid.

When a developmental abnormality in the genital tract is suspected, a complete urologic work up including intravenous pyelography must be carried out.

However, anomalies of the genital tract may cause no symptoms during the life-time of the woman. The defects may be discovered incidentally or remain undetected. Discovery is quite often made by accident at the time of laparotomy and then a complete exploration should be carried out including palpation of the renal fossae which might reveal absence of one kidney.

If clinical symptoms do occur they do so characteristically during the 4 episodes or mile-

stones of a woman's life—menarche, marriage, pregnancy and labour.

Haematometra, haematocolpos or haematosalpinx does occur in defectively developed Mullerian ducts that do not have adequate external communication. The fact that the patient does have periodic bleeding per vaginam does not exclude the presence of retained menstruum in one of the underdeveloped uterine horns.

As a rule, fertility remains unimpaired. The presence of two uteri would seem to favour fertility. The possibility of conception in a rudimentary horn which has no communication with the vagina does exist in the case where transperitoneal migration of spermatozoa or of a fertilised ovum to the rudimentary side occurs.

The bicornuate uterus is of little clinical significance unless pregnancy supervenes. Early diagnosis of congenital uterine and vaginal anomalies is of paramount importance in the proper management of the case. Examination early in the pregnancy will disclose gross asymmetry of the uterus on account of unequal enlargement of the uterine horns. The 3 most common complications are abortion, uterine inertia and foetal malpositions.

The abortion rate is high, varying between 25 to 50%. If one horn becomes pregnant then the non-gravid horn will show decidual changes of the endometrium, and abnormal bleeding frequently occurs from the decidua of this horn. The bleeding is often interpreted incorrectly as a sign of imminent abortion and the pregnancy inadvertenly terminated by hasty exploration of the gravid horn with a curette.

The indication for a plastic procedure of a double uterus associated with repeated abortion does not rest simply on making this anatomical diagnosis. Before surgery is undertaken the various endocrine and metabolic factors often associated with abortion must be ruled out.

It is best to handle pregnancies in patients with uterine anomalies by Caesarean section following minimal trial of labour. Breech and shoulder presentation are common, as are premature rupture of the membranes and abruptio placentae.

In those cases in which a vaginal septum causes dystocia, resection of the septum between clamps and ligation may be all that is necessary to effect delivery by the vaginal route.

### Surgical Management of Genital Anomalies of the Mullerian System

Conservative surgery in these cases is to be desired if specifically indicated. Operating on them because of the existence of an anomaly is discouraged by gynaecologists because in most instances no benefit is derived from the elective plastic procedures. Since the malformations are so variable it is not possible to lay down a rule-of-thumb plan of therapy. Provided that sound surgical principles are meticulously observed, the type of surgery is predicated on a thorough evaluation of the genital deformity and the associated renal status.

In the case of congenital absence of the vagina, construction of an artificial vagina is indicated in the woman who is already married or is going to get married soon All methods of constructing an artificial vagina have several technical problems, difficulties and complications: the need for dissection of an adequate space between bladder and rectum and avoidance of injury to bladder and rectum during the process of dissection, haemostasis, infection of the new tract, maintenance of the patency, of the development of painful granulations in the new vagina and other problems that may arise unexpectedly at one time.

The most important step in the operation is to dissect an adequate space between the bladder and rectum. All previous methods, until I published my own method in the British Journal of Obstetrics & Gynaecology, February 1960, consisted of making a transverse or vertical incision across the occluding connective tissue between bladder and rectum and then blindly pushing aside the connective tissue in the plane of dissection to form a tunnel. This manoeuvre causes much trauma and bleeding, injury to bladder and rectum is very liable and contracture of the canal very prone afterwards.

My method is based on the premise that vestiges of the Mullerian ducts are always present on each side of the mid-line in the space between the bladder and rectum. I have demonstrated this in studies of sections through

a pyramidal block of tissue removed from this area of aplasia.

The technique consists of dilating the regions in which the openings of the undeveloped Mullerian ducts are recognised as faintly outlined dimples on each side of the mid-sagittal plane between bladder and rectum, using Hegar's or rectal dilators. This procedure results in the formation of two canals separated by a septum in the mid-line. The septum is finally removed and the end result is a capacious tunnel.

The advantages of this method are: (1) The ease with which an adequate tunnel is made; (2) Lessening of the danger of bladder and rectum injury; (3) Minimising the bleeding; (4) Use is made of the embryological epithelium, exposed as a result of the dilatation, to supplement the epithelium creeping up into the tunnel from the vestibule or to complement the split thickness skin graft, if a graft is used to line the tunnel.

At the end of this paper I shall project slides showing the presence of the undeveloped Mullerian ducts in the space between bladder and rectum and also a full length film illustrating my operation.

### Imperforate Hymen

When the obstruction is due to imperforate hymen menstrual blood and cellular debris accumulate in and enlarge the vagina (haematocolpos). The back pressure may be sufficient to cause accumulation of blood in the uterus (haematometra) and in the tubes (haematosalpinx.) Occasionally the peritoneal cavity of the pelvis may be filled with this blood and debris.

The pain is cyclic in character and coincides with the menstrual molimina. Episodes of sudden severe pain may be initiated when blood spills into the peritoneal cavity.

The diagnosis is suggested by the fact that the patient has cyclic pain and menstrual molimina but no menstruation. Examination reveals the bulging of the hymen and perinaeum.

The hymen is excised or incised to establish drainage.

### Hermaphroditism

The gynaecologists' responsibility begins when the hermaphrodite is born and he must decide its sex. In these days mistakes are seldom made. At times, in later life the gynaecologist may be asked whether it is advisable to alter the adopted sex of a hermaphrodite whose sex has been wrongly diagnosed. As a rule it is never wise to change the sex of the individual.

It is often impossible to be sure of the gonadal sex of the individual without doing a biopsy of the sex glands, as identically the same intersexual appearances of the external genitals may be seen in individuals with either testes or ovaries or, for that matter, with both types of gonads. The male phallus with a complete hypospadias may look like a large clitoris; a cleft scrotum may resemble labia majora; the urethra may look like a tiny vaginal orifice.

To complicate matters, sex hormone studies have been found to be of no great importance in differentiating the sex of such individuals because one and the same gonad is capable of producing both male and female hormones. This is of practical importance, for the testes should not be removed in the case of pseudohermaphroditism in which the external genitalia are predominatly female and the psychology of the feminine type. In a number of cases of this group in which the tests were removed the patient suffered typical menopausal symptoms.

It is generally impossible to correct the physical faults of the hermaphrodite completely. However, the gynaecologist can be of great help in eliminating features that tend to emphasise the embarassing sexual confusion. It is, of course, easier to remove redundant organs or tissues than it is to try to build them up if they are lacking. In the case of masculine hermaphroditism it is often easier to amputate the hypertrophied phallus and construct an artificial vagina than to try to adapt the external genitalia to the male type. Correction of a complete hypospadias involves multiple operations, the end result of which is a phallus which is not of much value for coitus.

Plastic procedures are indicated if they contribute to the peace of mind of the hermaphrodite and make her life easier, because social and psychic problems are constantly in the mind of the hermaphrodite.

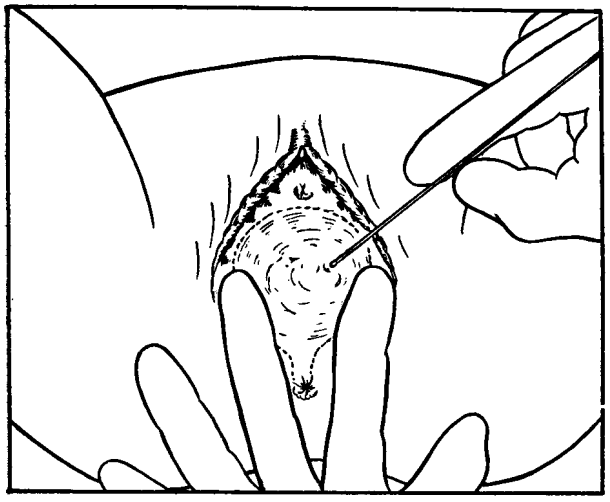

Fig. 1. A racket-shaped incision is made around the rim of the rudimentary vaginal pouch, the handle extending backwards over the fourchette to include a 1 cm. width of skin over the perinaeum.

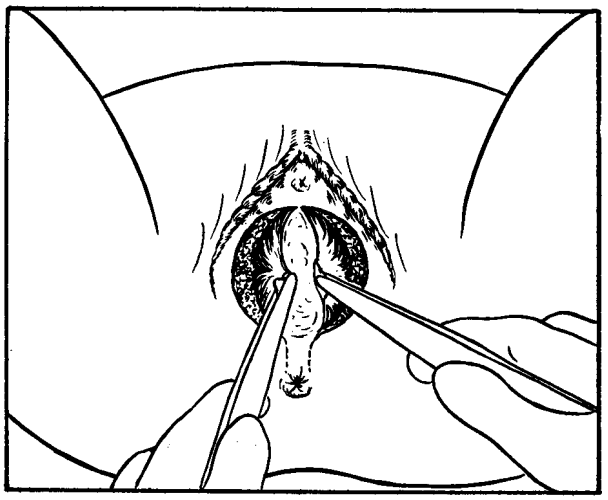

Fig. 3. Similar dissection is carried out in the right half-circle and the almost completely separated mucosa, still attached to the deeper tissues in the mid-line, is lifted outwards with tissue forceps and put on light tension.

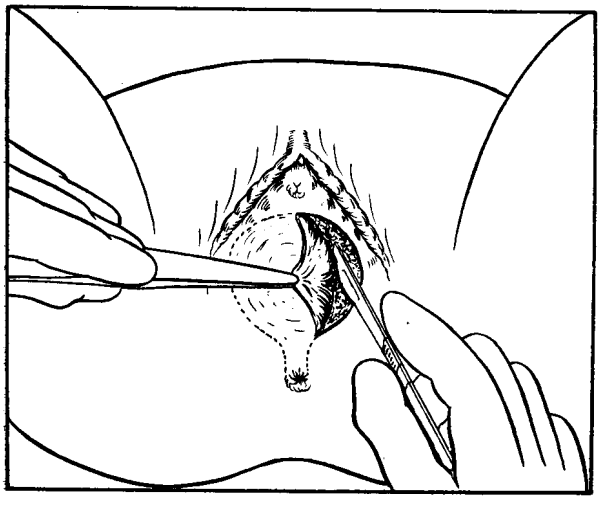

Fig. 2. Dissection of the mucosal flap is carried from the periphery of the left side of the circle to almost the level of the mid-sagittal plane.

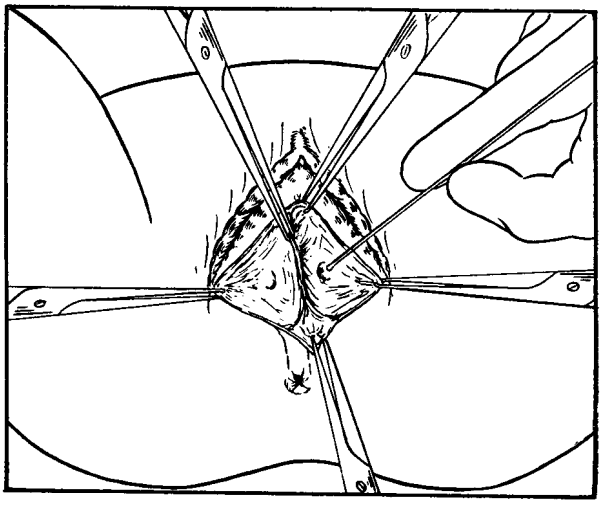

Fig. 4. The effect of this manoeuvre is to cause an outline to take shape in the dissected area which, without much stretch of the imagination, resembles that seen at the introitus in the case of a double-barreled vagina, except that the barrels seem to be choked with tissue. On either side, in the centre of the contralateral dissected areas a dimple, surrounded by fibrils of tissue disposed eccentrically, will become distinguishable.

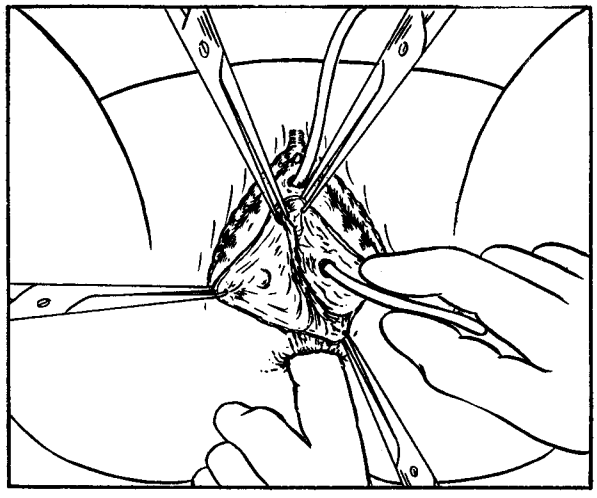

Fig. 5. A No. 4 Hegar's dilator is inserted into the centre of each dimple and pushed gently in the direction of the pelvic axis. If the right plane has been entered, this manoeuvre is easy and the dilator slides in to a depth of 10-12 cm. without inducing any bleeding.

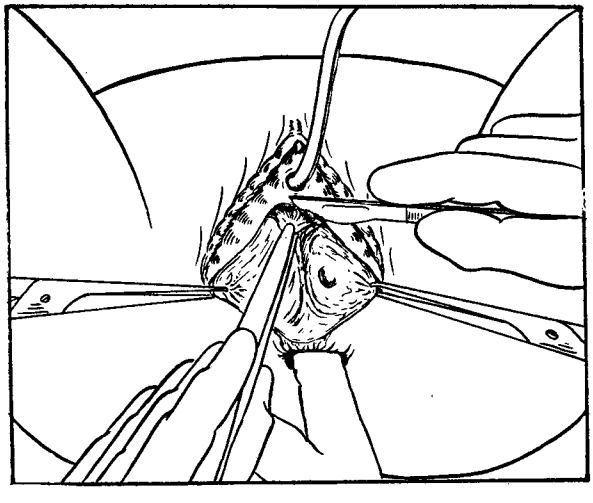

Fig. 6. The racket-shaped mucosal-cum-perinaeal handle skin-flap is then dissected off its attachment in the mid sagittal plane and allowed to hang down posteriorly away from the field of operation. This free mucosa and split thickness perinaeal skin will be available later as an isograft for covering the raw surface of the tunnel posteriorly after the septum between the two barrels has been excised.

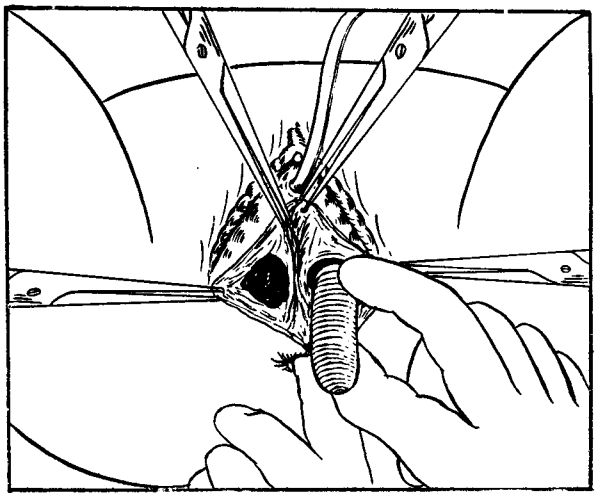

Fig. 7. The partially dilated canals on each side are gradually dilated by Hegar's dilators until the dilatation has been carried to No. 25 Hegar. The result is a man-made double-barreled canal with a relatively thick septum of connective tissue separating the barrels. The process of dilation is better orientated by identifying the position of the bladder with a sound and of the rectum by an assistant's finger inserted therein. However, the chances of bladder or rectal injury are much less by this method than by the orthodox method of transverse dissection.

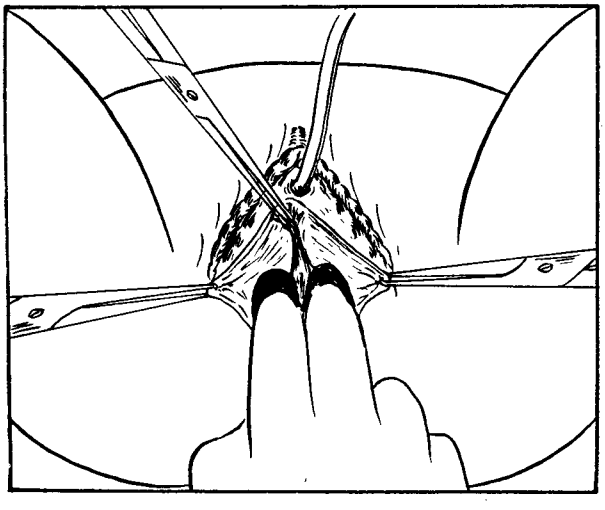

Fig. 8. When the dilatations have been completed, two fingers can be easily inserted into the new tunnel, one on each side of the septum, and the capacity of the space at least equals that of a normal vagina.

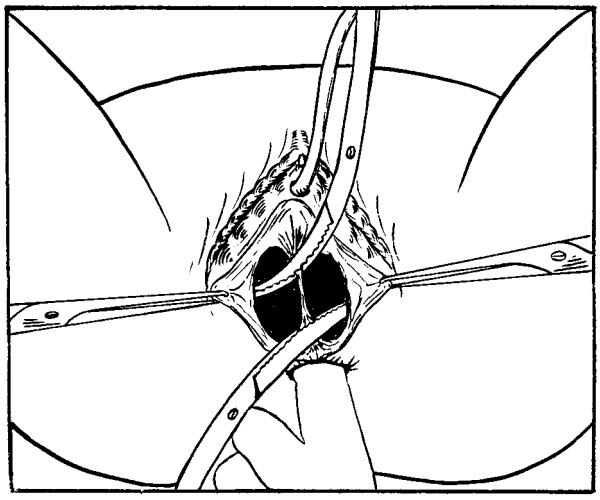

Fig. 9. The next step is to convert the double-barrel canal into a single chamber by resecting the relatively thick septum between the chambers. One long slightly curved forceps, such as is used to draw on the Round ligaments in ventri-suspension operations, is applied along the upper attachment of the septum, orientated by the bladder sound to ensure that the delicate bladder wall is not dragged down and included in the jaws of the forceps. A similar forceps is applied to the lower attachment of the septum, orientated by the assistant's finger in the rectum to avoid injury.

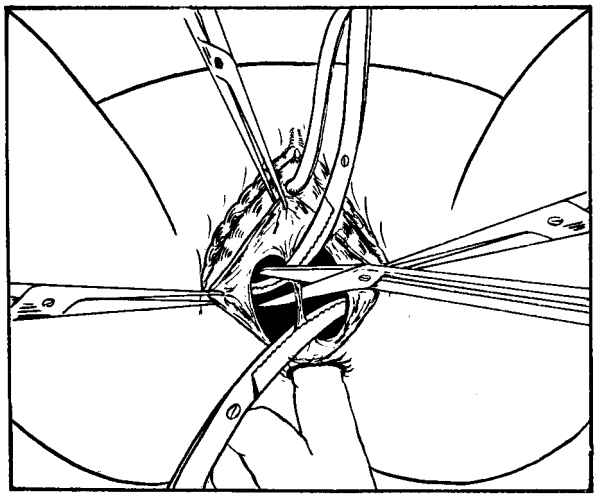

Fig. 10. The septum is then cut away flush with the clamps, using angulated scissors, and removed in one piece. The clamps are then removed and usually no bleeding ensues.

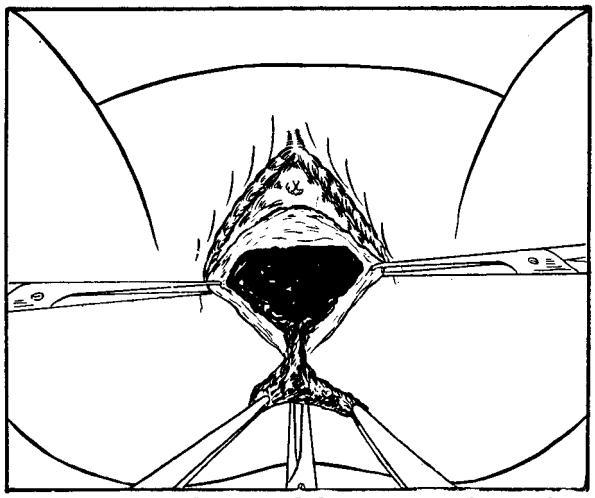

Fig. 11. When the bore of the new recess is examined it will be noted that, except at the areas anteriorly and posteriorly when the septum has been cut away, it is lined by smooth tissue which has a pale white sheen. The dome of the recess if formed by peritoneum reflected from the superior surface of the bladder to the anterior aspect of the rectum. This can be pushed upwards easily by the blunt dilator without danger of puncturing. The racket-shaped musocal-cum-perinaeal handle splint-thickness skin graft is then inserted into the tunnel over its raw area posteriorly and held in position by a few interrupted 000 plain cat-gut sutures.

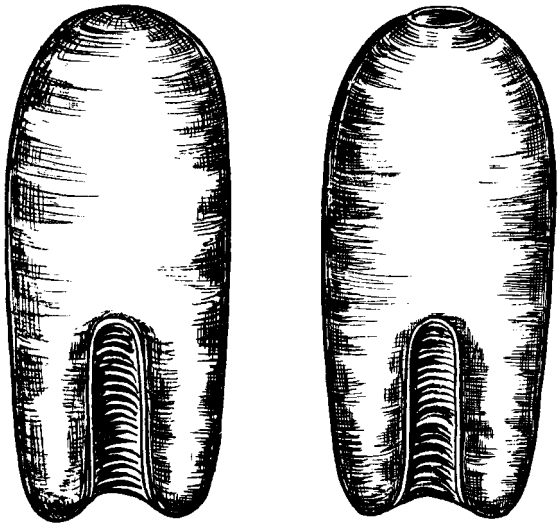

Fig. 12. Two types of mould: on the left, the usual type with a groove at its lower end to prevent pressure on the urethra.
On the right the type used when a functional uterus is present; a rigid tube draining the cervix can be threaded through the mould, thus allowing uterine discharges to escape.