Construction of a functional vagina: a new surgical approach to the Rokitansky-Küster-Hauser syndrome

by

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The Rokitansky-Küster-Hauser (RKH) syndrome is a genital malformation, consisting of rudimental and generally solid cornua of the uterus, vaginal aplasia and normal ovaries and oviducts. The clinical picture includes primary amenorrhoea, development of normal secondary sex characters at the usual age, occasional menstrual molimina, inability to have intercourses and sterility.

The RKH syndrome is less rare than is sometimes thought and our personal series includes 159 patients since 1966. The malformation is the result of arrested development of the müllerian ducts during the second month of pregnancy. The segment forming the cornua and cervix does not canalise. The same is probably true with respect to the cranial segment of the vagina, though here failure of the urogenital sinus to develop caudocranially may also be responsable.

The factors giving rise to this malformative syndrome are not known. It may be supposed that both extragenetic and genetic causes are involved.

The vulva of these patients is virtually normal, while the hymen is generally absent or vestigial. The hymenal pseudomembrane is often surrounded by petal-like fleshy excrescences. The meatus urinarius is abnormally placed in a mediovulvar site half way between the vestibulum and fourchette of the vulva. Its unusual size and funnel-shape mean that it may be mistaken for the introitus vaginae. While sometimes congenital, this defect is more often the result of attempted coitus per urethram. The vagina itself is entirely absent. It is represented by a short groove, which often has a fibromuscular cord on its median line. The corpus uteri is replaced by two comma-like and usually solid rudimentary cornua that run downwards lateromedially and posteriorly to the bladder and meet on the median line. A beginning of canalisation is sometimes noted. The oviducts are normal in appearance and nearly always in the usual sites. The ovaries are somewhat laterally and cranially placed, but are normal in shape and size.

The external appearance and general proportions of these patients are decidedly feminine and some subjects are very attractive.

The RKH syndrome is not hereditary and the familial history will prove negative.

Apart from amenorrhoea, the personal physiological history will be fully normal.

The clinical picture leading to the patient seeking medical attention is essentially based on amenorrhoea, inability to have intercourses and sterility. Amenorrhoea is nearly always uterine and hence clinically silent.

Rectal examination shows absence of the corpus uteri, while a more accurate investigation, under general anaesthesia reveals two cord-like formations that

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Clinical Exper. Obst. & Gynecology
Jan. 1974 Vol. I, pp. 3-8

sometimes appear to come together medially in a button the size of a hazel-nut. The adnexa, slightly increased in size, may also be palpated occasionally.

Inguinal hernia or the sequelae of an operation for its correction are not uncommon. The syndrome is variously accompanied by a number of extragenital abnormalities, particularly of the urinary tract. Cases with single or ectopic kidney or with renoureteral reduplication are described. Non urological malformations are less common: congenital dislocation of the hip, spina bifida, syndactyly, polydactyly or oligodactyly, supernumerary vertebrae and ribs, cleft palate, various congenital heart malformations, deafness, etc. Three patients of our series presented a Klippel-Feil syndrome.

Diagnosis is also assisted by suitable clinical and laboratory examinations. Coelioscopy and pneumogynaecography will give information concerning the internal genitalia. Intravenous pyelography and isotopic nephrography will be mandatory for the reasons already given. Skeletal radiography will reveal bone deformities. Pelvic arteriography will detect widespread arterial hypoplasia rather than offer information of practical value. The hormonal exfoliative cytological studies on specimens taken from the vaginal depression, the basal thermic curve and the urinary pregnandiol, oestrogens and gonadotropins determinations will be useful in showing normal ovary function. Female chromosome sex can be determined in the usual way (chromosome analysis, sex chromatin).

Differential diagnosis will be directed towards Morris syndrome, Turner syndrome, simple vaginal aplasia, pseudohermaphroditism, imperforate hymen, transverse vaginal septa and other complex malformative syndromes. Little difficulty is experienced when the clinical data are associated with chromosome, gonadic sex and hormone studies.

There are only relative indications for surgical management in RKH syndrome ([1]). It will be obvious that surgery will not establish menstrual flow or permit pregnancy, but only facilitate more or less normal intercourses. Psychological considerations are, of course, of great weight. In the case of unmarried subjects, the question of whether the operation should be performed before or after marriage may have to be considered.

The fact that a large number of surgical techniques have been proposed for the creation of a neovagina makes it clear that results are of uncertain quality and justifies the search for better methods ([2]). The main steps of our technique (in use since 1965) are:

1) Pfannestiel incision of the abdomen and transverse incision of the peritoneal fold.
2) Creation of an intervesicorectal passage by downward blunt (Fig. 1).
3) Insertion of a grooved sound in this passage to lift the pseudohymen, which is perforated by passing a straight thread-carrier along the sound (Fig. 2).
4) The ends of two Perlon threads are passed through the eye of the carrier. The other ends are attached to an acrylic olive (Fig. 3).
5) The sound and the carrier are withdrawn. The threads are passed subperitoneally and brought outside the anterior rectus muscles (Figg. 4 and 5).
6) Suture of the bladder peritoneum and the abdominal walls in layers.
7) Attachment of the two threads to a traction apparatus fitted on the abdomen (Fig. 6).

After the operation, progression of the olive is already sufficient to form a

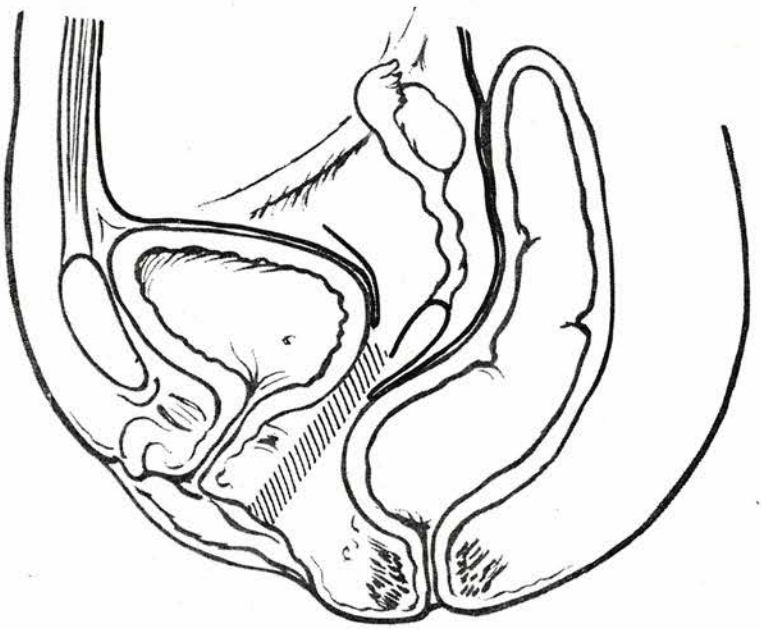

FIG. 1

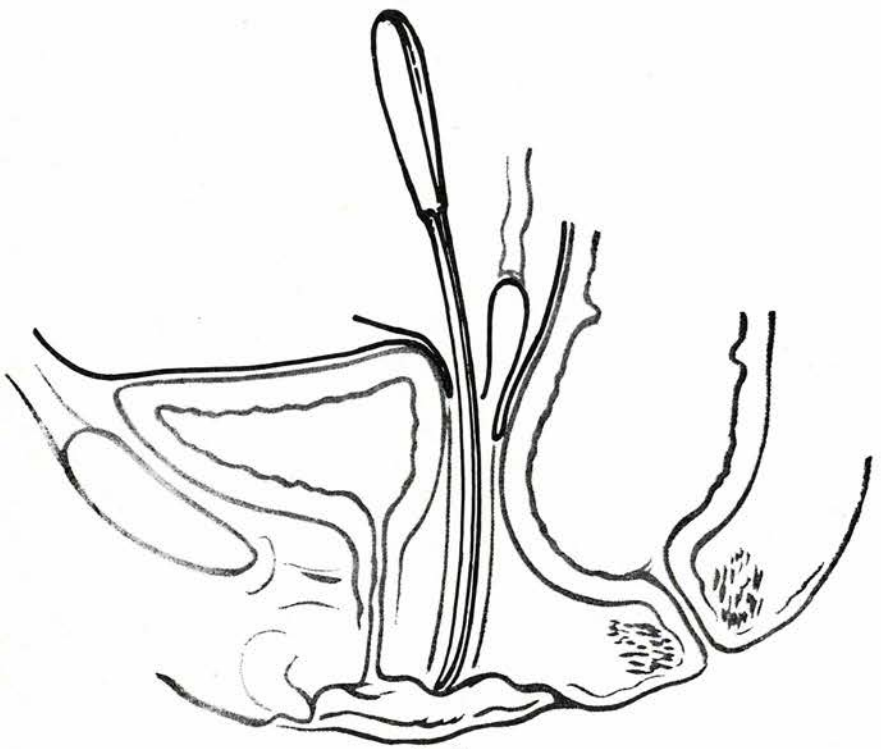

FIG. 2

Clinical Exper. Obst. & Gynecology
Jan. 1974 Vol. I, pp. 3-8

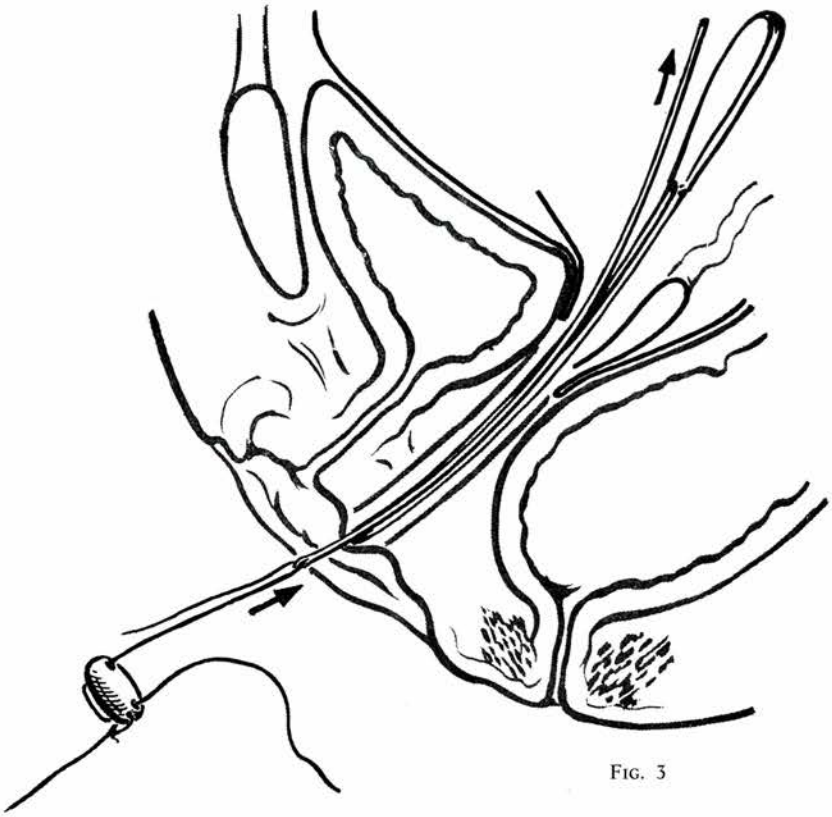

FIG. 3

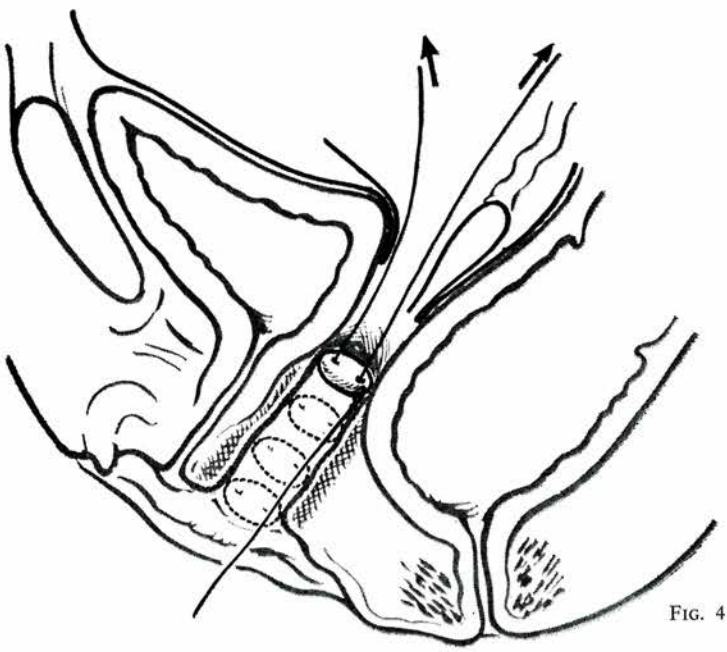

FIG. 4

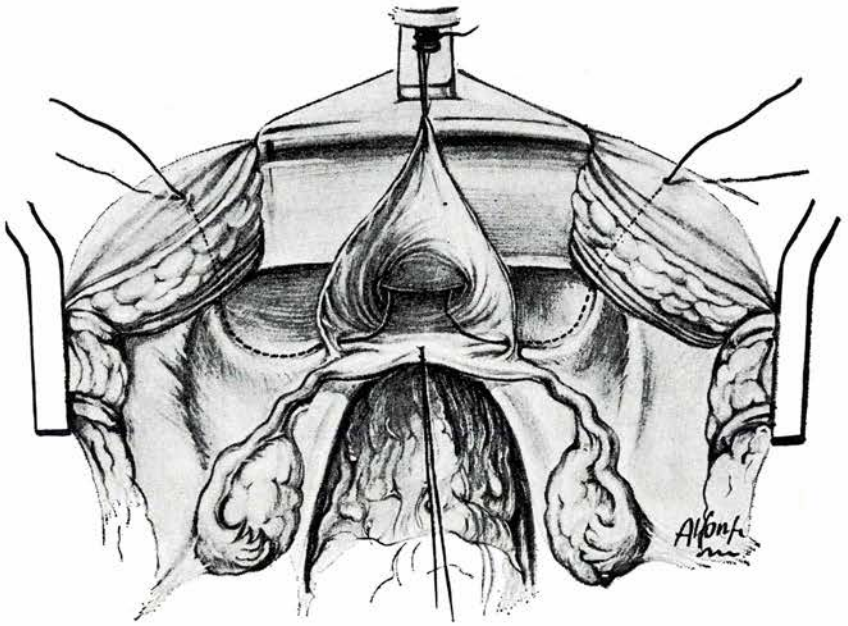

FIG. 5

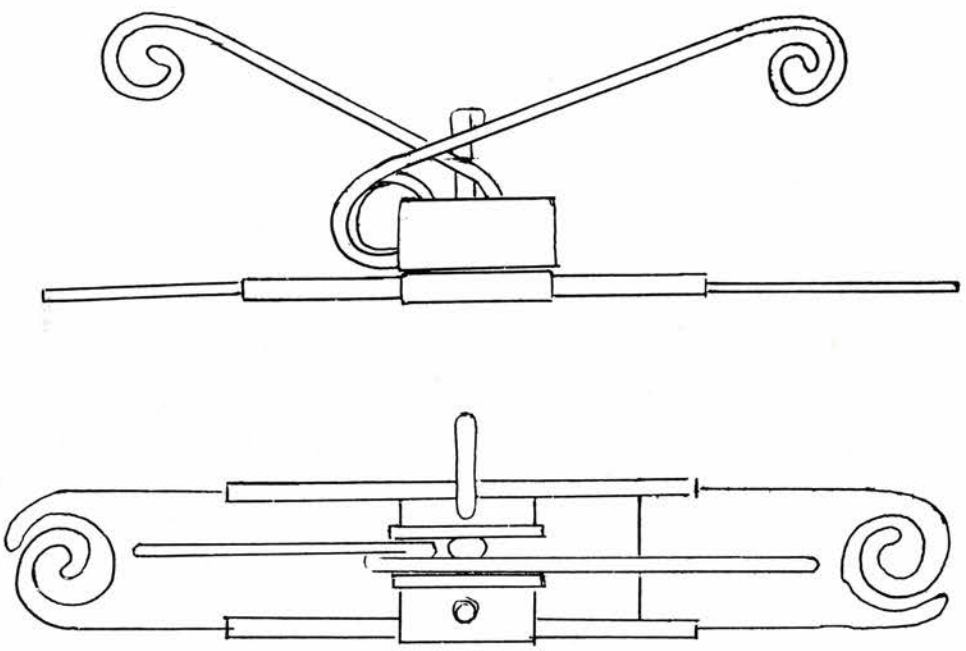

FIG. 6

Clinical Exper. Obst. & Gynecology
Jan. 1974 Vol. I, pp. 8-11

canal some 3-4 cm in length. Traction exerted every 24 hr will result in a final length of 9 cm.

Abouth 7-8 days after the operation, the traction apparatus is removed and the olive and threads are extracted via the vagina. Phalluses of gradually increasing size are then inserted.

This technique has been applied in 159 cases. A functionally successful result was achieved in all but two cases. These were patients who had married without revealing their deformity. The operation was initially successful, but marital differences resulted in abandonment of intercourse and reduction in the length of the vagina. Divorce followed in one case.

With our technique, the newly formed vagina is covered by the mucosa of the pseudohymenal canal as this is gradually stretched by increasing traction. Over a period of time, this new wall acquires the anatomical and functional features of a normal vaginal wall. The cytohormonal picture becomes almost normal; a virtually normal histological and histochemical pattern is also observed and pH values rangers between 5 and 7.

SUMMARY

The Author describs the aetiopathogenetics, anatomo-pathological, diagnostical and therapeutical aspects of the Rokitansky-Küster-Hauser sindrome.

A personal technique of surgical therapy applied on 159 cases is referred.

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The antimetabolic drugs in the treatment of gynaecological cancer

by

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In the study of gynaecological neoplasies chemotherapy has attained a validity of clinical application which, for certain drugs and with the appropriate route of administration and in suitable combinations, is decisive for the treatment of some neoplasms (ovarian or trophoblastic tumours; other uterine tumours, metastases, etc) they are effective although only as a palliative. From this a « combined therapy » has been developed which has been carried out extensively in our school and in which we have had considerable experience ([1, 2, 3, 4, 5]). Chemo-isotope-radio-surgical treatment appears to be especially suitable and to be capable of actual control of some neoplasms of the female genitalia ([6, 3, 4]) and in our

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