

CONGENITAL ABSENCE OF UTERUS AND VAGINA WITH SINGLE LEFT ECTOPIC (PELVIC) KIDNEY

(A Case Report)

by

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Developmental anomalies in the female genital system are fairly common, including the congenital abnormalities of the uterus. However, complete absence of the uterus is, according to Ward-McQuaid and Lennon (1950), rare, although they reported 3 cases in which it was almost completely absent and associated with bilateral inguinal hernia; the hernial sacs contained ovaries in one case and testicular tissue, along with undeveloped tubes and uterine rudiments in other two cases. Jarcho (1946) is of the opinion that in the so-called complete absence of the uterus, the organ is usually represented by at least a ribbon or unilateral tube of the uterine tissue. Congenital absence of the vagina is not a great rarity (Kanter, 1935) but the psychological and social aspects of the problem associated with the malady have led to an evolution of the surgical treatment, which in the words of Counsellor

(1948) constitutes a most interesting chapter in the field of gynaecology. It is important to note that congenital absence of the vagina may or may not be associated with absence of the uterus. Counsellor and Sluder (1944) did not report the absence of the uterus in a single case among the 35 cases operated by them for absence of the vagina.

The urinary and reproductive systems are very closely associated during development, and hence it is not surprising that congenital malformations in one system may be associated with abnormalities in the other system. Thompson and Pace (1937) reviewed 97 cases of ectopic kidney and observed that majority of solitary kidneys are normally located, simple ectopia is apparently associated with a relatively high proportion of congenital absence of the kidney of the other side. They found ectopic solitary kidneys in 8 out of 97 cases and concluded with the observation that genital anomalies are said to be present in about a third of individuals having renal anomalies. Palmer (1933) found a solitary congenital pelvic kidney in 4 of the 32 cases, which he reviewed. Shumaker

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(1938) quoted evidence that genital anomalies in the female occur especially frequently in those with unilateral renal agenesis, probably in about 70% of all females with congenital solitary kidneys. Schreiber and Smith (1946) reported a case of absence of the vagina, uterus, right kidney and ureter with ectopic (pelvic) left kidney. Bryan, Nigro and Counseller (1949) reported a series of 100 cases of congenital absence of the vagina, representing an incidence of 1 in 4,000 female patients in their series. Among 41 of these 100 patients who underwent urological examination, 20 were said to have had normal urinary tract, while 21 had abnormalities such as pelvic or solitary kidneys and the like. All these reports thus firmly establish that there is frequent association of genital and urinary anomalies. Bryan, *et al* (1949) further observed that only rarely in their series were the genito-urinary anomalies associated with abnormalities of other structures.

In Zenana Hospital, attached to S. M. S. Medical College, Jaipur, during the last 10 years, 10 cases of congenital absence of the vagina (including the present case) were treated operatively. In 5 cases, the uterus was also absent. Urological examinations were carried out in 6 cases, out of which 2 cases were associated with solitary ectopic pelvic kidney (including the present case) and double ureters were present on one side in 2 cases.

Case Report:

Mrs. S., a 20 year old girl was admitted to the hospital on 12.7.1966 with complaints of (a) primary amenorrhoea and (b)

inability to have sexual intercourse. She was, physically a typical female having fully developed secondary sex characters. She was married 2 months ago and reported a normal reaction to her husband. The buccal mucosal smear was positive for sex chromatin, which further confirmed that the patient was genetically a female.

The patient was fairly well nourished and the general examination of cardiovascular, respiratory and central nervous system did not clinically reveal any abnormality. The haemoglobin estimation, total and differential W.B.C. count and E.S.R. were within normal limits. Screening of the chest did not show anything abnormal.

On bimanual examination, the vaginal orifice was absent, only one finger could be just dipped in, and the uterus could not be palpated. Examination per rectum also failed to reveal the uterus, but a mass, about 4" x 3" in size could be felt high up in the pelvis on the left side, which was suspected to be an ovarian or a retroperitoneal tumour.

Intravenous pyelography could not be done due to shortage of x-ray films. However, pyelography done post-operatively (Fig. 1) showed the presence of a single left ectopic (pelvic) kidney.

The patient was operated on 18.7.66. An artificial vagina was constructed by the method advocated by Wharton (1938). A transverse incision 1½" long, was made in the region of the dimple between the two labia minora. By blunt dissection and with the help of gauze and fingers the space was deepened, by pushing the urinary bladder in front and the rectum behind. The process was continued till the pelvic peritoneum was reached. Complete haemostasis was attained by ligating the bleeding points. The peritoneum was opened, and through the opening an attempt was made to identify the mass felt earlier per rectum, higher up in the pelvis on the left side. However, this was not possible and the opening in the peritoneal pouch was sutured up.

An exploratory laparotomy was done by a midline subumbilical incision. The mass in the pelvis, on the left side was identified as an ectopic kidney. Biopsy piece was taken from the mass for histological con-

firmation. Both ovaries were normal in size and location, the tubes were rudimentary and the round ligaments were well developed. In the region of the uterus, a midline fibrous thickening was present. The right kidney along with the ureter was found absent. The abdomen was closed in layers, in the usual manner. A self retaining catheter was put into the urinary bladder.

A vaginal glass mould wrapped in thin gauze, 4" long and 2" in diameter was lubricated with antiseptic ointment, pushed into the artificially constructed vaginal space, and strapped firmly in position. The mould was changed every third day, but the gauze was replaced by a rubber condom. The patient developed B. Coli urinary infection 15 days after the operation and was treated with antibiotics. The patient was discharged from the hospital after one month. Patchy epithelialization had already taken place. The patient was provided with rubber covered vaginal moulds made up of gauze and thread, which were fairly firm and stiff. She was instructed to insert the moulds in the vaginal space everyday, after proper cleaning and lubrication with an antiseptic ointment. The patient reported back to the hospital after 3 months (20.11.66) for a follow up examination. Complete epithelialization of the vaginal space had taken place and the artificial vagina was found to be quite spacious. On bi-manual examination, the ectopic kidney could be felt on the left side of the pelvis. The patient was sent home and advised to use the vaginal mould for dilatation every day for another 3 months, and clean the vagina by plain luke warm water douches. On her next follow up visit to the hospital on 2.4.67, the patient was happy and relaxed in being able to have sexual intercourse, to the mutual satisfaction of the partners.

Comments

Patients with congenital absence of the vagina, but with normal ovaries and fully developed secondary sex characters, as they most often happen to be, are genetically, hormonally and psychologically typical fe-

males. The mere knowledge of the fact that they are incapable of cohabitation with the opposite sex, in spite of their otherwise normal attraction and reactions to the male partner, puts a heavy psychological strain, and such patients most often suffer from an inferiority complex. This realization has led to ingenious attempts to construct an artificial vagina, and it has been seen that such patients tend to marry if they find an opportunity to do so. The present patient, however, was already married.

All attempts to reconstruct an artificial vagina require:-

1. Provision of a space between the urinary bladder in front and the rectum behind. This has been achieved by:-
 - (a) A non-operative procedure introduced by Frank in 1938, subsequently modified by him in 1940 through which he was able to obtain 3½" as maximum depth of the vagina by using graduated tubes pressed against the perineum, downwards and inwards ½ hour everyday, for a specified period.
 - (b) Operative procedures which require a blunt dissection in the region between the urinary bladder and the rectum.
2. Provision of an epithelial lining for the artificially created space. Several procedures have been adopted to achieve this object:-

(a) Part of intestine may be brought down along with its epithelial lining. Baldwin (1904) brought down a double loop of the ileum. The procedure was associated with high mortality, and in addition the intestinal mucosa often secreted a very irritating and disagreeable discharge. Schubert (1911) utilized the lower segment of the rectum, but frequent damage to the rectal sphincter during the operation often resulted in a faecal fistula. Shirodkar utilises a segment of the sigmoid and claims excellent results. In spite of its results, this is a major operation associated with high mortality and complications.

(b) Whole skin grafts have been used with success for quite some time. Beck (1900) was the first to use a single stage pedicle graft from the thigh. A multiple stage procedure was adopted by Frank and Geist (1927), and Grad (1932). Jewett (1904) advocated the use of a labial graft, but this procedure became more popular when Graves (1908) combined the use of two labial flaps and two thigh flaps over a glass mould. The device, though practised with

success is more complicated than the modern simpler procedures. Williams (1964) devised a brilliantly simple technique by forming a tube of the skin in the region of the vaginal pit, which is richly endowed with nerve endings designed for response to sexual stimulation.

(c) Use of free skin grafting methods was first advocated by Kirschner and Wagner (1930) who used Thiersch graft on a rubber sponge prosthesis. McIndoe and Banister (1938) similarly used the graft over a vulcanite mould. Bonney and McIndoe (1944) performed about 30 and Counseller and Sluder (1944) about 35 such operations, using skin covered moulds. The patients may have regular menstruation after the operation.

(d) The space opened up by blunt dissection may be allowed to epithelialize by itself, while the space is held open by some substance. Dupuytren (1817) used a tampon to keep the space patent. Similarly, petroleum gauze was packed in the vaginal space by Wells (1935) and iodoform gauze was used by Kanter (1935). Wharton (1938) used a mould made of paraffin or balsa

wood, covered by a condom. However, soon afterwards Wharton (1940) advocated the use of Thirsch graft over the mould, since the graft helps to reduce the formation of scar tissue. Miller, Willson and Collins (1945) also prefer the use of a graft because, according to them, the vagina thus formed is more soft, pliable and better epithelialized.

3. Prevention of contracture poses an important problem in case of an artificial vagina. Most of the cases of stricture and shortening of the newly formed vagina result from an early removal of the vaginal mould. The vaginal mould should be used for a period of 3-6 months or longer, as suggested by Counseller (1948), and the patient should be advised to use vaginal dilators regularly thereafter till she is married. Improper epithelialization leading to scar formation, and perivaginal inflammation are important contributing factors towards the shortening and stricture of the artificial vagina.

Most of the post-operative complications can be avoided if due care is exercised during the operation, while others can be easily dealt with. The usual complications met with are:-

1. Early extrusion of the mould. The mould should

be replaced and firmly secured in the vaginal space.

2. Urinary infection is fairly common, and should be treated in the routine manner with antibiotics.
3. Recto-vaginal and urethro - vaginal fistulae usually occur as a result of pressure necrosis, caused by the mould. It is important to create the vaginal space in the normal direction of the vagina i.e. in the plane of the pelvis when placed in the normal anatomical position.
4. Haemorrhage and vaginal granulations. It is important to obtain complete haemostasis during the operation. Granulation tissue, in the areas where epithelialization has not occurred, should be removed with curette or diathermy loop.
5. During sexual intercourse there might be trauma to the ectopic pelvic kidney (Counseller, 1948). In such cases, with the kidney low down in the pelvis, it may not be possible to provide a proper depth of the vagina.

Out of the 10 cases of absence of vagina treated operatively in our hospital, 6 were operated by Wharton's method (1938), and in 3 cases, McIndoe's method (1938) of using skin graft over a mould was used, but only one case took the graft success-

fully. Williams method (1964) has been successfully used recently in one case and the patient remained in the hospital only for 10 days. According to our hospital records, urinary infection and early extrusion of the mould constituted the commonest complications. The mould was passed per rectum, after an enema, through a recto-vaginal fistula in one case. The fistula was repaired but the patient died due to congenital heart disease.

The techniques for operative reconstruction of the vagina, which are simple, and the available data on the subject dealing with the necessary precautions to prevent complications, which can be easily adopted, thus offer a lot of hope for the unfortunates born with absence or various forms of atresia of the vagina.

Summary

A case of congenital absence of the uterus and vagina with single ectopic (pelvic) kidney, normal ovaries and rudimentary fallopian tubes has been reported. In this case, an artificial vagina was constructed by the method advocated by Wharton (1938), with success.

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Fig. on Art Paper VIII