# UNICORNUATE UTERUS WITH ECTOPIC TERATOMATOUS OVARY AND ECTOPIC KIDNEY

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In the same week that the report of a case of true unicornuate uterus, with a review of the literature by Ogilvie[1] was received, we had operated on a patient in whom, in addition to the typical morphology of a right-sided unicornuate uterus, a dermoid cyst in an ectopic left ovary plus an ectopic left kidney were found. This combination of findings has not been reported before, and as only 4 cases out of a total of 53 recorded unicornuate uteri have to date been published from the British Empire,[1-4] the following case report, the first from South Africa, is presented.

## CASE REPORT

S.J., a married Coloured woman aged 20, was first seen on 18 July 1957. She had had one pregnancy, which had terminated in February 1957 with the spontaneous delivery of a stillborn child. The confinement had taken place at home and the immediate cause of the stillbirth was unknown. She could, however, state that she had been in labour for 34 hours, and that the head had been born first.

Her sole complaint was of slight postcoital bleeding since her last menstrual period 3 weeks before she presented herself at the out-patient department. Menstruation was normal in duration and amount, occurring monthly, and was associated with moderate dysmenorrhoea, the pain being felt in the hypogastrium on the right side only.

She had had no operations or serious illnesses in the past.

On examination, no local cause for the postcoital bleeding could be detected. There was no vaginal septum. The uterus was anteverted and found to be markedly dextroflexed and of normal size and mobility. A rounded left adnexal swelling, the size of a tennis ball and cystic in nature, was present.

The patient was admitted to hospital on 22 July 1957 for curettage and ovarian cystectomy. The operation was carried out the next day. Normal curettings were obtained and no endocervical lesion found. At laparotomy, performed through a mid-line subumbilical incision, the findings were as illustrated in Fig. 1—a true unicornuate uterus with normal right tube, ovary, and broad and round ligaments; complete absence of the left tube and broad ligament; an enlarged cystic left ovary at the pelvic brim and, leading from it to the left internal inguinal ring, a short, thick ligament. Between the attachment of this ligament to the ovary, and the attachment of the bladder peritoneum to the uterocervical junction, there was a thin falciform ridge of peritoneum, but no ligamentous structure was palpable in it.

The right kidney was palpable in its normal position, but the left kidney was at the pelvic brim behind the sigmoid mesocolon,

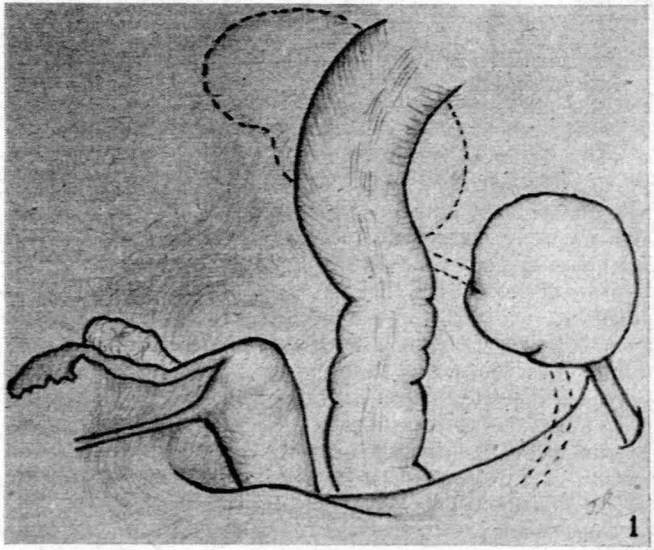


*Fig* 1. Diagrammatic illustration of the findings at operation.

as diagrammatically shown in Fig. 1. The pelvic course of the left ureter could be made out, and appeared to be normal.

A dermoid cyst 2½ inches in diameter, containing hair and

teeth, was removed from the ectopic left ovary and the remaining ovarian tissue conserved.

Histological examination confirmed the nature of the dermoid cyst. The curettings (obtained 30 days after the previous menstrual period) were of non-secretory endometrium and revealed no other abnormality.

The patient's post-operative course was uncomplicated, and she agreed to cystoscopy and retrograde pyelography, which was carried out 4 weeks later. Cystoscopy revealed some distortion of the trigone, the left ureteric orifice being more or less in the mid-line posteriorly, and the right-sided one being more laterally displaced, at about 9 o'clock. Catheters were introduced without difficulty, and the retrograde pyelographic findings are illustrated in Figs. 2 and 3.

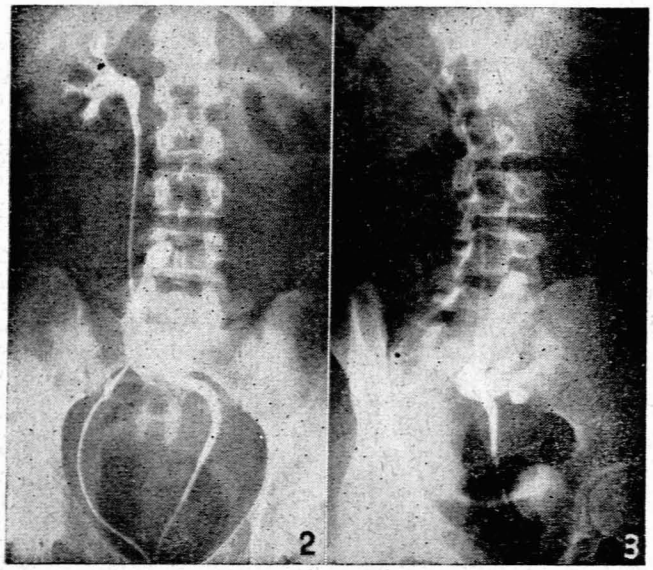

*Fig.* 2. Retrograde pyelogram showing the right kidney in normal position and the left kidney at the pelvic brim in the mid-line posteriorly, over the lumbosacral articulation. Ureteric catheters *in situ*.
*Fig.* 3. Retrograde pyelogram, oblique view showing the ectopic left kidney at the sacral promontory.

Hysterosalpingography carried out another fortnight later confirmed the presence of a right-sided unicornuate uterus with patent right Fallopian tube.

The patient was discharged with instructions to report to the antenatal clinic as soon as she became pregnant again.

## REVIEW OF LITERATURE

For a comprehensive review of the recorded cases of true unicornuate uterus the reader is referred to the recent excellent paper by Ogilvie already referred to above.[1] Of the cases reviewed by him, only one had been suspected clinically, being confirmed at Caesarean section;[5] 13 cases had been diagnosed at laparotomy for various indications, mostly unconnected with the uterus itself; and the remainder of the 53 cases he reviews were apparently found at autopsy. The circumstances under which the diagnosis of unicornuate uterus was arrived at are most interesting and reflect the lack of clinical evidence to suggest such a diagnosis.

With regard to obstetric history, most of the authorities quoted by Ogilvie are agreed that fertility is very little affected by congenital anomalies of the female genital tract, but that there is a somewhat higher incidence of abortion and of obstetric complications in late pregnancy and at delivery. All these aspects are covered very comprehensively indeed by Smith[6] in reviewing 141,946 consecutive patients at the New York Lying-in Hospital from 1899 to 1930. Although he arrives at an incidence of around 1 in 1,500 for cases of incomplete fusion of the Müllerian ducts (other than simple arcuate uterus or septate vagina) amongst this large series of pregnant patients, he encountered no record of a case of unicornuate uterus; this is an indication of the great rarity of the condition. Ogilvie found the obstetrical histories of the reported cases reviewed by him to be very incomplete, but of the 17 married women of whom details were available, 13 had been pregnant 35 times, producing 28 infants and 7 abortions. One in 5 pregnancies ending in abortion represents an incidence of about double that obtaining generally. However, 5 of the 6 women who aborted had also had full-time pregnancies. There therefore appears to be little impairment of fertility in the presence of unicornuate uterus. Schumacker[7] quotes Kussmaul[8] as having cited 14 pregnancies in 4 cases reported, one of whom, Chaussier's patient,[9] had 10 pregnancies.

Although DeLee and Greenhill are of the opinion that pregnancy in a unicornuate uterus tends to be uneventful,[10] our patient's delivery at home terminated in a stillbirth after a long labour, and from our review of the literature it would appear that complications are not uncommon. Birk's patient had 3 consecutive breech deliveries.[11] Tucker and Baker[5] carried out elective Caesarean section and sterilization for their patient, who had a deformed pelvis and had suffered 3 attacks of pyelitis related to her single kidney and ureter. Whittemore's patient[12] likewise was delivered abdominally, for she had conceived only after the construction of an artificial vagina; before this she had menstruated *per urethram via* a fistulous communication between cervix and bladder. At the time of publication she was pregnant for the second time. The patient of Cabanes and Jahier[13] in 5 pregnancies had presented with antepartum haemorrhage 3 times, once necessitating Caesarean section, and postpartum haemorrhage leading to manual removal of the placenta twice complicated delivery. One of the 2 cases reported by Daro, Gollin and Nora[14] had a postpartum haemorrhage after each of her two spontaneous deliveries. Taber[15] reports a case operated upon for a ruptured ectopic pregnancy in a rudimentary left tube suspended with the left ovary from the pelvic brim, who had a right-sided unicornuate uterus with normal right tube and ovary, and who had previously given birth to an infant of 3 lb. 6 oz. at 42 weeks, the small size of which he thought might have been due to the uterine anomaly. These 6 patients are included in Ogilvie's review of 28 viable pregnancies amongst the 13 women of whom obstetrical details are available; the remaining 7 apparently gave birth to 15 infants without recorded complications.

There are no typical menstrual symptoms related to unicornuate uterus. According to Ogilvie, amongst 15 cases where the nature of menstruation was recorded, 3 had never menstruated, 3 had scanty painful periods, 4 had heavy painful periods, and 5 had menstruated normally. Our patient gave a history of normal menstruation with only moderate dysmenorrhoea, *pain being felt only on the side of the cornu.*

Clinically, amongst the reports available to us, *a deviation of the uterus to one side* of the pelvis was noted in 3 cases[12, 15, 16] and in each of these the uterus appeared to be deviated to the right, as was also markedly the case in our own patient.

The various indications for operation in the cases diagnosed at laparotomy make interesting reading, as do the findings. The 3 Caesarean sections have already been referred to,[2, 12, 13]

as well as Taber's case, where the indication was a ruptured ectopic pregnancy in a rudimentary tube.[15] The patient of Daro *et al.*, also referred to above,[14] was operated upon too for a history suggestive of ectopic pregnancy. The tender sausage-shaped mass proved to be an intra-uterine pregnancy in a right-sided unicornuate uterus with normal right tube, ovary and broad ligament. The left ovary and short fimbriated end of the left tube lay at the pelvic brim, and on that side the broad ligament, kidney and ureter were absent. This pregnancy was one of her two which went to term and were complicated by post-partum haemorrhage.

Three cases were operated for appendicitis. Alexander[3] found bleeding from a ruptured follicle of the right ovary, a right-sided unicornuate uterus, and complete absence of the left tube, ovary, and round and broad ligaments, as well as of the left kidney and ureter. Schumacker[7] also encountered a normal appendix, and in his case there was a left-sided unicornuate uterus with normal ovary and tube, an ectopic right ovary with rudimentary tube at the internal inguinal ring, and an absent right kidney. Dannreuther's patient[16] did have a chronically inflamed appendix adherent to a normal right ovary and tube, in addition to a right-sided unicornuate uterus and complete absence of the left broad ligament, tube and ovary. The left ureter could not be felt and was absent on cystoscopy, but the patient refused a pyelogram.

Varino and Beacham[17] operated upon a parous patient for a prolapse and ovarian cyst. After repair of the former, laparotomy revealed a cystic right ovary allowing cystectomy, a right-sided unicornuate uterus, and complete absence of left broad ligament, tube, ovary, kidney and ureter. Ungerleider[18] describes a patient who underwent operation for menorrhagia and fibroids, at which she was found to have fibroids in a left-sided unicornuate uterus associated with normal left tube and ovary, the right ovary, tube and round ligament being fused into a common freely movable stump with no connection to the uterus whatever. Hysterectomy was carried out. An intravenous pyelogram confirmed the absence of the right kidney. This patient had previously been operated upon for a congenital club foot associated with a spina bifida and imperfect development of her right ilium. Guthrie and Wilson[19] describe a case operated upon by the late W. J. Mayo with the removal of a large abdominal tumour consisting of a haematocolpos distending the upper vagina, a right-sided unicornuate uterus and normal right tube and ovary. The left tube, ovary and kidney were absent, as was the lower end of the vagina. The patient had never menstruated but had experienced monthly colic. She was aged 30. Intercourse had taken place per urethram and had been difficult at first but quite satisfactory later!

Ogilvie's patient[1] was undergoing hysterotomy and sterilization for chronic nephritis, only the right kidney functioning and showing marked hydronephrosis on pyelography. She had reached the 23rd week of her first pregnancy. Findings at operation were a right-sided unicornuate uterus and absent left tube, ovary, broad and round ligaments, and kidney. The patient died 2 years later.

Neerhut[4] describes the findings in a patient who underwent laparotomy for an acute abdomen and died from complications unrelated to her genital anomalies. She had a left-sided unicornuate uterus with absent right broad ligament and tube, except for a small piece of the fimbrial end attached to an enlarged ectopic right ovary at the pelvic brim. Her right kidney and ureter were absent, but the adrenal on that side was normal in size and situation.

*Conditions on the defective side.* From the literature available to us, we have been able to extract complete particulars with regard to the ovary and tube on the defective side in 37 cases, including our own. In 16 cases the ovary and tube were completely absent on the side where no uterine horn had developed. In another 16 cases an ectopic ovary and rudimentary tube were present, the latter in almost all cases being no more than the fimbriated ostium. In 2 of these 16 cases the ectopic ovary and tube were found in hernial sacs; in the others the situation was either above the pelvic brim or just below it near the internal inguinal ring. In another 2 of the latter 16 cases ovarian and tubal elements were described as fused in one solid rudimentary structure. In the remaining 5 cases, including our own, an ectopic ovary was present on the defective side but any evidence of tubal rudiments was entirely absent. In practically all cases, a strong ligamentous structure is described in the reports of those cases where there is an ovary present on the defective side, linking it with the internal inguinal ring, as in our patient. This is variously designated 'ovarian' or 'round' ligament by the different authors. We like to think of it simply as the gubernaculum which has never had the opportunity of differentiating itself into the abovementioned 2 ligaments by reason of the fact that there has never been a uterine cornu for it to acquire an attachment to.[20] For the same reason we feel that the attenuated 'round ligament' described by some of the authors referred to as running from the internal inguinal ring to the point of attachment of the bladder peritoneal reflexion to the cervix is no more than a thin falciform ridge of peritoneum extending from the cervix to the side wall of the pelvis and representing a vestigial broad ligament. We took care in palpating it in our patient and were unable to feel any ligamentous structure in it. It is our opinion that, if anything, 2 ligaments should be palpable in the broad ligament, representing both ovarian and round-ligament components, that search should then reveal an unsuspected rudimentary uterine horn in the region of the vesico-cervical reflexion, and that any direct ligament between ovary and internal ring as short and thick as described in these cases, including our own, would then be incongruous.

*Genital and urinary anomalies.* The frequent association of congenital anomalies of the genital and urinary systems is well-known. Where a genital malformation is present, the majority of patients also suffer from renal agenesis on the defective side. Of the 54 cases of unicornuate uterus reported to date, including our own, 39 had complete absence of the kidney and ureter on the side which lacked a uterine cornu (27 on the left and 12 on the right side). One patient whose left kidney was absent had in addition her right kidney in an ectopic position at the pelvic brim.[14] This is the only reference to an ectopic kidney in association with unicornuate uterus, other than our own, and is the second case reported by Daro *et al.* It differs from ours in that the ectopic kidney was the only one. The patient had died from chronic nephritis, and the anomalies had been discovered at autopsy. Ogilvie[1] refers to 2 further cases with rudimentary and U-shaped kidneys respectively in association with a unicornuate uterus. Our patient with a normal right kidney and ectopic but otherwise normal left kidney on the same side as the genital defect, therefore appears to be unique in having escaped a gross defect of her urinary tract, yet brings the total of urinary tract

anomalies associated with unicornuate uterus to 42 out of the 54.

We have been unable to find any previous report of a case of unicornuate uterus with the additional finding of a dermoid cyst or other specific cyst or tumour of either ovary.

Many theories have been advanced to explain congenital anomalies of the female genital tract and the association with urinary tract defects. They are of absorbing interest, but no one theory describes all the degrees of malformation. For discussions on these, the reader is referred to the papers of Ogilvie[1] and Neerhut.[4]

SUMMARY

A case of true unicornuate uterus associated with a dermoid cyst in an ectopic ovary, as well as an ectopic kidney, both on the same side as the uterine defect, is reported. The case appears to be unique in respect of each of these associated findings. The literature is reviewed with particular reference to:

Pregnancy in unicornuate uterus.

Clinical findings which might suggest the diagnosis.

The indications for operation in cases diagnosed at laparotomy.

The particular findings with regard to morphology of the ovary, tube and broad ligament on the defective side.

Associated defects of the urinary system.

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