A CASE OF UTERUS DIDELPHYS WITH UNILATERAL GYNATRESIA

BY

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Developmental anomalies in the female genital tract can arise either from a failure of canalization or from a lack of fusion of the Müllerian ducts. The grosser examples of congenital abnormality are rare, and a recent case in which there was complete duplication of the genital tract with unilateral atresia and retention of the menses would seem to merit description.

Case Report

An unmarried girl aged 17 was admitted to hospital in August, 1948. Menstruation began at 15. The cycle was regular 5/28 and the loss normal. The only complaint was of indefinite backache for approximately one year, it being a little worse at the beginning of each period. Her doctor discovered an abdominal swelling and sent her to hospital. There were no other symptoms, micturition was normal, and the bowels were regular.

On examination the patient looked well and outwardly was normally developed. Abdominal examination revealed a tense, cystic, regular swelling arising from the pelvis and reaching to the umbilicus, more to the right than to the left of the midline and relatively immobile. It was dull to percussion and there was no souffle. Per rectum a cystic tumour could be felt anteriorly and to the right, largely filling the pelvis.

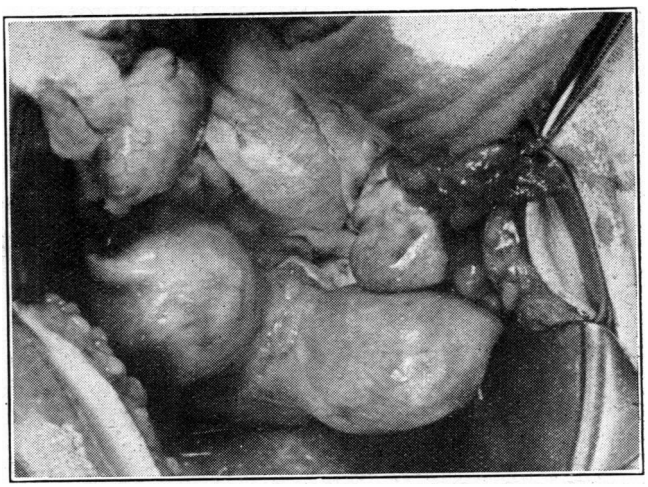
Examination under anaesthesia showed the vulva apparently normal and the hymen intact. On digital exploration the vagina was found displaced to the left and posteriorly by a cystic swelling, tense, bulging down the right anterior vaginal wall, and continuous above with the abdominal tumour. The lower pole of the swelling was a little softer than the rest. The vaginal vault and cervix could not be reached, and the uterus was not identified.

At laparotomy the pelvis was found to be occupied almost completely by a large, smooth, rounded swelling—which was largely extraperitoneal—rising to the umbilicus and presenting in the wound. It was identified as the enormously distended right vagina. Sitting on its summit were two smaller and quite separate swellings, each clearly a uterus. The left was of average size and consistence with normal appendages, the right softer, distended, and one and a half times as big, with normal attachments of the right tube, ovary, and round ligament. The right tube was distended to three times its normal size by an organizing haematosalpinx and was irreparably damaged. Right salpingectomy was performed—the right ovary being conserved—and the abdomen was closed.

With the patient in the lithotomy position a crucial incision was made in the lower extremity of the bulging right antero-lateral wall of the vagina. After excising a thin wall of tissue, approximately 36 oz. (1 litre) of dark "tarry" blood drained away, at first under pressure, and the abdominal tumour was rapidly reduced in size. The edges of the occluding membrane were excised and sutures inserted. Prophylactic chemotherapy was instituted, but the post-operative progress was uneventful, and vaginal discharge ceased on the sixth day. Urinalysis showed no gross abnormality in a catheter specimen. The blood urea was 38 mg. per 100 ml. Excretion pyelography showed the left renal tract within normal limits. The right side was not seen. On cystoscopy indigo-carmine excretion was satisfactory from the left side. There was no right ureteric orifice.

Six months later, examination under anaesthesia showed the left vagina and uterus to be normal; the right uterus had contracted down to a similar size, and the right vagina opened into the lower end of the left, on its right anterior aspect, immediately above the hymen.

A second laparotomy was performed, and showed two completely separate uteri, the left tube and both ovaries being normal (see illustration). The two vaginae were side by side.



Second laparotomy, showing two uteri completely separate, normal left tube and ovary, and right ovary.

After division of the right broad ligament and the vesico-rectal fold the peritoneum was divided immediately above its utero-vesical reflection and the bladder separated. The right ureter could not be identified and the right kidney was not palpable. The right uterine artery having been divided, mobilization of the vagina was completed. The right vagina was opened and right hemi-hysterectomy completed, together with the removal of all but the lower end of the vagina. The right round ligament was sutured to the remaining uterus, the peritoneum reconstituted so as to restore normal anatomical relationships, and the abdomen closed. Excision of the lowermost end of the right vagina was completed from the perineum and the cavity obliterated. Convalescence was uneventful.

Comment

There can be no doubt of the infrequency of complete duplication of the genital tract—uterus didelphys; complicated by unilateral atresia with menstrual retention and a haematocolpo-metrosalpinx the condition must be exceedingly rare, and very few cases can be found in the literature. Brown and Brews (1930), in a review of 50 cases of congenital retention of the menses, recorded such a case in a girl of 16 with an abdominal tumour to the umbilicus. Laparotomy was followed by "excision of the hymen," but the patient died of infection. Purslow (1922) described a probably similar case in a 16-year-old girl treated satisfactorily by removal of the affected uterus and appendages with vaginal drainage; the size of the tumour was not stated, but it was confined to the pelvis.

The exact nature of Wilson's (1925) case of a girl aged 14 does not appear to have been recognized, but probably there was complete duplication of the genital tract, and the pelvic tumour removed (together with both uterus and appendages) was presumably a haematocolpos. Simon (1928) reviewed 23 cases of haematometra and recorded one example in a patient with an "incomplete uterus didelphys." He further quoted Quénu and Le Sourd (1926) as reviewing eight cases of uterus didelphys with haematometra. However, there is a confusion of nomenclature pervading the literature, and the term "uterus didelphys" is variously employed. It usually implies persistence of the two Müllerian ducts in their entirety, but is reserved by some authorities for those extremely rare cases in which the

external genitalia are duplicated too, whilst others use it in cases in which duplication is incomplete.

The association of congenital abnormalities in other systems with defects in the genital tract has been commented on by many authors and was recently emphasized by Burton-Brown (1948). In particular, various developmental errors are to be found in the urinary organs—a fact which is not surprising, bearing in mind that the genital and urinary organs are so closely associated in their development.

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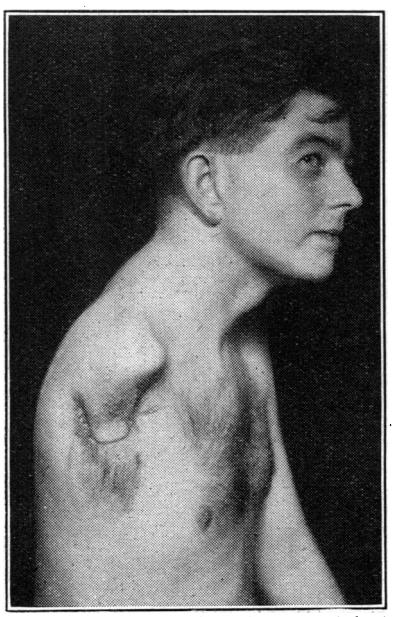
Medical Memoranda

Traumatic Amputation of the Upper Limb

The following case of avulsion of the upper limb is interesting enough to be placed on record.

CASE REPORT

A Civil Servant aged 26 was driving his motor-cycle on December 1, 1948, when he ran into the back of a vehicle carrying tree trunks and sustained almost complete avulsion of his upper limb and compound fracture of his right tibia and fibula. He was admitted to the Edgware General Hospital at 9.45 a.m. in a very shocked condition. A little blood was coming from a large wound in the right axilla. The arm was cold and there was no radial pulse. The right leg was externally rotated and markedly shortened. A blood transfusion was given and he was taken to the theatre at 6.30 p.m.



Photograph taken after the wound had healed.

On exploring the large axillary wound it was found that the arm was connected to the body by skin and a few shreds of muscle of the posterior axillary wall. The exposed muscles of the limb were blackened and oedematous. The axillary vessels, which were found to be torn across but not bleeding, were ligated. When the amputation was completed it was found that the glenoid fossa had come away with the arm. The skin flaps were trimmed and the wound closed with drainage. The fractured right leg was reduced and enclosed in plaster-of-Paris.

The patient made a slow but steady recovery and was discharged on January 29, 1949. The accompanying photograph shows the final result.

Several cases were mentioned by Paul Berger in his *Amputation Interscapulo-thoracique* published in 1887, among them that originally recorded by W. Cheseldon in *The Anatomy of the Human Body* in 1756, concerning a miller, Samuel Wood, whose right arm was torn off when caught in the machinery of his mill. References to other cases in which the arm was torn off with or without the scapula are given below.

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Unilateral Spinal Accessory Nerve Palsy caused by an Arm Sling

Spillane (1949) draws attention to the problem of the aetiology of unilateral spinal accessory nerve palsy. The following case may throw light on the causation of the palsy in some instances.

CASE REPORT

A man aged 29 slipped and fell while running over mud during the monsoon in India, the right arm being violently extended above the head in falling. The greater tuberosity of the right humerus was fractured, but as the position of the fragments was satisfactory the arm was immobilized only by means of a large arm sling and he continued his duties. After wearing the sling for six days the left shoulder, which bore much of the weight of the injured right arm, became sore.

The sling was worn intermittently for another nine days and was then discarded on account of a now persistent ache in the left shoulder, while the injured right shoulder was relatively comfortable. The aching in the left shoulder became more pronounced during the succeeding six days and then it suddenly became agonizing. The pain was present over the left side of the neck; it limited movement of the head and prevented sleep for two nights. At the time the condition was thought to be an attack of fibrositis. The pain diminished over the course of the next three to four days, but weakness of the left shoulder was now experienced. An ache in the shoulder remained, and this was aggravated by exercise. Any lifting, and even such acts as carrying a medium-sized book in the left hand or walking with the arm by the side, caused aching and made the patient conscious of weakness of the limb.

Wasting of the upper portion of the left trapezius was first noted two weeks after the so-called attack of fibrositis, and four weeks after this attack the trapezius had become "paper thin," the left shoulder had dropped 2 in. (5 cm.), and there was winging of the scapula, which at the same time was rotated so that the axillary border was nearly horizontal. It was impossible to raise the abducted arm above shoulder level. No wasting of the sterno-mastoid or other muscles became apparent and there were no sensory changes.

The patient continued to lead an active life; he found it more comfortable, however, to walk with the left hand on the hip or tucked inside the buttoned jacket so as to take some of the weight off the affected shoulder. The weakness of the shoulder gradually diminished, and four months after the "fibrositic" attack there was some noticeable increase in size of the left trapezius muscle, though pronounced wasting was still apparent on comparison with the opposite side. The inequality of the two muscles remained, though in diminishing degree, until 11 months after the "fibrositic" attack, by which