

THE DOUBLE UTERUS

A REVIEW OF TWENTY-ONE CASES

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The normally developed uterus results from the fusion of the intermediate segments of the paired Mullerian ducts to form a double-barrelled structure, followed by breakdown of the median septum to give a single uterine cavity (Koff, 1933). Failure of fusion at different stages of development gives rise to varying degrees of apparent duplication of the uterus, ranging from minor division (arcuate uterus) to complete non fusion (uterus didelphys). Persistence of the median septum results in the septate uterus.

True duplication of the uterus arising from two pairs of Mullerian ducts is found in extremely rare foetal monstrosities (Schiller, 1950).

AETIOLOGY

The cause of the double uterus is obscure. Many theories have been propounded such as defective germ plasm, hormone derangement and unfavourable environmental factors. An hereditary tendency has been suggested by Way (1945).

A striking parallelism exists between the various types of double uterus and the normal uterus in lower animals. As a rule the lower the species on the mammalian scale, the less the tendency to fusion. Thus the marsupials, carnivora, and some apes have uterus didelphys, uterus bicornis and uterus arcuatus in that order (Jarcho, 1946). The similarity is so great that Blair Bell (1934) regards the double uterus as an atavism rather than a malformation.

INCIDENCE

Smith (1931) studying the incidence of double uterus in the New York Lying-in Hospital over the period 1899-1930, gave an overall incidence of 1 in 1,500.

Hunter (1950) found that the frequency in gynaecological practice was about 1 in 3,000. In a survey of autopsy records at the London Hospital from 1907-1955 reported by Brews (1957) the incidence was 1 in 950.

COLLECTION OF MATERIAL

Twenty-one cases of double uterus were found in the operation records and autopsy reports from the Royal Victoria and Royal Maternity

Hospitals, Belfast, over the 10-year period of 1947 to 1956. As the source of material was highly restrictive any calculation of incidence was unlikely to be of value.

The diagnosis was based on findings at pelvic examination, laparotomy, hysterosalpingography and autopsy. The circumstances under which the cases were detected were as follows:—

In Labour	— 6 cases
Gynaecological investigations	— 6
Miscarriage	— 4
Antenatal examination	— 2
Autopsy	— 2
Appendicectomy	— 1

Hysterosalpingography had been carried out in only 4 cases.

CLASSIFICATION

The 21 cases are classified as follows:—

Corpus duplex cervix duplex —	9 (8 vagina duplex)
Corpus duplex cervix simplex —	9
Corpus arcuatum —	1
Corpus septum —	2 (1 vagina duplex)

The classification is based on that recommended by Monie and Sigurdson (1950) for its simplicity and clinical applicability (Fig. 1). The great majority (18 cases) were corpus duplex, corresponding to uterus bicornis (Read, 1955) or uterus bicornis bicorpus and uterus bicornis unicorpus (Way, 1945).

GYNAECOLOGICAL FEATURES

The great majority of cases were in the childbearing age. Except for a child of 5 weeks, the ages of the remaining 20 ranged from 22 to 51.

The average age of the menarche was 13½ years, corresponding with the figure for British girls (Wilson and Sutherland, 1949). Menorrhagia was a prominent complaint in 8 cases. Of these, 5 whose average age was 43, required hysterectomy, and one had a radium-induced menopause. Dysmenorrhoea was a symptom in 4 patients. Cervical polyps were found in 3 patients—2 endocervical and 1 endometrial in origin.

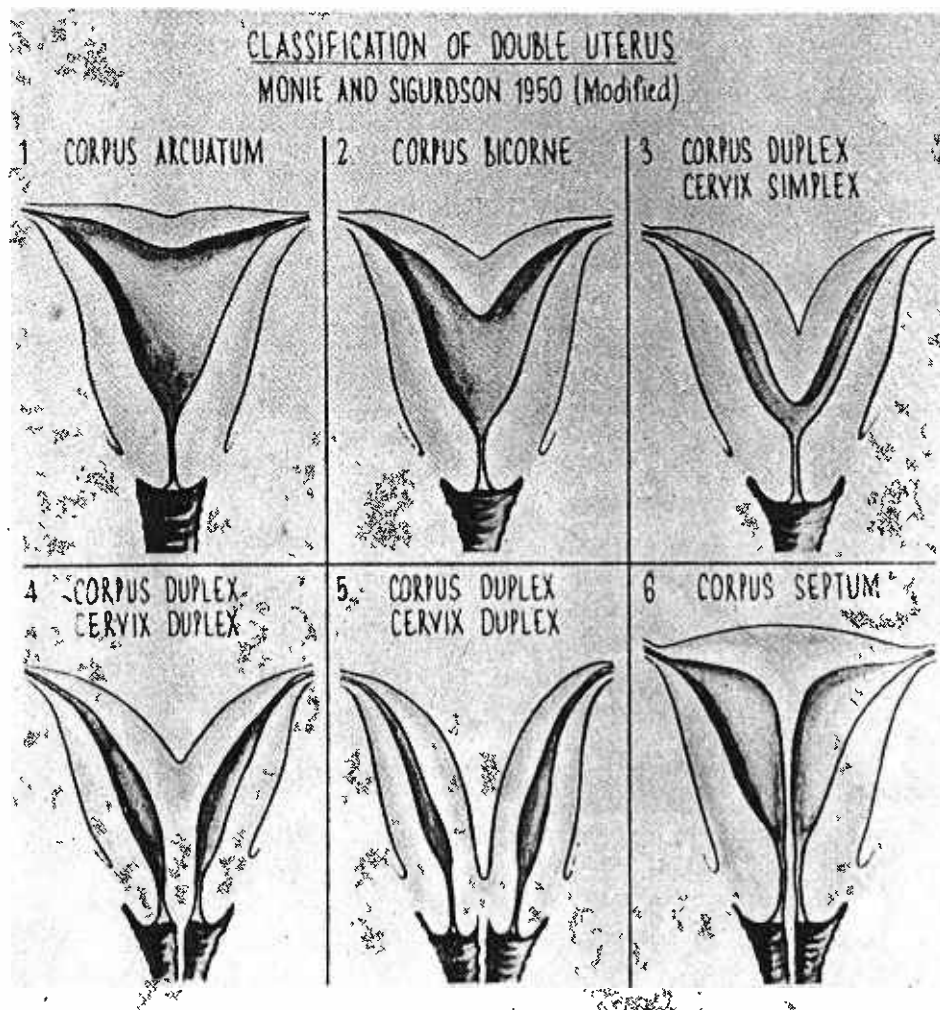


Fig. 1.

There were 2 cases of haematotrachelos due to unilateral cervical obstruction of corpus duplex cervix duplex. In each case surgical drainage was followed by pregnancy; one subsequently delivered 2 and the other 3 children.

Eighteen patients in the series were married. Of these, 16 accounted for a total of 59 pregnancies, an average of 3.7 per patient. Two women were in their 3rd pregnancies at the time of writing, and 7 others were under 30 years of age, in the prime of their reproductive epoch.

The two who were childless had been married for 2 years and 13 years respectively. No cause for the infertility had been detected in the first. The second gave a history of severe external dyspareunia and examination revealed the hymen to be intact.

ASSOCIATED ANOMALIES

There was one case of urinary tract abnormality in a 5 week old baby. At autopsy she was

found to have corpus duplex cervix duplex, with a double ureter on the left side. Intravenous pyelography performed in 3 patients revealed no abnormality.

There were 9 cases of septate vagina, 8 associated with corpus duplex cervix duplex and one with corpus septus.

On three occasions the rectovesical ligament was encountered at laparotomy. This ligament is a connective tissue fold extending from the rectum to the bladder, crossing the junction between the two horns of the uterus.

OBSTETRIC FEATURES

Of the 59 pregnancies, 18 terminated in miscarriage, an incidence of 30.5 per cent. This high incidence was probably due to the presence of intrauterine irregularities or septa which interfered with proper placentation.

Antepartum haemorrhage occurring in 7 patients (18 per cent) was thought to be due to bleeding from the decidua in the non-pregnant horn. There were no instances of

placenta praevia or accidental haemorrhage associated with pre-eclamptic toxæmia.

Breech presentation occurred 15 times (36.6 per cent). It is postulated that in these cases spontaneous version of the foetus was prevented by the abnormally shaped uterine cavity. Transverse lie occurred only once. Way (1945) draws attention to its frequency in minor degrees of failure of fusion.

There were 3 sets of twins, an incidence of 1 in 20.

LABOUR COMPLICATIONS

Of 39 successful pregnancies, 7 terminated prematurely (18 per cent). There was one case of prolonged labour and four of obstructed labour. Two were due to obstruction by the non-gravid horn and these were terminated by Caesarean Section. The other two cases of obstructed labour were due to vaginal septa which held up delivery in the second stage. In each case the septum was divided and delivery completed with forceps.

Forceps delivery was employed six times and Caesarean Section four times. Retained placenta was reported once.

COMMENTS

In the diagnosis of the double uterus, much depends on the individual interest and thoroughness in the procedure of gynaecological examination. For this reason, the condition is often missed: hence the difficulty in computing the true incidence of the anomaly. The use of autopsy material would rule out to a large extent this uncertain personal factor and the figure thus obtained should be reasonably accurate. On this surmise, the series reported by Brews (1957) was studied and the figure obtained was 1 in 950.

In women with double uteri, general sex development, puberty and maturation are essentially normal. Dysmenorrhoea is not usually a prominent symptom, and, if present, it may be one-sided due to unilateral retention of menstrual products (Phillips, 1936). Polymenorrhoea and menstruation during pregnancy arising from the non-gravid horn may also be encountered (Jarcho, 1946).

Menorrhagia appears to be the chief menstrual disorder. It may be explained by the increased area of endometrial shedding. It was incapacitating enough to require hysterectomy in 5 cases and the induction of menopause with radium in one other. Hunter (1950) found this symptom in 5 out of 32 cases of double uterus.

Haematotrachelos, encountered twice in this series, has been reported previously by Brews (1957). Haematometra and haematosalpinx may develop, with subsequent infection or endometriosis and resultant sterility (Strassman, 1952). As a rule, however, fertility is not impaired after the relief of cryptomenorrhoea (Brews, 1957).

An associated vaginal septum is usually symptomless but it may cause dyspareunia, or hinder the ascent of spermatozoa, and thereby possibly lead to infertility. In labour, a septum closely attached to the cervix may impede dilatation, and in the second stage it may hold up delivery. Most septa are symptomless and disrupt during vaginal delivery (Brews, 1957).

The peculiar association of congenital uterine malformations with urinary tract anomalies has long been recognised (Gruenwald, 1941; Collins, 1932; Alexander, 1947). Almost any variety of urinary tract anomaly may co-exist (Whar-ton, 1947).

The rectovesical ligament is a vestigial structure found in association with the double uterus. Its presence may complicate bladder reflexion during hysterectomy (Hunter, 1957) and its close relationship with the bladder and rectum increases the risk of injury to these viscera.

When considering the childbearing potentialities, an average of 3.7 pregnancies per patient among 16 out of 18 married women proves that fertility is not affected. Nine of the cases had a childbearing capacity of another 10 years.

These observations agree with the findings of Smith (1931) and Miller (1922). But the chances of pregnancy continuing to term are significantly reduced as there is a marked tendency to abortion. A high abortion rate was also reported by Strassman, (1952) and Miller, (1922). It is interesting to note that the 18 abortions in this series were confined to 7 patients, of which 2 were secondary habitual aborters. All 7 had one or more living children each, the maximum being four. This shows that given time, the prospects of successful pregnancy are not unfavourable.

During pregnancy, the endometrium in the non-pregnant horn undergoes decidual change. Amenorrhoea is the rule, but the non-pregnant horn may occasionally continue to discharge blood of a menstrual type and simulate threatened abortion and antepartum haemorrhage. Sometimes there may be a colourless discharge (Hunter, 1957) or passage of a uterine cast.

Malpresentation, premature labour, uterine inertia (Fenton and Singh, 1952), obstructed labour (Browne, 1956), and retained placenta

(Way, 1945) have all been reported. The majority of successful pregnancies in the present series passed through the antenatal period safely and 89 per cent delivered by the vaginal route.

MANAGEMENT OF THE DOUBLE UTERUS

Management can only begin after diagnosis. It is suggested that there is room for improvement in diagnostic methods. A pelvic examination should always be systematically performed, with due regard to careful inspection. This routine should be applied to all antenatal patients if cases are not to be missed. The presence of a vaginal septum or double cervix may provide the clue to diagnosis. A double uterus may be diagnosed when a retained placenta is being removed. Cases of unexplained infertility, habitual abortion, premature labour and persistent malpresentation should be considered for hysterosalpingography.

When considering the differential diagnosis of an obscure pelvic mass, it is worth remembering that one half of a double uterus may simulate a uterine fibroid or some adnexal swelling of tubal, ovarian or broad ligament origin (Macafee, 1958).

From the clinical survey, certain general principles in management may be formulated:—

- a. Conservatism should be the keynote.
- b. Symptoms should be treated as they appear.
- c. Major plastic surgery is rarely indicated, and then only under special circumstances.
- d. The possible co-existence of associated anomalies, particularly of the urinary tract should be routinely investigated.

Vaginal septa should be excised in the non-gravid state. If discovered in early pregnancy, it is advisable to wait until the fifth month before contemplating surgery so as to minimise the risk of abortion.

Excision of intra-uterine septa, first performed successfully by Ruge and Schroeder (1882), may be considered in cases of septate uterus with recurrent abortion and infertility.

In every case, a full urological investigation including intravenous pyelography should be carried out (Lash, 1955) and whenever possible, hystero-salpingography also.

PLASTIC UNIFICATION OF THE DOUBLE UTERUS

This operation, originated by P. Strassman (1907), has since been performed with com-

mendable success. His son, E. Strassman (1952) reviewing 128 cases collected from the literature, reported the following result:—

Pregnancy following operation	83
Miscarriage	10
Vaginal delivery	61
Caesarean Section	10
Outcome awaited	2

There was no instance of maternal death or uterine rupture. The incidence of full-term pregnancies rose from 3.7 per cent to 85.6 per cent and abortions fell from 70 per cent to 12 per cent. Dysmenorrhoea and menorrhagia were relieved.

There appears, therefore, to be a definite though strictly limited place for this operation in patients with histories of habitual abortion and infertility, particularly those who are over 30 years. Severe dysmenorrhoea and crippling menorrhagia associated with infertility may also call for operation (Strassman, 1952) if conservative and expectant measures have failed.

In women with intractable menstrual complaints whose reproductive days are over, hysterectomy is the logical line of treatment.

SUMMARY

1. Twenty-one cases of double uterus have been reviewed.
2. Anatomically, the double uterus is malformed and irregular in shape. Its possible complications are repeated abortion, premature labour, antepartum haemorrhage, malpresentation, uterine inertia, obstructed labour, retained placenta, menorrhagia and cryptomenorrhoea.
3. Despite these handicaps, fertility is not impaired and foetal salvage reasonably good.
4. Management should therefore be symptomatic and conservative.
5. Investigation with due regard to associated anomalies, particularly of the urinary tract, has been emphasised.
6. Plastic unification of the uterus has a place in the management of certain cases of intractable abortion and infertility.

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REFERENCES

- Alexander, H.D. (1947) True unicornuate uterus and total absence of left broad ligament, round ligament, salpinx, ovary, kidney and ureter. *Canad. med. Ass. J.*, 56, 539.
- Bell, W.B. (1934) *The Principles of Gynaecology*, Baillière, London, p. 20.
- Brews, R.A. (1957) Some Clinical Aspects of Developmental anomalies of the Female Genito-urinary Tract. *Proc. R. Soc. Med.*, 50: 199.
- Browne, F.J. (1955) *Postgrad. Obstet. and Gynaec.*, Butterworth, London, p. 116.
- Collins, D.C. (1932) Congenital Unilateral Renal Agenesis—*Ann. Surg.* 95, 715.
- Fenton, A.N. & Singh, B.P. (1952) Pregnancy associated with congenital Abnormalities of the Female Reproductive Tract. *Amer. J. Obstet. Gynec.*, 63, 744.
- Gruenwald, P. (1941) The Relation of the Growing Mullerian Duct to the Wolffian Duct and its Importance for the Genesis of Malformations. *Anat. Rec.*, 81, 1.
- Hunter, W. (1950) Double Uterus—*J. Obstet. Gynaec. Brit. Emp.*, 57, 721.
- Hunter, W. (1957) Pregnancy in the Deformed Uterus—*J. Obstet. Gynaec. Brit. Emp.*, 64, 243.
- Jarcho, J. (1946) Malformations of the Uterus—*Amer. J. Surg.* 71, 106.
- Koff, A.K. (1933) Development of the Vagina in the Human Foetus. *Contrib. Embryol., Carnegie Inst. Wash.*, 24, 61.
- Lash, S.R. & Lash, A.F. (1955) Management of Anomalies of the Genital Tract.—*Amer. J. Obstet. Gynec.* 70, 383.
- Macafee, C.H.G. Personal communication.
- Miller, N.F. (1922) Clinical aspects of uterus didelphys. *Amer. J. Obstet. Gynec.* 4, 398.
- Monie, I.W., & Sigurdson, L.A. (1950) A Proposed Classification for Uterine and Vaginal Anomalies. *Amer. J. Obstet. Gynec.*, 59, 696.
- Phillips, M.H. (1936) Unilateral Dysmenorrhoea. *Clin. J.* 65, 48.
- Read, C.D. (1955) *Gynaecology*. Churchill, London, p. 104.
- Ruge, P. (1884) *Ztschr. f. Geburtsh. u. Gynak.* 10, 141.
- Schiller, W. (1950) *Progress in Gynaecology*, Vol. II, Grune & Stratton, New York, p. 7.
- Smith, F.R. (1931) The Significance of incomplete fusion of the Mullerian ducts in pregnancy and parturition, with a report on 35 cases. *Amer. J. Obstet. Gynec.*, 22, 714.
- Strassmann, E.O. (1952) Plastic Unification of Double Uterus. A Study of 123 collected and Five Personal Cases. *Amer. J. Obstet. Gynec.* 64, 25.
- Strassmann, P. (1907) *Zentralbl. f. Gynak* 31, 1322.
- Way, S. (1945) Influence of Minor degrees of failure of fusion of the Mullerian Ducts on pregnancy and labour. *J. Obstet. Gynaec. Brit. Emp.* 52, 325.
- Way, S. (1947) A Further contribution to the study of the influence of failure of Mullerian duct fusion on pregnancy and labour. *J. Obstet. Gynaec. Brit. Emp.* 54, 469.
- Wilson, D.C., & Sutherland, I. (1949) The age of the Menarche. *Brit. Med. J.* ii, 130.
- Wharton, L.R. (1947) Congenital Malformations associated with developmental defects of the female reproductive organs. *Amer. J. Obstet. Gynec.* 53, 37.