THE CLINICAL SIGNIFICANCE OF CONGENITAL UTERINE ABNORMALITIES

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INTRODUCTION

This study, based on considerable clinical material, purports to show the congenitally abnormal uterus as a more significant factor in the complications of pregnancy and labour, and hence foetal mortality, than is generally conceded. This failure to appreciate their significance may be ascribed to the difficulties of physical diagnosis, especially in its less marked forms. However their recognition is dependent upon a knowledge of their clinical manifestations, which tend to show fairly distinctive patterns. They are often repeated in succeeding pregnancies and it is from a familiarity with these that the clinician will be alerted to the possibility of an unsuspected anomaly. An increased ability to recognise the anomalous uterus will allow the complications of pregnancy and labour to be anticipated and by their better management reduce their incidence and so effect an appreciable foetal salvage. Although the medical literature now contains many reports of pregnancy in association with congenital uterine anomalies they rarely contain sufficient material to form an authoritative study. Unfortunately the few comprehensive reviews make use of cases abstracted from the literature to form the major part of their study in addition to a small core of personal records.

Obviously any survey compiled in this way must give a distorted picture for it is only those reports of special clinical interest which appear in the literature and any such study must show an abnormally high rate of complications.

The material in the present study originated from 18 personal hospital records but to extend the scope of the investigation the records of a further six hospitals were examined. Unfortunately two of these were incomplete and statistically valueless leaving sixty-nine examples of congenital uterine anomaly with a total of one hundred and fifty pregnancies. This material was then subjected to a careful statisticaly analysis and the information obtained showing the incidence of abortion, foetal mortality, malpresentation, operative delivery, maternal morbidity and mortality, complications of pregnancy and labour, foetal anomalies and other miscellaneous data all of which were compared with certain normals and similar data from another study. The more academic aspects of the subject are also fully discussed and the relevant British, American and European literature has been extensively examined and the opinions therein collated and integrated with my clinical observations to give a composite and it is hoped objective picture.

HISTORICAL SURVEY

The curious paradox of the human uterus portrayed as a double chambered bicornuate organ appears in all the earliest extant anatomical treatises, and with minor corrections, persisted until the 15th century. The explanation for this anatomical error is not difficult to find when one remembers that all anatomical knowledge of this age was based on animal dissections, human dissection being expressly forbidden on religious grounds.

The Ebers papyrus (1550 B.C.) is probably the oldest surviving anatomical record, and contains a rather crude portrayal of a uterus which is bicornuate in outline and resembles that of a cow. This may appear surprising in view of the high degree of skill attained by the Egyptians of this period in the art of embalming which should have afforded the opportunity of observing the pelvic organs. It is, however, known that the actual evisceration of the cadavre prior to embalming was performed by bondmen drawn from the lowest caste of the community, who were, of course, completely illiterate. In an interesting dissertation Reuter (1914), puts forward an alternative explanation, in which he remarks that the bicornuate uterus was the hieroglyphic symbol for the uterus, and does not necessarily represent an anatomical concept. He does, however, agree

that it is improbable that the Egyptians profited from their knowledge of embalming as human dissection was forbidden by their religion.

Terra cotta donarii or votive offerings are frequently encountered during archaeological excavations of Roman remains; some of these take the form of a double chambered uterus, and are considered as additional evidence of the widespread misconception in uterine topography prevalent at these times. Sambon (1895) however considers they are symbols of fertility or multiple pregnancy which were placed in the temples as tokens of thanks for favours granted by the gods.

Hippocrates (460-370 B.C.) although writing extensively on the subject of gynaecological disease and making frequent mention of the uterus, does not give a description of the genital tract even in his treatise on Anatomy. The terms 'metre' and 'hystere' are both used and usually in the plural form, which suggests that he considered the uterus to have two or three cavities, this being the accepted view of his time.

There were, however, several centres among the ancient mediterranean civilisations where human dissection was practised and of these the most famous was the medical school at Alexandria. Unfortunately not a single treatise of her earlier teachers remains, all having been destroyed when that city was sacked by the Romans, and consequently any knowledge we have of this school is from the work of

subsequent writers. Early in the Christian Era the practice of human dissection slowly declined, and had ceased altogether by the time Claudius Galen came to the Medical School at Alexandria to study anatomy. Galen's fame as a gynaecologist rests solely upon his description of the female genital tract, and being denied the opportunity of dissecting the human cadavre, his anatomical knowledge was gained from dissecting animals. He therefore assumed the human uterus to be bicornuate and likened it to that of the cow or goat, but he did however correct the earlier conception that the uterus contained several cavities.

Cophon (early 12th century) is usually credited as producing the first anatomical treatise in Christian Europe, and as it was based on dissections of the pig (Anatomia Porci) the uterus is again shown as a bicornuate organ.

Claudius Galen's contributions to medicine were enormous and for thirteen centuries he was considered the outstanding authority in all branches of medicine, but it was just this blind acceptance of all his pronouncements which proved such an obstacle to further advancement in medical knowledge. Thus, even with the return of human dissection the earlier mistakes in uterine anatomy were not immediately corrected, for we have Mondino in his Anatomia Mundini (1311) portraying the uterus as having seven cells, as if he were trying to

strike a compromise between the Aristotelean and Galen concepts. As Mondino had dissected several cadavres, it is difficult to ascribe this to anything other than intellectual dishonesty borne through fear of being deemed heretical if expressing dissenting views. Most of the errors of the past thirteen centuries were corrected by Jacobus Berengaris da Carpi (1470-1530) who by many careful dissections attained considerable anatomical accuracy, although a fundal notch is to be found in his illustrations of the uterus. Similarly in the Fabrica Corporis Humani (1543) a faint septum can be seen, as though Vesalius were loath to abandon completely the traditional concepts of uterine anatomy. The first truly accurate anatomical description of the human uterus appeared in 1553, in a set of superb engravings by Bartolomeo Eustachio and the same century (1557) saw the first description of a bicornuate uterus. This, according to Morgagni (1761) was by Francesco Antonio Catti, a Neapolitan anatomist who was also the first to note ossification in the dura mata. Unfortunately, no further information is given by Morgagni, and a search of the literature including the monumental work of Felice la Torre on the Uterus through the Centuries (L'Utero attraverso i secoli 1917) and Viana and Vozza's Biography of Italian Men of Science (L'Obstetricia e la Ginecologia 1935) reveal no mention of Catti. Additional information has been obtained from the Wellcome Historical Medical Library who, in a personal communication (1956), suggest that the word 'dissertation' may imply that Catti was defending a thesis

at a University. This thesis may never have been printed but Morgagni may have seen the record in the manuscript annals which would explain why there are no other references. The final details regarding Gatti were kindly given by Professor Adalberto Pazzini, who in a personal communication (1956) states that he was also known as Francesco Gatta Lucano and it was under this name that his work 'Isagogae Anatomicae' was published in Naples in 1557. This information was gathered by Stefano delle Chiaje and presented to the Italian Academy of Science who published it in the records of their transactions.

The following description of a pregnancy in a rudimentary uterine horn was related by Pierre Dionis, surgeon to the Queen of France and Dauphine nearly 300 years ago. 'A young woman of twenty years fell pregnant in the second month of her marriage, her breasts becoming painful, morning vomiting being present with the usual bizarre desires and revulsions of pregnancy. However there was doubt in her mind as her periods continued regularly although in less amount and it was only when her belly began to grow that she was certain of her state. Movements were first felt at four and half months and at the fifth month the periods were suppressed, although there was occasional slight loss of blood.

1. Genital Tract with Ruptured Gravid Rudimentary Horn from Table 1 - Pierre Dionis 1683.

2. Table 1 - Fig.1.
Uterus Subseptus showing incomplete septum.

Fig.2. External Genitalia in (1) Sagittal Septum dividing introitus.

Eisenmann. From the Tabulae Anatomicae 1752.

TAB. I.

Fig. 1.

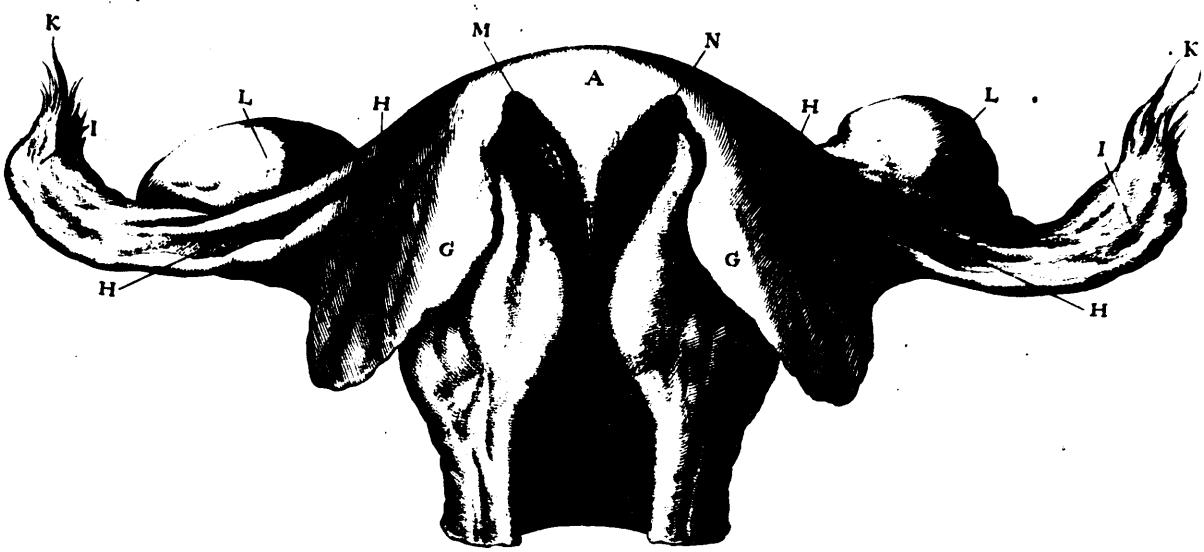
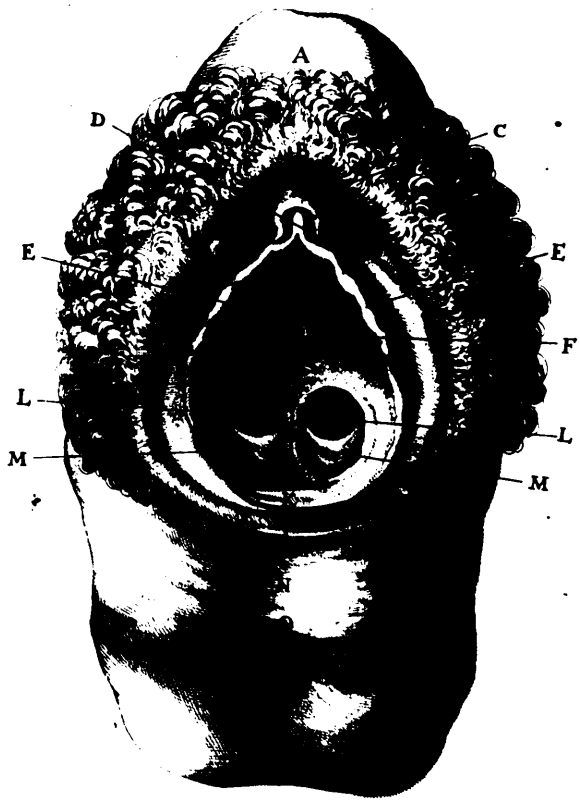


Fig. 2.



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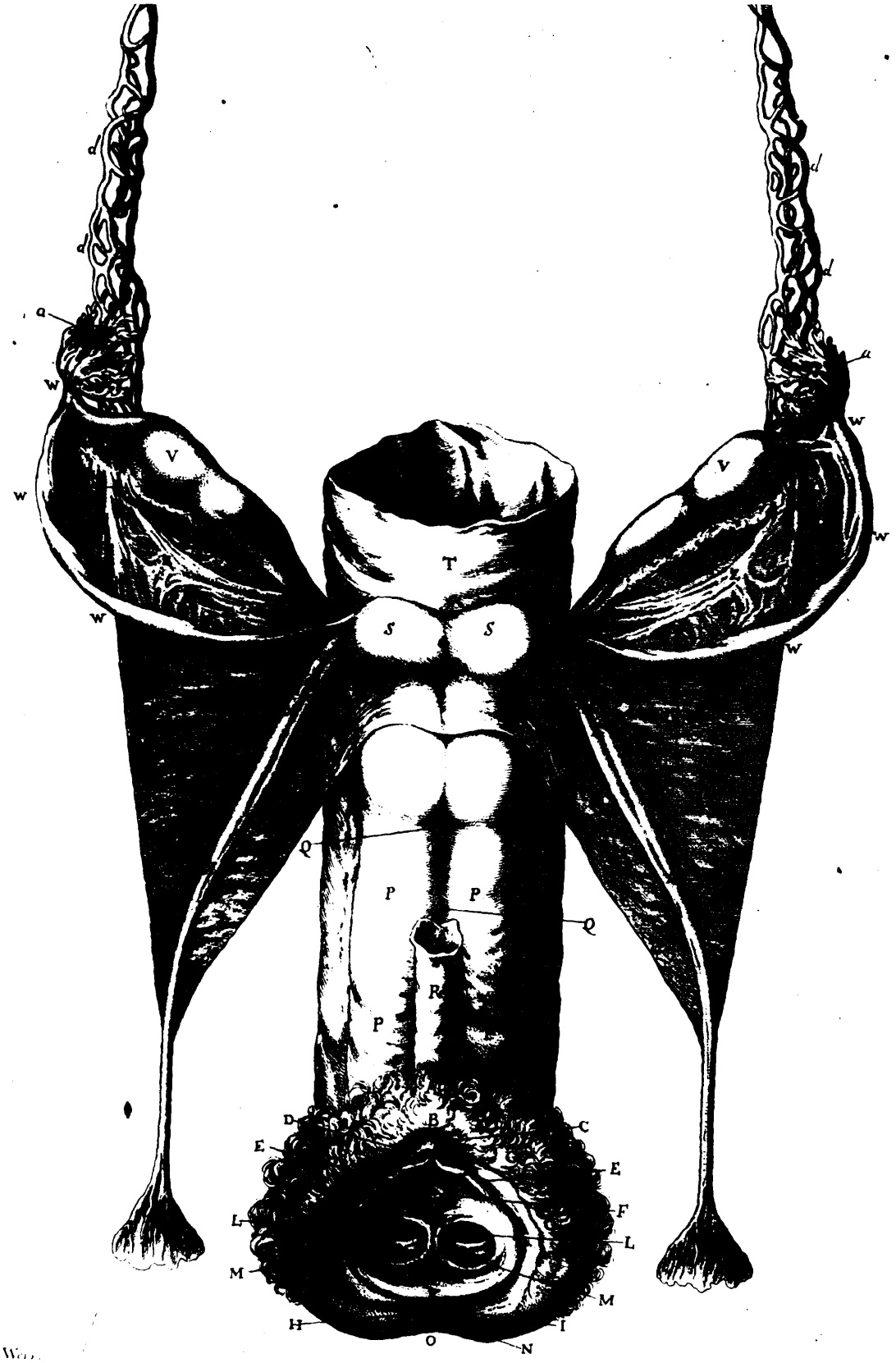

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5. Table 11 - Uterus Bicornis Bicorpus Bicollis with Septate Vagina.
Dissection showing internal and external genitalia.
From the Tabulae Anatomicae of Eisenmann 17

She continued to increase in size and feel movements as all women, but the movements were only felt on the left side and she appeared to be carrying higher than normal. On the evening of the 5th June 1681, she was seized by a severe pain in the belly which raged so fiercely that it was feared that she was going into premature labour. Movements now ceased and there was no further increase in size.

Twelve days after the first crisis she was again taken with severe abdominal pain which made her retch and writhe in agony. A surgeon of the court who lived nearby was called and in spite of all his remedies her agonies continued becoming so severe that it was impossible to warm her extremities. By five in the morning it was clear that she was becoming progressively weaker; she could only breathe with difficulty and her belly was visibly increasing in size. She was placed on a stool and shortly afterwards succumbed. A post-mortem Caesarean section was performed in the hope of saving the child but it was only possible to baptise it.

Word of the tragic death of this young girl, coming to the ears of the court, the Queen and Madame la Dauphine commanded M.Dionis to open the body and to try and ascertain the cause of so sudden a death.' From the post mortem description and accompanying diagram this was obviously a rupture of a gravid rudimentary horn.

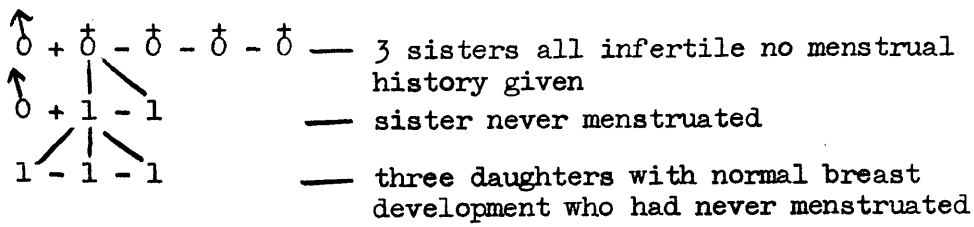
It is stated (Schumann 1936 et al.) that Mauriceau in 1688 reported a description of pregnancy in association with a bicornuate uterus, but reference to the original shows clearly that this was a case of tubal pregnancy.

In this country in 1773 John Purcell, Professor of Anatomy at Trinity College Dublin, published an account of the post mortem findings of a woman dying in the ninth month of pregnancy. Autopsy revealed a uterus with a pregnancy at term to which a normal sized uterus was attached at the level of the internal os and the two cervices were separated by a sagittal vaginal septum. There were only two ovaries and no communication was found between the uteri. This description and the accompanying diagram suggest a case of uterus bicornis bicorpus bicollis with a septate vagina with pregnancy in one horn. Although John Huxham published a tract in 1723, on congenital abnormalities of the genital tract all copies have been lost. Eisenmann(1752) of Strasbourg produced several beautiful engravings illustrating four different degrees of uterine malformations.

With the 19th century congenital malformations came to be studied and explained on an embryological basis, Merkel (1812) being among the first to do this. The classical contributions to embryology by Rathke and Muller give a much clearer understanding of the subject and Kussmaul (1895) was the first to give an exhaustive account of the subject in which both the embryology and physiology were discussed.

Excellent summaries of the literature are given by Saint-Hilaire (1831 and 1832) and Piquand (1912), the latter being the best historical account listing all cases since the 16th century.

The familial incidence of genital malformations was noted as early as 1870, when Phillips of Guy's Hospital reported two sisters who had never menstruated. The elder, aged 20 years, complained of lower abdominal pain at monthly intervals since her marriage four years previously. Breast development was normal and she was hirsute and of small stature, external genitalia appeared normal but the vagina was represented by a short cul de sac, in which there were three small apertures. Neither uterus nor ovaries could be felt on rectal examination. The sister presented similar features but with an even smaller vagina. Nelson(1861) describes three sisters with genital abnormalities and Squarey (1872), gives an account of a family with genital abnormalities through three generations, but unfortunately no mention is made of the actual type of abnormality.



Matthews Duncan (1882) described a case of a multipara who, apart from slight bleeding in the early months of pregnancy, had always passed through pregnancy and labour without trouble. At the ninth confinement a piece of chorion was noted to be missing when the placenta was examined and on digital exploration of the uterus the presence of a double uterus and two cervices was revealed; the right horn which had carried the pregnancy being much larger than the left. He comments upon the fact of repeatedly normal confinements being possible in the presence of such an abnormality. At the same meeting Braxton Hicks related two examples of double uterus and vagina, the former having a normal pregnancy and confinement and the latter requiring division of the vaginal septum before the head could be delivered. In a case similar to that published by Matthews Duncan, Henderson (1883) relates the history of a patient having had seven normal pregnancies uncomplicated apart from bleeding in the early months of pregnancy and a possible tubal abortion with the passage of a decidual cast. He reasoned that if these uterine abnormalities were compatible with normal confinement then reproductive life could quite easily be passed through without the abnormality being discovered. This led to the assumption that these anomalies were probably commoner than generally supposed, and that only by post mortem examination

could any idea of their true incidence be obtained and even then they could be missed. In the space of five years between 1878 - 1883 he found fifteen cases had been reported in the British Medical Journal of which 10 were cases of double uterus and vagina, 3 were cases of double uterus and single vagina, 2 were doubtful.

The M.D. thesis of Thibaud (1892 - 93) was on the subject of uterine malformations and gave the first reference to the similarity between the degrees of uterine abnormality and the uteri of different animal species. The point that these abnormalities were not just scientific curiosities but of clinical importance was stressed.

The historical aspects of surgical correction of congenital uterine abnormalities and the history and development of hysterosalpingography are included under the appropriate chapters.

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THE DEVELOPMENT OF THE OVARIES AND THE FEMALE GENITAL TRACT

THE DEVELOPMENT OF THE OVARIES AND THE FEMALE GENITAL TRACT

One of the more curious features in the developmental history of the vertebrates is the close embryological association between the reproductive and the urinary organs. Indeed, in many ways, as Felix (1912) and others have suggested, the two sets of organs can be regarded as constituting a single system. An attempt to explain this association would involve a detailed account of the genito-urinary apparatus of primitive vertebrates and their presumptive ancestors. It would also involve some phylogenetic speculation of doubtful value. Consequently, in this presentation the association of the two systems will be taken as an established fact in vertebrate morphology. Reference to the details of the association will be restricted to the embryological facts that throw light on the development and structure of the human female genital tract. The necessity for some reference to the development of the urinary system, however, gives an inevitable complexity to any attempt at the description of the development of the female reproductive system.

Another, and again unavoidable, complication in such a description arises from the existence of two sexes and from the presence, in each sex, of portions of the reproductive system of the other sex. An account of the development of this system in one sex, therefore, requires considerable cross

reference to the conditions that obtain in the reproductive system of the other sex. Such cross reference, however, has more than mere academic interest as a number of the pathological conditions and the phenomenon of pseudo-hermaphroditism find their formal explanation in terms of the developmental changes and alterations occurring in both male and female.

For convenience of presentation the subsequent description and discussion of the development of the female reproductive system has been divided into separate sections. These are (1) introductory survey; (2) the development of the ovaries; (3) the mesonephric (Wolffian) and the para-mesonephric (Mullerian) ducts and their derivatives; (4) the urogenital sinus; (5) the external genitalia; ~~and (6) hermaphroditism~~. Reference to abnormal development of different parts of the female reproductive tract will be made in relation to the relevant descriptions of normal development.

INTRODUCTORY SURVEY

The vertebrate urogenital system is, for the most part, derived from the "intermediate" mesoderm or nephrogenic cord. This is the intra-embryonic mesoderm which lies on each side of the early embryonic body, between the somites medially and the lateral plate mesoderm, laterally. The lateral plate mesoderm soon develops an extensive cavitation, which becomes

the intraembryonic coelom. Early in development the "intermediate" mesoderm or nephrogenic cord of each side is at first medial and eventually, with the growth changes in the embryo, dorsal to the corresponding half of this coelomic cavity. Growth of the intermediate mesoderm not only produces an elevation on the surface of the embryo, but also a ridge which projects into the coelomic cavity. This projection (the urogenital ridge) extends, in the earlier stages, along the greater length of the dorsal wall of the coelom lateral to the attachment of the mesentery of the developing gut. Each urogenital ridge is attached to the dorsal coelomic wall by a broad urogenital mesentery.

In the urogenital ridge the three successive excretory systems, known as pronephros, mesonephros and metanephros differentiate. Since there is a marked cranio-caudal gradient in embryonic differentiation, the pronephros develops in the more cranial part of the ridge, to be succeeded, in its intermediate part, by the mesonephros. Finally, the most caudal part of the ridge provides the blastema from which most of the metanephros, or definitive kidney, will be derived. The details of the development of these successive kidneys do not require consideration here, though in later sections some reference must be made to certain aspects of their embryology. The excretory duct systems of the successive kidneys, however, must now be briefly considered.

Pronephros. Each pronephros is formed by about seven pairs of transient tubules which begin to appear in ten somite (late third week) embryos in the nephrogenic cord of the future cervical region. They reach their greatest development in the fourth week and are found opposite the seventh to the fourteenth somites. The duct from each pronephros (called the pronephric or segmental duct) is formed by extension of the distal end of each pronephric tubule caudally where it meets and fuses with the immediately succeeding tubule. In this fashion each pronephros develops a single excretory duct which grows caudally extending beyond the pronephros and eventually reaching and opening into the dilated terminal part of the endodermal hind gut, or cloaca. The pronephros itself is probably non-functional in the mammalian embryo. The pronephric duct, however, has a most significant part to play in the subsequent development of the urogenital system.

Mesonephros. Immediately behind the pronephros, as this vestigial kidney is involuting, a second generation of kidney tissue differentiates from the nephrogenic cord. It is still a debatable point whether this second kidney is a completely separate organ or represents merely a later and continuing development of the pronephros(Frazer,1950). For descriptive purposes this second kidney is conveniently regarded as a new formation and is called the mesonephros. Like the pronephros it is formed by a series of tubules. In the mesonephros, however, these tubules are more complicated than those in the earlier organ

and there are many more of them. They also differ from the younger stage in that there is evidence that they function as excretory organs. Like the pronephric tubules, however, the mesonephric tubules come to open into the segmental or pronephric duct. As a consequence of its new attachment by these tubules to the mesonephros, this duct is now called the mesonephric or Wolffian duct. The mesonephros projects into the dorsal portion of the corresponding half of the coelom as the mesonephric ridge.

Metanephros. The excretory part of the metanephros develops from the portion of the nephrogenic cord (metanephric blastema) caudal to the mesonephros; its collecting tubules develop from the ureteric outgrowth, which arises from the mesonephric duct close to its junction with the cloaca.

At a stage between 4 and 5 mm. (thirty-one to thirty-two days) a hollow outgrowth, the ureteric bud, arises from the postero-medial wall of the meso-nephric duct near its junction with the cloaca. This bud grows dorsally and at the same time its origin migrates to the posterior and, later, to the postero-lateral wall of the mesonephric duct. It will later come to be directed cranialwards as its duct (the ureter) continues to elongate. This elongation is due to active cranial growth of the bud, to growth at its site of origin due to caudal migration of the cloaca, and to interstitial growth. Shortly after its appearance the cranial end of the ureteric bud enlarges and comes into contact

with the metanephrogenic tissue (blastema) of the caudal one-fourth of the nephrogenic cord. Further growth of the bud results in its cranial portion and the blastema being displaced cranially and dorsal to the caudal portion of the mesonephros. Here it is situated retroperitoneally, but anterior to the umbilical artery. The later changes which occur in the cranial part of the ureter and the associated metanephrogenic blastema have as their result the formation of the definitive kidney. They do not require further consideration here.

DEVELOPMENT OF THE OVARIES

Although the sex of the embryo has been determined at the time of fertilization, it is impossible to distinguish between female and male embryos until relatively late in development (17 mm. i.e. at about forty-four days). Embryos earlier than this are often referred to as being in the "indifferent" or "neuter" stage. During this stage the gonads arise in intimate association with the nephric system. The first sign of their appearance is a ridge-like thickening (gonadial or germinal ridge) on the ventro-medial border of each mesonephros. Each germinal ridge consists histologically of a dense blastema of mesodermal cells covered by the mesothelium of the related part of the mesonephric ridge. It is difficult, indeed almost impossible, on histological grounds alone to determine the nature and origin of the cells of the gonadial ridge. Most of the cells of the blastema are

are presumably mesodermal cells of the mesonephric ridge. The mesothelium overlying each gonadial ridge receives the name of the germinal epithelium and its initial origin, too, is clearly from the general coelomic lining. There are, however, two complicating factors in ovarian development which are the cause of a lack of agreement between investigators of the details of the histogenesis and pathology of the ovary. These factors are (1) the primordial germ cells and (2) the relationship, or attempt at relationship, between the mesonephros itself and the gonad. Of these two the problem of the origin, nature and fate of the primordial germ cells deserves, indeed requires, special consideration.

Primordial Germ Cells. A discussion on these cells involves a brief consideration of a biological point of some general significance. As is well known, Weismann (1904), largely for theoretical reasons, considered that an individual organism consisted of two fundamental subdivisions, (1) the soma, or body generally, and (2) the germ plasma, sequestered in the soma, and responsible for the future of the race. Weismann considered that there was evidence for an early segregation of the germ plasma - indeed, that it was probably derived directly from the parental germ plasm. In some organisms (Hegner, 1914; and Bounoure, 1939, 1940) there is good evidence that such a segregation occurs, though not exactly as Weismann postulated. In vertebrates the evidence for such a segregation in the very

early stages of development is still conflicting. In the later stages, however (even in human embryos, see Politzer, 1928a and 1928b, 1930, 1933; and Witschi,1948), there is significant evidence that the primordial germ cells arise outside the gonads, in the region of the caudal part of the wall of the yolk sac and the hind gut, and migrate subsequently to the region of the medial aspect of the mesonephric ridge where they settle down. The fate of these primordial germ cells is one of the most urgent problems of gonadial histogenesis. For some investigators they represent the ancestors of all the subsequent definitive germ cells. Many others, however, consider that the primordial germ cells are transient, disappearing and playing no part in the later development of the gonads. Even the question of where they settle down is at issue. They have been described as reaching the germinal epithelium where, by some mysterious process, they become indistinguishable from other cells of this epithelium but reveal their origin, and true nature, in later stages of ovarian development by proliferating to form cocytes. Other embryologists consider that though some primordial germ cells may reach the germinal epithelium, many remain in the matrix of the ovarian stroma to form the definitive germ cells in later stages. It must suffice here to say that the authors are impressed by the increasing evidence that the primordial germ cells are genetically concerned in the history of the subsequent generations of definitive germ cells.

At the 14 mm. stage (about forty-one days) the blastema of each gonadial ridge becomes subdivided into sex cords by the development of connective tissue trabeculae (Fig.28,A). These cords are at first connected to the overlying germinal epithelium and, so far as is known, they are initially of the same structure and appearance in each sex. After the 25 mm. stage (about fifty-two days), however, a developing ovary can clearly be distinguished from a developing testis owing to the development in the latter of the tunica albuginea. This is a dense fibrous layer which separates and sex cords completely from the germinal epithelium. In the developing ovary, with no definite tunica albuginea being formed, separation of the sex cords from the germinal epithelium is incomplete. Many embryologists consider that this epithelium will continue to contribute cells to the ovarian sex cords throughout foetal life and, in post-natal life, until the menopause.

In the further development of the ovary a rete (ovarii) also appears. Normally, however, it is never as well formed as that in the testis. It can, nevertheless, occasionally establish an imperfect union with certain of the mesonephric tubules. The sex cords in the ovary become much fragmented and it is from the resulting small groups of these cells that the primordial ovarian follicles arise. The primordial ova, at least in part, become encapsulated within these follicles. The encapsulating cells derived from the sex cords of the blastema probably become the

pregranulosa cells. In marked contrast to the testis the coelomic germinal epithelium continues to contribute cells to ovarian sex cords for a long period, possibly into post-natal, or even adult, life. The ovarian interstitial cells appear to arise from the stromal mesenchyme of the orginal gonadial ridge. The cells of the theca interna may also have a stromal origin (Gillman,1948). The absence, in normal development, of an established continuity between the ovarian sex cords and mesonephric tubules is, of course, the explanation of why the mesonephric duct in the female does not become the definitive sex duct. Normal development provides no duct in continuity with the ovary. Consequently the ova, when ripe, can only rupture through the overlying coelomic mesotherlium.

The Mesonephric (Wolffian) and Paramesonephric (Mullerian) Ducts and their Derivatives

In the female embryo the mesonephric ducts undergo a marked atrophy, but not until they have given origin to the ureters and contributed to the wall of the urogenital sinus. The paramesonephric ducts which appear in both sexes, one on each side of the body, attain complete development only in the female and can be regarded as the female sex ducts. In association with viviparity the paramesonephric ducts have, in mammals, become specialized. Their cranial portions remain separate and become the uterine (Fallopian) tubes, their intermediate regions fuse to a varying degree in different mammalian species to give origin

to the uterus, while their caudal portions unite and contribute to a greater or lesser extent to the development of the vagina.

In the human female embryo and mesonephric duct commences to degenerate at the 30 mm. stage (about fifty-seven days). The upper part, as has already been stated, may form a temporary connection with the rete ovarii by way of certain of the mesonephric tubules. In any case, some of these mesonephric tubules (Kobelt's tubules), together with the related part of the mesonephric duct, persist normally, but to a variable extent, as the epoophoron (parovarian, Organ of Rosenmuller). Other closely related mesonephric tubules lose their connection with the mesonephric duct caudal to these tubular remnants normally degenerates slowly but completely. It can, however, still be identified up to almost the 70 mm. stage (about ninety-four days). As is well known, however, this portion of the mesonephric duct may persist, in whole or in part, into post-natal and even adult life as Gartner's duct. Cysts arising from this duct may be found in the mesosalpinx, along the lateral margin of the uterus, being rarely included in its muscular substance, or in relation to the antero-lateral wall of the vagina. Cysts of this origin have a mucosal lining of variable nature - usually cuboidal or columnar epithelium but occasionally ciliated or squamous. The most caudal part of the duct can give origin to a vulval cyst related to the hymen, the periurethral region, the labium minus or, even, the clitoris.

In the human embryo each paramesonephric (Müllerian) duct arises at the 10 mm. stage (about thirty-seven days) (Koff, 1933; Gruenwald, 1941) as an invagination of the coelomic mesothelium on the upper and lateral part of the mesonephric ridge and close to the cranial extremity of the gonadial part of this ridge. The invagination extends into the underlying mesenchyme lateral to the cranial extremity of the mesonephric duct. The invagination itself will persist as the abdominal ostium of the uterine tube. In later stages the margins of the ostium develop into the fimbriae to the tube. Shortly after the invagination has appeared its caudal tip becomes a solid bud of cells. This bud grows and extends in the mesenchyme lateral to, and parallel with, the mesonephric duct. With increase in length of the bud, it becomes canalized in its upper part and, as growth continues, the lumen thus formed becomes continuous with that of the primary invagination and extends gradually towards the growing tip as the latter progresses caudally along the lateral aspect of the mesonephric duct. When the paramesonephric duct, thus formed, has extended as far caudally as the caudal end of the mesonephros it crosses ventral to the mesonephric duct and thus takes up a position medial to this duct, and close to the paramesonephric duct of the opposite side. Continuing to grow in a caudo-medial direction the two paramesonephric ducts meet in the middle line and eventually fuse at or about the 25 mm. stage (about fifty-two days).

As these ducts extend caudo-medially they carry with them, as a covering, an extension of mesoderm from the lower end of the mesonephric ridge. This extension, together with mesoderm on the dorsal aspect of the developing urogenital sinus forms a mass of tissue which cranially constitutes a partition (sometimes called the urogenital septum) across the lower part of the abdominal cavity. Later each half of the partition lateral to the paramesonephric duct will become part of the broad ligament. The fusion between the caudal portions of the two paramesonephric ducts is at first partial and there is a septum between their lumina. In normal development, however, this septum has disappeared by the 56 mm. stage (about eighty-four days), and the fused ducts come to possess a common cavity called the utero-vaginal canal. This canal is lined at this stage with cuboidal epithelium and it is blind caudally. With further development the caudal tip of the utero-vaginal canal makes contact with the dorsalwall of the endodermal urogenital sinus and produces there an elevation, the Müllerian tubercle, which projects into the sinus. Proliferation of cells in the most caudal part of the uterovaginal canal results in the appearnace of a solid vaginal cord which by (interstitial?) growth progressively increases the distance between the utero-vaginal lumen and the urogenital sinus. From this solid cord most of the vagina will be derived. At about the 63 mm. stage (about ninety days), however, there is, apparently, a contribution to the caudal end of the mesodermal vaginal cord from the endoderm of the urogenital sinus at the position of Muller's tubercle. According to Koff(1933)

the endodermal contribution arises as bilateral evaginations ("sino-vaginal bulbs") from the urogenital sinus. The evaginations are closely related to the caudal extremities of the mesonephric ducts. Other investigators (e.g. Mijsberg,1924; Kemperman,1931) consider that the bulbs are, in fact, of mesonephric derivation. Whatever their origin the bulbs, by cellular proliferation, become solid and later fuse, the Mullerian tubercle disappears and the solid vaginal cord, now broader in the transverse plane, can better be called the vaginal plate. The cells of the whole of the plate gradually become squamous in nature and no obvious distinction between that part of it of mesodermal (paramesonephric) and that part of endodermal (uro-genital sinus) epithelium can be established. Koff, however, considers that about the caudal one-fifth of the definitive vagina is of sinus origin.

While those changes have been progressing the portion of the urogenital sinus just cranial to the site of origin of the sinovaginal bulbs becomes narrowed and elongated to form the urethra. At the same time the caudal extremity of the still solid vaginal plate moves, at least relatively, caudally, as the lowermost portion of the urogenital sinus elongates in the sagittal plane to form the vestibule. With continuing development the vaginal plate becomes canalized from above, as the utero-vaginal canal extends caudally, and from below, to some extent, by the breaking down of that part of the solid epithelium formed from the sino-vaginal bulbs.

Canalization, however, is not completed until the 162 mm. stage (about twenty-one weeks), or later.

While these junction between the future body and cervix of the uterus can be recognised at the 40 mm. stage (about sixty-two days) the uterine portion of the utero-vaginal canal is not effectively separated from the vaginal portion until the 150 mm. stage (about twenty weeks). Meanwhile a gentle curve in the sagittal plane develops at the junction between the two parts of the foetal uterus and a more pronounced curve between the developing cervix and the future vagina. From the time of establishment of a distinction between the two and into early post-natal life the cervical portion of the developing uterus is larger than its body.

Hymen If, as seems likely, Koff is correct in attributing the origin of the lower fifth of the vagina to the sino-vaginal bulbs, then the hymen must be considered to be the partition which persists, to a varying degree, between the dilated and canalized fused bulbs and the urogenital sinus proper. On this interpretation the hymen is formed by a deep layer of vaginal epithelium and a superficial layer of urogenital sinus epithelium (both epithelia of endodermal derivation) with an interposed layer of mesoderm. Obviously, however, if the vagina is, as has been maintained by a number of investigators (Bloomfield and Frazer,1927; Hunter,1930: von Lippmann,1939) purely of utero-vaginal origin then the deep surface of the hymen is covered with mesodermal (paramesonephric) epithelium. A mesonephric origin for the sino-vaginal bulbs

would give a mesodermal epithelium also on this deep surface.

Abnormalities in the Development of the Uterine Tubes, Uterus and Vagina.

The bilateral origin of the female reproductive tract explains many of the commonly occurring developmental anomalies. Further, the association in development of this tract with the kidneys and mesonephric ducts is frequently revealed or emphasized by teratological manifestations that simultaneously involve both the reproductive and urinary systems.

THE UROGENITAL SINUS

In the late somite human embryo the caudal part of the hindgut, to which the allanto-enteric diverticulum is attached, is slightly dilated to form the cloaca. This is, of course, lined by endoderm, and it is separated from the amniotic cavity by the cloacal membrane on the superficial aspect of which is situated the shallow ectodermal depression called the external proctodaeum. At about the 4 mm. stage (about thirty-one days) the pronephric (later mesonephric) ducts join the cloaca, one on each side at about the junction of its middle and anterior thirds. A little later the angle, in the cranial part of the cloaca, formed between the allantois and the hindgut projects into the cloaca as a coronally orientated urorectal septum. This septum extends caudally and by the 16 mm. stage (about forty-three days) normally reaches and fuses with the endoderm of the cloacal membrane. The cloaca is thus divided into a small dorsal part,

the primitive rectum, and a larger ventral part, the primitive urogenital sinus. The junction of the urorectal septum and the cloacal membrane is the primitive perineum. The portion of the membrane in front of the junction is the urogenital membrane and that behind it is the anal membrane. When the primitive urogenital sinus is completely separated from the rectum it can be subdivided into a part cranial to the level of the openings of the mesonephric duct and a part caudal to this level. The former is called the vesico-urethral canal and from it in subsequent development the bladder and primitive urethra will be derived. The caudal part of the primitive urogenital sinus is called the definitive urogenital sinus. Initially the vesico-urethral canal has the shape of an elongated cylinder, continuous cranially, with the allantois and slightly flattened ventro-dorsally. It should be noted that there is no sharp demarcation between the allantois and the apex of the future bladder. As the infra-umbilical portion of the abdominal wall is established the bladder segment of the vesico-urethral canal gradually dilates. The definitive urogenital sinus soon becomes flattened from side to side. At the same time it elongates ventro-dorsally. Proliferation of the endodermal cells of the urogenital membrane, with possibly some also of the adjacent ectodermal cells, encroaches on the cavity of the sinus so that it is more or less obliterated by a loosely arranged cellular mass.

By the 16 mm. stage (about forty-three days) the definitive urogenital sinus has become divided into an upper pars pelvina and a lower pars phallica. The latter is markedly elongated antero-posteriorly and is flattened coronally. The urogenital membrane separates it from the ventral part of the ectodermal cloaca. In the female the para pelvina of the urogenital sinus probably contributes to the caudal end of the urethra. As the projection of Muller's tubercle is into this part of the sinus its endodermal wall also contributes to the development of the lowermost part of the vagina. The subsequent history of the para phallica of the urogenital sinus is most easily considered in association with the development of the external genitalia.

The External Genitalia

In the early, indifferent, or "neuter" stage of development, the area around the external aspect of the cloacal membrane shows three prominences. One of these, the genital tubercle, is situated in front, between the ventral margins of the membrane and the infra-umbilical abdominal wall. The other two, the future labial swellings, are placed flanking the membrane, one on either side. The anterior part of the cloacal membrane, after the urorectal septum has reached this membrane, is the urogenital membrane and the ectodermal depression superficial to it, which is the ventral part of the original proctodaeum, or ectodermal cloaca, can now be called the urogenital sulcus.

This sulcus is separated from the pars phallica of the urogenital sinus by the urogenital membrane.

As the pars phallica of the sinus becomes elongated in the sagittal plane it is drawn into, or encroaches upon, the genital tubercle. The cavity of the pars phallica is more or less obliterated by a cellular proliferation. The portion of this proliferation that becomes related to the genital tubercle is the urethral plate which, however, in the female has no significant part to play in normal development. Shortly after the 16 mm. stage the dorsal part of the urogenital membrane breaks down and the resulting communication between the phallic portion of the endodermal urogenital sinus and the ectodermal urogenital sulcus is known as the primitive urogenital orifice.

As the 25 mm. stage the external genitalia in the two sexes are very similar. Shortly after this stage the genital tubercle shows a caudal flexion in the female embryo and can be identified as the clitoris. It is not, however, until about the 50 mm. stage (about eighty days) that the sex can, with assurance, be determined from external inspection. As the clitoris is not encroached on to any appreciable extent by the urethral plate, there is no female equivalent of the penile portion of the male urethra. In subsequent development the

the labial folds extend towards each other in front of the anus and behind the urogenital orifice until eventually they meet and fuse in the posterior commissure. The lateral part of each fold then increases in size to form the labium majus of this side. Unlike the male, the urethral folds in the female do not fuse but persist as the labia minor so that the urogenital orifice is unclosed. Thus, in the female, the phallic portion of the urogenital sinus, and a considerable portion of its pelvic portion are exposed to the exterior as the vestibule. Into this vestibule the female urethra and the vagina open separately. The entrance to the vagina is, of course, incomplete because of the presence of the hymen. The line of junction between the ectoderm of the urogenital sulcus and the endoderm of the pars phallica of the urogenital sinus cannot be identified with any assurance in the later stages of development. It is frequently considered to be represented by the free edges of the labia minor but it is probably on the medial sides of the mucocutaneous folds. The greater vestibular (Bartholin's) glands, which are the female homologues of the bulbo-urethral glands, arise as outgrowths of the endodermal lining of the vestibular glands

develop in the anterior part of the urethra. They have been interpreted by Johnson (1920) as equivalent to the glands of Littre of the male. The paraurethral glands of Skene arise from the urogenital sinus. Both of these groups of glands correspond to the prostate of the male.

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COMPARATIVE ANATOMY

A definite parallelism exists between the various forms of uterine anomaly in human beings and the normal uterus of animals lower in the developmental scale, and a wider knowledge of this parallelism leads to a clearer concept of human malformations and a better understanding of their management, Schumann (1936).

Failure of fusion of the Mullerian ducts in women assumes one of a definite number of forms, and as pointed out by Schumann, each of these forms has its direct analogue in the normal form of the uterus in one or more of the lower orders of mammals. Schumann also remarks that the lower the species in the mammalian scale the less the tendency to fusion of the two sides of the genital tract.

Indeed the similarity is often so great that Blair-Bell(1934) has suggested the use of the word "atavism" in preference to "malformation" because the anomalous uterus often conforms exactly with that of one of the lower orders. In other words, women may have uteri which are found to be counterparts of the uteri of rodents, carnivores, insectivores, or sub-human primates.

According to Wilder(1923) the uterus is an adaptive organ, essentially a localised enlargement, which develops as needed. Thus, in viviparous sharks, such as the squalus, the expanded lower portion of each Mullerian duct becomes enlarged to form a

uterus in which the embryos are retained until they have almost attained adult form. The same is true of certain salamanders. However, in none of these instances is the organ more than a container or brood cavity. There is no placenta nor is there any other direct connection between the embryo and the uterine wall.

The same is true also of certain lower mammals such as monotremes and marsupials, in which there is no placenta and the young are produced in a very immature state. It is only in the higher or placental mammals that the walls of the uterus become differentiated for nutrition of the embryo, and the uterus thereby becomes a physiologically active organ.

In mammals, starting with the lower species, all the theoretical intermediate evolutionary stages have their counterpart, ranging from double uterus to single uterus. Thus, as Walter (1939) states, monotremes have two separate uteri without vaginae, whereas marsupials have two separate uteri with separate vaginae. Among placental mammals a double uterus is found in certain rodents, such as the mouse, hare, marmot and also in elephants, certain bats and the aardvark. A beginning of fusion between the two uteri is evident in pigs, cattle, certain rodents, certain bats and carnivores. A two-horned uterus is characteristic of ungulates, cetaceans, insectivores and some carnivores, while a single uterus, the uterus simplex is found in apes and humans.

A more detailed consideration of each of these species will serve to illustrate further the resemblance between the normal uteri of lower animals and anomalous uteri encountered in human beings.

Monotremes: In the egg-laying mammals or monotremes (e.g. the ornithorhynchus or duckbill of Australia and Tasmania; and the echidna or spiny antater) the Mullerian ducts tend to fuse posteriorly while the anterior portions remain separate. In the monotremes, the uterus opens into a urogenital sinus (Neal and Rand)(1939). The so-called uterine portions are in reality oviducts. As described by Wilder, they are short thick-walled, of rather large calibre, and entirely distinct from one another.

Marsupials: The marsupials or didelphians are represented by the opossum and the kangaroo. In the marsupials a vagina occurs and this may be regarded as representing an important stage in the evolution of the Mullerian ducts. In the opossum and kangaroo, the uteri and lateral vaginae are connected with a central vaginal pouch, which has a more or less perfect longitudinal septum and ends blindly at the summit of the urogenital sinus (Blair-Bell), (1934).

Rodentia: The orders rodentia (rabbits, squirrels, guinea pigs, rats, gophers) offers examples of the transition from the primitive types of uterus to the more advanced forms(Owen).

Two distinct uteri, with separate cervices, are found in the lower species, such as the viscacha and rabbit. In the agouti, the uterus is double but has a single cervix. In rodents, the vagina, from being septate or double in lower forms becomes single in the higher members of the order. In pigmy musks Owen (1868) states that "the cornua of the uterus are unequal in size: the right was the largest in the specimen examined."

Carnivora: The next important step in development is found in the order carnivora (cats, dogs, weasels, bears, raccoons, seals) where the two uterine bodies fuse in their lower parts to form a bicornate uterus.

Ungulata: The order ungulata (Hoofed animals) also offers examples of the uterus bicornis. According to Patten, in the sow the fusion of the Mullerian ducts is carried only a short distance beyond the cervix to form a typical bicornate uterus. In his text on developmental anatomy, Arey (1940) cites the uterus of sheep as a typical example of bicornate uterus.

Primates: The uterus simplex, the highest form of uterine development, is found in the primates, which includes monkeys, baboons, apes and man. According to Neal and Rand (1939), a sub-order of primates, the lemuroids (including lemurs and tarsiers), have two-horned uteri (uterus bicornis). However, some of the leumuroids have arcuate or cordiform uteri.

Although, as noted by Perrigard (1942), the uterus unicornis is normal in birds, Blair-Bell believes that this anomaly cannot be considered an atavism, but rather the result of the total suppression of one Mullerian duct.

Congenital absence of Uterus: Although small or infantile uteri are quite common, complete congenital absence of the uterus is quite rare and it is very improbable that in the few cases reported there was absolute absence of all vestiges of the organ. Nevertheless, this seems to have occurred in the case of apparent congenital absence of uterus reported by Bowles and Burgess. (1939) Following the removal of an appendix in an eleven year old Korean girl, it was noted that the broad ligament of one side swept across the region where the uterus should have been and fused with its counterpart from the opposite side of the pelvis. No thickening was found that could be interpreted as the uterine body. The ovaries were normal, but vaginal examination revealed no apparent cervix.

Congenital Absence of the Vagina: As noted by Blair-Bell a rudimentary uterus which is imperforate and consists mainly of fibrous tissue, is associated with absence of the vagina. Wharton (1943) attributes absence of the vagina to inhibition of the development of the Mullerian ducts. In this condition the growth of the ducts ceases very early in embryonic life. The ducts remain separate and do not fuse to form the uterus, cervix, or upper vagina. Anderson (1931), however, takes a view opposite to that given by most writers. He is of the opinion that the absence of the vagina and uterus is usually caused not by lack of fusion of the Mullerian ducts but by premature fusion.

For obvious reasons, abnormalities of the vagina are more frequently observed and reported than are internal malformations of the genital tract. Hence malformations of the vagina, such as atresia or complete or partial vaginal septum, are not considered rare. However, Reel (1943), in a report of four cases of congenital absence of the vagina encountered during a twenty-three year period, considers total absence of the vagina as probably the rarest congenital maldevelopment of the female genital tract. Various mechanical procedures and a variety of surgical techniques have been developed to correct some of these abnormalities or to restore a degree of normalcy to others.

Grave anomalies of the uterus are often accompanied by other serious errors of development, including not only vaginal atresia, but also such conditions as cloacal defects, atresia ani, vulvovaginal anus, malformations of the abdominal wall, hernia, and other defects Wharton (1943). Occasional reports, such as those of Benjamin and Danforth (1940) and Siegel, describe bizarre combinations of developmental abnormalities associated with uterine anomalies. In Siegel's recent paper (1944), the congenital anomalies noted in a twenty-one day infant consisted of:- absence of right kidney and ureter, uterus didelphys, imperforate anus, imperforate vagina, pseudohydrocolpos, urethrosigmoid (pseudo-hermaphrodismus feminus externus). Such a conglomeration of congenital anomalies is obviously incompatible with life.

Milder combinations of malformation also appear. For example, Ladd and Chisholm (1940) recently reported a case in which a double uterus and vagina was associated with duplication of the sigmoid and rectum with two external openings.

Uterine Anomalies Associated with Anomalies of the Urinary Organs

Since the embryonal predecessor of the Mullerian duct also contributes to the development of the urinary tract Reel(1943), it is not surprising that uterine anomalies should often be associated with renal agenesis. Indeed, many authors have called attention to the frequent coincidence of congenital anomalies of the genitalia and anomalies of the urinary tract. Hinman (1940), in a critical review of congenital bilateral absence of the kidneys, states that renal agenesia is usually associated with gross malformation of other organs. In the female, the derivatives of the Mullerian ducts are most often affected.

Unilateral renal agenesis is frequently associated with uterus unicornis, but it occurs in association with symmetrical uterine anomalies as well as with unilateral defects.

A study of the literature shows that in many cases of uterine or other genital malformation there has been no attempt or adequate urological examination. Conversely, in cases of abnormalities of the genital system. It should be regarded as axiomatic that in all cases in which the genital tract is anomalous, the urinary tract must be studied thoroughly.

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AETIOLOGY

AETIOLOGY

The literature devoted to abnormal development is extensive, but as Needham (1942) has stated "the part devoted to teratomata is marked by an unusual degree of unscientific speculation, inaccurate description and logical mistakes". Therefore it is as well to set forth an account of the modern views on the genesis of congenital malformations as a whole before considering those of a particular organ.

INCIDENCE

Mall (1910) found that 50 per cent of aborted embryos were pathological. He states that in every hundred pregnancies, eighty can be expected to end in the birth of a normal individual, seven to be aborted as ova showing pathological change, and one to result in a monster at birth. The remaining twelve "normal" embryos are by no means all normal, for there are constantly to be found in them, especially in the younger specimens, minor changes which must be viewed as the forerunner of real monsters. The level of demarcation between the normal and pathological ovum is by no means sharply defined, and in all probability a number of so called normal embryos are slightly deformed or abnormal. Malpas (1937) in 13,964 births at the Liverpool Maternity Hospital, found the incidence of congenital malformations to be 2.1 per cent but obviously this must be higher when one considers the not infrequent postmortem findings of some congenital malformation unsuspected in life.

Congenital anomalies may be attributed to:

1. Genetic factors.

11. Environmental factors.

1. Genetic factors

There is no doubt that the presence of an abnormal gene in either the male or female germ cell before fertilization is a very frequent cause. When these abnormalities first appear in a family they are due to a mutation which may be defined as "a change within the genetic constitution resulting in a discernible difference in behaviour or structure which is perpetuated by heredity".

Haldane (1935) was able to show, from his analysis of haemophilia that mutations are frequent in human genes. Such gene mutations may therefore account for the sudden appearance of a congenital malformation in hitherto unblemished families. The mutation may, however produce an abnormality which is incompatible with life, and will not therefore be handed down, though as more than one germ cell in the parent may be affected, there is always the possibility that the abnormality or one similar to it will appear in a sibling. Gunther and Penrose (1935) have calculated that the mutation rate of the gene in man may be as high as 1:60.000 per individual per generation.

There are many examples of congenital malformation which are hereditary and obey normal Mendelian principles, examples of which are polydactyly, transmitted as a Mendelian dominant, harelip as a recessive character and haemophilia and colour blindness as sex linked characteristics. Gruneberg (1944) has shown that many abnormalities of rodents such as hydrocephalus, cleft palate, harelip, polydactylism and brachydactylism are hereditary.

11. Environmental factors

The importance of environmental influence in the causation of congenital malformations is strongly suggested by experimental evidence, comparative embryologists having produced practically every known deformity by slightly varying the environment of the embryo. Mall is of the opinion that this is the reason for the very high incidence of malformed embryos to be found in association with tubal pregnancy, and concludes that the majority of human congenital abnormalities arise from faulty implantation of the ovum followed by malnutrition of the embryo. This is of special interest when one recalls that reports on congenital uterine malformations suggest a higher incidence of abnormal offspring, which if Mall is correct in his contention, might arise from faulty implantation and poor decidual reaction in the maldeveloped uterus.

Defective diet in animals will produce abnormalities in the offspring. Hale (1937) showed that a diet deficient in vitamin A fed to sows would produce various eye defects in their litters, which if in turn mated and fed on a high vitamin A diet produced normal litters, proving that a dietary defect and not an heredity one was the cause. Similar results were obtained by Warkany and Nelson (1940) who fed rats in early pregnancy on a riboflavin free diet, and produced congenital abnormalities in their young including cleft palate, deformities of the extremities, syndactylism and hare lip. There is however no proof that these animal experiments have any bearing on the causation of congenital human abnormalities.

Diabetes Mellitus Oakley and Peel (1949) reported an incidence of 6.3 per cent congenital abnormalities of a serious nature in children born to diabetic mothers when over a similar period the general incidence was 0.94 per cent.

Endocrine Factors The influence of sex hormones in embryonic development is now well established and in the lower types the sex of the individual may be modified or even reversed, by administration of suitable hormones when the embryo is in the neuter stage. Hormonal influence on sex reversal is well known in cattle - the so called free martin condition (Lillie, 1917) where a genetic female calf is a co-twin with a male. The former's primary chromosomal sex is female (XX chromosome) while that of its co-twin is male (XY chromosomes).

During development the placental circulation of the twins anastomose so that the embryonic sex hormones pass freely from one embryo to the other. Since the testis develops before the ovaries, the male sex hormones are produced earlier than the female hormones and by passing into the circulation of the genotypic female calf, influence her sexual differentiation in the male direction. Hence her genital organs show many male characteristics; the ovaries in particular are inhibited and show testis like characters and do not produce ova so that at maturity such a female is sterile.

Rubella The work of Gregg (1941) and Swann (1949) showed the high incidence of congenital abnormality in the children born to mothers who suffered from rubella in early pregnancy. The most common abnormalities were congenital cataract, congenital malformations of the heart and deaf mutism. The earlier the pregnancy at the time of the infection the more liable the child was to be affected. Toxoplasmosis is another infection which when occurring in pregnancy can produce abnormalities in the foetus.

CONGENITAL UTERINE MALFORMATIONS

Although many theories have been advanced in explanation of these anomalies, only two, heredity and endocrine influence, have any experimental or clinical evidence in their support. It will be seen that the majority of these theories, which are summarised below, concern themselves with explaining a failure of fusion of the two Mullerian ducts on mechanical grounds.

Constitutional or Germinal Defects: Eck (~~Saunders,~~ 1936) considered they arose from some defect in germ plasm and that most commonly they are found in broadly built women of stable temperament.

Persistence of the Vesico-Rectal Fold: Ancel and Villemin (Puddicombe, 1929). This is a vascular fold which is found only in women, and is almost constant, according to Polak, Okinczwe, Huet and Patacki (1923), when the bodies of the uterus are unduly separated. This fold extends from the upper part of the rectum to the bladder and is produced by a branch of the superior haemorrhoidal artery, and is mentioned as being present in 52 of the 600 cases of double uterus reported by Dubreuil and Chambardel (1927-1928). Eden and Lockyer considered it of allantoidal origin or a relic of the terminal intestinal mesentery but on occasion may arise from foetal peritonitis. While conceded by many authors as an aetiological factor, Basset (1933) found five cases of double uterus

in which there was no vesico-rectal fold and Nagel, (Puddicombe,1929) and others consider this fold the result and not the cause of a double uterus.

Abnormal Shortness of the Round Ligaments: Newton (1924). Here, it is postulated that fusion of the two Müllerian ducts is prevented by the shortness of the two round ligaments. He also refers to Pick, who found tumours in thirty cases and claims these have an aetiological bearing.

Blair Bell (1934) attributed the failure of fusion to deficient action of the decussating subperitoneal muscle fibres, which normally form the myometrium and, secondarily, to deficient formation of the uterosacral muscle fibres, with a short genital mesentery as a contributing factor.

Formative disturbances of the intestinal tract and developmental errors of the ventral abdominal wall are aetiological factors according to Felix and Rosenstein (1929). Keibel and Mall (1908) were able to collect similar theories from the literature to account for these disturbances, such as hydronephrosis, distension of the bladder, and rectum; anomalies in the formation of the anterior abdominal wall, such as herniae, cleft pelvis, and foetal peritonitis.

The Importance of the Wolffian Duct in the Genesis of Uterine Malformation. Gruenwald (1941). This author believes that the Wolffian Duct takes part in the formation of the vagina, and in this way ectopic ureters opening into the vagina can be explained on an embryological basis (vide case No.SM19).

Similarly arrested development of the Wolfian duct causes agenesis of the ureter and absence of Mullerian duct in an area corresponding to the defect in the Wolfian duct. For example unilateral or unicornuate uterus is associated with absence of the kidney on the same side.

Anderson (1943) states that 80 per cent of rudimentary horns are on the right side, and explains this by stating that the left Mullerian duct advances ahead of the left during its earlier development and reaches its mesenchymal blood supply first. Hence by a slight mistiming the left duct may fail to obtain its blood supply and will fail to develop.

Endocrine Influence: A large amount of work by endocrinologists and experimental biologists has clearly shown that the so-called sex hormones can have a marked influence on the phenotypical expression of sex. During early development the embryo shows morphological ambisexuality, and as described earlier, mesonephric and paramesonephric duct systems are established in both genetic females and genetic males, and their normal development can be markedly changed by the administration of the relevant hormone at the appropriate time in their development.

Basically the effects of the oestrogenic hormones in the genetic male embryo is to stimulate the essentially female structures while inhibiting the essentially male structures,

while the effect of the androgenic hormones in the genetic female embryo is to stimulate the male structure whilst inhibiting the development of the female structures respectively Burns (1942), Raynaud (1942), Jost (1948). However the effects of the androgenic hormone on the Mullerian ducts is not as clear as the effect of oestrogens on essentially male embryonic structures and depends especially on timing, dosage and on species differences, Willier (1955). In the embryos of mammals complete suppression of the Mullerian ducts is not obtained but the vaginal region may be inhibited, for example in the opposum Burns (1942), and in mice Raynaud (1942). Paradoxically the administration of large doses of androgenic hormone may stimulate the Mullerian system, Moore (1947), Burns (1949). These experimental facts gained from animals are not easily reproduced in the human species and although E.Davis and E.Potter (1948) administered testosterone to women in early pregnancy, it was given in too small doses and too late to modify the differentiation of the somatic sex of the foetus. Furthermore as far as testosterone is concerned it has not been shown that it crosses the placental barrier. ~~J.~~ Zander and ~~H.A.~~ Muller (1953) have reported the case of a woman operated upon for a mammary sarcoma at the sixth month of pregnancy, and who from that time was treated by testosterone which attained a total of 3.2 Gm.

At birth the child, which was a female, showed hypertrophy of the clitoris as well as a scrotal like swelling of the labia majora characteristic of a female pseudohermaphrodite. The urinary 17 ketosteroids showed a normal value which excluded a hyperplasia of the suprarenal cortex. During the first few days after birth the swelling of the clitoris and prepuce visibly regressed, and three months later had returned to normal. Zander and Muller were of the opinion that the infant's condition at birth could be correlated with the administration of testosterone to the mother, and in support of this quote Jost, who by administering testosterone to the female rat in pregnancy, was able to produce male intersexes states in the female embryos. On the other hand Kerckhove (1954) reported a case where 6.85 Gm. methyl testosterone were administered perlingually, without any change being found in the female offspring. These contradictory observations obviously do not permit any decisive conclusion to be reached concerning the administration of male hormone in pregnancy. Snoeck and Ghilain (1955) are doubtful if the intramuscular injection of testosterone has any effect and in the case of Zander and Muller, suggest that the child was a male and not female intersex. A very interesting case was reported by Brentnall in 1945, of a woman who commenced to develop signs of virilism at the third month of pregnancy, manifested by the growth of hair on the face, trunk, chest, and deepening of the voice.

Delivery was effected by Caesarean section, when a new growth of the ovary was found and which on histological examination proved to be an arrhenoblastoma of a very cellular type. The child was a female and on examination was seen to have a hypertrophy of the labia and clitoris, absence of the vagina. The urethra opened at the base of the clitoris and on the fourth day a blood stained discharge could be seen issuing from it; this being taken as presumptive evidence of the presence of a uterus. It would appear reasonable to assume some connection between the presence of the arrhenoblastoma and the genital defect, the hormonal effect of the former inhibiting the normal development of the vagina which derives from the sinovaginal bulbs. As the signs of virilism did not make their appearance until the end of the third month one would expect the uterus to be normal in this case or at the most to have a persistance of the septum between the two Mullerian ducts, this normally disappearing at the 84th day. On the other hand the lower 1/5th of the vagina and vestibule do not complete their development until the 160 mm. stage or 21 weeks and it is this part of the genital tract which one would expect to be affected where the embryo came under the influence of the androgenic hormone at the end of the third month. The same objection can be put forward here as in the case of Zander and Muller, namely that the infant is in reality a male pseudohermaphrodite, but, although the genetic sex has not as yet been determined, the bleeding per urethram in the first few days of life is strong persumptive evidence of the presence of a uterus capable of responding to the maternal oestrogens.

Female Pseudohermaphroditism or Congenital Adrenogenital Syndrome in Females

The great majority of female pseudohermaphrodites arise from a congenital hyperplasia of the suprarenal cortex which affect the foetus while in utero. These subjects at birth show a normal Mullerian tract having undergone complete differentiation, (tubes, uterus and vagina) the ovaries are well developed and the Wolfian ducts have undergone the normal involution. In contrast, the organogenesis of the urogenital sinus is profoundly disturbed and appears to have been suppressed. Certain experiments in animals Jost (1950) tend to show the existance of an anterior posterior gradient in sensitivity of the foetal genital tract to injected androgens. Snoeck and Ghislain (1955) are of the opinion that this differentiation of these particular segments of the genital, which in timing exposes them to the full influence of the endocrine action of the foetal cortical androgens while still undifferentiated. This state is to be found normally in the foetus of 106 mm. that is to say after the twelfth week, and MacNeil (1947) has shown that at the fourth month of gestation the foetal suprarenal cortex shows in its fascicular zone areas of lipoid. Rosa (1955) has confirmed these findings in female embryos of four and a half months. Although it is agreed that in the adult it is from the deeper part of the reticular zone that androgens arise it cannot be excluded that it is the same in the foetus.

With Bishop (1954) one can agree that it is from the eleventh to twelfth week in hyperplasia of the suprarenal cortex that the effects of excessive androgen production make themselves felt. On the other hand, the development of the definitive urethra and lower 1/5th of the vagina which arise from the urogenital sinus is only completed in the course of the fifth month, and it would appear to be between these two periods that the inhibitory influence of the suprarenal androgens is manifested.

It would appear that the uterus in female pseudohermaphrodites is either normal or absent and in his book on Intersexuality Young (1936) gives no example of the uterus being congenitally malformed although it was absent in several and failures of development in the urogenital sinus were frequent. Broster(1938) found only one case of a bicornuate uterus in thirty-three cases of the adrenogenital syndrome, and here in addition to a penile hypospadias a small testis was found in place of the ovary, which suggests that this was a male pseudohermaphrodite or true hermaphrodite. A similar case to this was reported by Blacker and Lawrence who at autopsy in stillborn infant found an ovary, unicornuate uterus and Fallopian tube on one side and on the other an ovotestis, enlarged Wolfian duct representing epididymis and vas deferens, and a malformed Fallopian tube (Mullerian duct).

These findings would appear to confirm the opinion that in foetal adrenal hyperplasia the effects of the excessive secretion of androgenic hormone does not occur before the twelfth week,

SUMMARY

(1) It would appear from experimental evidence that androgens can suppress the normal development of the Mullerian ducts in animal embryos but the effects differ according to the animal species, time of administration and dosage.

(2) It is possible that administration of male hormone to pregnant women may have an effect on the foetus but the evidence is conflicting, and considerably more work is required before any definite pronouncement can be made.

(3) In congenital adrenal hyperplasia or female pseudohermaphrodites, the effect of the excessive production of androgens is not felt until the twelfth week of gestation, by which time the uterus is normally developed and only that part of the genital tract still undifferentiated, namely, the lower 1/5th which is derived from the urogenital sinus, will have its normal development inhibited.

HEREDITY

Many examples of genital abnormalities having hereditary and familial tendencies have been reported, and as long ago as 1870 Phillips of Guy's Hospital recorded two sisters with absence of the vagina and Squarey (1872) traced the pedigree of a family through five generations showing a genital defect. More recently Weissenberg (1928) described a Russian family with two sisters showing atresia of the vagina. Both were married, had normal breast development and external genitalia. Delbet (1940) also records the hereditary absence of the uterus in a French family with five cases in three generations. The condition was transmitted through normal sisters of affected women, to their daughters or granddaughters. These women, all married, were of feminine aspect with large breasts and hips, but they never menstruated and one had an axillary breast.

Imperforate hymen is another genital abnormality which can apparently be inherited, McIlroy and Ward (1930) describing the condition in three sisters who were otherwise normal. A somewhat similar condition 'imperforate' is widespread in a particular strain of mice, in which fifty-one females appeared with imperforate vaginae from forty-two mothers, Gowen and Heidenthal (1940). When the vagina was opened surgically ten out of twenty-two produced litters, some of the females of which were imperforate. The various crosses suggest that two recessive factors are necessary to produce the condition and Chase (1944) found that certain inbred strains regularly produce 6.6 per cent in each generation.

Polymastia. From the numerous records of polymastia or multiple breasts in the literature, it is evident that the condition is much more frequent than commonly supposed. It is of course an atavistic tendency or regression to the condition found in man's primate ancestors, with many young at birth. The Phoenician goddess Astarte was endowed with several breasts and the statue of Diana of Ephesus in the Vatican museum shows extreme polymastia, which was regarded as a sign of fertility. The lemurs and higher primates normally have one pair of breasts, but the lemurs also have an inguinal pair of 'anchoring nipples' for attachment of their young, which are two in number. Axillary breasts are normal in the flying lemur. Leichtenstern (1878) cited 291 cases of super accessory breasts, and of these there were three cases of twins. There are early records showing inheritance of this anomaly Peterquin (1837), describing a ~~anomalies described a~~ family in which the father had an extra mamma on the left side under the normal one, his parents and wife were normal, but his three sons had an extra mamma on the right side, his two daughters on the left side, thus the condition was transmitted to all his children. His daughters had six and four children respectively and all were normal. This would appear to be a change in dominence. Five cases of polymastia in four generations of a family were described by Klinkerfuss(1924). They were all females and all in the left axilla except for one,

who had one in each axilla. Polythelia, another atavism, shows a strong tendency to be inherited, and is apparently as common in males as females. Marie (1893) cites a family history where there were twenty cases of polythelia in four generations. The girl had an extra nipple below the normal one on the left side and her eleven siblings, her father, five of his brothers, the grandmother and paternal great grandmother were similarly afflicted. Koopman who made a study of polythelia found it to be commoner in men. An analysis of Graham Bell's well known experiments in breeding multinippled sheep with the intention of producing a higher frequency of twins was made by Castle (1924). He found that the character supernumerary nipples was strongly inherited (evidently dominant), selection having produced a strain with generally six, and occasionally seven or eight, from one with never more than four. Plum (1938) however from an analysis of crosses in pigs found that the character is recessive not dominant.

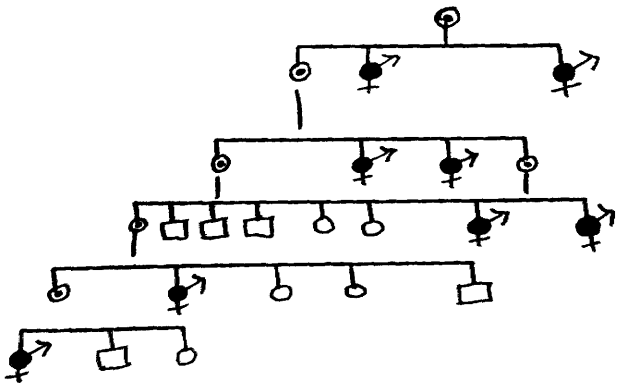
Intersexuality.

From a study of some sixty cases of the adrenogenital syndrome, Broster (1934) came to the conclusion that heredity plays a part. In a quarter of the cases there was a history of hirsutism, which was twice as common on the female side.

Similar conditions were found in two cases of twins and one case of mother and her four year old daughter.

O'Farrell (1935), found five intersexes among forty members of the third generation of a family whose ancestors were all normal. In many cases female pseudohermaphrodism is associated with hyperplasia of the adrenal cortex, they showing male secondary sexual characteristics, hypertrophied clitoris and apparent absence of the vagina.

Biggs and Rose (1947) collected nine families of pseudohermaphrodites, in four of which both male and females were affected. Pettersson and Bonnier (1937) found a Swedish family with six cases of male pseudohermaphrodites in three generations. Phenotypically they were female but did not menstruate, and have no ovaries or uteri. Several were operated for inguinal herniae when internal testes were discovered.



It is clear from this pedigree that the condition is transmitted by normal females to half their offspring, the intersexes representing transformed males.

Heredity as a Factor in the Genesis of Uterine Malformations

In view of the many examples already quoted of genital anomalies whose pedigrees show hereditary and familial tendency, it is surprising that prior to Stanley Way (1945) no one appears to have considered congenital uterine malformations on a similar basis.

Blair Bell (1909) was so impressed by the analogy between the various forms of congenital human uterine malformation and the phyllogenetically older uteri of various mammals, that he suggested the word atavism in preference to malformation. He does not, however, appear to have considered the possibility that, like other atavisms, congenital uterine malformations might have a hereditary and familial basis. Makela (1916), appears to have been the first to describe a pedigree where uterine malformations were present in two generations. The mother had a bicornuate uterus, and her four children all died shortly after birth. Autopsy revealed abnormalities of the lungs, diaphragms, and uteri in all four. However it appears that the author published this case more as an example of a rarety than of a case illustrating the genetic basis of uterine anomalies. Jarcho (1946) in his monograph on uterine malformations describes a case where the mother had a unicornuate uterus and her daughter who died shortly after birth, was found at autopsy to have a cordiform uterus whose outline is beautifully illustrated by hysterosalpingography. The significance of this appears to have been entirely missed, as Jarcho doesn't comment upon it.

Stanley Way (1945 and 1947) by his contributions to the subject has shown conclusively that there is a familial and hereditary tendency. In thirty-seven cases of congenital uterine abnormality he was able to demonstrate five examples of familial tendency. He investigated the children where possible by hysterosalpingography, stating that even in the neonatal child the hymen could be easily dilated to allow the nozzle of the syringe containing the radio-opaque substance to be introduced into the cervical canal. However this method was later abandoned owing to the frequency with which the radio-opaque substance was extravassated and could be seen in the uterine and ovarian veins.

Hunter (1950) found only one case among his series of thirty-two uterine malformations. It is of course extremely difficult to arrive at the true incidence because of the few opportunities of investigating the relatives. Even so Way has shown that the relatives of women known to have uterine malformations, frequently give a bad obstetric history, and suggests that whenever possible they should be investigated for similar deformities. In support of his contention he relates the case of a woman, known to have a bicornuate uterus, who attended his antenatal clinic. Her mother had had eleven pregnancies all abnormal, including three placenta praevias, four postpartum haemorrhages, and one internal version, and her eldest daughter had died in childbirth from shock and antepartum haemorrhage occasioned by placenta praevia and internal version. Unfortunately the mother

refused to have an hysterosalpingography and the daughter left for the States where she was lost sight of, and no record was obtainable of her delivery.

Way also is of the opinion that repeated placenta praevias is suggestive of a uterine anomaly and in support of this quotes the case of Andrews and Nicholls(1941) of a woman who had three successive placenta praevias, delivery in the last being effected by Caesarean section at which a hitherto unsuspected subseptate uterus was discovered, although the authors do not appear to have noted the significance of this.

Cartledge and Hancock (1942) reported the pedigree of a family through four generations in which sixteen were breech deliveries, seven normal vertices, and six unknown. Although no allusion is made to it by the authors, it would be of interest to conjecture whether some form of uterine anomaly existed in these cases to explain the high incidence of breech delivery and whether this does not represent a further example of the familial tendency of uterine malformations. Unfortunately there is no record of any uterine investigation.

AETIOLOGY

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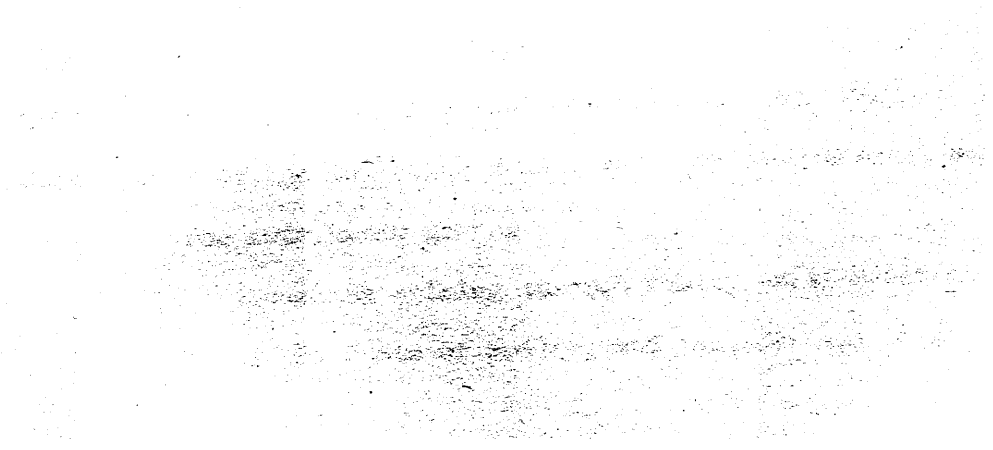
ANATOMICAL CLASSIFICATION



CLASSIFICATION

The lack of a generally accepted classification has led to considerable confusion in assessing cases reported in the literature. Not uncommonly the same term is used to describe different anomalies and even more frequently the same anomaly is known by several different designations and such terms as didelphys, pseudodidelphys, duplex and bilocularis are used interchangeably. This variable nomenclature renders many of the published reports valueless as it is often quite impossible to decide, unless there is an accompaning diagram, the precise form of anomaly being described. The present position is clearly summarised by Hunter (1950) who says 'the descriptions are frequently lacking in precision and the loose confused terminology renders many of the published conclusions of little value.'

Kaufmann (1922) was the first to attempt a proper classification in which he proposed a scheme with four basic groups, each representing a distinct phase in the abnormal development of the Mullerian ducts, and each with several sub-groups. The four basic groups were:-

(1) anomalies arising through faulty juxta-position of the ducts of Müller, and includes all forms of double uterus.

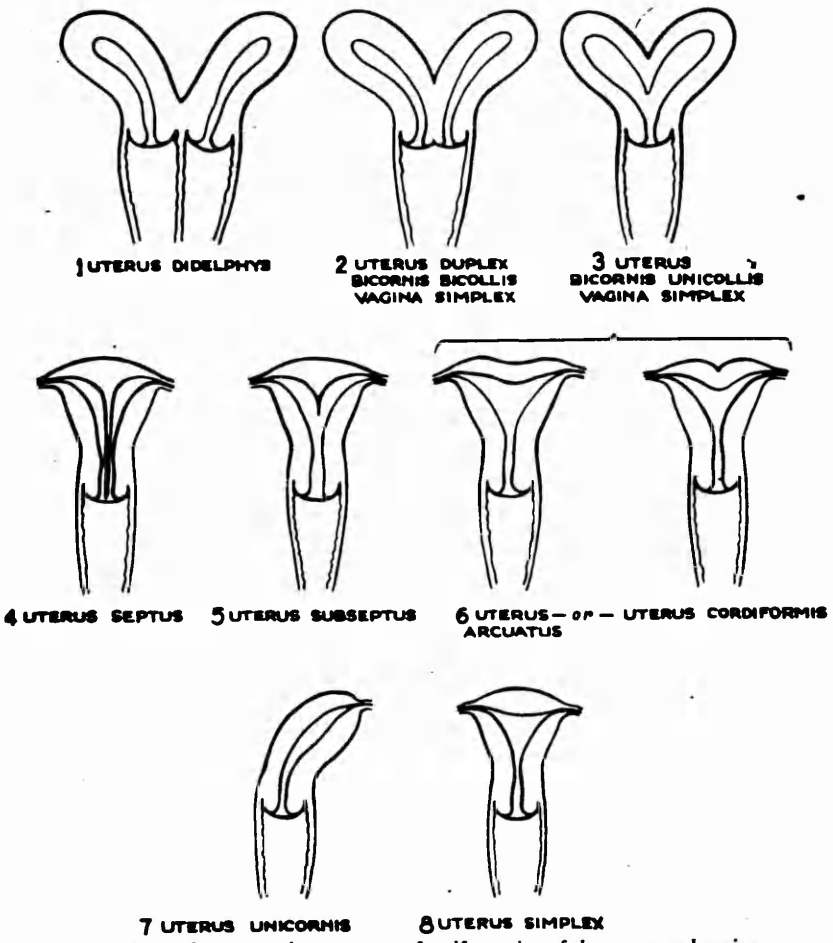


FIG. 2. Normal uterus and seven types of malformation of the uterus and vagina.

4. Jarcho's Classification of Congenital Uterine Anomalies (1946)

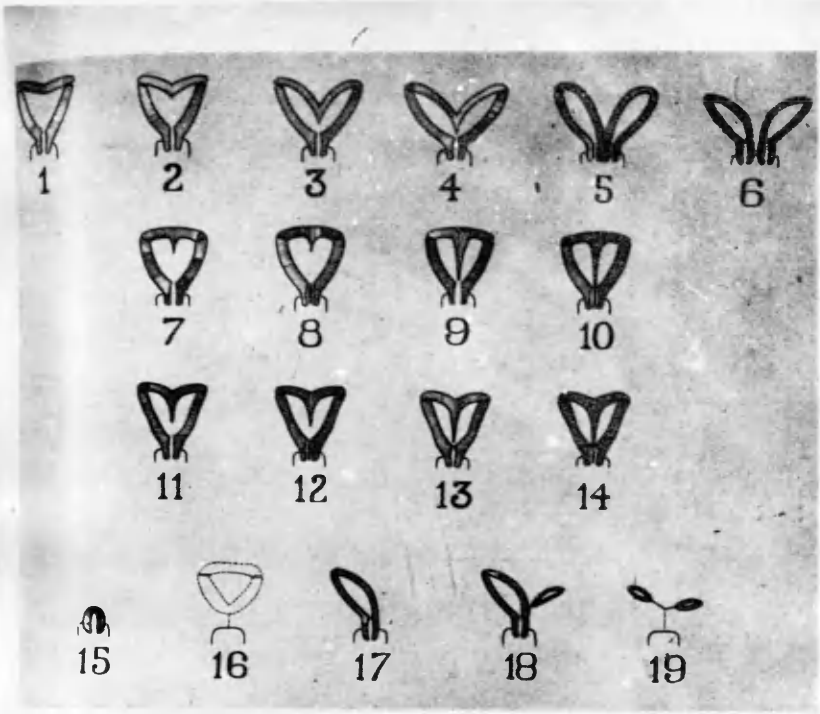


FIG. 1.

UTERINE DEFORMITIES

1. Uterus arcuatus.
2. Uterus cordiformis.
3. Uterus bicornis unicorpus unicollis.
4. Uterus bicornis bicorpus unicollis.
5. Uterus bicornis bicorpus bicollis—pseudo-didelphys.
6. Uterus bicornis bicorpus bicollis—didelphys.
7. Uterus simplex subseptus unicorpus unicollis.
8. Uterus simplex subseptus unicorpus bicollis.
9. Uterus simplex septus bicorpus unicollis.
10. Uterus simplex septus bicorpus bicollis
11. Uterus cordiformis subseptus unicorpus unicollis
12. Uterus cordiformis subseptus unicorpus bicollis.
13. Uterus cordiformis septus bicorpus unicollis.
14. Uterus cordiformis septus bicorpus bicollis.
15. Absence of body of uterus.
16. Absence of cervix.
17. Absence of semi-uterus (uterus unicornis unicorpus unicollis).
18. Rudimentary uterine horn—unilateral.
19. Rudimentary uterine horn—bilateral.

F.H.

5. The more precise classification suggested by Hunter (1950).

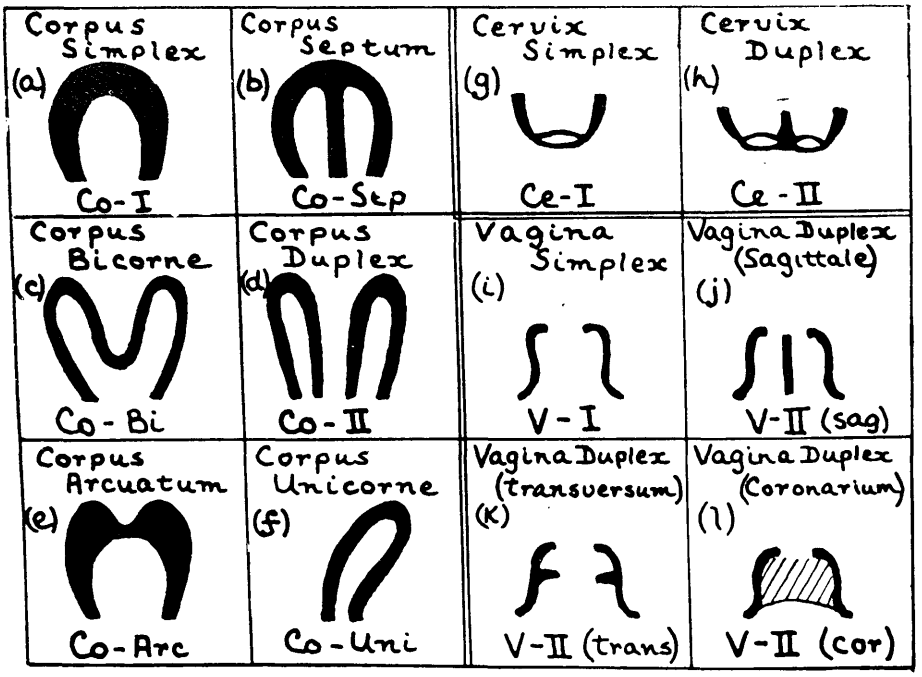


Fig. 1.—Outlines illustrating the principal terms used in Table I.

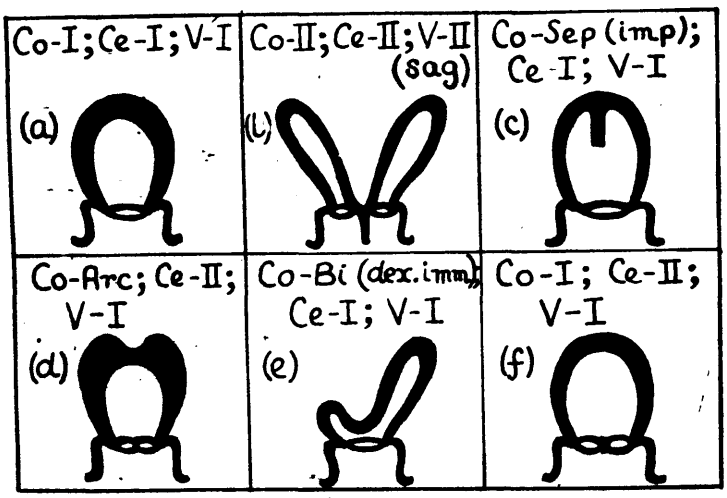


Fig. 2.—Various cases as they would be classified according to the scheme in Table I.
(a) Uterus (corpus simplex et cervix simplex) et vagina simplex.
(b) Uterus (corpus duplex et cervix duplex) et vagina duplex (sagittale).
(c) Uterus (corpus septum imperfectum et cervix simplex) et vagina simplex.
(d) Uterus (corpus arcuatum et cervix duplex) et vagina simplex.
(e) Uterus corpus bicorne (dextrum immaturum) et cervix simplex et vagina simplex.
(f) Uterus (corpus simplex et cervix duplex) et vagina simplex.

6 & 7. Classification of Monie and Sigurdson (1950). Anatomically accurate with an ingenious method of diagrammatic representation.

Fig. 1 and 2.

A bicornuate uterus with a rudimentary horn would come under Type 111.

Others, like Way (1945) and Hunter (1950) feel that a more precise classification is required and should incorporate, whenever possible, the form of uterine fundus, cavity and cervical canal. In this direction the classification of Monie and Sigurdson (1950) is the most comprehensive and anatomically precise. Here,in addition to a classification which shows the type of uterus, cervix and vagina there is an ingenious system of abbreviations and diagrams showing the different forms of anomaly in simple form.

Unfortunately, most systems are too complex or over-simplified and although the more complex systems are more suitable especially for the compilation of statistical data, they tend to be cumbersome and for that reason not generally acceptable. At the present time the classification used by Jarcho, although far from perfect is the most widely adopted and the one in which the majority of cases are reported in the world literature and for this reason it has been adopted in this thesis.

ANATOMY

The muscular development in the majority of these uteri is normal although the medial walls, where there are separate horns tend to be less thick and in septate uteri there is sometimes a visible and palpable groove extending from the fundus down the posterior wall in the midline. The consistency and colour are usually normal and the cavity especially in the uterus didelphys and pseudodidelphys are fusiform. The distance from the external os to the apex of the uterine cavity measures 2.5", and the capacity on an average 6 ml. which approaches that of the normal uterus, Hunter (1950). Karczma(1954) in a careful radiological study of congenital uterine malformations came to the conclusion that:

(a) each half presents a reduced cavity

(b) 6.25% remain relatively symmetrical

(c) 93.75% are asymmetrical in shape and size.

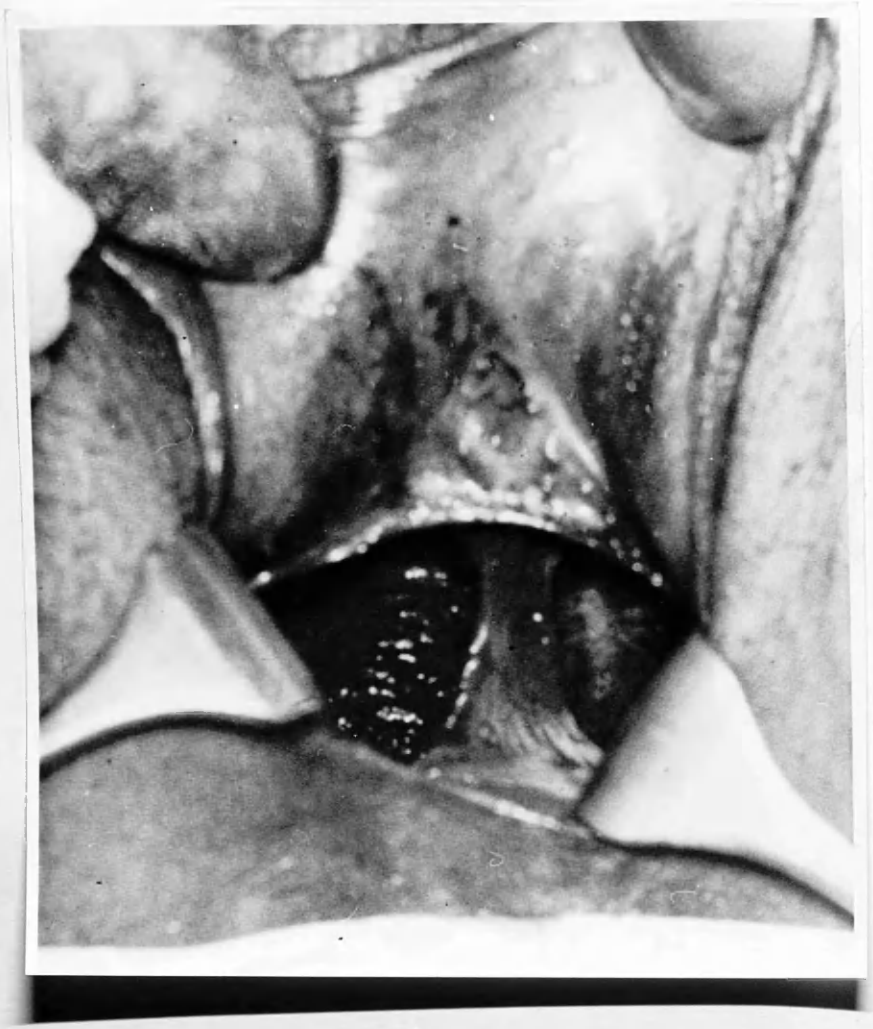
In 16 cases where there were separate cavities, bicornuate or septate, which were measured by a hysterometer, a difference of 1 - 1.5 cm. was found in the length of the horns. The normal position is one of anteversion and in the minor degrees of external deformity anteflexion also. However in the major forms of the anomalous uterus the horns diverge at an angle of 60 - 80 degrees and are almost straight in contrast to less severe forms where the angle between the horns varies between 30 - 150 degrees.

The broad and round ligaments especially in the uterus didelphys tend to be shorter and thicker (Blair Bell 1909). The Fallopian tubes are frequently tortuous, undulating and are usually directed upwards to continue the curve of the uterine wall, a condition which is normal in some of the lower animals.

Recto-vesical and sigmo-vesical peritoneal septum. This has already been discussed under the section on aetiology and embryology, (Page).

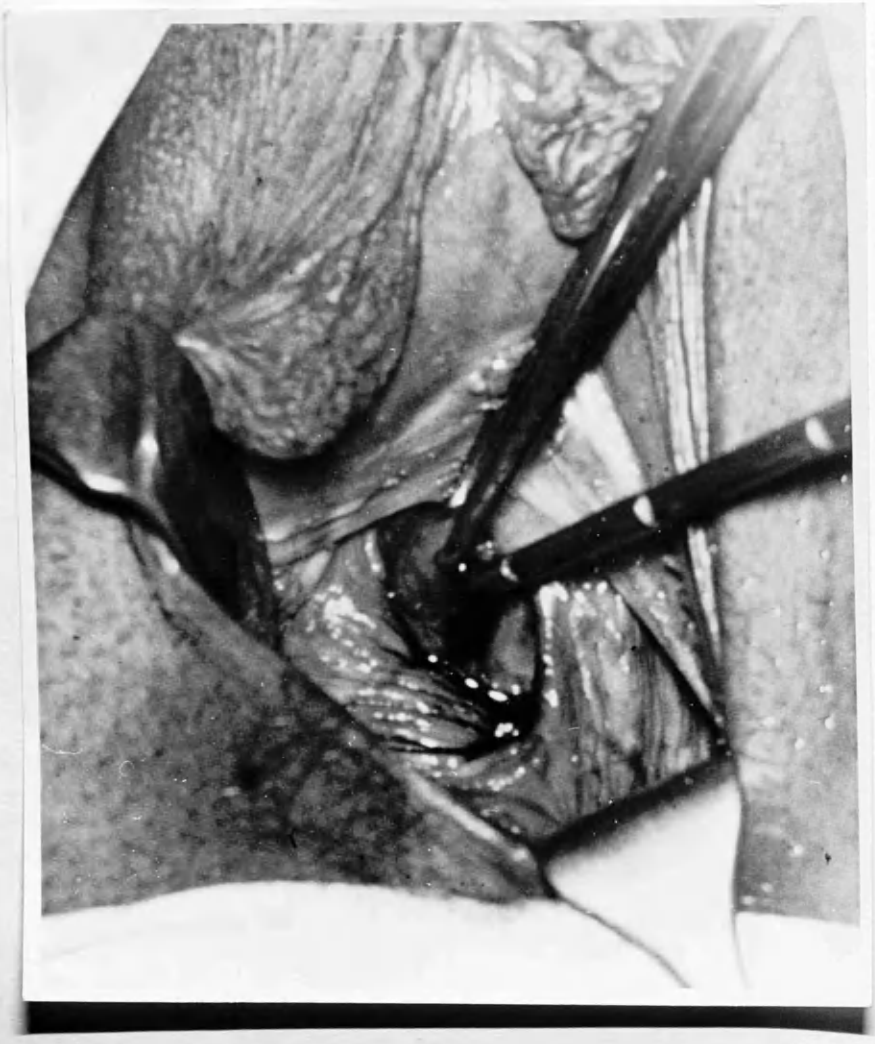
Cervices - may be equal in size, both underdeveloped or one normal in size and the other so underdeveloped that the only evidence of its existence is a small papilla in the vault of the vagina. Again the medial margins of the cervices may be fused giving a wedge shaped single cervix with two canals, or an apparently single cervix, may be partitioned by a sagittal septum.

The two cervices are usually divergent from above down and give the impression of being firmly bound together at the level of the internal ora which is in fact the probable level of closest union between the two uteri. The cervical canals also diverge from the internal ora to the external ora. When the minor difficulty of bringing the cervix into the axis of the vagina has been overcome, no undue difficulty is experienced in dilatation of the canal. In the case of a double uterus having a single cervix it differs in no way from the normal.

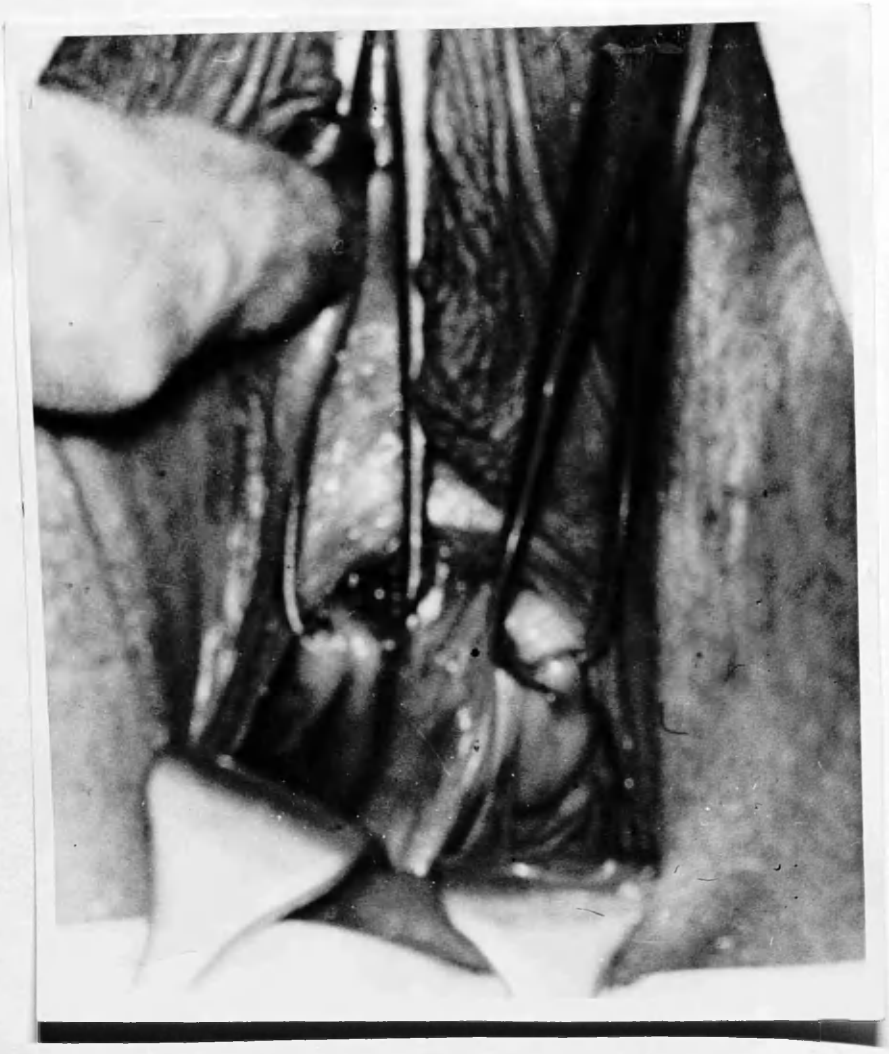


8. Vagina divided sagitally by thick median septum - from Case SM 2.

Where one cervix is atretic a haematometra may form in the attached horn, similarly where one vagina ends blindly, a haematocolpos may form.



9. Right Cervix exposed SM 2.



10. Exposure of both cervices separated by vaginal septum.
SM 2.

Gross forms of congenital anomaly of the genital tract are usually accompanied by serious developmental errors in other parts of the body which are inconsistent with survival, and for this reason are rarely encountered in the adult, Keevil (1943) and Hunter (1950). Extreme examples of marked maldevelopment of the genital tract are usually reported in detail and the more outstanding of these are recorded below:

(1) Duplication lower spine, pelvis, lower limbs - the two medial legs being underdeveloped, - and all the pelvic organs, in an adult monocephalic ileodelphic monster which survived and reached adulthood. Pregnancy occurred but was terminated at $3\frac{1}{2}$ months for severe hyperemesis gravidarum, when an apparently normal foetus was removed, Wells (1888).

(2) Double uterus, vagina, sigmoid colon and rectum. Ladd and Chisholm (1943) and a further case by Robinson (1946).

(3) Double uterus with double vulva and vagina, urethra and bladder, Von Engel (1887). A further example was reported by Gemmell and Paterson (1913) in which there were successive full-time pregnancies and labour in each uterus.

(4) Double uterus with duplication vulva, vagina, bladder and rectum, Lesbré (1927).

(5) Duplication vulva, vagina and bladder, and rectum with duplication uterus in a two year old child, Ombrédanne (1936). An earlier example was reported by Suppiger (1876) in a newborn child.

(6) Uterus didelphys with imperforate vagina and rectum, sigmoido-urethral fistula and absence right kidney and ureter, in an infant which lived 21 days, Siegel (1944).

(7) Bicornuate uterus with three Fallopian tubes, two from one horn and one from the other which contained an ectopic pregnancy, Thorek et alia (1950).

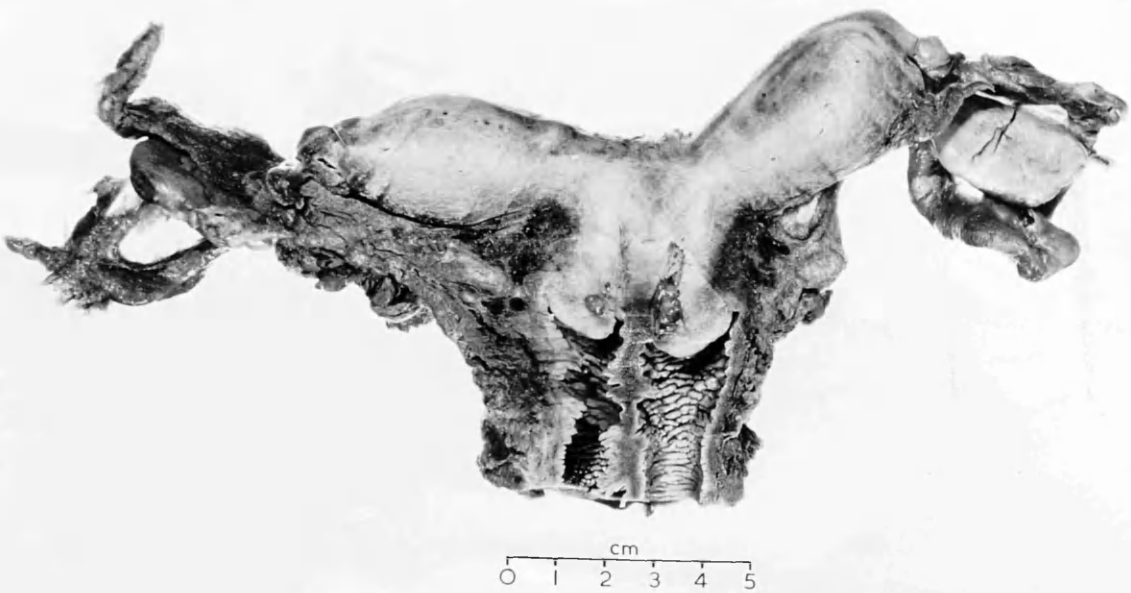
(8) Three distinct uterine cavities with three Fallopian tubes, Kelso (1956). (The existence of this anomaly is based on hysterosalpingography alone and its interpretation is more imaginative than factual).

(9) Complete Agenesis of Mullerian Ducts in an adult, Grogan and Blacker (1956).

The true double uterus in the anatomical sense should arise from two sets of Mullerian ducts, in which each uterus has two Fallopian tubes and two ovaries. Other than in monsters which have not survived, there does not appear to be an authentic example of such a malformation, Hunter (1950), for although Crossedale (1894) described such a case there is insufficient evidence to support his claim.

Uterus Didelphys

In this, the most extreme form of genital anomaly, the duplication of the uterus, vagina and vulva are absolutely distinct, and frequently the bladder is also duplicated, and the pelvis of the "split" variety. A strong fold of peritoneum runs between the two uterine bodies, connecting the rectum and bladder, the recto-vesical ligament. Piquand (1910) in a search of the literature was only able to collect nineteen or twenty examples which he considered authentic and only a further half dozen examples have been added since.

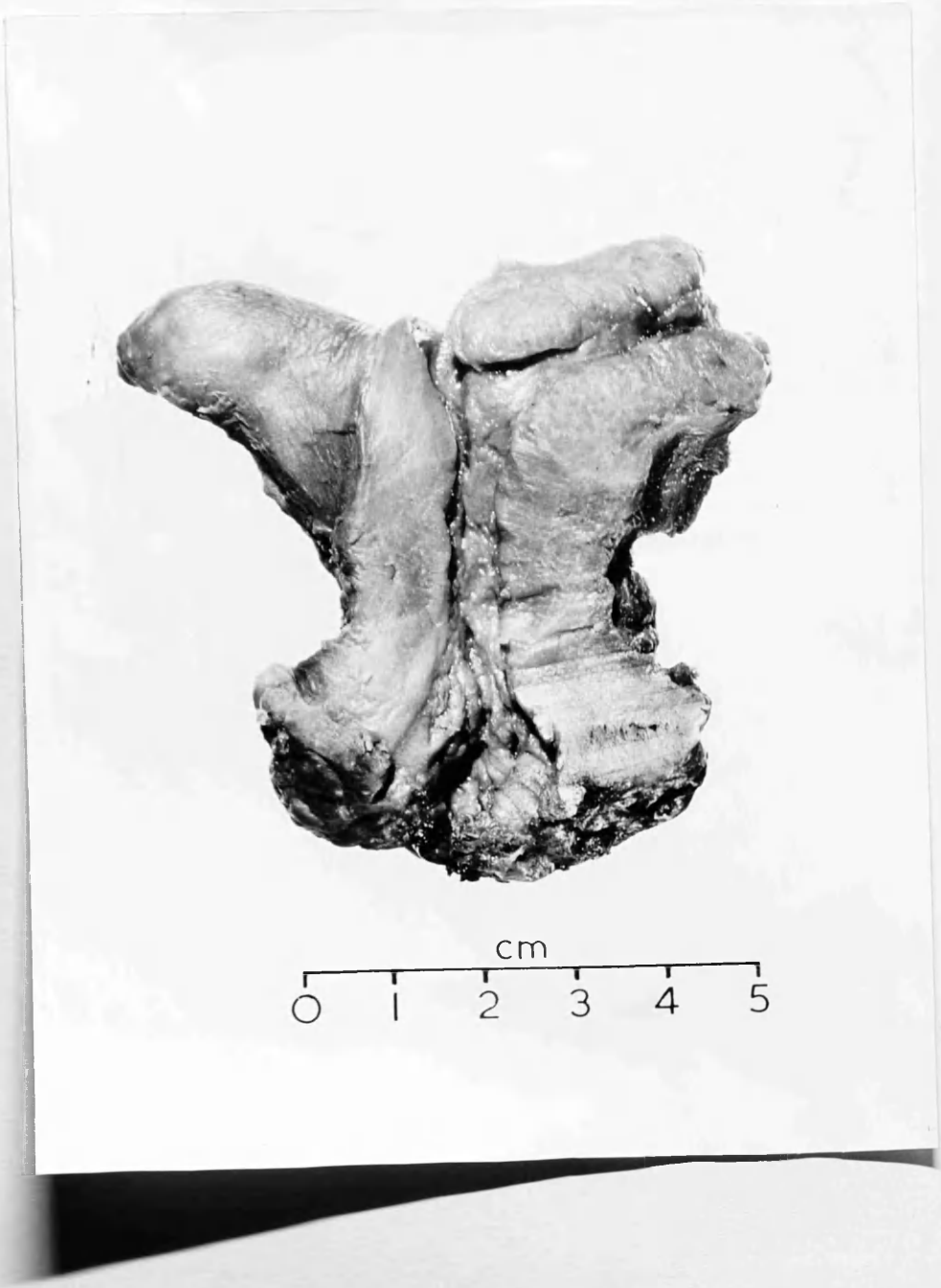


11. Uterus Pseudodidelphys
(Bicornis bicorpus bicollis vagina septate).

Coronal Section to show two uterine bodies and cervices; the latter separated by vaginal septum.

Gordon Museum.

equal in size, but more rarely one or both may be underdeveloped to a degree which leads to the formation of a haematometra in the attached uterine horn or horns. Failure of canalisation between the upper four fifths and lower one fifth of the vagina, on one or both sides of a septate vagina may cause unilateral or bilateral haematocolpera.



12. Bicornuate Uterus(bicornis bicorpus unicollis).

Specimen opened to show junction of the two horns above the level of the internal os.

Gordon Museum

BICORNUATE UTERUS Type 111

UTERUS BICORNIS BICORPUS UNICOLLIS

Here fusion of the two Mullerian ducts is complete to a point just above the internal ora, resulting in the two horns being unduly separated; there is usually a single vagina but occasionally it may have a longitudinal or transverse septum. As there are completely separate uterine horns it will be subject to the same mechanical disadvantage as the uterus pseudodidelphys. Unfortunately the term "bicornuate" is frequently applied to any uterus with two cavities whose bodies are not completely fused throughout their length. This does not entirely arise through descriptive inaccuracy but because of the clinical difficulty of deciding between the lesser degrees of bicornuate uterus and septate forms with a notched fundus.

Munro Kerr considers "bicornuate" should strictly speaking be confined to the arcuate and uterus bicornis unicollis. The uterine horns may be equal in size, both underdeveloped or one normal in size and the other rudimentary. Rarely one or both horns may be solid and the site of endometriosis

UTERUS BICORNIS UNO LATERIS RUDIMENTARIS

Type 111

Jarcho considers this a form of bicornuate uterus in which one horn is underdeveloped as a result of hypoplasia of the corresponding Mullerian duct and, according to Anderson (1943), it is the right horn which is so affected in 80% of cases. The degree of development of the affected horn is very variable, from being only slightly smaller than normal on the one hand to the other where it is so small that its presence is easily overlooked and frequently confused with the much rarer uterus unicornis.

The condition may be bilateral and if there is an associated failure of canalisation, the rudimentary horn or horns will remain solid. The attachment of the rudimentary to the normal horn is usually at the level of the internal os and consists of a distinct band of tissue which is rarely canalised or if a canal exists, ends blindly Munro Kerr (1940). In 80% Perrigard (1942) was unable to find evidence of communication between the rudimentary and normal horn and vagina thus confirming the earlier observation of Piquand (1910) that a patent canal exists in only15% of cases and only where

there is a minor degree of underdevelopment present and the anomaly approaches the uterus bicornis unicollis.

The Fallopian tube of the affected horn is commonly small but the ovary approaches normal. Unless in the more marked forms of atretic horn normal cyclical bleeding will occur from the lining endometrium and in the non-communicating type of anomaly will lead to the formation of a haematometra, Franks (1931).

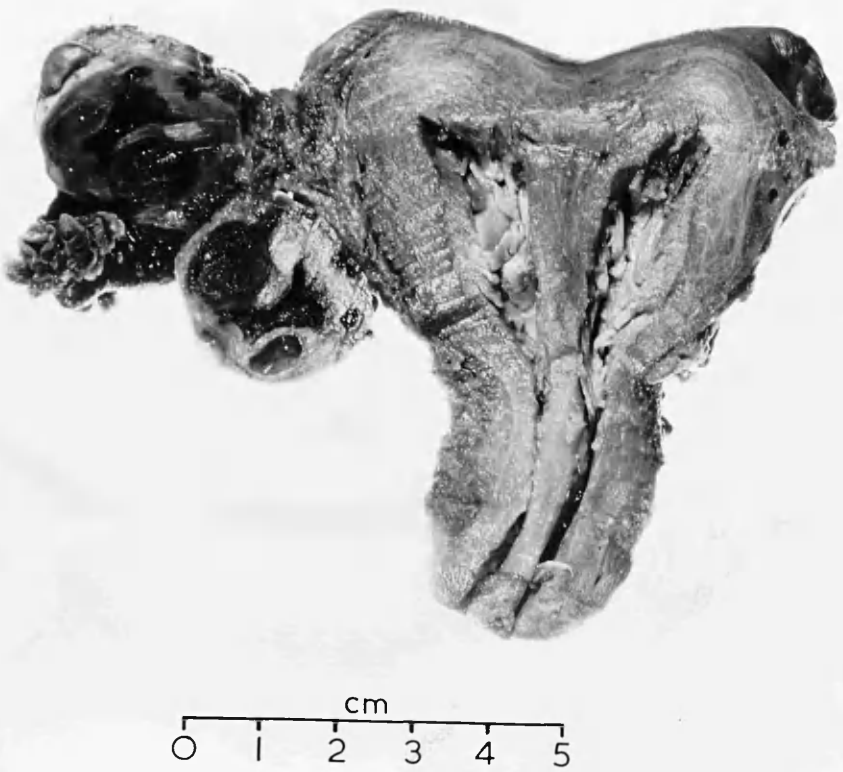
UTERUS ARCUATUS

Syn. uterus bicornis unicorpus

Falls (1939) defined this, as a form of bicornuate uterus in which normal fusion of the two horns in embryonic life stopped just short of completion, forming an organ which in the non-pregnant state can with difficulty be differentiated from the normal, but which during pregnancy exhibits marked irregularity of contour. Externally, this form of anomally is marked by a fundal concavity and where the line of demarcation is deeper and more definite, giving rise to a heart-shaped fundus, the malformation is generally referred to as a uterus cordiformis. On external examination only slight indentation or arching of the fundus may be felt. Such a uterus frequently contains a complete or incomplete septum.

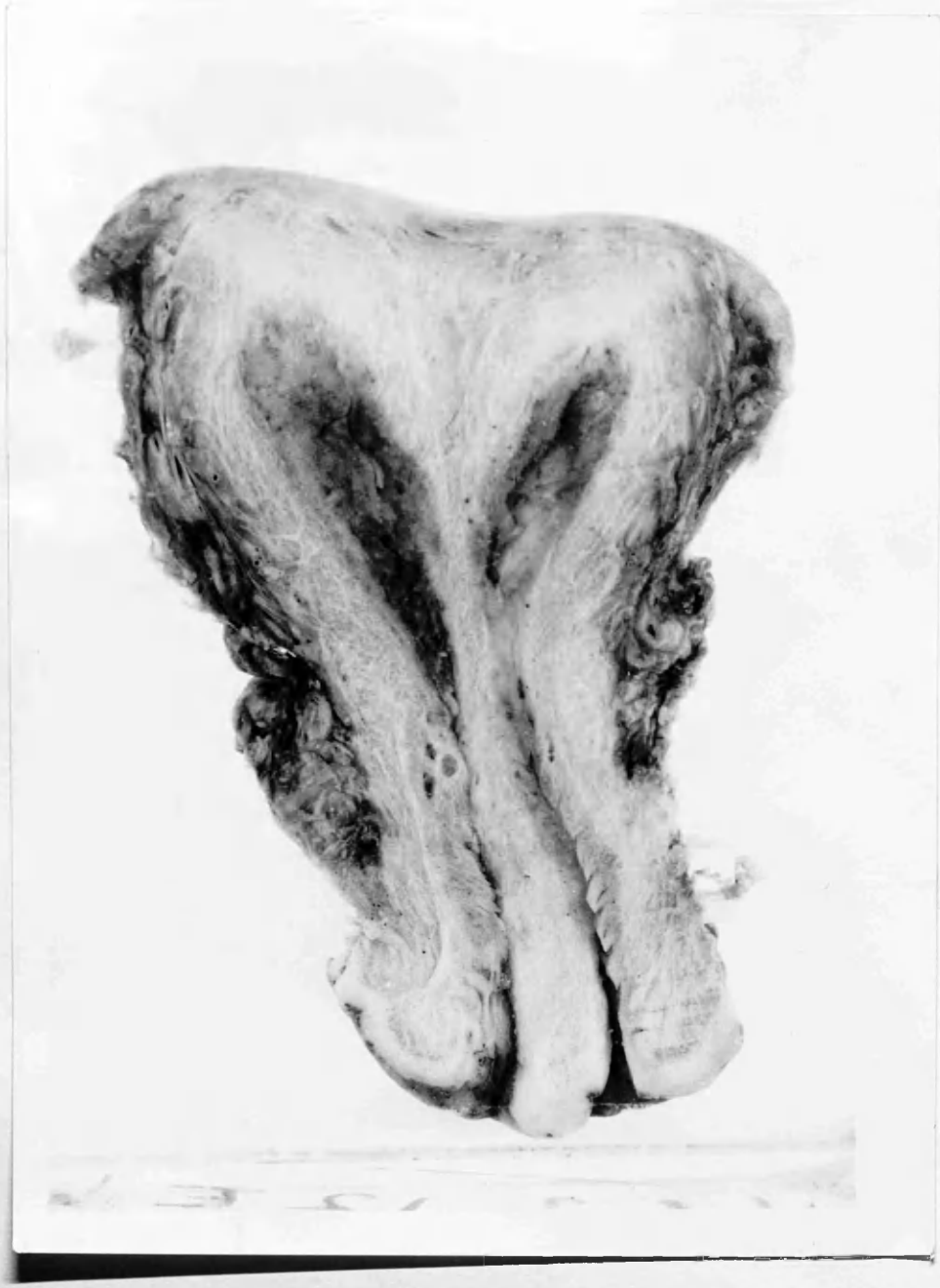
Way (1945) considers the arcuate uterus to be present when the fundal depression does not involve the whole depth of the myometrium, this type although quite common does not interfere with pregnancy. That form of anomally normally designated as arcuate is termed uterus bicornis unicorpus by Way.

Perrigard (1942) and Schauffler (1941) consider this the commonest form of congenital anomally and Falls (1939), in 7,553 consecutive deliveries at the Research and Educational Hospital, Chicago, found 155 examples, an incidence of 3.8 per cent.



13. Septate Uterus (cordiformis septus bicorpus bicollis). Septum is complete and divides uterine cavity and cervix. Note central depression of fundus.

Gordon Museum.



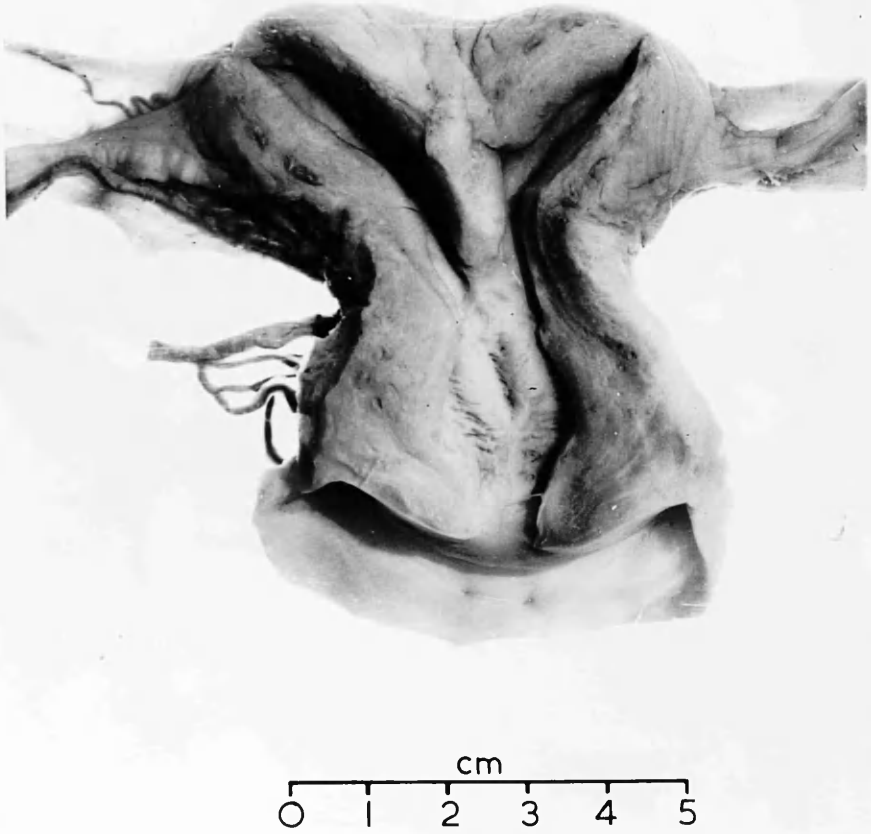
14. Septate Uterus - similar to (13)
but with less pronounced fundal depression.

Gordon Museum

UTERUS SEPTUS

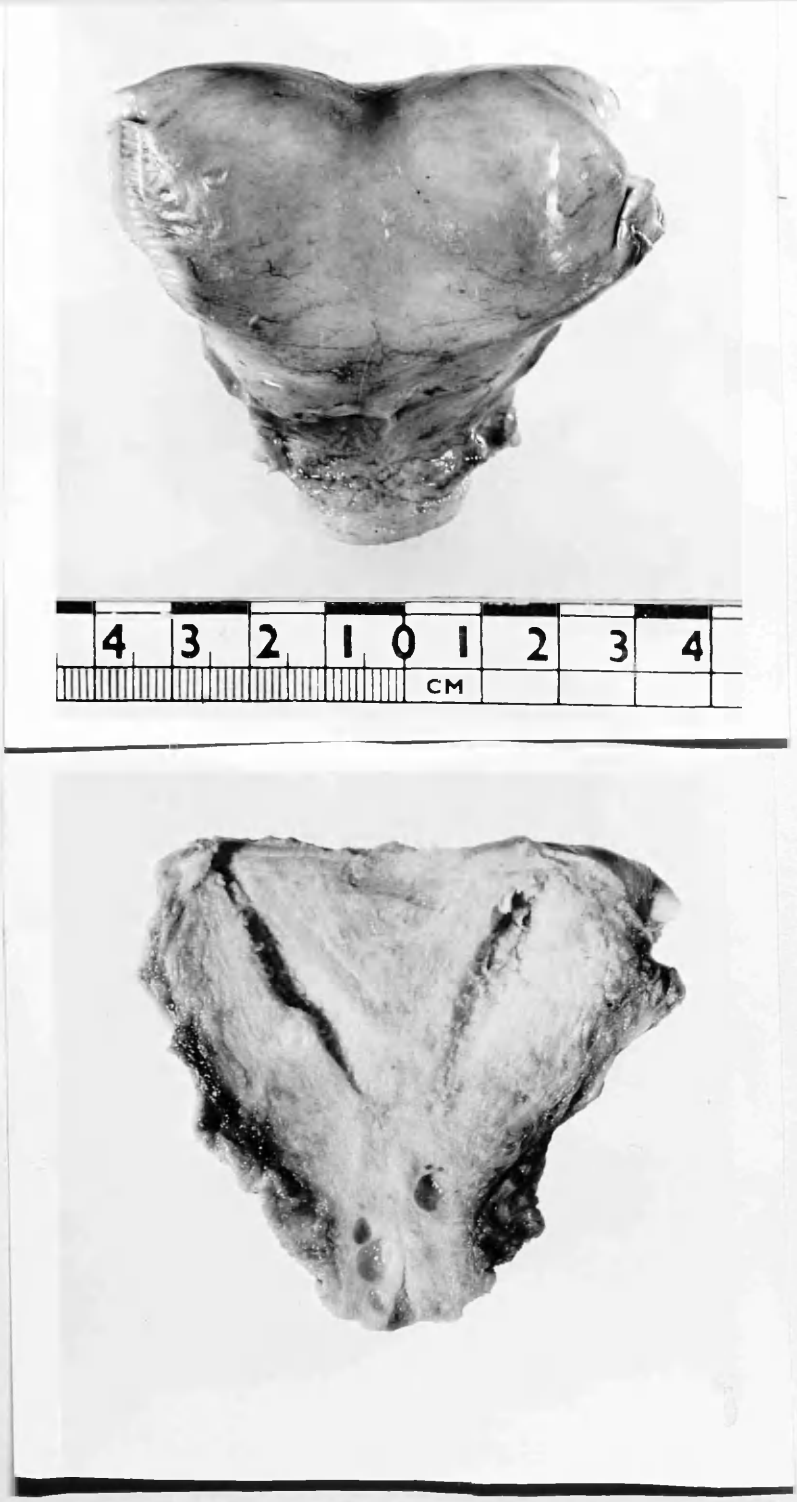
Syn. (uterus bilocularis)

Failure resorption of the medial septus results in this form of anomally, where externally the uterus appears normal. Internally a septum extends from the fundus usually to the external os dividing the cervix, occasionally even beyond, forming a vaginal septum. The uterine septum is normally thin but may be very thick, especially if one half of the uterus is less well developed than the other. One half of the divided cervix may be imperforate and cause the formation of a haematometra Munro Kerr (1949).
Munro Kerr and Way (1945) among others consider the outline of the fundus should have a smooth uninterrupted outline with no sulcus present, while Jarcho considers a mild fundal depression not inconsistent with this form. This latter form Munro Kerr would classify as a form of bicornuate uterus. It is this ambiguity which gives most weight to the adoption of a classification in which the cornu, body, and cervix are separately designated.



15. Subseptate Uterus (cordiformis subseptus unicorpus unicollis).
Septum is incomplete, from fundus to internal os.
Faint fundal depression.

Gordon Museum



16 & 17. Subseptate Uterus

a. well marked fundal depression.

b. specimen opened showing thick incomplete septum.

Author's specimen.

UTERUS SUBSEPTUS

Similar to the septate form except that the septum does not extend as far as the internal os. Rarely there is a central deficiency in the septum which is fenestrated. (Injury to the septum in labour may cause a similar appearance). Morphologically there is little difference between this anomaly and the arcuate uterus even if one restricts the term to a uterus with a smooth fundal outline, the difference no longer pertains in the pregnant state for inequality of growth in the two halves will produce a fundal depression.

UTERUS UNICORNIS

This very rare anomaly arises through complete suppression of one Mullerian duct, Schumaker (1938) and should not be confused with a uterus bicornis unicollis in which one horn is rudimentary and so small as to be easily overlooked. Munro Kerr is of the opinion that nearly all reported cases of uterus unicornis are actually examples of a bicornuate uterus where the presence of the second horn is missed. Of great interest is the frequency with which the uterus unicornis is associated with renal agenesis on the same side as the suppressed Mullerian duct. Neerhut (1954) produced an interesting paper showing the dependance of the Mullerian duct on the Wolffian system for its normal development. He was able to show experimentally that interruption of growth in the Wolffian duct at any point caused cessation of growth at the corresponding point in the Mullerian duct. Schumaker found some major genital defect in 70 per cent of cases of renal agenesis and Ogilvie (1957) found renal agenesis reported in 70 per cent of cases of uterus unicornis.

Commonly both ovaries are present but on the defective side tends to be longer, narrower and more vertical than normally, the blood vessel entering by the upper pole and it commonly occupies an ectopic position, either above the pelvic brim or in the region of the internal inguinal ring.

When present the round ligament is poorly developed and attached to the cervical portion of the uterus. A rudimentary Fallopian tube may be present representing the fimbrinated end and may be solid or canalised and Taber (1950) reported an ectopic pregnancy in this site. All that remains of the broad ligament is the remnant of the lower position. Anderson (1943) states that in 80 per cent the defect is on the right but ~~recently~~ Ogilvie (1957) found that in 64% of cases the agenesis was on the left.

THE HELICOID UTERUS

Sheares (1953) described a uterine anomaly which he termed the helicoid uterus. He observed, in studying a large series of hysterosalpingograms and by examining patients under anaesthesia, that the helicoid uterus was deviated to one side of the midline, and one-half of the uterus appeared to be more anterior, and better developed. In 8 of these cases a partial septum was found by using a uterine sound. He attributes this condition to an inequality in the rate of growth of the two Mullerian ducts. A study of 48 infertile couples with this anomaly showed the incidence of pregnancy to be 11.8 per cent, which was only one-third of the results obtained in the follow-up of a control group.

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CONCOMITANT MALFORMATIONS

Congenital defects of the genital tract are not infrequently associated with developmental anomalies in the other somatic systems, of which the renal, skeletal, and gastro-intestinal respectively, are most commonly found, Woolf and Allen (1953). In view of the close developmental association between the mesonephric and paramesonephric systems, it is hardly surprising that defects of the renal tract are the anomalies most commonly associated with genital malformations. Neerhut (1954) in a series of carefully controlled experiments, showed the complete dependance of the growing Mullerian duct on the Wolffian system for its normal development and that interruption of growth at any point in the latter would result in a cessation of growth at a similar point in the Mullerian duct. Although this would explain the frequent association of renal agenesis and uterus unicornis it does not explain renal agenesis with a normal genital tract - or the converse - a normal renal tract with a uterus unicornis.

Mijsberg (1925) view that the mesonephric duct contributes to the formation of the vagina, is not generally accepted, but Gruenwald (1941) has shown that the closeness of the relationship between the mesonephric and paramesonephric ducts can explain such abnormalties as the ureter opening into the vagina or uterus.

All forms of congenital urinary tract malformation are to be found, but clinically the most important are:-

(1) unilateral agenesis especially where the solitary kidney is in an ectopic position.

(11) horseshoe, congenital cystic kidneys and valves causing back pressure symptoms.

(111)ureteric anomalies especially where there are ectopic openings into uterus or vagina.

(1V) ectopic vesicae and congenital fistulae.

Incidence

Campbell (1951) estimates that anomalies of the upper urinary tract are present in approximately one third of cases of major genital anomaly, and Jones (1957) in a series of 150 congenital anomalies of the uterus found eight with a proved renal malformation, (5.3 per cent) but considers this errs on the low side. The association of renal and Mullerian agenesis occur together in at least 70 per cent of cases Schumaker (1938) which closely approximates the findings of Biet and Legros (1955) based on 20,000 autopsies, that where there was a single kidney it followed in 66 per cent of cases that there was a hemi-uterus and this in turn equals the 41 genital anomalies found by Ballowitz (1952) in 77 cases of unilateral kidney. It is not only with the 'hemi-uterus' that renal anomalies are found,Woolf and Allen (1953) in their interesting paper on concomitant lesions, showed that renal agenesis and ectopic position of the solitary kidney while occurring most commonly

with those genital malformations which arise as a result of ipsilateral errors of Mullerian development, may be found with any form of genital anomaly. Their findings in thirty-five cases are given in detail below.

Table 1

The Uterine Anomaly Arising as a Result of Imperfect Development of Mullerian Ducts

Uterine Anomaly	Number of Cases	Number in which 1 kidney absent	Abnormal Position	Normal Side
Uterus Unicornis	4	3	1(pelvis)	4
Rudimentary or imperfect development one horn	7	5	2(pelvis)	7
Bilateral development both horns, cervices, vaginae but with one vaginae imperforate	4	4	0	4
TOTAL: 15	12	3	3	15

Table 11

The Anomalies of the Uterus Arising as a Result of Imperfect Development of Both Mullerian Ducts				
	Number	Both Kidneys	One Kidney	No Kidneys
Bilateral development both horns with single or double vaginae	8	6	1	1
Complete absence uterus, vagina present	4	4	0	0
Complete absence uterus and vagina	5	2	2 (1 pelvic)	1
Bilateral development uterus with cloaca	3	2	1	0
TOTALS:	20	14	4	2

Pregnancy and labour in an arcuate uterus associated with bilateral fused pelvic kidneys was recently reported by Reinberger and Mackey (1955).

Clinical Significance

The presence of a renal anomaly may be of paramount importance in view of the impaired renal function so often present and the increased hazards to which it is liable in the event of pregnancy. Absence of the kidney on one side would appear to be the commonest renal anomaly and when the solitary organ is situated in the pelvis it is very liable to be damaged during labour, and Everett (1947) reported two deaths which occurred post-partum from renal failure following renal compression during labour, and emphasizes the danger of a pelvic kidney. Even more significant are the findings of Guttierez (1933) that regardless of the position, the single organ is nearly always potentially affected with some chronic disease like nephritis, pyelitis, pyelonephritis or other types of associated pathological process predisposing to urinary stasis. Lack of drainage plus infection lead to hydro- and pyonephrosis and stone formation, terminating in uraemia.

The normal anatomical relations of the ureter may be altered thereby increasing the chances of damage during operation; Counsellor and Huder (1944) reporting fifteen cases of vaginal aplasia in which five had a related anomaly of the urinary tract, draw attention to the importance of this in vaginoplasty operations.

Ectopic ureters opening into the vagina or uterus usually give rise to symptoms which in childhood are diagnosed as nocturnal enuresis and the finding of the ectopic ureteral opening may tax the skill of even the most competent urologist.

The presence of some congenital anomaly of the genital tract should indicate the necessity of investigating the urinary tract and vice versa, and although this has been emphasized by Schumaker (1938) Jarcho (1946) amongst many other authorities, it is still the exception rather than the rule in the published records. Where a genital anomaly is known to exist a careful investigation of the urinary tract must be undertaken and where present careful assessment of the functional value made especially in regard to the potential dangers when associated with pregnancy.

Suprarenal Gland Guttierrez (1933) has pointed out the double origin of this gland, the cortex arising from the Wolffian and the medulla from the neurogenic system, and if one kidney is missing one would expect absence of the cortical element of the same side, but this does not appear to have been noted in any of the cases recorded in the published literature.

Ectopic vesicae

Gross anomalies of the bladder are frequently associated with gross malformations in other systems and are usually incompatible with life, although the rare case reported by Gemmell and Patterson (1913) with duplication of the vulva, there was also duplication of the bladder. Not uncommon are urinary defects associated with the congenital adrenogenital syndrome or female pseudohermaphroditism, ranging from minor degrees of hypospadias to vesico-vaginal fistulae.

Other Associated Malformations

Jones (1957) in reporting an incidence of 7.3 per cent of concomitant anomalies other than of the renal tract, considers this to be fairly accurate as the defects are usually obvious and easily detected, these included vestibular anus, left sided caecum, herniae and skeletal defects. The presence of the last mentioned, with genital anomalies is well recognised and as early as 1901 were noted by Piquand, especially the form known as 'split pelvis' in which there is wide separation of the symphysis pubis of as much as 4" and which may be associated with ectopic vesicae. An increased incidence of contracted pelvis has been reported by Smith (1931) but not confirmed by other investigators. Ungerleider (1952) and Tucker and Baker (1953) have each reported a case of pelvic asymmetry in association with uterus unicornis and renal agenesis

arising through a defect of the ilium. The pelvic inlet was elliptical in shape, with anterior rotation of the whole pelvis on the defective side, where the ilium articulated with the transverse process of the 5th lumbar vertebra. The authors consider this anomaly arises through failure of the ilium to descend caudally and articulate with the 1st sacral vertebra. In both cases a lumbar scoliosis was present on the defective side and also spina bifida occulta. This latter condition has frequently been reported in all degrees and forms as a concomitant lesion, and worthy of inclusion is the case described by Woolf (1953) of pelvic kidney, rudimentary horn, hemivertebrae and the Klippel-Feil syndrome. The associated deformities in a case of vestibular anus reported by Burton Brown (1948) were a septate uterus, a horse-shoe kidney and bilateral congenital dislocation of the hips; the latter is of particular interest in view of the well known tendency for it to be inherited.

Congenital Anomalies of the Gastro-Intestinal Tract - are most commonly of the type arising from persistence of the cloacal membrane, recto-vaginal fistulae or vaginal anus Woolf (1953), and vestibular or vulva anus which is commoner than the first type and may have good sphincter control and be discovered for the first time at confinement, Munro Kerr (1944).

The presence of a cyst of Gartner's duct should suggest the possibility of an associated congenital malformation of the genital or renal tracts, Bleier (1955) who describes three such cases and reviews a further six abstracted from the literature. In two of the cases the cysts communicated with the vagina through a canal in the cervix but in the third there was no such communication. All three were accompanied by some malformation of the urogenital system, unilateral renal agenesis in the first, congenital dilatation of the ureter in the second, and in the third bilateral duplication of the kidneys and ureters, double uterus and interventricular septal defect.

CONCOMITANT MALFORMATIONS

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INCIDENCE

When one recalls that these malformations are not infrequently discovered for the first time at post mortem and that their presence is not incompatible with a normal reproductive life, it will be realised that their true incidence cannot be accurately ~~be~~ determined. However, their importance lies in relation to their effect on pregnancy and the nearest approach to the true incidence is to be gathered from hospital records extending over a number of years, but even here a considerable number of cases will be missed making the estimated incidence on the low side. The estimated incidence shows a wide variation which to a certain extent will depend on the efficiency of the clinician and his ability to recognise the condition, and this is well brought out in Smith's (1931) study of 27, 703 consecutive deliveries over a five year period, where the incidence of all types of congenital uterine anomaly was 1/1458. The figures from the same hospital for a twenty-five year period was considerably lower 1/7040 (114, 243 deliveries) the difference being ascribed to the author's personal interest in the condition and his ability to recognise it, and similarly Stanley Way considered the increased incidence between his first and second studies

in 1945 and 1947 respectively, to better diagnostic ability.

Individual reports covering one or two cases usually advance our knowledge of uterine malformations, but do not give a true index of their frequency, Jarcho (1946). Thus in a review of the literature between 1922 - 36 Beaver and Abbott (1937) found 246 cases reported, but they readily admit that because of the voluminous literature, other cases may have been overlooked, and in addition only those cases adjudged to have sufficient clinical interest are reported, and these often without adequate detail. It is now conceded by most obstetricians that these anomalies are much more frequently encountered than was formerly considered, Jones (1950) and is accounted for by the increased interest in the condition and improved methods of diagnosis. Even so, contemporary figures still show a wide variation especially in relation to any particular form of anomaly but this, at least in part, arises from the confused terminology.

Kimura (1930) recorded 52 examples of uterine anomaly (excluding the arcuate type) in 36, 785 gynaecological Out-Patients, and more recently Hunter (1950) reported an incidence of 1/3000 in 33,274 consecutive admissions to the Newcastle Infirmary, (only those uteri with separate cavities included).

Since these anomalies rarely give rise to symptoms prior to pregnancy, it is obvious that only a small proportion will find their way to the gynaecological clinic, and any incidence estimated on gynaecological material must be well below the true figure.

The reported incidence amongst obstetric patients also shows a considerable variation, and all too frequently the various studies are confined to one particular form of anomaly, and the evaluation of the published data is extremely difficult because of the great confusion in terminology and classification.

Taylor (1943) makes the incidence for all types of genital anomalies to be 1/1,500 obstetric and 1/2000 gynaecological patients and Baker (1953) reports an overall incidence of 0.3 per cent for all deliveries between 1st January, 1950 and 31st December, 1952, but of these only 9 were personal case records - a further 118 having been abstracted from the literature More significant are the figures of the Royal Victoria Hospital, Montreal, where between the years 1942-53, there were 39,190 deliveries, with 41 proved congenital malformations of the uterus - an incidence of 1/954, Philpott (1954).

This excludes the arcuate form of uterus which was considered so common as not to be specifically indexed, so, if included would raise the incidence. Fenton and Singh (1952) reported an incidence of 1/633 compiled on the largest number of individual cases (62) so far published, which compares with 1/700 reported by Jones (1957).

Of special significance are the findings of Fly and Pratt (1956) who examined 4,000 uteri removed by hysterectomy for conditions other than malignancy at the Mayo Clinic between 1950 - 1955 and found five with some form of uterine duplication - an incidence of 1/800.

It should be noted that the incidence when estimated in relation to pregnancy will reflect the true incidence only if fertility and conception rates are the same in the congenitally malformed and normal uterus. Recent investigations (Hunter 1957, et alia) show that the conception rate is adversely affected in the malformed organ and therefore the incidence when calculated in relation to pregnancy will be lower than the true value.

A much higher incidence is recorded by Moore (1941) who, from his personal records estimates the incidence of all forms anomalous uterus and vaginae to be 1/500-600.

Pfleiderer (1929) reported 93 instances of genital anomaly over a period of twenty years at the Tubingen Frauenklinik which were composed of:-

27 arcuate uteri

16 septate uteri with double or single vaginae

15 uterus bicornis unicollis

15 uterus bicornis bicollis with or without double vaginae

14 solid rudimentary uteri with solid vaginae

6 uterus bicornis with rudimentary horn.

An analysis of the 17 cases studied at post mortem by Masson and Kaump (1933) showed the following:-

3 uterus didelphys

4 uterus duplex with septate vaginae

3 rudimentary uteri with absence vagina.

2 showing absence Fallopian tube and ovary

1 uterus unicornis

1 hemihypoplasia uterus with atresia vagina

1 rudimentary uterus with atresia vagina

1 unilateral atresia Fallopian tube.

King's College Hospital

Incidence congenital defects uterus over 10 years (1944-1954)

A

Obstetric Incidence:
12 congenital uterine anomalies
in 10,840 deliveries
= 1:903

Year	Uterine Abnormalities	
1954	Bicornuate uterus	Neonatal death. Premature Labour.
1953	Bicornuate uterus	Lower segment Caesarean section
1952	Bicornuate uterus	Premature live birth, Neonatal death (also 1954)
1951	Bicornuate uterus	Lower segment Caesarean section
1951	Impacted horn	Caesarean section
1950	Cordiform uterus	Normal delivery
1949	Bicornuate uterus and septate vagina	Lower segment Caesarean section (also 1953)
1948	Double uterus	Normal delivery
1948	Bicornuate uterus	Lower segment Caesarean section(also 1951)
1948	Bicornuate uterus	Normal delivery
1948	Bicornuate uterus	Lower segment Caesarean Section(also 1951)
1944	Bicornuate uterus	Normal delivery.

King's College Hospital

B Incidence for Gynaecological patients

13/4372 patients
= 1: 336

excluding 1 left idiopathic megaloureter
2 infantile uteri.
= 11/4372
= 1: 397

Analysis Cases (1) Obstetric: 3 of Type 1
9 of Type 111

(11) Gynaecological:

Year	Uterine Abnormalities
1954	Rudimentary horn
1954	Absence of ovaries and uterus
1954	Congenital absence of vagina and uterus
1954	Congenital bifid uterus and renal pelvis
1952	Congenital abnormality of genital tract
1951	Bicornuate uterus. Previous normal delivery 1948.
1950	Bicornuate uterus (Had miscarriage 1949)
1948	Double uterus and cervix and septate vagina (Maternity 1949 and 1953)
1948	Absence of vagina and uterus (also 1954)
1945	Double vagina and cervix
1944	Cordiform uterus. Admitted for investigation of miscarriages and again in 1953 for adenomyoma of uterus

Central Middlesex Hospital

For the Years 1945 - 1954 Inclusive

13 congenital malformations of the uterus
in 19,031 deliveries

Obstetric Incidence: 1/1,464

Analysis of the Cases

Type	Number
1	2
111	10
V	1
	13 Total

Incidence in Relation to Type

Type Anomaly	Philpott	Baker	Present Series
1	10%	38%	30%
111	50%	46%	45%
Others	40%	16%	25%

In all three studies Type 111 is the commonest malformation almost equalling the sum of the other two. There is a relatively high incidence of Type 1 in Baker's series and of Type V in Philpotts.

"Double" Uterus - Smith (1931) gives the incidence of all types "double" uterus as 1 in 1,500, but does not define the term except to state that simple forms bicornuate and arcuate uterus, are not included. He also gives the frequency of uterus didelphys as 1 in 28,000 pregnant women.

Bicornuate Uterus - (in its less marked forms) and subseptate uteri. Way (1945 and 47) found ten cases in 3,894 deliveries - an incidence of 1 in 389. This figure is artificially high because of selection of cases - and Way gives the corrected incidence as 1 in 1,223 patients booked for hospital confinement.

Arcuate Uterus - Falls (1939) gives the frequency of this type anomaly as 38 per 1,000 deliveries or 3.8 per cent in over 7,553 deliveries.

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CLINICAL SIGNIFICANCE

Prior to pregnancy it is rare for the congenitally malformed uterus to give rise to symptoms which will bring them to the notice of the gynaecologist.

Spasmodic dysmenorrhoea is relatively common and according to Miller (1923) of diagnostic significance in whose series the incidence was 20%. These views are endorsed by Hunter (1957) who adds that the condition may be severe and resistant to treatment and not relieved by pregnancy. Unilateral dysmenorrhoea should suggest the presence of a uterus didelphys and if persisting throughout menstruation may indicate a unilateral haematometra - Galloway (1947). Even without the formation of a haematometra, a rudimentary horn may cause severe and incapacitating dysmenorrhoea Munro Kerr (1949), presumably by interfering with the normal action of the uterine muscle.

The onset of the menarche was not delayed in the investigation by Hunter (1950) which agrees with the findings in the present study where the average was $14\frac{1}{2}$ years and although the report by Granberry (1938) gives the slightly higher age of 15 years as the age of onset and in some as late as 26 years.

The presence of a vaginal septum may cause dyspareunia and can also act as a mechanical barrier to conception (SM 2) and the chances of fertilisation may differ in the two halves of the vagina. Hymeneal rupture with extension of the laceration into the septum can cause profuse haemorrhage (WM 1).

There would appear to be a definite tendency to menorrhagia, so much so that Jarcho (1946), Steinberg (1952) and Philpott (1954) among others, consider it may be of diagnostic significance. It has been ascribed to the increased endometrial surface - Masson and Rieniets (1933), to the increased incidence of fibromyomata - Beningo (1954) and to a hormonal imbalance - Steinberg (1952) of which anovular menstrual cycles are another manifestation and are commonly found in association with the congenitally malformed uterus. Menstrual bleeding at fortnightly intervals from alternate horns of a double uterus has been reported by Sprigg (1895).

Examples of all the more common diseases of the uterus have been described in association with these anomalies, Hunter (1950). An increased incidence of fibromyomata has been reported by Beningo (1954) but in the five cases described which were all of the uterus bicornis unicollis type, the average age of the patients was thirty-six and in light of this, his findings lose much of their significance. Endometriosis and adenomyosis have also been recorded and interesting reports on the association of this condition with the congenitally malformed uterus are given by Limage (1953) and Leverton (1953), the former describing endometriosis of the appendages associated with pregnancy in a uterus bilocularis unicorpus, and the latter a uterus didelphys solidus with gynaetresia, adenomyosis and pelvic endometriosis.

Gynaetresia

Haematocolpos occurs most commonly when there is an occlusive membrane lying behind the hymen although there may be transverse diaphragms at any level. Where the vagina is divided by a longitudinal septum the condition may be unilateral or bilateral and may rarely be associated with an ipsilateral or bilateral haematometra.

As early as 1900 Wells was able to collect seven examples of unilateral and two of bilateral haematometra, in addition to several pyometra. Haematometra occurs most commonly in association with a rudimentary horn and gives rise to increasingly severe dysmenorrhoea maximal during menstruation. If the condition is unrelieved a haematosalpinx may form leading to drainage into the peritoneal cavity and peritonitis (SM 20) or the condition may be self limiting, the lining endometrium being destroyed and eventually the haematometra becomes organised and if not fixed by adhesions may undergo torsion or prolapse into the Pouch of Douglas in which condition it may cause obstructed labour. The diagnosis is difficult and not usually made before laparotomy and the condition is most commonly confused with a fibroid or ovarian cyst. Haematometra may also be found with the uterus bicornis bicollis and the uterus septus, usually where there is atresia of one cervix or hemi-cervix, rarely from injury as in the case of postpartum gynaetresia associated with a uterus septus with single vagina, which was reported by Weisl (1950).

An unusual example of haematocolpos in which the lower half of the vagina was absent was reported by Henderson (1954) and the very rare condition of hydrometrocolpos is fully discussed in a paper by Lide and Coker (1952).

Malignant Disease

The increased susceptibility of the congenitally malformed organ to malignant change is well recognised yet Von Franque (1930) in a review of the literature from 1770 to 1930, found only nine examples of malignant change in association with the uterus. More recently Fly and Pratt (1956), collected a further eight cases from the literature for the period 1930 - 56 and to this they added three of their own, a total of twenty cases.

Adenocarcinoma of the Corpus Uteri

The same authors from 691 uteri removed at the Mayo Clinic between 1945 - 55 for adenocarcinoma of the body found two with a congenital uterine deformity, an incidence of 1:345.

Carcinoma of the Cervix

In the same ten year period from 633 uteri removed for carcinoma of the cervix, including carcinoma in situ, there was one example of a congenital duplication of the uterus, an incidence of 1:633.

Over the same period the incidence of congenital deformities in uteri removed for non-malignant conditions was 1:800, it would therefore appear that malignant change does more commonly occur in the congenitally malformed than in the normal uterus. Rightly the authors indicate that the number of cases so far reported is too small for any definite conclusion to be drawn.

Infertility

There still exists considerable difference of opinion regarding the effect of the anomalous uterus on fertility and conception. Masson and Kaump (1937), Campbell (1933) and Meaker (1934) feel there is little if any difference between the malformed and normal uterus, while Luikart (1936) and Schauffler (1941) consider the chances of conception are actually favoured in the congenitally malformed organ. Jarcho (1946) reported a lowering of the conception rate through mechanical difficulties such as the presence of a vaginal septum, and Steinberg (1952) concludes that as anovular cycles are commoner in these patients the fertility rate may be lowered, but does not believe that the uterus itself has any effect. Siegler (1944) holds the opposite opinion and Bernhard in Seitz and Amreie (1944) quotes Phillip who in forty-six examples of double uterus found primary infertility in 43.5%, the uterus pseudo-didelphys showing the highest rate with 50%, uterus septus and bicornis unicollis in only 20%. The investigation of Fenton and Singh (1952) and Baker (1953) did not reveal any lowering of fertility, 2.16 pregnancies per patient in the latter series. Philpott (1954), while admitting that his series is too small on which to speak authoritatively, feels that conception and fertility are not materially altered.

This was also the opinion of Hunter (1950) in his first report but later (1957) as a result of a careful investigation and analysis of fifty-one cases of double uterus compared with thirty-eight controls in which the uterus was normal he found that:

(a) few had been pregnant

(b) the total number of pregnancies was less

(c) there were fewer viable pregnancies.

Five times as many patients with a double uterus failed to become pregnant as in the control group but after the first pregnancy fertility in the two groups was similar. Karczma (1954) holds similar views maintaining that the chances of conception are less until the first conception, but once pregnancy is achieved fertility in the malformed and normal organ is equal.

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CLINICAL SIGNIFICANCE IN RELATION TO PREGNANCY

The clinical significance of the congenitally malformed uterus is principally in relation to its effect on pregnancy and will differ according to the anomaly. The normal classification based on developmental anatomy, loses much of its value in pregnancy for the changes consistent with this state make exact identification of the precise form of anomaly impossible. In addition this classification gives no indication of the obstetrical significance or gestational capacity. It is therefore convenient when considering the significance of these anomalies to arrange them into several broad groups each with a fairly distinct clinical pattern indicative of their obstetric significance.

(1) In this group are included all those forms of 'double uterus' where the point of fusion between the two horns is at or below the level of the internal os, and includes the uterus bicornis bicollis and the more marked forms of bicornuate uteri. In pregnancy the non-gravid half usually remains in the pelvis and is not pulled up into the abdomen with the growing pregnant horn and in this position can interfere with descent of the presenting part.

(11) Where the level of fusion of the two horns is above the level of the internal os, the non-pregnant horn is pulled up into the abdomen with the growing pregnant half. Included in this group are the less marked forms of bicornuate uterus, arcuate and subseptate forms.

(111) Those forms which arise from aplasia or hypoplasia of one horn frequently associated with an ipsilateral failure of development of the Wolffian duct, and includes the rudimentary horn and unicornuate uterus.

Group 1

The non-pregnant horn hypertrophies (being subject to the same hormonal influence as the pregnant half) and when the latter rises into the abdomen the former usually remains in the pelvic cavity, and, where it may cause obstruction to the advance of the foetus in labour. Hunter (1950) has pointed out that posterior rotation of the non-gravid horn may cause axial rotation of the pregnant half; the broad ligament being carried forward towards the midline and anterior abdominal wall. This will add to the technical difficulties of Caesarean section as the vascular broad ligament commonly presents in the abdominal wound.

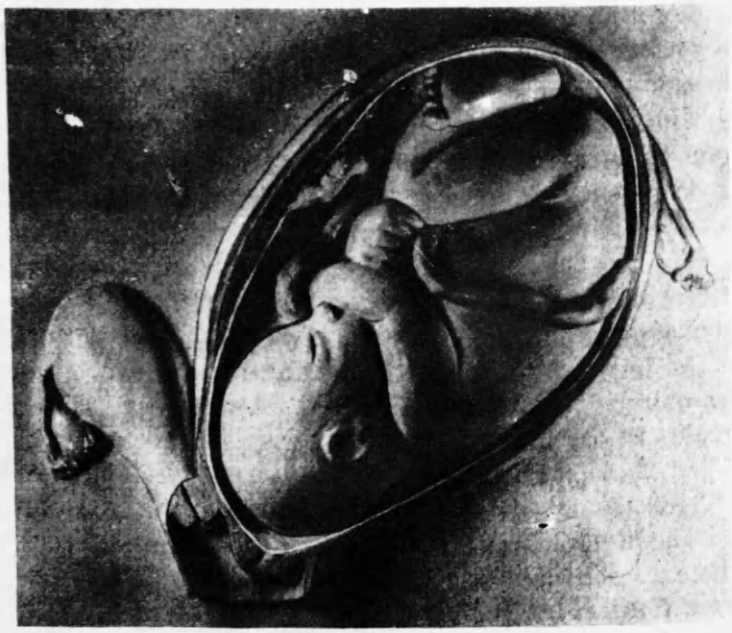

Fig. 3. — *Uterus Didelphys* (10 months pregnancy). The full-term pregnancy causes left lateral deviation. The right horn, somewhat hypertrophied, is also displaced.

18. Uterus Pseudodidelphys.
Pregnancy at term - lateral
flexion gravid half and hypertrophied
non-gravid horn.

Falls 1954.

Torsion of the pregnant horn is associated with symptoms suggestive of concealed accidental haemorrhage, Herd (1945). Horn (1924) and Caswell (1926) both describe cases of torsion at six months cyesis followed by 2 years later by breech delivery of a living child, and in both, the gangrenous pregnant horn was removed by subtotal hysterectomy. More recently Hunter (1957) reported a minor dextro-rotation of the uterus with anterior sacculation of the lower part of the pregnant left horn which was accompanied by rotation of the non-pregnant horn into the Pouch of Douglas, causing an oblique lie of the foetus.

The pregnant horn usually deviates to one side of the abdomen, decidual changes take place in the non-pregnant horn which may be associated with an intermittent discharge of clear fluid during pregnancy, or there may be a discharge of blood similar to menstruation, sometimes at monthly intervals. Until proved otherwise any bleeding during pregnancy should be regarded as a threatened miscarriage or antepartum haemorrhage.

A decidual cast complete or incomplete may be discharged from the non-pregnant horn before, during or after pregnancy, and Corbett (1945).

The pregnant horn in this Group 1 is usually fusiform in shape and has a lower segment which is small and funnel shaped and indistensible, in contrast to the lower segment of the normal uterus. This tapering lower segment may interfere with descent of the presenting part in late pregnancy, favours frank breech presentation, and will also predispose to disordered uterine action. Where the vertex presents it is usually well flexed and may show moulding when still above the brim, presumably in an attempt to accommodate itself to the lower segment. It is commonly high and may be displaced towards the side of the abdomen on which the back lies, and it may also be difficult to fit into the pelvic brim. The foetal trunk may be displaced laterally and lying obliquely in the flanks. The foetal back can either be towards the medial aspect of the pregnant horn or towards the mother's flanks, the legs being closely applied to the ventral aspect of the foetus, Hunter (1957).

Cervices Rarely the cervix of the non-pregnant horn is driven down in front of the foetal head or breech to appear at the vulva. Hunter (1950) considers this to indicate that the non-pregnant horn has prolapsed below the presenting part. A cervical septum may tear or obstruct the advance of the presenting part Arthur(1948) or a uterine septum above the level of the internal os may tear and the leading part of the foetus present at the non-pregnant cervix.

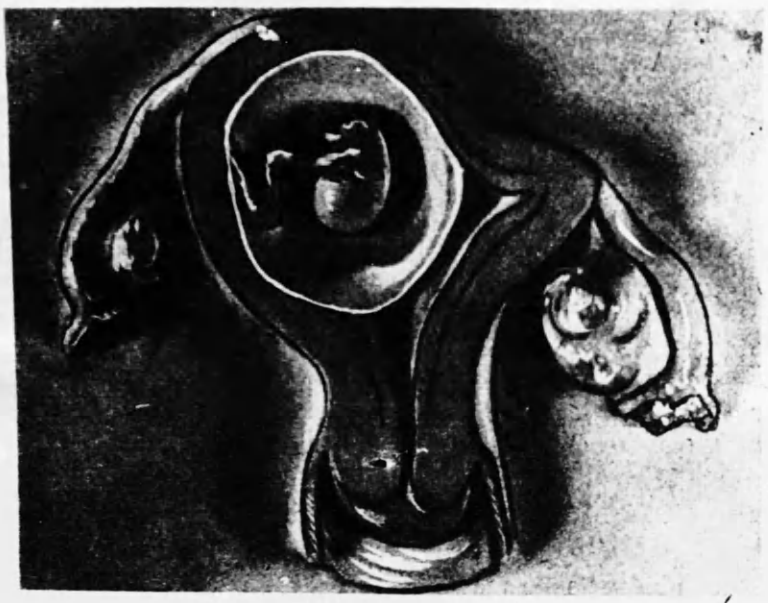

Fig. 1. — *Uterus Arcuatus* (8 weeks pregnancy). Pregnancy in the right horn produces enlargement and softening simulating interstitial ectopic pregnancy. By contrast, the firm left horn often is mistaken for a fibromyoma.

19. Arcuate Uterus.
Early gestational changes.

Falls 1954.

Vaginal Septum - This may delay advance of the foetus through the narrowed vaginal canal. If not divided, will usually tear and the laceration is liable to occur in the groove between the septum and the anterior vaginal wall, with the risk of bladder injury in addition to brisk haemorrhage from the septum. Occasionally obstructed labour has been reported.

Group 11

In this group the non-pregnant horn is lifted up into the abdomen as the pregnant half rises out of the pelvis. The pregnant horn grows more rapidly than the non-pregnant half, producing an asymmetrical uterine enlargement of which the non-pregnant side is of firmer consistency and may be mistaken for a fibroid, Falls (1954). This asymmetrical growth of the uterus is not uncommonly associated with pain and the signs of a threatened miscarriage, these symptoms arising through over stretching of the musculature, especially at the site of imperfect union of the two horns which appears resistent to expansion of the myometrium, Macdonald (1949). The clinical appearance closely simulates interstitial and angular pregnancy.

The inequality of growth between the two sides makes the fundal deformity more marked, but as term approaches and the fundus broadens it again becomes less marked while the presence of the bulky intra-uterine septum forces the foetus to take up an oblique lie, Way (1947).

Premature labour is common, Falls (1954) Hunter (1950) and Way (1947) and has been attributed to a hyper-irritability of the myometrium from Falls (1954) and the reduced gestational capacity Karczma (1954).

Falls, while agreeing that premature labour is usual, has found postmaturity not uncommon in the arcuate uterus and induction of labour often necessary, but response may be unsatisfactory. Eventually when labour does commence the contractions may be weak and ineffectual and a long labour is to be expected. (Although statistically not of value, it is interesting to note that the one case of postmaturity in the present investigation occurred in association with an arcuate uterus.)

Premature separation of the placenta is common Way (1945), especially when inserted on the septum, and this has been ascribed to the poor decidual reaction in the latter Ritschel(1949).

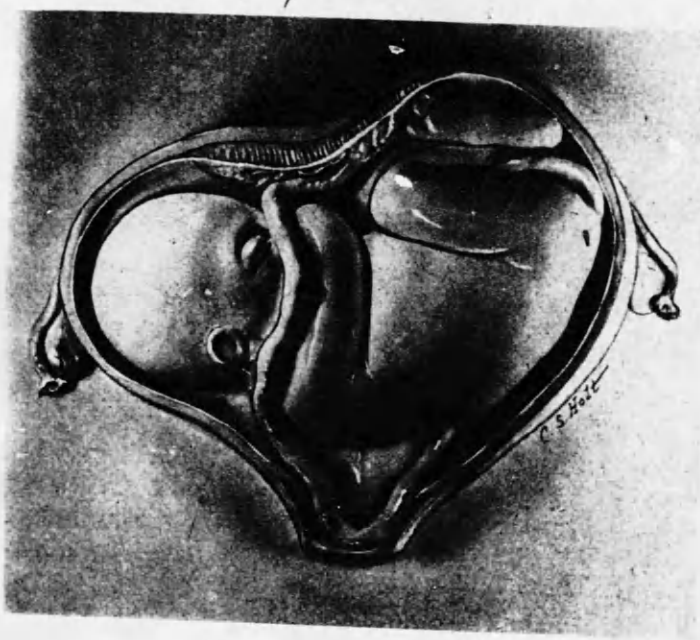

Fig. 2. — *Uterus Arcuatus* (1st stage of labor). Transverse presentation with prolapsed cord and arm and malformation of uterine wall and placenta result in fetal anoxemia.

20. Arcuate Uterus.

The foetus adopts a transverse or oblique position to accommodate itself to the shape of the uterus. The dorso inferior position is always suggestive of the presence of a minor degree of uterine anomaly.

Falls 1954.

The septum may also prevent the normal high insertion of the placenta and so favour placenta praevia, which in turn increases the chances of transverse lie, Way (1945).

Retention of the Placenta

Way (1945) considers this to arise from the high incidence of cornual insertion of the placenta which favours retention of the placenta even in the normal uterus. Falls (1954) warns against the dangers of severe post-partum haemorrhage which occur when the placenta separates and is retained in the opposite horn. Hunter finds postpartum haemorrhage commonest with the lesser degrees of bicornuate uterus and considers this to be caused by mechanical interference with normal retraction from the hypertrophied non-pregnant horn, with its extreme attachment to the pregnant horn. Where the placental site is situated on the septum control of haemorrhage from the torn sinuses it may be unsatisfactory in view of the inefficient myometrium in this area, Hunter (1957). The presence of a septum favours transverse lie unless the foetus is very small, but when labour commences the soft breech may be forced into the pelvis with resultant breech delivery, Way (1945), Falls (1957).

RUDIMENTARY HORN

Uterus bicornis uno lateris rudimentarius

According to Munro Kerr (1949) this variety of malformation is not uncommon and is of considerable importance clinically especially in relation to pregnancy.

In the non-communicating type, which accounts for 80%, Perrigard (1942) the spermatozoon travels up the normal horn to reach the Fallopian tube where fertilisation of an ovum from the ipsi- or contralateral ovary occurs, and implantation of the fertilised ovum in the rudimentary horn then takes place via the Fallopian tube of the atretic horn. The course of pregnancy in a rudimentary horn is variable but the poorly developed musculature already weakened by the eroding trophoblast Hoff (1906) is usually incapable of accommodating the growing ovum Cherry (1929) with consequent rupture of the gravid horn. This accident occurred 40 times in the 246 cases reviewed by Beaver and Abbot (1937) and 45 times in the 100 cases collected by Werth (1906) and, Tien (1949) in a more recent review of the literature gives the incidence as 60%. The time of rupture according to Falls (1933) will depend on the degree to which the muscle can hypertrophy to accommodate the growing ovum but even more important as Munro Kerr (1949) has pointed out

is how rudimentary the horn is, for, naturally, the more rudimentary the horn, the less will be its resistance to the chorionic villi and growing ovum. This is well illustrated in the case described by Duff (1914) in which rupture occurred at the 5th week and where the true condition was not recognised even at laparotomy when it was considered to be a tubal pregnancy, and only after the ruptured sac had been examined microscopically was it proved to be an extremely small rudimentary horn. At the other extrme are the cases where pregnancy proceeds to the later months of pregnancy and Robinson (1946) describes a case of this type where the rudimentary horn did not rupture until the 38th week when a live child was obtained. Rupture usually occurs between the fourth and fifth months Munro Kerr (1949) and as Jarcho points out, in this respect closely resembles interstitial pregnancy. The clinical features also closely resemble those found in extra-uterine pregnancy, recurrent attacks of abdominal pain, tenderness, faintness, haemorrhage and vaginal discharge and occasionally the decidua of the non-gravid horn may be passed in the form of a cast. Less commonly rupture and collapse occur without any premonitory symptoms but where the pregnancy is of a few months and has advanced beyond the first two months, a history of recurrent abdominal pain and slight vaginal discharge is nearly always found.

If term is reached 'spurious labour' occurs and the foetus dies and may become mummified or undergo adipocerous change or very rarely calcification to form a lithopaedion. Should the foetus and its sac become infected from the adjoining bowel a local peritonitis sets in with abscess formation and slow disintegration of the foetus which may be discharged piece meal through a fistulous opening with the bowel, bladder or anterior abdominal wall. Today the condition is usually diagnosed before pregnancy reaches the later months Munro Kerr (1949) who refers to the paper by Mahfouz (1932) in which several examples with illustrations are recorded. Although rupture of the gravid horn is the usual termination, less commonly missed abortion occurs from repeated small haemorrhages which cause detachment of the placenta with death of the foetus and this was the termination in one of the cases in the present series (G 5) and a similar happening is reported by Liu (1949) in his case to be followed by normal pregnancy in the other horn. A rare complication is torsion of the gravid horn.

Pregnancy in the normal horn may terminate without any abnormal features and the rudimentary horn remain unrecognised. But in the cases which do come under ones notice, the rudimentary horn has become displaced into the Pouch of Douglas causing obstruction to the passage of the child, more especially if there exists a haematometra in it, Munro Kerr (1949).

Bannister (1937) recorded a case in which there was coexistent pregnancies - one in the rudimentary horn and the other in the normally developed half. The former was removed and the latter proceeded normally. True tubal pregnancy in the underdeveloped Fallopian tube of a rudimentary horn is reported by Taber (1950) and the role of the rudimentary horn as a cause of abdominal pregnancy together with a case reported are discussed by Simpson (1951).

Diagnosis is seldom made prior to laparotomy and the condition is commonly mistaken for some form of ectopic pregnancy. The gravid horn can usually be felt apart from the uterus in the early months of pregnancy but later the non-gravid horn may be taken for a fibroid or ovarian cyst and the key to the diagnosis is the position of the round ligaments - Munro Kerr (1949), for both swellings will lie medial to these structures but prior to laparotomy it is unusual to appreciate the exact position of these structures. The same author emphasizes the great danger of mistaking the gravid uterine horn for a fibromyoma complicating intra-uterine pregnancy for, should the pregnancy be allowed to continue in the hope that the myoma will be lifted out of the pelvis, the patient's life may be endangered by rupture of the gravid horn. Hysterosalpingography and pneumoperitoneum may be employed as diagnostic aids and occasionally straight and oblique views of the abdomen will reveal the outline of the amniotic sac Snow (1952).

Uterus Unicornis

The number of cases of this rare anomaly so far reported is insufficient to base an opinion as to its effect in pregnancy. The advent of pregnancy would not appear to be uncommon for Ogilive (1957) in a review of the literature, calculated that at least one in three became pregnant. Similarly Schumaker (1938) although finding a number of miscarriages reported in association with this anomaly considered the number too small on which to base an opinion. Although Caesarean section was frequently the method of delivery where pregnancy had reached viability, the indication was not the anomaly per se but some associated condition such as the presence of a pelvic deformity, Tucker and Baker (1953), and the presence of an artificial vagina, Whittemore (1942). Cabanes and Jahier (1938) reported a case with antepartum haemorrhage in three of the five pregnancies and postpartum haemorrhage and manual removal of the placenta in two. All five infants were live born but one died a few hours after birth. Three consecutive breech deliveries occurred in Birk's (1931) patient.

In the present study there were two examples of uterus unicornis, (SG 1 and QC 18) both primigravidae. The first was delivered of a mature anencephalic infant as a breech and in the second, pregnancy was terminated by Caesarean section because of severe pre-eclamptic toxaemia, with the delivery of premature twins which survived.

The principal importance of this anomaly is the frequent association of contra-ipsilateral renal agenesis - that is on the same side as the suppressed Mullerian duct, Ogilvie (1957) putting this occurrence as high as 70%. The single kidney may be ectopic in position and if in the pelvis may be a cause of dystocia and itself be damaged from trauma as reported by Varino (1942).

Reference has already been made to Guatierez's (1933) statement that the single organ is nearly always potentially affected with some chronic disease and to other associated malformations (see associated malformations).

Investigation of the renal tract in the two cases presented in this study showed one to be normal (SG 1) and the other to have ipsilateral renal agenesis (QC 18).

GESTATIONAL PHYSIOLOGY AND PATHOLOGY

Falls assumes the myometrium in the congenitally malformed uterus to be defective on account of the embryological maldevelopment and considers this is incompatible with normal blood circulation and innervation. Under these circumstances a normal decidual reaction is unlikely which in turn leads to defective nidation and placenta formation and consequently affects the normal development of the embryo. Thus a combination of factors are produced which will promote foetal death and or expulsion of the foetus. Falls agrees that while his assumptions are difficult to prove they are equally difficult to disprove. A similar view is held by Hunter 1957 - who states that diminished vascularity of the myometrium leads to placental insufficiency, foetal anoxia and then foetal death before or during labour. He considers these risks are increased if the placental site is on the centre of the fundus in the arcuate uterus or on the medial wall of bicornuate uterus, for in these areas the myometrium tends to be especially thin (Madonald, 1949). The work of Ritschel (1949) on the histology of the uterine septum supports the view that the myometrium is incapable of a normal decidual reaction. Histological examination shows high grade muscle hypoplasia similar to the connective tissue. The surface has no normal endometrium and is separated from the uterine cavity by a narrow epithelial isthmus which has no apparent function.

The vascular supply decreases towards the periphery of the septum. Both Falls and Hunter found the placentae in a large proportion of cases to be abnormally thin and large in area, and Falls states that not infrequently the placenta is heart-shaped. In the present investigation the placentae although occasionally bipartite or heart-shaped were otherwise normal.

Abortion and Premature Labour.

Falls (1956) claims that the myometrium in the malformed uterus is hyper-irritable and in support of this states that Braxton Hicks Contractions are more powerful and more frequent and considers this to be the chief factor in the increased rate of abortion and prematurity, in addition to which are the deformed uterine cavity, defective nidation, and increased incidence foetal abnormalities. The work of Rudolph (1940) in which certain uterine dysfunctions are explained on an embryological basis, would appear to give support to Fall's claim of a hyper-irritable myometrium. Rudolph claimed that even the anatomically normal uterus, may exhibit different physiological properties in its 'two halves' because of its bilateral origin.

Intra-Uterine Asphyxia

That the foetal heart sounds may display a peculiar irregularity, inexplicable on any discernible change in local conditions, has been noted by Falls (1951) and Hunter (1950) and is considered as arising from faulty placentation and possibly from an abnormal uterine circulation and innervation (Falls), - which will tend to keep the foetus on the verge of hypoxia, - it may become so acute that intra-uterine death results, and for which no cause is found.

Placenta Praevia

It has been suggested that a poor decidua may well delay implantation which in consequence will take place nearer to the internal os.

Accidental Haemorrhage

This also can be explained on the assumption of a deficient decidual reaction and poor placentation.

Pre-eclamptic Toxaemia

Ernst (1941), Jarcho (1946) and most of the earlier writers considers the toxaemias of pregnancy to be commonly associated with the congenitally malformed uterus, but this theory has not been substantiated by any recent investigator apart from Falls (1956) and Jones (1951).

Although Falls found the incidence of toxaemia to be impressive, no figures are given but he is of the opinion that premature separation of the placenta and toxaemia are part of the same process, and that while the former is associated with haemorrhage into the decidua spongiosa and into the decidua compacta in the latter, both in some way arise from a combination of a primary circulatory defect and defective myometrium.

Uterine Inertia

Primarily the myometrium is poorly developed favouring uterine inertia, the increased frequency malpresentation, premature rupture membranes and poor application of the presenting part to the lower uterine segment will all tend to increase the chances of a slow labour. In the double uterus the lateral flexion of the pregnant horn and in the lesser bicornuate forms, the septum will increase the chances of incoordinate uterine action.

Multiple Pregnancy and Superfoetation.

Berkeley, Bonney and MacLeod (1938) give the incidence of twins as 1:12 pregnancies compared with 1:89 for the normal uterus. Recent studies have not however been able to confirm these findings, there being no examples in Hunter's series (1950-57) while Jones (1957) found the incidence in the anomalous and the normal organ to be the same.

In the present study there were three sets of twins, 2 in Type 111 and 1 in Type V11 - an incidence of 1:50 pregnancies, and although this is considerably higher than normal it would require to be based on a much larger series to be statistically of value. Both foetuses may be in the one horn or one in each and Brown (1956) considers the radiological appearance of both vertices at the same level with widely divergent trunks suggestive of the latter condition. He adds that where both foetuses are in the one horn, it inclines towards one flank but later in pregnancy because of the bulk it fills both flanks.

Superfoetation.

This interesting phenomenon was studies by Bainbridge(1934) who collected several examples of which the following are particularly well documented.

(1) Delivery at term followed three and a half months later by delivery of a second mature foetus.

(2) Delivery from alternate horns of mature infants at an interval of four months.

(3) Delivery of a mature white infant from the left horn followed two months later by delivery of a black infant from the right horn.

It is difficult to prove that these are true examples of superfoetation but the third case must count at least as superfecundation. Apart from their interest as medical curiosities they present fundamental material for the investigation of various physiological aspects of pregnancy and labour. Of particular interest in this respect is the case reported by Colaco (1949) in which there was a twenty-one day interval between deliveries from the two horns of a uterus pseudo-didelphys. Following attempted external version the patient commenced in labour and delivered from one horn, during which time the other horn remained quiescent apart from a few feeble contractions. This according to Colaco would suggest that the onset of labour is chiefly dependent upon some intrinsic and not purely endocrine factor. There was neither lochia nor lactation until the second delivery was completed,

phenomena previously noted by Paulin (1905) and quoted by Munro Kerr. As a similar inhibition occurs if the placenta is left in situ after removing the foetus of an abdominal pregnancy it is probably the presence of the placenta which gives rise to the phenomena already noted. The following case reports would appear to support Colaco's contention that the onset of labour in the two horns are independent and depend upon some intrinsic rather than central mechanism:

(1) Abortion at the fifth month from the left horn of a 'double' uterus, the pregnancy in the right continuing to term when delivery was effected by Caesarean section because of uterine inertia and early rupture of the membranes, Karczma (1954).

(2) the shedding of a complete decidual cast from the non-gravid horn ten days prior to the onset of labour, Corbet (1945).

(3) an additional report of the deliveries of twins from alternate horns of a 'double' uterus with an interval of days between the two labours, Brody (1954).
There is not however always a long interval between the two deliveries for in the example reported by Brown (1956), five minutes after the first infant was delivered the cervix of the second horn was found to be fully dilated.

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COMPLICATIONS OF PREGNANCY AND LABOUR

TERMINATION OF PREGNANCY PRIOR TO VIABILITY

The chances of pregnancy reaching viability are much less in the congenitally malformed uterus than in the normal organ, and in the great majority of studies, termination of pregnancy prior to the 28th week, rates high as a complication of pregnancy.

Taylor (1943) from his personal records reported the incidence of abortion as 45 per cent and collected the figures given below from several other sources for comparison:

Author	Incidence per cent	Type of Anomaly
Findley (1926)	39.6%	chiefly uterus bicornis bicollis
Miller (1922)	28.3%	
Schauffler(1941)	53%	study confined to lesser forms of bicornuate and arcuate anomalies.

Jarcho (1946) found that 44% of all pregnancies in his study terminated prior to the 28th week and this was not confined to any particular form of anomaly. More recent reports include those of Philpott (1954) with 28.6%, Karczma (1954) with 51%, and Lash and Lash (1955) 24%.

The variation in the estimated incidence will in part be explained by the relative proportion of the different types of anomaly in each series, and in part to the lack of unanimity in classification.

In the present study 62 of the total 150 pregnancies were lost prior to viability, an incidence of 41.3%. Seven of these were not attributable to the anomaly per se but to the high operative interference rate associated with these malformations (this is described more fully under Statistical Analysis) and if allowance is made for these the incidence falls to 36%.

Aetiology

The cause of the high rate of abortion is unknown but there are several features in the congenitally malformed uterus which detract from its value as a gestational organ. Karczma (1954) considers the uterine musculature deficient and incapable of sufficient hypertrophy to accommodate the growing ovum and agrees with Falls (1954) that abortion usually occurs later (16th to 20th week) than with the normal uterus. Although agreeing that the poorly developed uterine musculature is primarily at fault, both Way (1945) and Falls (1954) contend that this leads to a deficient decidual reaction with consequent faulty implantation of the trophoblast.

Hyperirritability of the uterine muscle, as shown by frequent and powerful Braxton Hick's contractions, is considered to be a further factor by Falls.

(2) Mechanical interference with the growing ovum by the deformed uterine cavity, especially in the arcuate and subseptate forms of uterus with which the highest abortion rate is usually associated, is generally accepted as a major factor. Certainly resection of the septum is not uncommonly followed by successful pregnancy but it can be equally well be argued that the high abortion rate associated with this form of uterus arises from implantation on the septum.

(3) Congenital foetal abnormalities are more frequently reported than with the normal organ and it would appear reasonable to assume that there are an increased number of abortions from defective ova.

(4) A further factor in the high foetal loss prior to viability is the increased rate of operative interference found in association with the anomalous uterus.

Hunter (1957) found abortion most commonly associated with the major forms of uterine deformity, especially where the fundal recess of the pregnant horn was constricted as in the bicornuate uterus whether or not the cavity is divided as far as the internal cervical os; it is least where the conformation of the cavity approaches normal.

Pregnancy may not be equally stable in both horns and in illustration of this Bainbridge (1924) quotes Goutermann whose patient had three normal deliveries at term from the right horn and aborted four pregnancies from the left. In these cases both horns were equally well developed. In Hunter's series(1957) the abortion rate for the 'double' organ was 21%, compared with 16% for a control group with normal uteri, and of those women with a double uterus 39% had had at least one abortion compared with 29% in the control group. In the arcuate and subseptate anomalies Way (1947) gives the incidence as 26%.

Incidence Relative to Type of Anomaly

The risk of abortion varies in relation to the type of anomaly and would appear to be least where the uterine cavity approximates the normal and highest in the arcuate and subseptate forms.

Baker (1953), Fenton and Singh (1952) and Way (1945) agree with Hunter that the risk of abortion is greatest when the ovum implants itself on the septum. A comparison of the incidence relative to the form of anomaly in Baker's and the present series is given below:

Type of Uterine Anomaly	Baker's Series	Present Investigation	
		Gross	Corrected
1	8.2	28	25
111	27.1	43	39
V	-	55	54
Others	21.1	-	-

It should be noted that Baker uses only three divisions, the bicornuate, arcuate and subseptate forms being included in Group 111.

Fenton and Singh (1952) state that the abortion rate is three times that for the normal uterus and in their investigation was 16.56% compared with the normal 5.6%. The results of this and Baker's analyses closely correspond except for the abortion rate which in the latter was only 7.1% (corrected), thus closely approximating the figure for the normal uterus but this is exceptional and difficult to explain.

A more recent investigation by Jones (1957) gives the findings which correspond more closely with those of my own investigation.

Uterus bicornis bicollis	32.4%
Bicornis Unicollis (includes arcuate and subseptate forms)	33.8%
Simple septate uterus (septum from fundus to internal os)	22.2%

Again the difference in terminology does not allow for an exact comparison of the two series.

Habitual Abortion

Defining habitual abortion as the occurrence of two or more spontaneous abortions without obvious pathogenesis, Halbrecht (1951) goes on to add that the congenitally malformed uterus is more often the cause than was formerly recognised. This opinion is based on the results of an investigation by hysterography of 56 cases of habitual abortion in which 22 or 40% showed some developmental defect of the uterus, predominantly the bicornuate, subseptate and arcuate forms. Way (1945) in an investigation of 12 cases of recurrent abortion discovered 2 examples of subseptate uterus. Halbrecht (1951) does not deny that pregnancy and labour may be perfectly normal with the malformed uterus and considers the outcome of these pregnancies depends chiefly on accident. In abortion at the 4th - 6th or premature labour at the 7th - 8th month the principal role is the reduced capacity of the uterine cavity to accommodate the growing ovum. The placental site is of less importance in these cases than in early abortion in which the anatomical and histological and in particular the deficient vascularization are the decisive factors.

Palmer (1953) agrees that the congenitally anomalous uterus to be an important factor in habitual abortion and was

present in 25% of the women he investigated for habitual abortion and because of this urges proper investigation of recurrent abortion as the uterine anomaly can at least in part be corrected by surgery.

TERMINATION OF PREGNANCY PRIOR TO VIABILITY

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PAINFUL SACCULATION OF THE PREGNANT HORN IN THE ARCUATE AND MINOR FORMS OF BICORNUATE UTERUS

Clinical Significance of Sacculation of the Pregnant Horn Of the Arcuate and Minor Forms of Bicornuate Uterus.

The anatomical and physiological changes in the arcuate and lesser degrees of bicornuate uterus consequent upon pregnancy have already been considered under Gestational Physiology; and it will suffice to reiterate the cardinal points. The gravid horn or side, enlarges and softens more rapidly than the non-gravid half and in consequence the uterus assumes an asymmetrical outline. The poorly developed musculature of the pregnant horn becomes over-stretched causing pain and tenderness over the site of implantation in addition to which there is a tendency to abort from the hyperirritable uterine muscle, Falls (1954).

The clinical importance of the condition is the facility with which it simulates other more serious conditions and for which laparotomy may be required before the correct diagnosis is reached. The following case recorded by Way (1945) illustrates the principal features of the complication and here the patient was admitted to hospital at the eighth week of her third pregnancy complaining of severe lower abdominal pain and slight vaginal bleeding. On pelvic examination a soft very tender mass could be felt to the left of the uterus but not apart from it and this in view of the history was considered sufficient evidence for the provisional diagnosis of ectopic pregnancy. At laparotomy both Fallopian tubes

were noted to be normal and the uterus to have an elongated swelling in the region of the left cornu. The abdomen was closed and the pregnancy allowed to proceed normally. The symptoms subsided rapidly, and although the asymmetrical outline of the uterus was still present at the subsequent antenatal visits the pain and tenderness had entirely disappeared two weeks post-operatively.

As in this case the softened and tender pregnant horn is usually mistaken for a tubal pregnancy or when more advanced for an interstitial, angular or rudimentary horn pregnancy. The firmer non-gravid half may be taken for a fibroid or more rarely an ovarian cyst. Ectopic pregnancy is the commonest misdiagnosis but where the pain is felt in the right iliac fossa, a provisional diagnosis of appendicitis is not uncommon.

Although there is little to be found in the literature regarding this condition, it would not appear to be so very uncommon if only to judge from the number of patients with a uterine anomaly who are subjected to laparotomy in early pregnancy. In the present study there were five examples of this complication and, in four the correct diagnosis was only made at laparotomy, the provisional diagnosis having been ectopic pregnancy in four including one treated conservatively, and appendicitis in one other. Rubovits (1951) writing on the subject of sacculation of the pregnant uterus described three types:-

(1) A transitory asymmetry of the uterus often designated in American literature as Piskacek's sign, and usually most marked at the 16th to 24th week of pregnancy after which time it tends to resume its normal shape. Rudolph (1940) considered this form of sacculation to be physiological and points out that it disappears after escape of the amniotic fluid. To this group Rubovits assigns angular pregnancy.

(2) Diverticula or sacculation in the midline or cornu of the uterus, the former arising in the theoretically weakened uterine wall at the point of fusion of the Mullerian ducts, and the latter from excessive penetration of the chorionic villi which leads to herniation of the weakened musculature.

(3) Sacculation of the pregnant horn of the bicornuate and arcuate uterus.

Clinically, the sacculation produced by the growing ovum in the gravid half of the bicornuate or arcuate uterus closely resembles angular pregnancy which was first described in this country by Munro Kerr in 1907 and of which the characteristic features are(I) pain, (2) lateral distension of the uterus in the cornual region, and (3) a tendency to abort.

Although the two conditions closely resemble one another, they are entirely separate clinical entities for angular pregnancy occurs in the normally developed uterus, implantation occurring over or just external to the opening of the interstitial portion of the tube and the ovum develops towards the uterine cavity. The pain and tenderness of the sacculation may in both conditions be very severe but symptoms in the angular pregnancy must commonly attract attention between the 12th - 20th week Munro Kerr (1949) while in sacculation of the malformed uterus this would appear to be earlier, from the 6th - 12th weeks. In both conditions where abortion does not occur, the symptoms tend to regress spontaneously, by the end of the second trimester in the former and earlier in the latter although here a fundal depression is usually palpable throughout pregnancy.

Undoubtedly, the close resemblance between the two conditions has led to confusion and difficulty in differentiating between them and the recent work of Granjon and Beau (1950) has attempted to show that angular pregnancy as a clinical entity does not exist. These authors investigated six cases in which the diagnosis of angular pregnancy had already been established and by hysterosalpingography were able in each to show evidence of a congenital uterine malformation. They concluded that if a more careful examination was made, cases diagnosed as angular pregnancy would be found to be pregnancy in the congenitally malformed uterus.

Fitzgerald (1952) while agreeing that many cases labelled as angular pregnancy are in reality examples of pregnancy in the deformed uterus, is of the opinion that angular pregnancy as a clinical entity does exist. He points out that mistakes in diagnosis often arise through making a diagnosis on the shape of the uterus alone. He adds that examination of the uterus after expulsion of its contents will very often reveal the presence of a hitherto unsuspected malformation and in support of this he quotes several personal case records. Finally he makes a plea for hysterosalpingography to be performed in all cases so that the diagnosis may be confirmed.

A personal case record McNeil (1957) would appear to substantiate Fitzgerald's claim that angular pregnancy is a clinical entity; this showing all the features of an angular pregnancy yet in no way appeared to differ from the case (SM 17) recorded in this thesis of sacculation of the gravid horn in a bicornuate uterus. Laparotomy was performed in both because of uncertainty of the diagnosis and in each the swelling lay medial to the round ligament but in the former an hysterosalpingogram performed at a later date showed a normal uterine cavity.

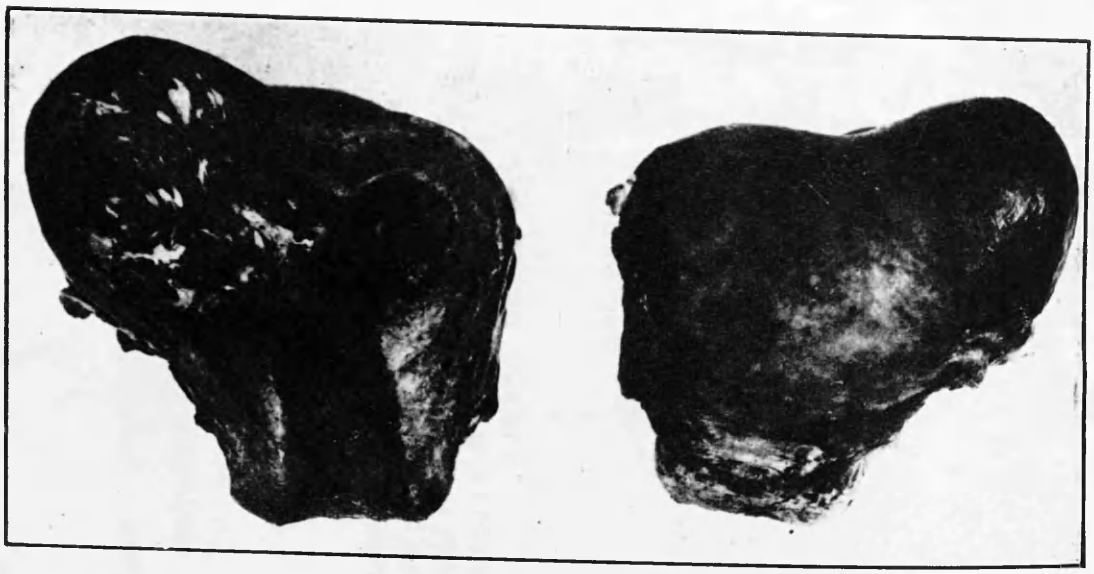

FIG. 336.—ANGULAR PREGNANCY.

Uterus removed by sub-total hysterectomy after spontaneous delivery of a living child. The placenta (*not* accreta) is incarcerated in the dilated left cornu of the uterus, and attempts at manual removal failed. The pregnancy was complicated by slight hæmorrhages, but the uterus was not tender, nor was there any pain (Gibberd).

21. Angular Pregnancy or Pregnancy in the Congenitally Malformed Uterus?.

Munro Kerr and Chassar Moir - "Operative Ostetrics" (1949)

It is possible to discern a fundal notch and from it a short fleshy septum extending into the uterine cavity suggestive of an arcuate or subseptate uterus.

It is suggested but difficult to prove because of the lack of radiological evidence, that many examples of pregnancy in the anomalous uterus are reported as the angular type and from an examination of museum specimens this would appear to be particularly true of those cases supposed to cause retention of the placenta and in which great difficulty is experienced in manual removal. One such case reported by Gibberd of which an illustration appears in Munro Kerr's and Chassar Moir's 'Operative Obstetrics' (1949), is now in the Gordon Museum of Guy's Hospital strongly suggests that this is an arcuate or minor degree of bicornuate uterus for the fundus shows a definite midline depression and the uterus a short wedge shaped septum separating the two horns. It would appear possible that the changes consistent with pregnancy in the anomalous uterus are a not infrequent cause of pain in early pregnancy and certainly a careful examination should be made in an attempt to exclude this possibility in any woman complaining of persistent or severe lower abdominal pain in early pregnancy.

The treatment of interstitial pregnancy is laparotomy with resection of the pregnant horn, but both angular pregnancy and sacculation of the gravid horn of the malformed uterus can usually be treated conservatively but there is no way at present

in which these can be definitely differentiated apart from laparotomy. Such conditions as tubal pregnancy, ovarian cyst or intrauterine pregnancy complicated by appendicitis can be excluded by proving the uterus asymmetrical in outline and usually this can be decided on careful bimanual examination if necessary under an anaesthetic. The abnormal shape of the uterus is more apparent during a contraction which can be produced by gentle massage of the uterus for although Hyams (1953) recommends the administration of an oxytocic drug in sufficient quantity to make the uterus contract it would not appear to be without risk. The position of the round ligament in relation to the swelling has not proved a help in diagnosis and where doubt of the diagnosis remains especially where the symptoms increase the only way of settling this is by laparotomy.

Foetal Malpresentation

With an incidence of 38 per cent, foetal malpresentation was the commonest complication of pregnancy in the present investigation and this is in line with the results of other investigations where malpresentation is found to be one of the commonest if not the commonest complication of pregnancy. Although only a 26 per cent incidence it was the commonest complication in Baker's (1953) series, Karczma (1953) 55.8%, and Fenton and Singh (1952) 28.7% and these figures are based on the results for 11 types uterine anomaly. The incidence of malpresentation for a particular form of anomaly have been published by Findley (1926) on the uterus pseudo-didelphys, Way (1945 and 1947) less marked degrees uterine anomaly, Falls (1939) arcuate uterus and Hunter (1950 and 1957) the double uterus - and their findings are tabulated below:-

Type of Anomaly	Malpresentation		Incidence	Author
	Breech	Oblique and Transverse		
Pseudodidelphys (Bicornis bicollis)	25%			Findley
-"-	25%			Hunter
Lesser degrees Bicornuate and Arcuate	11%			Falls
	9%	27%		Way

Aetiology

Several factors tend to favour foetal malpresentation of these the most important is mechanical, and differs according to the type anomaly with its distorted anatomy. In the uterus pseudodidelphys and the more severe forms of bicornuate uterus the pregnant horn is often fusiform and the lower segment funnel shaped small and indistensible Hunter (1950 and 1957). This will tend to interfere with descent of the presenting part in late pregnancy and with its ability to accommodate the vertex and hence will favour breech presentation. In general the uterus bicornis bicollis favours breech presentation and the lesser degrees bicornuate arcuate and septate forms an oblique or transverse lie, Hunter (1950), Way (1947) Falls (1955), but where the non-gravid horn is prolapsed in front of the presenting part, the foetus may be forced to take up an oblique lie Munro Kerr (1949). Rarely extreme lateral flexion of the pregnant horn may be the cause of oblique lie, Hunter (1957).

The presence of the bulky intra-uterine septum as in the arcuate and subseptate uteri bulging into the uterine cavity, will force the foetus to take up an oblique position Way (1945) but where the foetus is small, the soft breech may be forced into the pelvis when labour commences, with resultant breech delivery.

Where there is a complete septum dividing the uterus into separate cavities, hypertrophy of the non-pregnant half may bulge cause bulging of the septum with consequent oblique lie of the foetus, Luikart (1936) Falls (1956).

Rarely where there is a partial or fenestrated septum the foetus may become engaged around it, resulting in a persistent transverse lie, Titus (1950), Hunter (1957) states that one in every nine transverse lie is found in association with a deformity of the uterine cavity, and that the foetal back more often overlies the pelvis than across the fundus. Falls (1956) found an increased incidence of face and brow presentation, although he agrees the evidence for this is less well documented and for a further condition of opisthotonos or foetal hyperextension which he first described in 1916. This latter condition is usually associated with a bipartite placenta and it is not usually possible to correct the malpresentation antenatally.

The increased incidence of premature labour, multiple pregnancy, placenta praevia and foetal abnormalities are additional factors which will tend to favour foetal malposition.

The correction of breech and oblique presentations to a vertex is less likely to be successful than in the normal uterus and Falls (1956) considers this should arouse suspicion

of a congenital uterine malformation and attributes this to the hyper-irritability of the uterine musculature and persistent or recurrent malpresentation is usually associated with the arcuate and subseptate uteri. Hunter (1950) and Karczma (1954) while agreeing that version is frequently difficult or impossible, consider this to be caused by the reduced uterine volume and point out that this will also attribute to the increased incidence of malpresentation. A further contributory factor mentioned by Way (1947) is the increased rate of fundal implantation. He goes on to say that persistence of an oblique lie does not call for elective Caesarean section, as spontaneous correction frequently occurs early in labour.

Incidence Malpresentation in Relation to Type Anomaly

Total 41.4% all deliveries

There were 33 breech and three transverse presentations or 38% of all deliveries and 3.4%.

Breech	33	14	13	5	1
Transverse	3	1		2	
	36	15	13	7	1

Incidence Malformation Related to Type Anomaly

I	III	V
42%	37%	29%
3%		12%
45%	37%	41%

Prematurity

The proportion of women who go to term is considerably less than those with a normal uterus, and this in part is accounted for by the higher incidence of malpresentation, multiple pregnancy, placenta praevia, accidental haemorrhage, foetal anomalies and operative interference. In addition, there appears to be an intrinsic tendency to premature labour which Falls (1956) considers arises from a hyper-irritability of the myometrium and Karczma (1954) to the reduced gestational capacity of the uterine cavity.

Baker (1953) found the viability rate to be only 77.2% where the incidence for the normal uterus was 93.1%, (the standard of viability being 1,500 Gm.) and Fenton and Singh (1952) in their series using the same standard give the rate as 66.4%. The term birth rate in both series being 57.5 and 56.1 respectively where the figure for the normal uterus was 85.0, making premature labour the commonest complication of labour in both series. Miller (1923) from a study of the uterus pseudodidelphys found only 42% of his cases had a spontaneous delivery at term and that premature labour is most often associated with the most marked forms of uterine anomaly and this is confirmed by Lash and Lash (1955) and Jones (1957) but in the present study prematurity was found to be commonest in the arcuate and subseptate uteri.

It should be noted that Type V in the studies of Lash and Lash and Jones differs from the first study in representing the completely septate organ and not the arcuate and subseptate forms which it indicates in the Jarcho terminology and this will in part account for the difference. The incidence of prematurity in Baker's study relative to the form of uterus, statistically showed no difference.

Way (1945) who confined his investigation to the subseptate and lesser degrees of bicornuate uterus reported a lower incidence of prematurity, namely 9%.

Falls (1954) who has written extensively on the arcuate uterus states that while premature labour is common, not infrequently this form of uterus is associated with postmaturity and although of no statistical significance, the one example of postmaturity in this study, was (G 3) associated with this form of uterus. Falls explains this paradox on the grounds of the deficient development of the uterine musculature and to the frequency with which the presenting part remains high even at term; this however is also put forward as the reason for prematurity in these uteri.

FOETAL MALPRESENTATION

With an incidence of 41.4%, foetal malpresentation was the commonest complication of pregnancy in the present study and this is in line with the results of other investigations where malpresentation is found to be one of the commonest if not the commonest complication of pregnancy. Although the incidence was only 26% it was the commonest complication in Baker's (1953) series, 55.8% is reported by Karczma and Fenton and Singh (1952) 28.7% and these figures are based on the results for all types of uterine anomaly. The incidence of malpresentation for any particular form of anomaly have been reported by Findley (1926) or the uterus pseudo-didelphys, Way (1945 and 1947) less marked degrees of uterine anomaly, Falls (1939) arcuate uterus and Hunter (1950 and 1957) the double uterus - and their findings are tabulated below:-

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The presentation was abnormal in 41.4% of all deliveries of which 38% (33) were breech and 3.4% (3) oblique lie.

Presentation Related to Type of Uterus:

Presentation	Number	Type of Uterus			
		1	111	V	V11
Breech	33	14	13	5	1
Oblique Lie	3	1	-	2	-
Total:	36	15	13	7	1

Expressed as a percentage:

	Type of Uterus		
	1	111	V
Breech	42%	37%	29%
Oblique Lie	3%	0	12%
Total:	45%	37%	41%

It will be noted from these tables that the incidence of malpresentation although highest with the Type 1 uterus, shows little variation for all three types of anomaly. It does show however, that the most marked form of anomaly (Type 1) favours breech presentation to oblique lie in a ratio of 14:1 but with the less marked forms of anomaly this ratio decreases to 2:1.

Dystocia and Obstructed Labour from the Non-Pregnant Horn.

This complication is most likely to occur with the most marked forms of anomaly where the point of union of the two horns is at or below the level of the internal os, the non-gravid horn undergoing axial rotation posteriorly into the pouch of Douglas as the pregnant horn rises out of the pelvis into the abdomen, Hunter (1950). As a cause of dystocia, Taylor (1943) in a review of the literature found an incidence of 4.5% in the 271 cases he was able to collect. Baker (1953) in his comprehensive report on pregnancy in association with congenital malformations of the uterus found this to be the second commonest complication of labour, yet in the present investigation there is no record of this complication and examination of the records of Queen Charlotte's Hospital going back to 1914 this complication is noted as having occurred seven times including twice in one patient in succeeding pregnancies. However it is more from an anticipation of dystocia than its occurrence which prompted delivery by Caesarean section often before the onset of labour. In more than half of the cases the prolapsed horn was the site of fibromyomata which suggests that they may have a bearing on the aetiology of the condition. Dystocia from this cause, Falls (1956) states, was commonly reported by the older obstetricians but he, personally has not been impressed by this as a complication of congenital uterine malformations and his records show that where Caesarean section was necessary it was not because of this complication. There were no instances of this complication in the series reported by Jones (1957), who agrees with Falls that as a cause of dystocia, the non-gravid horn is chiefly a theoretical danger.

Where the non-gravid horn does become incarcerated in the Pouch of Douglas below the presenting part, it will if unrelieved cause obstructed labour and rupture of the uterus, this having been reported by Hall (1933). According to Berkeley, Bonney and Macleod (1938) the treatment of choice in this complication is Caesarean section, for although the non-pregnant horn may occasionally be pushed up from below, it is never pulled up spontaneously in labour.

Acosta-Sison(1952) states that the 'double uterus' may be a cause of dystocia or even rupture under the following circumstances: (1) pregnancy in a rudimentary horn, (2) where the non-gravid horn is below the presenting part it will obstruct the passage of the child at delivery, and (3) where the pregnant horn is connected at right angles to its fellow. A further factor increasing the risk of uterine rupture is the weakened musculature of the medial walls, Puddicombe (1929). Yet in spite of all the theoretical risks of uterine rupture, Milne (1946) in a review of the relevant literature found it to be a rare occurrence.

Uterine Inertia in First and Second Stages of Labour

There are many factors which should make uterine inertia a common complication of these anomalies and of these the more common are the maldeveloped uterine musculature, misdirected uterine forces, disturbed polarity from the attachment of the non-gravid horn, and high incidence of malpresentations. Yet in the present study this complication occurred only twice on the usual standard of a labour lasting more than 48 hours.

COMPLICATIONS OF THE THIRD STAGE OF LABOUR

Third Stage of Labour

A major disadvantage of the congenitally malformed uterus is the associated high rate of complications in the third stage of labour. In the series published by Baker (1953) it was the third commonest complication of labour with a 5.5% incidence of postpartum haemorrhage and 0.9% postpartum atony compared with 0.4% and 2.9% respectively for the normal organ. These figures are unusual for it is postpartum atony or more properly retention of the placenta which is the commonest complication and more representative are the figures of Fenton and Singh (1952) with 9% and 5.2% respectively compared with the same standards used in Baker's series. Karczma (1954) reported the very high figure of 34.6% for the combined retained placenta and postpartum rate but this in part is explained by the authors advocation of manual exploration of the uterus in the third stage.

Aetiology.

There are several factors which tend to interfere with the normal third stage of labour and these may be divided into extrinsic and intrinsic, the former being the higher rate of uterine inertia, placenta praevia, accidental haemorrhage, and operative interference associated with the congenitally malformed organ and the latter the deformed uterine cavity and maldeveloped uterine muscle interfering with normal retraction and abnormal

implantation from deficient decidual reaction causing imperfect placental separation. Hunter (1950) reporting the incidence of retention of the placenta as 11%, from a study confined to the double organ, suggests the non-pregnant horn may interfere with the normal third stage in the same way as a full bladder. Way (1947) in a small series devoted to the minor degrees of bicornuate and arcuate uterus gives the incidence of retained placenta as 15% and considers this may arise from the high rate of cornual insertion of the placenta which even in the normal organ favours retention of the placenta. He adds that if the placenta is partially inserted into the septum it nearly always fails to separate and Hunter warns of the difficulty which may be experienced in controlling haemorrhage from the torn sinuses in this area ~~and~~ because of the inefficient myometrium. Falls (1954) considers that the placenta after separation may be retained in the opposite horn and by interfering with normal retraction be a cause of severe postpartum haemorrhage.

It is generally agreed that abnormalities of the third stage are commonest with the minor degrees of uterine deformity and this Hunter (1950) ascribes to inefficient retraction caused by the mechanical interference of the non-pregnant horn with its extreme attachment to the pregnant uterus. Halbrecht (1951)

on the other hand is of the opinion that the placenta is retained partially or completely by constriction of the circular muscle fibres of the uterine horn. Williams(1952) considers cornual implantation secondary to a congenital uterine malformation, an important cause of atonic non-detachment of the placenta. The uterine deformity is usually of minor degree with cornual implantation of at least part of the placenta. He considers the cornual contraction ring described by Joyce and Lennon (1948) as a separate entity.

Complications of Third Stage of Labour Related to Form of Anomaly

In the present study there was some complication of the third stage in twelve pregnancies, the placenta having been removed manually in eight of these because of simple retention and in two, for postpartum haemorrhage and retained placenta. A further two cases of postpartum haemorrhage occurred after the placenta had been expelled. Therefore the incidence of retained placentae was 11.4% (10) and

of postpartum haemorrhage 4.5% (4)

When related to the type of uterus it will be noted that support is given to the contention that complications of the third stage occur most commonly with the less pronounced forms of uterine anomaly.

Type of Anomaly	1	111	V	Total
Retained placenta alone	1	0	7 *	8
Retained placenta and postpartum haemorrhage	0	1	1	2
Postpartum haemorrhage after delivery placenta	1	1	0	2
	2 +	2 +	8 =	12

* Two manual removals of the placentae in this group were performed routinely following internal podalic version; they have been included for in both cases the placentae were found to be morbidly adherent.

From the above table it will be seen that complications of the third stage of labour are twice as common with Type V as with the sum of Types 1 and 111 or that 66% of complications occurred with the group which includes the arcuate, subseptate and minor degrees of bicornuate uterus.

A comparable study in which the incidence of manual removal of the placenta is related to the form of uterus is that of Jones (1957).

Type of Anomaly	Simple Septate	Bicornuate	Double
Incidence 9.6% Manual Removal of Placenta	9.6%	9%	4%

The incidence of manual removal will vary according to the operative delivery rate and on the grounds to be fulfilled before this operation is undertaken. Philpott and Ross (1954) in their series removed the placenta only if there was a definite delay in placental separation and give the incidence as 20% for the anomalous organ and 3% in the normal uterus. (In the present study where there was simple delay the placenta was not removed until between $1\frac{1}{2}$ to 2 hours had elapsed and in the majority Crédé's expression was attempted once only.)

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DIAGNOSIS

Familiarity with the clinical manifestations to which the congenitally anomalous uterus may give rise is undoubtedly the key to their diagnosis, and if the condition is constantly kept in mind the number unsuspected will be greatly reduced.

In the present study there are several examples where the patient has passed through several pregnancies without the presence of a uterine malformation being suspected, and this is illustrated by case SM 5 where the patient had had three manual removal of the placentae without the condition having been suspected, yet the sequence of events is typical and it should be emphasized that in no condition is a careful history more important to making a diagnosis. The history will frequently be more revealing than physical examination and this is especially true where there have been previous pregnancies.

Arbitrarily the history may be divided into three groups, namely:

(1) Family,

(11)Gynaecological,

(111)Obstetrical.

(1) Reference has already been made under aetiology to the part played by heredity in the genesis of these malformations and although little appears to have been written on this aspect of the subject, there is sufficient evidence to show that uterine anomalies may be inherited and that several siblings may be affected. Way (1945 and 1947) in an investigation of 37 examples of congenital uterine anomaly was able to show a familial relationship in five, and these are summarised below:-

(a) Sisters with bicornuate uteri.

(b) Mother and daughter with bicornuate uteri.

(c) Mother with subseptate uterus who had two daughters in one of whom the uterus was bicornuate and normal in the other.

(d) Further example of mother and daughter with bicornuate uteri.

(e) A patient with a pregnancy in a bicornuate uterus whose sister had died in childbirth from shock following internal podalic version for severe antepartum haemorrhage with placenta praevia. Of considerable interest is the obstetric history of the sisters' mother who had had eleven pregnancies all of which were abnormal and included placenta praeviae on three occasions, post-partum haemorrhage in four and internal podalic version once. Unfortunately the mother refused to submit to a hysterosalpingogram and the daughter went to the U.S.A. during her pregnancy and no record of her delivery was obtainable.

In the present study there were two examples in which a familial relationship was demonstrated, in the first of which there were two sisters with bicornuate uteri (SM 6 and 7) and in the other (SM 12) the stillborn child of a woman with a bicornuate uterus, was found at postmortem examination to have a uterus bicornis bicollis and septate vagina.

The patient whose mother or sisters have a bad obstetric history may have a congenital anomaly of the uterus and therefore whenever such a history is obtained it should alert one to the possibility of a uterine anomaly.

(An attempt is now being made to trace the relatives of women with known uterine anomalies, so that the form of uterus may be determined. In this way it is hoped to obtain further evidence regarding the familial incidence of these anomalies).

(11) It has already been said that it is rare for symptoms to arise prior to pregnancy and although disorders of menstruation are considered commoner in the congenitally malformed organ in only one is the history so characteristic as to suggest the diagnosis namely the formation of a haematometra in a rudimentary horn. A history of increasingly severe unilateral dysmenorrhoea is typical of the condition. Delayed onset of puberty, considered as typical of the anomalous uterus by Jarcho (1946) was not confirmed in the present investigation.

The woman with a vaginal septum may complain of dyspareunia or rarely attempts at coitus or at the insertion of a tampon sometimes cause severe bleeding from the lower edge of the vaginal septum.

OBSTETRIC HISTORY

The information elicited from the obstetric history may be more helpful in alerting one to the possibility of a uterine malformation than the findings on physical examination. This is especially true of the minor degrees of anomaly which are rarely detected on pelvic examination but commonly give rise to complications in pregnancy often repeated in subsequent pregnancies. Late abortion premature labour and unexplained stillbirths especially if recurrent are common to all types of congenital uterine malformation, repeated breech presentation of the major forms while oblique and transverse lie and complications of the third stage of labour are typical of the minor anomalies. Bleeding in the early months of pregnancy from the non-gravid horn is considered an important diagnostic point by Philpott (1954) but in the present study this was not confirmed. Placenta praevia occurring in successive pregnancies, although rare was noted as early as 1891 by Schwartz, whose patient had placenta praevia in three successive pregnancies. This occurrence is also reported by Andrews (1941) and here also placenta praevia was found in three consecutive pregnancies and in the last of these pregnancy was terminated by Caesarean section when the uterus was found to be of the subseptate variety, but the author does not appear to have noted the significance of this.

The importance of the obstetric background is well demonstrated by Aaron's (1953) investigation of 46 women giving a history of unexplained stillbirth, premature labour or habitual abortion. Hysterography revealed some malformation of the uterus in 28 or 60% made up as follows:

Bicornuate uterus	15
Arcuate	7
Cordiform	3
Unicornuate	1
Abnormal length cervix.	2

Similarly Halbrecht (1951) in an investigation of primary infertility found the presence of a congenital malformation of the uterus in 18%.

Physical Examination

It is claimed by Schwarz (1918) and Schumann (1936) that these anomalies are frequently found in association with an usually broad pelvis and in women constitutionally of heavy build and phlegmatic temperament. Eck (1936) in an analysis of hospital patients, found they were commoner in those of Russian and Italian ancestry but in the present study the only phenotype which appeared significant as an association of these anomalies was hirsutism and minor degrees of female pseudohermaphroditism.

Concomitant Malformations

Congenital defects of the uterus are not infrequently associated with developmental defects in other systems especially the renal and lower genital tract and less commonly the gastro-intestinal and skeletal systems. Therefore the presence of a congenital defect should always arouse the suspicion of an associated anomaly of the uterus and necessitate a careful investigation of the genital tract. Developmental defects of the vulva, hypospadias, vestibular anus, recto- and vesico-vaginal fistulae, cysts of Gartner's duct, haematocolpos and vaginal septa, renal anomalies and aberrant ureters are among the commoner associated malformations. The finding of an aberrant ureter opening into the vagina suggesting the possibility of a uterine anomaly which was confirmed on hysterography as in SM 19 illustrates this association of anomalies in which the presence of the one lead to the investigation and detection of the other.

Vaginal Examination

Prior to pregnancy these anomalies are rarely diagnosed on vaginal examination, unless there is a vaginal septum which will alert the examiner to the presence of a malformation higher in the genital tract or where two cervices are detected. Sometimes the vaginal septum is flattened against the vaginal wall when its presence may be missed. Similarly where one of the two cervices is hypoplastic and perhaps little more than a dimple in the vault of the vagina and it is easily overlooked, especially if speculum examination has been omitted.

In those anomalies arising from aplasia or hypoplasia of one Mullerian duct such as the uterus unicornis or rudimentary horn, the cervix is placed eccentrically in the vaginal vault and Wolff (1953) considers this may be an aid to diagnosis and Jeffcoate (1957) state that the uterus which leans well to one side and cannot be straightened should always be suspected of being unicornuate.

In the bicornuate uterus the second horn may occasionally be felt on bimanual examination but is commonly mistaken for a single uterus with a fibroid or conversely a laterally placed interstitial or subserous fibroid may be mistaken for the second horn of a 'double' uterus, Hunter (1950).

Because of this confusion in diagnosis if a fibroid is palpable in a patient whose age is not in keeping with the findings, or if a single fibroid is found in a patient complaining of infertility, Halbrecht (1951), the possibility of a uterine anomaly should be considered and hysterosalpingography performed.

In the less marked forms of bicornuate and arcuate uteri identification on bimanual examination becomes increasingly difficult although it is suggested that an unusually broad fundus is suggestive of these anomalies. Obviously in the septate form of anomaly with a normal external contour, identification on pelvic examination is impossible and even at laparotomy it will escape detection.

During Pregnancy

Pregnancy facilitates the diagnosis of these anomali by (1) the increasing disparity in both size and consistency between the gravid and non-gravid horns and (11) by the occurrence of complications which should suggest the presence of the anomalous uterus. It is axiomatic of these anomalies that with the most marked forms pregnancy is likely to approach normal and conversely, it is with the minor degrees of anomaly that complications are most liable to occur. Consequently the former are more likely to be detected on physical examination and the latter have attention drawn to them through the complications to which they give rise.

In the 'double uterus' a fundal depression or cleft can usually be felt with the non pregnant horn lying to one side and a little in front of or behind the gravid horn but in the less marked forms only an asymmetry of outline is palpable. It is in the latter that confusion in diagnosis is liable to occur especially in early pregnancy because of the change in consistency in the gravid half which becomes increasingly soft compared to the firm non-pregnant half which is easily mistaken for a fibroid. Pain and tenderness are a common feature of the gravid half which is then liable to be mistaken for an interstitial or angular pregnancy or less commonly a fibroid undergoing necrobiosis. In the 'double' uterus the non-pregnant horn may be mistaken for a fibroid and later when the gravid horn rises out of the pelvis the non-gravid horn may prolapse into the pouch of Douglas where it may be mistaken for an ovarian cyst or even the head of a twin, Tombleson (1897) quoted by Hunter.

Where difficulty is experienced in distinguishing between the non-pregnant horn and a fibromyoma or ovarian cyst careful palpation will usually reveal some variation in shape and consistency from intermittent contractions Karzcma (1954) and occasionally the non-pregnant horn may be felt to contract.

Because of the changes which occur in the congenitally malformed uterus, it is never possible to decide with any accuracy the exact form of malformation during pregnancy. Philpott (1954) considers bleeding from the non-gravid horn in the early months of pregnancy typical of the 'double uterus' but agrees that until proved otherwise this should be treated as a threatened miscarriage. Pain is a frequent symptom in early pregnancy with the minor degrees of anomalous uterus and although this should be kept in mind it is often difficult if not impossible to make a diagnosis without resorting to laparotomy.

The asymmetry of outline becomes less pronounced in the last trimester of pregnancy but even at term it is usually possible to feel a fundal depression during a uterine contraction. Persistent or recurrent oblique or transverse lie without discernible cause should always raise the question of a uterine malformation.

The following phenomena have been noted as manifestations of pregnancy in the congenitally malformed uterus and if observed may be of value diagnostically; irregularity and slowing of the foetal heart before the onset of labour Falls (1956), a high head at term showing moulding Hunter (1957), the foetus in an attitude of opisthotonus, Falls (1956) passage of a decidual cast from the non-gravid horn with pregnancy continuing Corbet (1945), extreme lateral flexion of the pregnant uterus Acosta-Sison (1952), twin pregnancy with both vertices at the same level but wide divergence of the trunks.

In the present study the majority of anomalies were first discovered at evacuation of the uterus for retained products of conception and it follows that when performing this operation this possibility should be kept in mind. Also when performing manual removal of the placenta the uterine cavity should be carefully explored for the presence of an anomaly especially of the arcuate or subseptate variety.

The use of a uterine sound in the diagnosis of these anomalies in the non-pregnant state has been emphasised by Schauffler (1941) who points out that much valuable information can be obtained from this simple procedure. On passing the sound through a single cervical canal it may impinge against the point of union of the medial walls of the two horns. By gentle probing and measurement from the external os some indication may be obtained of the degree of fusion. The tip of the sound is then deflected to right and left and after negotiating the central buttress, may be insinuated into two separate cavities, Hunter (1950).

When there is a double cervix, a sound may be passed through each canal and if a metallic click can be obtained when the two instruments are brought together, there is a communication between the two cavities. The presence of a septum in association with a uterus simplex can only be confirmed by the passage of a sound or finger into the uterine cavity or when the uterus is opened at Caesarean section.

Supportive evidence of the presence of a uterine anomaly may be had from the shape of the amniotic sac which if re-constituted by the method recommended by Turpin (1938) will outline the shape of the uterine cavity. Less certain but still suggestive of the anomalous uterus is a heart shaped or bipartite placenta, Falls (1954).

The presence of an unsuspected developmental uterine anomaly may lead to grave error in diagnosis for normal curettings from

one uterine horn does not exclude the possibility of a carcinoma in the other or a viable pregnancy may be disturbed if the passage of a decidual cast from the non-pregnant horn is taken as evidence of incomplete abortion. A further diagnostic pitfall is attempting the assessment of cervical dilatation in labour on the cervix of the non-pregnant horn.

Hysterosalpingography

Hysterosalpingography or the visualisation of the uterine cavity by means of a radio-opaque substance is of inestimable value in the diagnosis of uterine anomalies but does not supplant clinical acumen and the necessity of a careful history and thorough physical examination.

Although Cary (1914) was the first to publish an account on the use of a radio-opaque substance to determine the patency of the Fallopian tubes, Dimier (1913) had experimented with a similar substance to outline the uterine cavity but his results were not published until 1916. Rubins (1914) was another early investigator in this field, and an account of his work on the use of a radio-opaque substance in the diagnosis of uterine tumours in rabbits appeared a few months after the publication of Cary's work.

In early experiments first sodium bromide then collargol but the risks attendent on their use was too great and it was not until 1926 that the relatively safe iodised oil (Lipiodol) was introduced. Forsdyke (1925) introduced a standard technique for hysterosalpingography and Jarcho (1927) by showing that it was relatively harmless provided certain elementary precautions were taken, did much to popularise its use as a diagnostic aid.

A disadvantage of lipiodol was the oil medium with the rare but definite risk of oil embolism and it has been reported as

causing granulomata but these disadvantages have now been overcome with the introduction of a water soluble media.

Such anomalies as the uterus septus and subseptus with normal uterine contour can only be revealed by hysterogram but even when the diagnosis can be made without this procedure it should not be omitted as it adds a definite element of exactness as to the type of malformation.

Although an invaluable diagnostic aid the interpretation of a hysterosalpingogram requires a certain amount of caution and it should be borne in mind that only the uterine cavity or cavities is revealed and not the outline of the uterus otherwise those anomalies twith two cavities such as the bicornuate and septate uterus may easily be confused. Similarly when only one horn of a double uterus fills it may be mistaken for a uterus unicornis. On the other hand filling defects produced by fibromyomata may simulate the 'double uterus' very closely especially where the tumour presses on the fundus of the uterus as in the case recorded by Jarcho (1928).

Apart from these minor drawbacks this procedure has proved of the greatest value and should be a routine procedure in the investigation of repeated abortion, infertility, unexplained stillbirth, malpresentations and repetitive abnormalities of pregnancy and labour so that the presence of a hitherto unsuspected bicornuate uterus may be revealed, Way (1947). The results of Halbrecht's (1941) investigation of 107 women complaining of infertility or habitual abortion would appear significant for hysterosalpingography showed a congenital malformation of the uterus in 18 per cent of the first

group and 50 per cent in the second.

Utero-tubal Insufflation

This procedure does not appear to be an aid in diagnosis for Jurgens (1947) in seven known congenital uterine anomalies, found only one which gave an abnormal Rubin test.

INVESTIGATION AND CONFIRMATION

SUSPECTED CONGENITAL UTERINE ANOMALIES

TIME AND CIRCUMSTANCES OF DIAGNOSIS

A comparison of some of the more recent studies on congenital uterine malformations will reveal considerable variation as to the time and circumstances of diagnosis. Fenton and Singh (1952) and Baker (1953) state the diagnosis of the anomalous uterus should present little difficulty to the diligent clinician while Philpott (1954), considers that in spite of statements to the contrary, the diagnosis is rarely made at the first examination. In Baker's study the diagnosis was made at routine examination antenatally in 59.1% which is nearly three times the 21.6% reported by Philpott. This difference is explained to some extent by the higher proportion of the Type 1 anomaly in the former and also because this series consists largely of cases abstracted from the literature with only a small number of personal records. Certain studies like Karczma (1954) in which the diagnosis had been made at gynaecological examination prior to pregnancy in 31.2%, at hysterography for infertility or unsuccessful pregnancy in 59.4% and only in 9.4% of cases at antenatal examination cannot be used in comparison because of selection of material.

ANALYSIS OF DIAGNOSIS IN PRESENT STUDY

Time of Diagnosis	Type of Anomaly 1	III	V	VII			
Antenatally	6	5	1	0	=	12	= 17%
Intrapartum	2	0	0	0	=	2	= 3%
Caesarean Section	0	4	2	1	=	7	= 10%
Manual Removal Placenta	1	1	5	0	=	7	= 10%
Laparotomy	0	8	0	1	=	9	= 13.5%
Infertility Investigation	2	7	3	0	=	12	= 17%
D & C for Retained Products of Conception	6	6	5	0	=	17	= 25%
Others	1	0	0	0	=	1	= 1.5%
Gynaecological							
Defloration Haemorrhage	1						
Haematocolpos	1	-	-	-	=	2	= 3%
Total:	20	31	16	2			

The time and circumstances of diagnosis from three studies are given for comparison in the table below.

	Philpott	Present Study	Baker
Normal antenatal examination	21.6%	17%	59.1%
Infertility Investigation	4.8%	17%	
Curettage for Retained Products of Conception	14.8%	25%	
At time of delivery	48%	23%	38.6%
Laparotomy	9.6%	13.5%	2.4%

In the present study the low rate of diagnosis at antenatal examination represents the percentage of the total number of anomalies discovered in this way, and does not take into account the 32 cases diagnosed prior to their attendance at the antenatal clinic. Therefore of the remaining 37 anomalies in which the diagnosis was unknown at the time of their first antenatal attendance, the time and circumstances of diagnosis will be as follows:

Antepartum	...	12 (32.4%)
Intrapartum	...	9 (24.3%)
Manual Removal of Placenta	...	7 (18.9%)
At Laparotomy	...	8 (21.6%)
At Postmortem	...	1 (2.7%)

If the time of diagnosis is related to the form of anomaly:

First Diagnosed	Type of Uterus		
	I	III	V
Antepartum	60%	28%	12%
Intrapartum	20%	22%	25%
Manual Removal of Placenta	10%	6%	60%
Laparotomy	-	45%	-

These results which are reflected in all similar investigations, clearly support the axiom that the more marked the form of anomaly, the more likely is a diagnosis to be made on physical examination and conversely, the minor degrees of anomaly are usually suspected from the symptoms to which they gave rise and are rarely diagnosed on physical examination.

DIAGNOSIS AND INVESTIGATION AND CONFIRMATION OF SUSPECTED CONGENITAL UTERINE ANOMALIES

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THE SURGICAL CORRECTION OF UTERINE MALFORMATIONS

THE SURGICAL CORRECTION OF UTERINE MALFORMATIONS

This section is a consideration of those operative procedures designed to convert the malformed uterus into a single cavity organ either by (a) reunification of its two horns or (b) by removal of the intrauterine septum. Pathological conditions arising in consequence of a genital malformation and requiring surgical intervention for their relief have already been considered.

Historical Review

Although Barthelemy Savard, a Paris surgeon, described the incision and drainage of a haematocolpos in in the 17th century,(Ricci), it was not until the latter half of the 19th century that surgical correction of the malformed uterus was attempted. This took the form of converting the septate uterus into a single cavity organ by excision of the septum and was first accomplished by Paul Ruge (1884) and three years later by Schroder.

This type of operation was performed under direct vision using the vaginal aproach, and in Ruge's case was that of a woman who had had two spontaneous abortions. One year postoperatively after a normal pregnancy and labour, she was delivered successfully of a living child at term. Within the next few years many more records of this type of operation were contributed to the literature with occasional minor deviations of technique such as that advocated by Rockey (1916) in America, where clamps were applied to the septum and left until it had sloughed away. The idea of plastic unification of the two horns of a bipartite uterus where this was a bar to conception or caused repeated abortions or premature labour, was conceived by Paul Strassmann in 1903 and successfully realised by him in 1907. The success of his operation is well illustrated by the patient's obstetric history before and after operation, for although only twenty seven years old, she had had eight pregnancies without a living child, all having terminated in abortion or premature labour and in addition she suffered from dysmenorrhoea and dyspareunia. Eighteen months after operation a living child was delivered at term to be followed by a further five deliveries in succeeding years.

The operation was performed vaginally, the two uterine bodies being delivered through an opening in the vesico-uterine pouch and the bridge of tissue joining the two halves divided down to their common internal os. The two uteri were then opened by a saddle shaped incision extending from the left to the right cornu so allowing the two bodies to be united, the lateral angles of the incision becoming the centre of the suture line. They were approximated by two layers of interrupted catgut sutures, the first transfixing the whole wall on both sides and the second the serosa superficially. The unified organ was then returned to the abdominal cavity and the peritoneum of the utero-vesico pouch utilised to fix the uterus in position and at the same time to peritonise its anterior surface. From this time, the operation gained in popularity and was practised in most of the leading clinics here, and in Europe, but only within recent years have reports of the operation appeared in the American literature due particularly to the efforts of Paul Strassmann's son, E.O.Strassmann.

In this country, Munro Kerr (1920) reported the successful plastic reconstruction of a uterus bicornis unicollis. The patient had had two miscarriages and as the two uterine horns were symmetrical they were joined after resecting the middle portion.

Eight months later she became pregnant and went into spontaneous labour at term and after a second stage lasting two and a half hours it was decided to effect delivery by forceps. The pelvic brim was slightly flattened and considerable difficulty was experienced in delivery and as a result the child died from intracranial haemorrhage on the twelfth day. The uterus was manually explored after delivery of the placenta in order that the condition of the cicatrix could be determined. This was felt as a strong ridge of tissue running vertically along the anterio and posterior wall of the uterus. Two years later the patient was delivered successfully of a second pregnancy; this time by Caesarean section, the uterus being removed as the patient dreaded further pregnancy. Bonney (1944) in conjunction with MacIndoe, was the first to perform a successful plastic operation on a girl with absence of the vagina in association with a double uterus with no cervix; the two uterine bodies terminating blindly in a mass of tissue. The artificial vagina was constructed in the usual way, by opening up the plane between the rectum and bladder and inserting an obturator with a skin graft. The two uterine bodies were united from above, in the manner of the usual Strassmann operation,

and an artificial opening made between them and the vagina into which a rubber drain was inserted to keep the channel patent. The skin graft took and the artificial cervix remained patent after removal of the drainage tube and some months later the patient commenced to menstruate normally.

INDICATIONS AND CONTRAINDICATIONS TO OPERATION

Although this thesis is concerned with malformations of the uterus, it is necessary to mention briefly the various congenital anomalies of the vagina which may require surgical correction.

Type of Abnormality	Operation
(1) Aplasia of the vagina, Complete or incomplete.	Artificial construction of vagina from skin grafts.
(11) Persistence of cloacal membrane giving rise to various forms of fistulae vesico-vaginal, recto-vaginal.	Closure of fistulae with or without transplantation of ureters, colostomy.
(111) Haemato- or hydrocolpos.	Excision of occluding membrane.
(1V) Longitudinal vaginal septa and transverse diaphragms, where these interfere with fertility, or are a cause of dyspareunia, or in labour where they may interfere with descent of presenting part.	Excision of septum.

Although the indications for plastic unification of the uterus vary according to the different authorities, all are agreed that the presence of a double uterus is not in itself an indication for operation. The literature offers hundreds of examples of women with these congenital malformations who have had normal pregnancies and labours and who have been completely unaware that anything was wrong, the abnormality being discovered accidentally at operation or autopsy. It cannot therefore be too strongly stressed that very careful assessment of the case is necessary before uterine metroplasty is performed and one must be certain that it is the uterine anomaly which is the cause of the complaint and not some additional lesion.

Strassmann (1952) gives three conditions which should be fulfilled before a metroplastic operation is performed:

(a) Hysterosalpingogram to determine exact size and position of uterine horns and of cervices in uterus didelphys. Patency of at least one tube should be assured for if bilateral tubal obstruction present then obviously any metroplastic operation is unjustifiable.

(b) Intravenous pyelography and in doubtful cases retrograde pyelography to investigate the renal tract.

(c) There should be an interval of at least six months between operation and the occurrence of pregnancy so that a firmly healed uterine scar is obtained.

Martius (1937) gives three definite reasons for operation and these are:

(1) Dysmenorrhoea.

(11) Sterility.

(111) Habitual Abortion.

The first indication dysmenorrhoea, is a very valid reason for surgical interference and is often the first symptom which will bring the patient to the doctor. Even so Steinberg (1955) is of the opinion that dysmenorrhoea which is solely caused by uterine malformations is caused either by stenosis of the cervical os of one of the horns or by haematometra in a rudimentary horn but Strassmann (1952) among other authorities is not in agreement with this. They are of the opinion that incoordinate uterine action is often the cause of severe dysmenorrhoea and can be cured by uterine metroplasty.

The second indication, sterility is not generally accepted and Steinberg (1952) Pfleiderer (1929) stating that sterility is rarely the lot of the woman with a genital malformation whilst Schauffler (1941) is of the opinion that on the contrary, the fertility and sex urge are distributed amongst these women in the same generous proportions as their organic sex equipment. Brody (1954) has pointed to the one definite exception to this rule, where a septate vagina exists with a double uterus and cervix and where intercourse takes place only on the side which connects with a possibly underdeveloped half of the uterus (see case SM2) here removal of the vaginal septum will remedy the condition, otherwise one has to search for other causes of sterility. The third indication listed above, habitual abortion, is the main indication and this opinion is held by the vast majority of workers in this field. For this reason the gynaecologist should perform a hysterosalpingography routinely on all cases of habitual abortion to exclude a uterine malformation. Palmer (1953) found an incidence of 25 per cent of uterine malformations in his reported series of habitual abortion and concludes that metroplasty is indicated in cases of bifid uterus where three consecutive abortions have occurred at the same time of gestation, or if two, provided the last happened in spite of maximal medical treatment.

E.C.Strassmann and Ospelt (1941) add a further indication for operation, menometrorrhagia which does not respond to conservative treatment. They consider the menometrorrhagia to be caused by 'disturbance of shedding of the endometrium in the two horns' and that operation gives good results. Steinberg feels this must be a very rare indication and certainly in our series menometrorrhagia was a rare symptom if other causes than the uterine anomaly were ruled out.

Although obstetrical complications are one of the commonest findings with uterine malformations, metroplasty is rarely indicated since it is usually possible to circumvent these complications by Caesarean section.

There is a minority opinion who consider that metroplasty is never indicated for bipartite uterus since many normal deliveries are reported of patients with these malformations Rheman (1934) Granberry and Faust (1938). Particularly unwarranted are operations designed solely to prevent any subsequent complications Hannes (1932). Contrary to these opinions Kustner (1929) felt that because of the various reports of ruptures of the pregnant bicornuate uteri at the locus minoris resistentiae, namely the saddle, prophylactic metroplasty had a definite place in modern gynaecology.

Operative Procedure

When Strassmann originally described this operation in 1907, he mentioned two possible ways of attacking the problem; namely, by the vaginal route or by laparotomy. Today laparotomy has become the method of choice, except in cases of uterus didelphys with two cervices. As it is not always feasible to unite the two canals from above it is preferable to precede the laparotomy with a plastic fusion of the two cervices per vaginam.

Strassmann accordingly, subdivided his cases into two groups. Group 1 includes those malformations of the uterus that have a common muscular coat (uterus septus, subseptus, arcuatus, and some mild forms of uterus bicornis which present only a small saddle formation at the fundus). Group 2 comprises uteri with divided musculature and pronounced "horn" formation such as uterus didelphys, uterus bicornis unicollis, and uterus duplex bicornis. The classification of the different types of uterine malformations from hysterosalpingogram occasionally offers considerable difficulties.

Procedure for Group 1

The vagina is adequately prepared by several applications of a germicide solution. After opening of the abdominal cavity, the peritoneum in front of the uterus is divided by a circular incision from one round ligament to the other to obtain a flap with which to peritonealize the suture-line later. An incision is now made on top of the fundus from side-to-side, starting about 1 cm. mesially of the tubal origin to the corresponding place on the other side. Von Mikulicz-Radecki cautioned about "avoiding the intramural portion of the tube, lest inflammatory occlusions of the tube occur." With a sharp scalpel all three coats of the uterus are divided, until the uterine cavity is reached. Bleeding from this incision is moderate and can be easily controlled by using suction with a small cannual-tip. With four Allis forceps applied, the inner aspect of the uterine cavity is now exposed. With a straight instrument, the depth and width of the half-cavities are explored to gain an idea of the thickness and course of the dividing septum. Then, with long straight and heavy scissors or a scalpel the septum is cut away as close to the uterine wall as possible. Very little bleeding is encountered in this procedure. The septum may be quite thick and tough so that a sharp instrument is imperative. After the septum

is removed, the cavity should be explored with the finger to ascertain that no remnants of the structure have been left. Puppel (1926) warned against leaving any parts of the septum as "spurs", since it might jeopardize the value of the operation. A uterine dressing forceps is now inserted downward through the cervical canal, and under guidance of the finger in the cul-de-sac, pushed through into the vagina. A one-half inch iodoform gauze strip is then led through the cervix, the dressing forceps withdrawn, and the uterine cavity packed with as much of the iodoform gauze as it will conveniently hold. This will aid haemostasis and at the same time, prevent a growing together of the two wound edges which has previously been reported.

The suture line should run perpendicular to the line of incision, and to this end, the Allis clamps are inserted at the mid points of the opening and pulled anteriorly and posteriorly respectively, in order to approximate the two halves. The uterus is to be closed in two or three layers, using chromic catgut No 0 on a medium-sized, cutting needle. Particular care should be exercised in placing the sutures, lest an artificial internal adenomyosis be created. However, H.Kustner (1951) in a report of this operation, as well as Skutsch (1952) stressed the fact that the endometrium ought to be closely approximated to obtain a smooth scar. This technique is at variance with the closure after a Caesarean section, where a suture through the endometrium is carefully avoided. All sutures of one layer

are left untied until the entire row is placed; then, they are tied and cut except the ones near the corner which serve to hold up the uterus. The final row takes in the serosal coat and the uppermost layer of the uterine muscle and, after all sutures are in place, the uterus assumes a shape which more or less can be compared with the shape of a rhomboid. Finally, the peritoneal fold is drawn over the uterus as far back as possible and "tacked" on with interrupted fine catgut sutures to the top as well as the posterior aspect of the uterus, thus leaving only a small part of the suture line exposed. Louros (1935) proposed to add, in cases of retroflexion, a suspension operation of the Gilliam type which may enhance the value of the procedure.

The packing in the uterine cavity is removed on the fifth postoperative day at which time it is assumed that the wound edges are sufficiently epithelialized to prevent them from growing together. A hysterography is done about two months laster to visualize the plastic effect.

Procedure for Group 2

The different steps for the operation are quite similar to those described for Group 1 with a few exceptions. If one is dealing with a uterus didelphys or bicornis bicollis, it is recommended to start the operation from the vagina, after putting the patient in lithotomy position. Depending upon the access to the two cervices, a Schuchardt incision

might be necessary in order to facilitate exposure. Then, the dividing septum between the two cervices is cut with heavy, straight scissors, to unite the two halves. Immediately after that, a laparotomy is done. The uterine incision is also placed horizontally starting from 1 cm. mesially of the origin of one tube, going across the saddle and up the mesial aspect of the other horn; this incision should likewise go through all three coats. The cervical canal should then be explored to see whether all of the dividing septum has been removed. The sutures in cases of this group should be placed in such as way that both front, as well as both hind halves are brought together and a drain should be inserted into the vagina. The closure is effected in a similar fashion with two or three layers of interrupted chromic catgut.

Modifications of Strassmann's Metroplasty

The classical method originally described by Strassmann (1907) has undergone some modifications by certain authors. Garcia-Pastor (1948) proposed that the original procedure be reserved for cases of bipartite uterus which exhibit a complete symmetry of the two halves. However, if one side be larger than the other, he prefers amputation of the smaller horn and implantation of the round ligament into the remaining half. A similar procedure had previously been advocated by Bottaro, as quoted after Rodrigo(1937).

In later years, Strassmann himself augmented his original operation by describing "the implantation of an unattached horn" in those uteri, where the isthmus of one horn was missing and thus no communication existed between the corpus and the cervix. This modified procedure has since been repeated by Colombino and Bartoszewski. A still different approach was tried by Eisaman (1944) who performed "stripping the peritoneum from the superior surfaces of the uterine horns, approximating the organs and anchoring them with silk sutures". Rodrigo also describes the morcellement of the uterine septum by means of a biopsy punch forceps, thus removing it piecemeal through the cervix. Luikart who operated on a puerperal uterus applied two "stomach clamps through the open cervix" and thus removed the septum. Strictly speaking, the two last mentioned procedures are really not a metroplasty, but rather a "septectomy". Jones and Jones (1953) performed a similar operation; they believed that "the literature contains no case of a non-communicating horn of a septate uterus"; however, this was already reported by Strassmann in 1912. The wedge-shaped excision from the fundus, as proposed by Kakuschkin (1924) is rejected by Strassmann (1928). The technique employed by Jones and Jones differs decidedly from that recommended by Strassmann; it follows more closely the method originally described by Kerwin (1925). Jones and Jones employ a "section" of the uterus in midline and dissect out

the septum in that fashion. Strassmann opposed this type of surgical approach, because he felt that it would leave the uterus with too large a scar (Jung,1931). Mateos Fournier (1952) proposed to suture the uterus in a zig-zag manner, because he believed it to be stronger and less apt to rupture in subsequent pregnancies.

Method of Delivery After Strassmann's Metroplasty

The question arises, whether a patient who has undergone a plastic operation on her uterus should be delivered vaginally or by Caesarean section. A great many European physicians have delivered their women by the vaginal route and resorted to Caesarean section only if dystocia occurred or other obstetrical indications existed warranting surgical intervention. A uterus which has been subjected to a metroplasty should be treated in the same manner as one after a previous abdominal delivery or myomectomy. By the same token that the old adage "Once a section, always a section" has been superseded today by judicious adaptation to the particular patient, the same has become true of patients after a metroplasty. Provided there is no obstetric contraindication, these patients should be allowed to go into labour spontaneously under constant observation of the attending physician. As soon as the first signs of dehiscence of the scar become apparent, the patient should be delivered abdominally. Many workers have thought it justifiable to advocate a "prophylactic" low-mid or outlet forceps to ease the strain upon the weakened uterus.

For 45 years no untoward effect, such as rupture of the uterus, has been reported in the literature after Strassmann's operation. It was not until very recently that Sindram and Sikkel (1953) reported a case of rupture of the uterus after Strassmann's metroplasty. The paper by Sindram and Sikkel may radically change the management of patients who have undergone plastic surgery of the uterus.

Strassmann (1956) the son of the originator of the operation which bears his name, reviewed the world literature on the subject and found a total of 128 cases from fifty-one surgeons, including five cases of his own and seventeen operated upon by his father. Three years later Steinberg (1955) again reviewed the world's literature and collected 142 cases, but was of the opinion that there were many more examples which had not been reported upon. The indications for operation are approximately the same in both series as can be seen from the following tables.

	Strassmann		Steinberg (only 107 had sufficient information to be used)
Habitual Abortion	36	43	45.7% a. 34.5% b. 11.2%
Dysmenorrhoea	33	40	40%
Menometrorrhagia	22	26	5.6%
Sterility	13	16	10.3%
Dyspareunia	9	11	
Premature labour	3	4	

Steinberg further divided the cases of habitual abortion into (a) those with two or more abortions or premature labour and (b) those with one abortion or premature labour.

Additional indications found by Steinberg were:

Operated upon during pregnancy ...	7.5%
Acute conditions within the abdomen	3.7%
Prophylactic operation	2.8%
Previous dystocia	0.9%
Others	1.8%

Steinberg also states that in many of his cases in the review had two or more indications and this probably accounts for the disparity.

Postoperative Results

Strassmann, 83 postoperative pregnancies in 128 cases.

Dysmenorrhoea relieved	34
Living baby, vaginal delivery ...	61%
Living baby, Caesarean section ...	10
Abortions	10
Stillbirths	1 died in utero prior to Caesarean section for placenta praevia
Neonatal deaths	
Postoperative deaths	1
Pregnant at time of report ...	1
Pregnancy not obtained or reported	

Postoperative results in the 142 cases

Dysmenorrhoea relieved in 31

61 (57) became pregnant 37 had living babies delivered per vaginam

14 delivered by Caesarean Section

4 miscarriages

2 stillbirths

2 neonatal deaths

1 postoperative death

1 pregnant at time of report

15 pregnancy not obtained nor reported.

The one death was from peritonitis from accidental rupture of the pyosalpinx at operation and occurred in the pre-chemotherapy days.

Functional results Strassmann states that high percentage menstrual disorders seem to clear up after operation, and this includes severe dysmenorrhoea as well as menometrorrhagia. In no case was a remark found indicating that these conditions persisted afterwards(sic) and in 34 instances complete relief recorded, and all of the authors cases were completely relieved of dysmenorrhoea. In two of his cases also menometrorrhagia subsided completely postoperatively.

Strassmann gives the results of 64 pregnancies in fifty cases of double uterus reported by Phillips (1948) with the results of pregnancy in 128 patients before and after operation and the results are very striking.

Phillips Series

64 pregnancies in 50 cases uterine duplication of which:

25 per cent went to term

16 per cent premature labour

59 per cent miscarriages.

In Strassmann's series of 128 patients selected for surgery there were:

110 preoperative pregnancies of which:

4 or 3.7 per cent went to term

17 or 15.3 per cent were premature

77 or 70 per cent were abortions

12 or 11 per cent outcome unknown.

Postoperative there were 83 pregnancies in the same 128 patients of:

71 or 85.6 per cent went to term

10 or 12 per cent miscarriages

2 or 2.4 per cent unreported upon.

These figures are even more striking when compared together as below:

COURSE OF PREGNANCY

	Term	Premature	Miscarriage	Unknown
Unselected cases double uterus (64 pregnancies)	25%	16%	59%	0
Cases selected for surgery Preoperatively(110 pregnancies)	3.7%	15.3%	70%	11%
Cases selected for surgery Postoperatively(83 pregnancies)	85.6%	0%	12%	2.4%

In summary Strassmann believes that with proper indications the operation provides a breeding space twice the size that was present before, and that this more than quadruples the chances of carrying a pregnancy to term, while it reduces the chances of miscarriage.

In the authors experience sterility can be relieved by metroplastic operation, when no other cause can be found but the uterine malformation.

Of the few British figures available those for the Chelsea Hospital for Women are given below. The earliest recorded was in 1927 where a successful metroplastic operation was performed on a woman who had had three unsuccessful pregnancies, two premature labours and one miscarriage, the latter following laparotomy for suspected ectopic gestation when the anomaly was first noted. Six months after plastic unification of the two horns the patient again became pregnant and was delivered spontaneously at term after a labour lasting 11 hours 40 minutes.

Although forty-five cases of congenital malformation of the uterus were admitted to the Chelsea Hospital for Women between the years 1935-1956, only twelve had plastic operative treatment of which eleven were vaginoplasties in each of these the uterus was absent and in the remaining one already mentioned case (MacIndoe and Bonney) vaginoplasty was combined with utriculoplasty.

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CASE RECORDS

CASE HISTORIES

The following case reports serve to illustrate many congenital uterine malformations and the different clinical conditions to which they may give rise. These case histories are from patients I have examined during the past seven years.

St.Mary Abbots. *

Type	I	III	V	Total
Number of anomalies	2	10	6	18
a viable births	4	13	9	26
b abortions	1	9	2	12
c mature infants	3	11	7	21
d infant deaths	0	4	4	8
e normal deliveries	4	6	2	12
f operative deliveries	0	7	7	14
g Caesarean section	0	4	2	6
h abnormal pregnancy	5	22	9	36
i abnormal labour	2	8	12	22
j maternal deaths	0	0	0	0
k maternal morbidity	0	0	0	0
l foetal abnormalities	0	0	2	2
m miscellaneous operative procedures	1	17	11	29

Erratum - should face page 267.

Guy's Hospital 1950 - 55

Total number obstetric patients

Number Congenital Uterine Anomalies

Statistical Analysis

Type Abnormality Number	I 3	III 5	V 1	VI 1	Total 10
a. Viable births	5	6	1	1	13
b. Abortions	3	11	6	0	20
c. Infants (more than $5\frac{1}{2}$ lb.)	1	3	1	1	6
d. Foetal deaths	3	2	1	0	6
e. Non-operative deliveries	1	5	1	1	8
f. Operative deliveries	4	1	0	0	5
g. Caesarean Section	0	0	0	0	0
h. Complication Pregnancy	7	10	2	2	21
i. Complication Labour	4	2	0	0	6
j. Maternal mortality	0	0	0	0	0
k. Maternal morbidity	1	3	0	0	4
l. Foetal abnormalities	0	0	0	0	0
m. Miscellaneous Operative deliveries	5	11	3	1	20

Uterus Bicornis Bicorpus Unicollis — Type 111

G1 13353 1953

Pain in Early Pregnancy

Premature Labour

Postpartum Haemorrhage

Mrs.J.Austin, aged 25.

Menstrual Type K 12 6/29 normal loss, moderate dysmenorrhoea.

Married 4 years, voluntary infertility.

Para 1. No miscarriages. L.M.P. 25th June, 1952. E.D.D. 2nd April, 1953.

a - 1
b - 0
c - 0
d - 0
e - 1
f - 0
g - 0
h - 2
i - 1
j - 0
k - 1
m - 1

At the third month of her pregnancy the patient was seized by acute pain in the right lower quadrant of the abdomen, which was associated with slight vaginal bleeding. Ectopic pregnancy was suspected and the patient was admitted to a hospital where laparotomy was performed and a bicornuate uterus was found with the pregnancy in the right horn. The abdomen was closed and the pregnancy proceeded normally.

Spontaneous delivery of a living male infant at the 37th week, weighing 5 lb. 7 oz. Postpartum haemorrhage of 26 oz. membranes noted to be ragged and on the 5th day of the puerperium a cotyledon of placenta passed.

Congenital Absence Anal Canal, Rectovaginal Fistula.

G2 1954
B5303 Bicornuate Uterus, Vaginal Septum.

Normal Pregnancy, Vaginal Septum Preventing Delivery.

Division of Septum, Forceps Delivery.

Mrs.T.Moss, aged 24.

Left colostomy aged 4, unsuccessful attempt to restore bowel continuity aged 16, followed by urinary incontinence, recurrent urinary infection and bladder calculi. Permanent suprapubic cystostomy.

Menstrual Type K 14 3/27 regular, painless.

Para 1 Gravida 2, one miscarriage at 26 weeks gestation in 1950.

1954 - forceps delivery of living male infant weighing $8\frac{1}{2}$ lb. at 38th week gestation, after division of vaginal septum which was preventing delivery of the head. Previously an examination under anaesthetic had revealed a thick median vaginal septum, short vagina, single cervix and bicornuate uterus.

Type 111

a - 1
b - 1
c - 1
d - 0
e - 0
f - 1
g - 0
h - 1
i - 1
j - 0
k - 1
l - 0
m - 1

G3 1955 Arcuate Uterus.

B6741 Infertility.

Transverse Lie, Normal Delivery.

Mrs.V.Jeffries, aged 23.

Menstrual Type K 14 5-6/28 regular, moderate loss, severe dysmenorrhoea. Married $2\frac{1}{2}$ years, attended infertility clinic where abnormality not noted. Advised only.

Para 1 Gravida 1. Became pregnant two months after visit to infertility clinic. At 34 weeks the lie of the foetus noted to be transverse and at the same time uterine fundus noted to be markedly arcuate. External cephalic version easily performed.

Normal delivery of living male infant, weighing 7 lb. after episiotomy. Duration of labour 18 hours. Medical induction as three weeks postmature by dates. Third stage normal.

Type V1

a - 1	h - 2
b - 0	i - 0
c - 1	j - 0
d - 0	k - 0
e - 1	l - 0
f - 0	m - 1
g - 0	

G4 1950 Uterus Unicorpus Unicollis Subseptus.

A8232 Repeated Unsuccessful Pregnancies.

Metroplastic Operation.

Mrs.Olive Butler, aged 28.

Menstrual Type K 10 5/28 regular.

Rheumatic Fever twice, mitral stenosis, appendicectomy.

Para 1 Gravida 7, six miscarriages between 16th - 24th week.

The first pregnancy in 1943 was complicated by pre-eclamptic toxaemia which necessitated surgical induction of labour by artificial rupture of the membranes, at the 36th week. Spontaneous delivery of a female infant, weighing 6 lb. 3 oz. which died on the 9th day postpartum. No post mortem.

This was followed by five miscarriages, in all of which bleeding occurred throughout the pregnancies.

In 1952 during evacuation of the uterus for retained products of conception a uterine septum was felt, and this was confirmed by hysterosalpingography. In view of the repeated unsuccessful pregnancies uterine metroplasty was decided upon. Laparotomy revealed a uterus normal in size and shape, and on opening the uterus the septum was excised with a wedge shaped portion of the fundus, and the edges of uterine incision accurately opposed by a series of interrupted catgut sutures, which in turn were buried by a continuous catgut suture.

The patient made an uninterrupted convalescence.

G4 1950

A8232 /continued.

Eight months later she again became pregnant and when 20 weeks pregnant was admitted to hospital complaining of severe lower abdominal pain and there was marked tenderness over the laparotomy scar. Suspecting dehiscence of the uterine scar the abdomen was opened but the region of the scar was perfectly sound without any trace of the previous operation. The abdomen was closed and the patient discharged well two weeks later. When 26 weeks pregnant she was again admitted to hospital with a temperature of 104° due to a pyelitis and aborted the following day.

Type V

a - 1	h - 2
b - 6	i - 0
c - 1	j - 0
d - 1	k - 0
e - 1	l - 0
f - 0	m - 3
g - 0	

G5 1953 Pregnancy in Rudimentary Horn

A15109 of Bicornuate Uterus.

Mrs. Dorothy Harrild, aged 24.

Menstrual Type K 13 5/28 regular, slight premenstrual dysmenorrhoea. Married $2\frac{1}{2}$ years.

Para 0 Gravida 1. At the 12th week of pregnancy the patient had slight painless red loss per vaginam, and was advised to rest at home. The bleeding subsided with rest but recurred three weeks later and was not associated with lower abdominal discomfort. She was admitted to hospital and the uterus was found to be just palpable per abdomen and therefore not compatible with her dates. She was observed for twelve days in the ward, at the end of this period a vaginal examination was performed when the uterus was felt to be the size of a 10 week pregnancy and there was a tender mass lying in front of the uterus. Laparotomy was performed and a bicornuate uterus was found with the right horn/rudimentary and gravid. The right horn and tube were removed but the ovary conserved. There did not appear to be any communication between it and the left horn, and on opening the specimen a macerated foetus of approximately twelve weeks size was seen.

G5 1953

A15109 /continued.

During the postoperative period no note was made of a decidual cast having been passed.

The patient made an uneventful recovery.

Type 111

a - 0	h - 1
b - 1	i - 0
c - 0	j - 0
d - 0	k - 1
e - 0	l - 0
f - 0	m - 1
g - 0	

G6 1954 Uterus bicorpus bicollis, vagina simplex.

GB 5239 Premature Labour, Breech Extractions.

Pre-Eclamptic Toxaemia.

Mrs.Betty Humphries, aged 25.

Menstrual Type K 16 7/29 moderate loss, regular, slight dysmenorrhoea.

Para 2 Gravida 2.

1954 - Surgical induction of labour at 34th week for pre-eclamptic toxaemia. Delivery of a stillborn male infant weighing $3\frac{1}{2}$ lb. by breech extraction with forceps to aftercoming head. Labour lasted 51 hours, uterine inertia treated by intravenous pitocin drip when both horns seen to contract independently. Third stage normal, pregnancy in left horn.

1955 - Premature labour at 34th week pregnancy. Breech extraction of a living female infant weighing 4 lb. 15 oz. Third stage normal, pregnancy in right horn.

Type 1 - vagina simplex.

a - 2	h - 3
b - 0	i - 1
c - 0	j - 0
d - 1	k - 1
e - 0	l - 0
f - 2	m - 1
g - 0	

G7 1954 Uterus Bicornis Bicollis Vagina Septate.

A1159 8 Repeated Miscarriages.

Prolapsed Cord, Breech Extraction.

Mrs.Rosina Murray, aged 27.

Menstrual Type K 12 5-6/28 regular, loss normal painless.

Para 1 Gravida 3, two miscarriages one at 10 weeks and the other at 12 weeks.

Abnormality discovered at evacuation of uterus for retained products. The first pregnancy was in the right and the second in the left horn.

The third pregnancy proceeded normally until the 20th week when admitted to hospital as a threatened miscarriage. This however settled with rest. During the antenatal period the presentation remained a breech in spite of attempted external cephalic version. Spontaneous labour at 36th week, breech extraction for prolapsed cord. Living female infant weighing 7 lb. 4 oz. Duration of labour 16 hours 30 minutes. Third stage normal.

Type 1

a - 1	g - 0
b - 2	h - 1
c - 1	i - 1
d - 0	j - 0
e - 0	k - 0
f - 1	l - 0
	m - 3

G8 1951 Uterus Bicornis Unicollis

A4481 Repeated Unsuccessful Pregnancies.

Mrs.Barbara Hackett, aged 26 years.

Menstrual Type K 15 4/28 regular, slight loss only, moderate premenstrual dysmenorrhoea. Married 9 years.

Para 2 Gravida 7 - Five miscarriages between the 12th - 27th week pregnancy. Two viable births, the first terminating in premature labour at the 32nd week, the infant dying after nine hours from atelectasis, and the second at 28th week. The viable pregnancies being the second and seventh.

Type 111

a - 2	g - 0
b - 5	h - 2
c - 0	i - 0
d - 2	j - 0
e - 2	k - 0
f - 0	l - 0
	m - 2

G9 1950 Bicornuate Uterus.

A13405 Repeated Miscarriages.

Two Normal deliveries.

Mrs.Joyce Pettingell, aged 25 years.

Menstrual Type K 14 7/28 regular, loss normal, painless.

Para 2 Gravida 6 - four miscarriages at fifth month.

Hysterosalpingogram after the third miscarriage revealed a bicornuate uterus. Following the four miscarriages the patient had two normal deliveries at term, weighing 6 lb. 5 oz. and 6 lb. 12 oz. respectively, both males. Progesterone had been implanted after the first missed period after both these pregnancies.

Type 111

a - 2	g - 0
b - 4	h - 4
c - 2	i - 0
d - 0	j - 0
e - 2	k - 0
f - 0	l - 0
	m - 6.

G10 1950 Uterus Bicornis Bicorpus Bicollis vagina septate

B2946 Repeated Unsuccessful Pregnancies.

Mrs.P.Wilson, aged 21.

Menstrual Type K 10 7-9/28 regular, loss moderate, slight dysmenorrhoea.

Para 2 Gravida 3. The first pregnancy terminated in a miscarriage at six weeks, and following evacuation of the uterus for retained products of conception, the abnormality was discovered. The second pregnancy, one year later, resulted in a stillborn male infant weighing 2 lb. 9 oz. after premature labour at the 32nd week with a prolapsed cord and forceps delivery.

In the third pregnancy premature labour occurred at the 35th week and after a spontaneous labour a male infant weighing 3 lb. 10 oz. was born but died after 48 hours from prematurity.

Type 1

a - 2	h - 2
b - 1	i - 1
c - 0	j - 0
d - 2	k - 0
e - 1	l - 0
f - 1	m - 1
g - 0	

West Middlesex Hospital

	1	111	V	Total
	4	3	3	10
a	5	4	3	12
b	3	2	8	13
c	4	3	0	7
d	0	0	1	1
e	2	2	3	7
f	3	2	0	5
g	0	1	0	1
h	6	3	10	19
i	6	4	3	13
j	0	0	0	0
k	1	1	0	2
l	0	0	0	0
m	4	4	1	9

West Middlesex Hospital

Uterus Bicornis Bicollis - Septate Vagina.

Forceps Delivery.

Postpartum Haemorrhage.

WM,1951

Mrs.Jean Weir, aged 23.

Type 1 Menstrual Type K 11 3/27 regular, normal loss, painless.

a - 1
b - 0
c - 1
d - 0
e - 0
f - 1
g - 0
h - 0
i - 2
j - 0
k - 0
l - 0
m - 0

Laceration vaginal septum on first coitus with profuse haemorrhage requiring vaginal packing when abnormality first noted.

Para 1 Gravida 1

Low forceps delivery for foetal distress of living male infant weighing 7 lb. 7 oz. Post partum haemorrhage of 30 oz. after delivery placenta.

Pregnancy in right horn.

Uterus bicornis bicollis. Trans Vaginal diaphragm.

Haematocolpos.

Forceps.

WM 2 1951.

Mrs.Mary Clelland, aged 21.

Type 1

a - 1
b - 0
c - 1
d - 0
e - 0
f - 1
g - 0
h - 0
i - 1
J - 0
k - 0
l - 0
m - 1

Menstrual Type K 18 6/26 loss heavy, regular, moderate dysmenorrhoea. Admitted to hospital aged 18 for urinary retention - found to have a haematocolpos, occluding membrane excised and vagina drained.

Para 1 Gravida 1.

Pregnancy in right horn. Vaginal examination at antenatal clinic revealed a transverse diaphragm at junction upper $\frac{1}{3}$ and lower $\frac{2}{3}$ vagina. Diaphragm had a central aperture which admitted two fingers. Above this two cervices were felt, the right in continuity with the pregnant horn - the non pregnant horn being the size of a normal uterus.

Premature labour at 35 weeks gestation. Transverse diaphragm tightly stretched over vertex preventing its descent. Diaphragm incised and a female infant weighing 5 lb. 9 oz. delivered by forceps.

Uterus bicornis bicollis - septate vagina

Congenital absence left kidney

Antepartum haemorrhage

Breech delivery.

WM3 1953

Mrs.Cora Manson, aged 29

Type 1

a - 2
b - 1
c - 2
d - 0
e - 1
f - 1
g - 0
h - 4
i - 1
j - 0
k - 1
l - 0
m - 2

Menstrual Type - K 12 cycle 6/26 regular, normal loss, moderate dysmenorrhoea.

Para 1 Gravida 3.

1948 - Spontaneous miscarriage at ten weeks gestation. Anomaly discovered at curettage.

1948 - Unstable presentation. Spontaneous delivery of a female infant weighing 6 lb. 2 oz. Third stage normal. Pyelitis in puerperium. Investigation urinary tract revealed absence left kidney.

1953 - Pregnancy in right horn. Breech presentation, failed external cephalic version. Antepartum haemorrhage (unclassified) assisted breech delivery of male infant weighing 8 lb. Third stage normal.

Uterus Bicornis Bicollis - Septate Vagina

Repeated miscarriages following normal delivery.

WM 4 - 1951

Type 1 Mrs.Amy Cooke, aged 30.

a - 1 Menstrual Type K 14 cycle 5/30 regular, normal loss, painless.
b - 2
c - 0 Para 1 Gravida 3.
d - 0
e - 1 1948 - Spontaneous delivery after division vaginal
f - 0
g - 0 septum (when anomaly first noted) of a female
h - 2
i - 1 infant weighing 5 lb. 1 oz. Third stage normal.
j - 0
k - 0 1949 - Spontaneous miscarriage at 10 weeks pregnant.
l - 0
m - 1 1951 - Spontaneous miscarriage at 8 weeks pregnant.

Uterus Bicornis Unicollis

Normal delivery at term

Previous Miscarriage and Premature Breech Delivery.

WM 5 1954

Mrs.Eileen Woods, aged 31.

Type 111

a - 2
b - 1
c - 1
d - 0
e - 1
f - 1
g - 0
h - 2
i - 1
j - 0
k - 0
l - 0
m - 1

Menstrual Type K 11 Cycle 3-6/21-30 irregular, loss heavy, painless.

Para 2 Gravida 3.

1951 - Persistent breech presentation, failed version, premature labour at 32nd week. Assisted breech delivery of a male infant weighing 4 lb. 5 oz. Third stage normal.

1952 - Spontaneous miscarriage at twenty weeks gestation. Hysterosalpingogram revealed bicornuate uterus.

1954 - Normal delivery at term of a female infant weighing 6 lb. 9 oz. Third stage normal.

Uterus Bicornis Unicollis

Spontaneous delivery

Post partum Haemorrhage

Manual Removal Placenta

WM 6 1951

Mrs.Eve Dakes, aged 19 years.

Type 111 Menstrual Type K 13 Cycle 4/27 regular, loss normal, moderate dysmenorrhoea.

a - 1
b - 0 Para 1 Gravida 1.
c - 1
d - 0 Breech presentation - turned successfully at 34th week
e - 1
f - 0 to a vertex presentation.
g - 0
h - 1 Spontaneous delivery of a female infant weighing
i - 1
j - 0 7 lb. 6 oz.
k - 1
l - 0 40 oz. post partum haemorrhage. Manual removal of
m - 2
placenta from right horn when anomaly discovered.

Velamentous insertion cord.

Uterus Bicornis Uno Latere (closed)

Rupture Gravid Rudimentary Horn

Hemihysterectomy

Normal Pregnancy. Caesarean Section.

WM 7 1954

Mrs.Joan Wallis, aged 25.

Type 111

a - 1
b - 1
c - 1
d - 0
e - 0
f - 1
g - 1
h - 1
i - 0
j - 0
k - 0
l - 0
m - 1

Menstrual Type K 12 Cycle 4/28, regular, moderate loss, moderate dysmenorrhoea.

Para 1 Gravida 2.

1951 - Rupture gravid left rudimentary horn at 16th week pregnancy. Haematoperitoneum and collapse. Hemihysterectomy performed with removal rudimentary horn. Pin-hole communication with vagina. Decidual cast passed on 4th day post-operatively.

1954 - Normal pregnancy - male infant weighing 8 lb. 4 oz. delivered by lower Segment Caesarean Section in view of past obstetric history.

Uterus Septus

Premature Labour

Repeated Miscarriages

WM 8 1953

Mrs. Adele Corrigan, aged 26.

Type V

a - 1
b - 2
c - 0
d - 0
e - 1
f - 0
g - 0
h - 2
i - 1
j - 0
k - 0
l - 0
m - 0

Menstrual Type K 11 Cycle 5-8/30-35 irregular, loss heavy, moderate dysmenorrhoea.

Para 1 Gravida 3

1947 - Premature spontaneous labour at 35th week pregnancy with delivery of a female infant weighing 4 lb. Third stage normal.

1951 - Spontaneous 10 week miscarriage. Hysterosalpingogram.

1953 - Spontaneous 12 week miscarriage.

Uterus Septus

Repeated Miscarriages

WM 9 1953

Mrs.Joan Stevenson, aged 23 years

Type V

a - 0
b - 2
c - 0
d - 0
e - 0
f - 0
g - 0
h - 2
i - 0
j - 0
k - 0
l - 0
m - 0

Menstrual Type - K 14 Cycle 6/28 regular, normal loss, painless.

Para 0 Gravida 2

1952 - spontaneous miscarriage at ten weeks pregnant.

1953 - Spontaneous miscarriage at twelve weeks pregnant

Hysterosalpingogram revealed septate uterus.

Uterus Septus

Repeated Unsuccessful Pregnancies

Metroplastic operation

Premature Labour, Accidental Haemorrhage

Stillbirth

Premature Labour, Antepartum Haemorrhage Type 11

Placenta Praevia. Live Birth.

WM 10 1950

Mrs.Martha Scott, aged 31

Type V — Menstrual Type K 15 Cycle 6-8/28 regular, normal loss, moderate dysmenorrhoea.

a - 2
b - 4 — Para 2 Gravida 6
c - 0
d - 1 — 1941 - miscarriage at 20 weeks gestation.
e - 2
f - 0 — 1943 - miscarriage at 22 weeks gestation.
g - 0
h - 2 — 1945 - Miscarriage at 20 weeks gestation.
i - 2
j - 0 — Hysterosalpingogram revealing septate uterus.
k - 0
l - 0 — 1947 - Miscarriage at 22 weeks.
m - 1

1948 - Laparotomy with excision of septum which extended from fundus to internal os.
Right side uterus better developed than left.

1949 - Became pregnant 5 months after metroplastic operation. Accidental antepartum haemorrhage, premature labour at 30 weeks gestation.
Male stillborn infant weighing 3 lb. 2 oz.
Third stage normal.

WM 10 1950/continued

1950 - Premature labour at 34th week pregnancy. Antepartum haemorrhage due to Type 11 placenta praevia - normal delivery of female child weighing 4 lb. 13 oz. Third stage normal. Manual exploration uterus - small tag of septum felt in region fundus.

St.George's Hospital

	1	111	V	V11	Total
	3	2	3	1	9
a	3	3	1	1	8
b	0	1	2	0	3
c	3	1	1	1	6
d	0	0	0	1	1
e	0	0	0	0	0
f	3	3	1	1	8
g	2	0	1	0	3
h	2	4	3	1	10
i	2	5	1	0	8
j	0	0	0	0	0
k	3	0	0	0	3
l	0	0	0	1	1
m	3	1	3	2	9

Uterus Unicornis

Persistent Breech Presentation

Anencephalic

SG 1 1953
8816

Mrs.Jean Wells, aged 32.

Type V11

a - 1
b - 0
c - 1
d - 1
e - 0
f - 1
g - 0
h - 1
i - 0
j - 0
k - 0
l - 1
m - 2

Menstrual Type K 13 7/24-26 regular, loss heavy, moderate dysmenorrhoea.

Abnormality first noted at laparotomy for appendicitis in 1939 - left ovary and Fallopian tube absent - no rudimentary horn seen on left side.

Gravida 1 Para 1.

Became pregnant six months after marriage in August 1953. Persistent breech presentation - external cephalic version unsuccessful. Breech delivery at term of a female anencephalic infant weighing 6 lb. No hydramnios noted. Third stage normal. Investigation urinary tract normal. Hysterosalpingogram.

Uterus Bicornis Unicollis

Repeated Breech deliveries

Prematurity.

SG 2 19
40439

Mrs.Gertrude Turner, aged 41

Type

a - 3
b - 0
c - 1
d - 0
e - 0
f - 3
g - 0
h - 3
i - 3
j - 0
k - 0
l - 0
m - 0

Menstrual Type K 12 Cycle 3/26 regular, normal loss, no dysmenorrhoea.

Para 3 Gravida 3

Premature breech delivery at 35th week female infant weighing 4 lb. 6 oz. Third stage normal.

Premature breech delivery at 34th week. Third stage normal. Male infant weighing $3\frac{1}{2}$ lb.

Breech delivery at term of a male infant weighing 8 lb. 4 oz. Third stage normal.

Uterus Bicornis Unicollis

Miscarriage.

SG 3
47523

Mrs.Nicole Prior, aged 22

Type 111

a - 0
b - 1
c - 0
d - 0
e - 0
f - 0
g - 0
h - 1
i - 0
j - 0
k - 0
l - 0
m - 1

Menstrual Type K 11 Cycle 5-6/28 regular, normal loss, severe dysmenorrhoea.

Para 0 Gravida

Admitted to hospital as incomplete miscarriage - two months amenorrhoea followed by heavy bleeding per vaginam. Two days after admission passed a decidual cast from non-pregnant horn and on the following day a further cast containing chorionic tissue. Abnormality discovered during digital curettage for removal retained products conception.

Uterus Unicorpus septus Bicollis Septate Vagina

Persistent Breech Presentation

Caesarean Section

SG 4
47768

Mrs. Gladys Morrison, aged 24 years.

Type 1

a - 1
b - 0
c - 1
d - 0
e - 0
f - 1
g - 1
h - 1
i - 0
j - 0
k - 1
l - 0
m - 1

Menstrual Type K 12 Cycle 3/28 regular, normal loss, moderate dysmenorrhoea.

Para 1 Gravida 1

Abnormality first noted on vaginal examination at antenatal clinic when two cervices separated by vaginal septum were felt.

Persistent breech presentation, external cephalic version attempted but unsuccessful.

Classical Caesarean Section at term. Longitudinal sulcus noted running from fundus to supravaginal cervix, dividing uterus into a large half on the right and a much smaller half on the left. Male foetus weighing 6 lb. 7 oz. delivered from right horn. Uneventful post-operative period.

Investigation urinary tract normal.

Uterus Bicornis Bicollis Vagina Septate

Persistent Breech Presentation

Lower Segment Caesarean Section

SG 5
96751

Mrs.Morris Potter, aged 19 years.

Type 1

a - 1
b - 0
c - 1
d - 0
e - 0
f - 1
g - 1
h - 1
i - 0
j - 0
k - 1
l - 0
m - 1

Menstrual Type K 12 Cycle 6/27 regular, loss normal, moderate dysmenorrhoea.

Para 1 Gravida 1

Two cervices felt on vaginal examination at antenatal clinic. Persistent breech presentation - external cephalic version unsuccessful. Minor degree pelvic contraction present on X-ray pelvimetry. Lower segment Caesarean Section performed - living male infant delivered from right horn after some difficulty in extracting head. Birth weight 7 lb. 11 oz. Notifiable pyrexia in puerperium. No investigation urinary tract.

Uterus Bicornis Bicollis septate vagina

Forceps delivery

Manual Removal Placenta.

SG 6
87946

Mrs.Phyllis Wilks, aged 29.

Type 1

a - 1
b - 0
c - 1
d - 0
e - 0
f - 1
g - 0
h - 0
i - 2
j - 0
k - 1
l - 0
m - 1

Menstrual Type K 13 Cycle 5/27 loss normal, no dysmenorrhoea.

Para 1 Gravida 1

Low forceps delivery for foetal distress of a living male infant at term weighing 6 lb.

Manual removal of a morbidly adherent placenta from right horn when abnormality first discovered.

Investigation of urinary tract.

Uterus Subseptus

Persistent Breech Presentation

Lower Segment Caesarean Section

SG 7
87394

Mrs.Gladys Wells, aged 24 years.

Type V

a - 1
b - 0
c - 1
d - 0
e - 0
f - 1
g - 1
h - 1
i - 1
j - 0
k - 0
l - 0
m - 1

Menstrual Type K 14 Cycle 3/30 loss heavy, severe dysmenorrhoea.

Para 1 Gravida 1

Persistent breech presentation, external cephalic version unsuccessful. As pelvis appeared adequate patient allowed to go into normal labour. Foetal distress occurred shortly after membranes ruptured at 2/5ths dilated. Female infant weighing 8 lb. 2 oz. delivered. During the operation a septum was felt extending from fundus for a $\frac{1}{3}$ of the way to the internal os. Infant delivered from the right side. Post-operative recovery uneventful.

Urinary tract not investigated.

Uterus Subseptus

Miscarriage

SG 8
38933

Mrs.Diana Stockton, aged 30 years.

Type V

a - 0
b - 1
c - 0
d - 0
e - 0
f - 0
g - 0
h - 1
i - 0
j - 0
k - 0
l - 0
m - 1

Menstrual Type K 11 Cycle 6/24 loss heavy, moderate dysmenorrhoea.

Para 0 Gravida 1

Admitted as incomplete miscarriage. Abnormality discovered during digital curettage for retained products conception when central septum felt extending from fundus to within 1" of internal os.

Uterus septus

Miscarriage

SG 9
121265

Mrs.R.Christie, aged 23 years.

Type V

a - 0
b - 1
c - 0
d - 0
e - 0
f - 0
g - 0
h - 1
i - 0
j - 0
k - 0
l - 0
m - 1

Menstrual Type K 12 Cycle 6/28 regular, loss normal, severe dysmenorrhoea.

Admitted to hospital with incomplete miscarriage. Abnormality discovered at removal retained products conception by digital curettage. Central septum extending down to internal os felt.

Repeated Normal Pregnancies in Association With a Uterus Bicornis Bicollis

S.M.1.

M.B., aged 35 years.

Type	
a - 3	Menstrual Type K 13 Cycle 5/28 regular, normal loss, no dysmenorrhoea.
b - 1	Married 14 years, gravida 4.
c - 2	
d - 0	Para 2, both spontaneous deliveries at term, labour
e - 3	
f - 0	and puerperium normal.
g - 0	
h - 5	One spontaneous miscarriage at the third month,
i - 1	
	cause unknown.
	Slight vaginal bleeding in all pregnancies which
	ceased at the third month.

Present Pregnancy

First seen at the antenatal clinic with a history of 12 weeks amenorrhoea, when a vaginal examination revealed the remains of a vaginal septum on the upper third of the anterior vaginal wall. Two softened cervices were palpable on either side of the septum, and a uterus enlarged to the size of a 12 weeks pregnancy was felt to be in continuity with the right cervix, with a small uterine body attached to the left cervix.

SM 1/continued

Pregnancy proceeded normally and the patient was admitted to hospital two days before term in early labour, when a pelvic examination was performed to exclude the possibility of the non-pregnant horn having prolapsed.

Labour proceeded normally and at the end of 5 hours 35 minutes a healthy male infant weighing $5\frac{1}{2}$ lb. was delivered. Third stage of labour and puerperium uneventful.

Intravenous pyelography showed a normal urinary tract without any malformation. Vaginal examination before discharge confirmed the diagnosis of a uterus bicornis bicorpus bicollis (pseudodidelphys).

Unfortunately the patient failed to return to the clinic for a hysterosalpingogram to be performed.

Commentary

This case is an illustration of the aphorism, "That with the more severe forms of congenital uterine malformation, pregnancy and labour usually approach normal." Here the patient had passed through three pregnancies without any difficulty arising apart from slight bleeding per vaginam in the early weeks of pregnancy. This bleeding was probably from the non-gravid horn and is common in this type of malformation although difficult to prove without speculum examination.

Vaginal Septum as Cause of Infertility.

Premature Labour and Division of the Septum

In a Uterus Bicornis Bicorpus Bicollis

with Septate Vagina.

SM 2

Mrs.H.R., aged 26 years. Married three years.

Type 1 Menstrual Type 8/28 days, regular, loss heavy, severe premenstrual dysmenorrhoea.

a - 1
b - 0
c - 1
d - 0
e - 1
f - 0
g - 0
h - 1

The patient was first seen at the gynaecological clinic, having been referred there by her private practitioner, with a complaint of infertility. She had been married three years and had not taken any precautions against conception occurring. Intercourse was regular and satisfactory to both parties.

On vaginal examination the vulva appeared normal but there was a fleshy median sagittal vaginal septum, extending from the vault for 2/3rds the length of the vagina, and in apposition to the right lateral wall of the vagina. Two cervices were felt separated by the vaginal septum and on bimanual examination two uterine bodies could be made out.

SM 2/continued

Although there was a double vagina, the one to the right of the midline was only a potential space, but its walls could be easily separated digitally. Accordingly the patient and her husband were instructed to attempt intercourse in this vagina.

Pregnancy and Labour

The patient was seen again five months later when she gave a history of twelve weeks amenorrhoea.

Vaginal Examination

The right vagina was now almost as capacious as the left. Both cervices were softened and the uterine body in continuity with the right cervix was enlarged consistent with a twelve weeks cyesis. Arrangements were made for routine antenatal care and hospital confinement. There was never any bleeding from the non-pregnant horn and the pregnancy proceeded normally until the 34th week when the patient commenced in labour.

Examination in Labour

The vertex was deeply engaged, with the position R.O.L. No foetal or maternal distress, membranes intact, regular strong uterine contractions. On pelvic examination the septum was pushed over to the left, right cervix, os 3/5ths dilated, taken up, foetal head midcavity.

SM 2/continued

The left cervix was conical, small and was felt high up and to the left of the head. The os admitted a finger tip and the uterine body on this side was not defined.

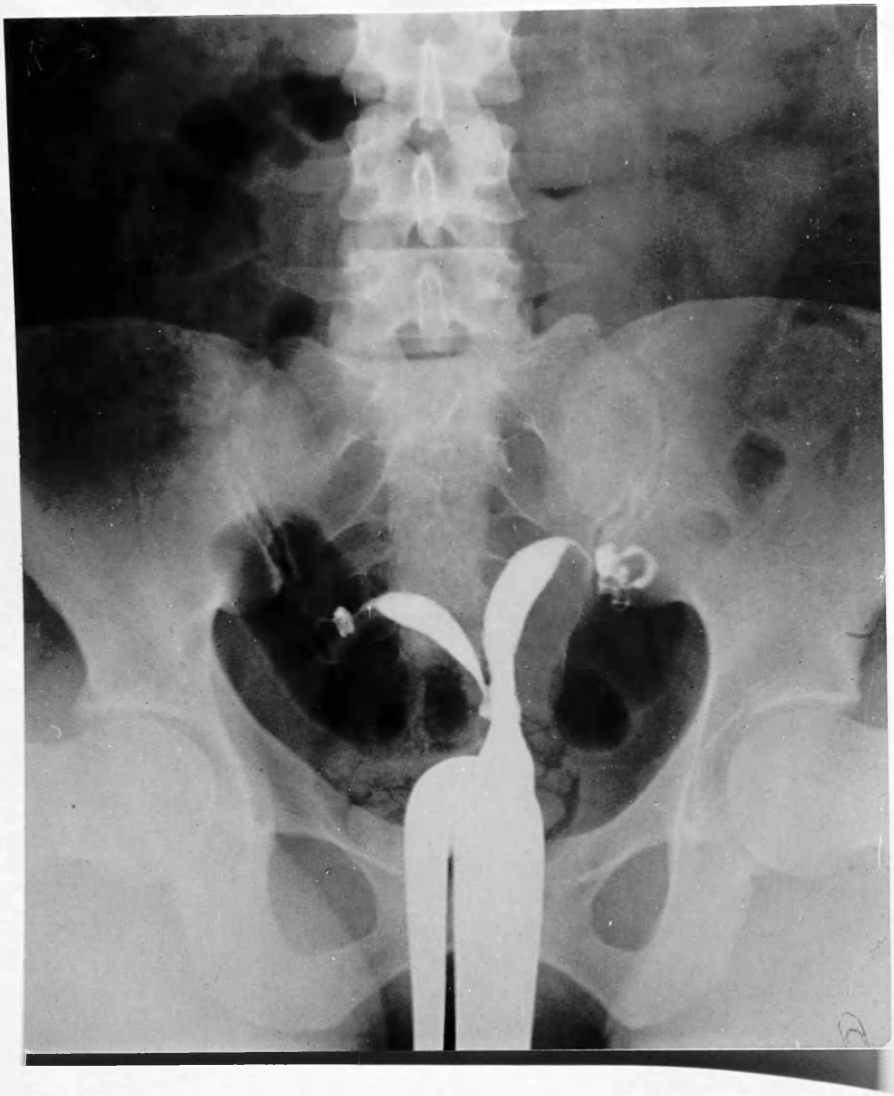
Labour progressed normally and as the head was distending the perineum, the vaginal septum was noted to be tightly stretched over the left half. The septum was then divided between artery forceps and this was quickly followed by the birth of the head followed by the rest of the child, a female weighing 7 lb. Following the third stage of labour, which was normal, the artery forceps were removed from the septum, without any haemorrhage other than a slight ooze.

Total duration of labour - 17 hours 10 minutes.

Second stage - 1 hour 6 minutes

Third stage - 45 minutes

Both mother and child were discharged well from hospital on the 10th day of the puerperium. Pelvic examination at this stage found the vagina to be single with the remains of the septum, complete only in the region of the cervices. The right uterine body was bulky and larger than the left which in size could be compared with a six weeks pregnancy.



22. Uterus pseudodidelphys.

Hysterogram from Case SM 2.

The recently gravid left horn larger than the right.

SM 2/continued

The right os was patulous and the left closed.

Lochia had not been abnormal in quantity.

Later investigation of the urinary system was normal.

Commentary

This case shows that mechanical difficulties may be the cause of infertility in these cases, and here, following the adoption of the other vagina conception occurred.

It was felt at the time that the vaginal septum would not have delayed the birth of the head and its division between forceps was to prevent it being torn with subsequent severe haemorrhage.

Normal Pregnancy and Labour Associated

with a Bicornuate Uterus.

SM 3

Mrs.D.E., aged 28 years

Type 111 Menstrual Cycle 5/28 regular, premenstrual dysmenorrhoea.

a - 1 Married 6 years. Involuntary infertility.
b - 0
c - 1 Attended infertility clinic and routine hystero-
d - 0
e - 1 salpingogram showed a uterus bicornis unicollis.
f - 0
g - 0 Attended antenatal clinic on 23rd July,1951.
h - 0
i - Vaginal examination revealed the right horn to be
j -
k - enlarged consistent with a ten week pregnancy -
l -
m - small left horn.

Pregnancy and labour were normal.

No bleeding from non-gravid half uterus.

Normal delivery of a girl, 7 lb. 8 oz.

Puerperium normal apart from a breast abscess.

No investigation of urinary tract.

Repeated Miscarriages in Association

with a Bicornuate Uterus

SM 4

Mrs.A.L., married 6 years.

Type 111 Menstrual Type K 12 Cycle 3-5/31 days, regular, always moderate dysmenorrhoea.

a - 0
b - 3
c - 0
d - 0
e -
f -
g -
h -
i -
j -
k -
l -
m -1

Obstetric History - Gravida 3 Para 0

Two spontaneous miscarriages at the tenth and twelfth week pregnancy respectively.

Present Pregnancy

The patient was first seen when she attended the antenatal clinic with a history of eight weeks amenorrhoea. Vaginal examination revealed a softened cervix and uterus enlarged consistent with an eight weeks cyesis.

In view of the previous miscarriages arrangements were made for the patient to be confined in hospital, but two weeks afterwards she was admitted to hospital with a history of slight painless bleeding per vaginam of 36 hours duration. In spite of rest and sedation the threatened miscarriage became inevitable and a fresh foetus with sac were passed 48 hours after admission.

SM 4/continued

As fairly heavy bleeding per vaginam continued, retention of some products of conception was suspected and evacuation of the uterus under anaesthetic decided upon.

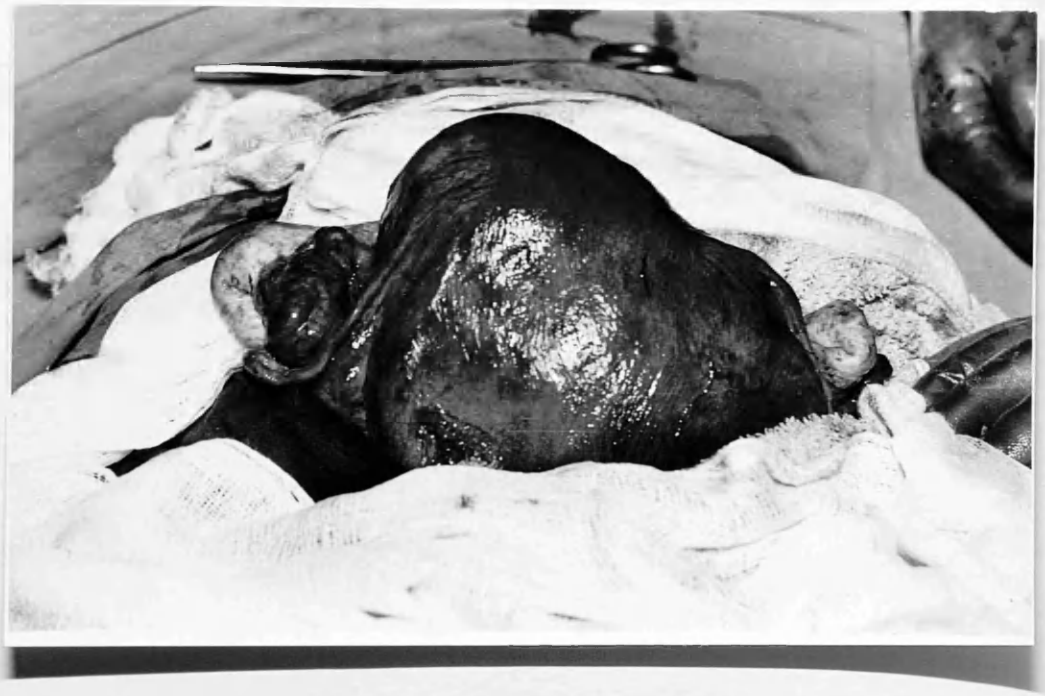
Examination under anaesthetic and removal retained products conception - vagina normal. Cervix single, os patulous and admits one finger. Bimanually a bicornuate uterus was felt, the left horn being much larger than the right. A sound was passed 4" into the left horn and 2" into the right; there was no communication between the two sides. A piece of placental tissue could be felt in the left horn and this was separated by digital curettage and removed by ovum forceps. Investigation of the urinary tract at a later stage showed this to be normal.

Commentary

Repeated miscarriages are commonly associated with these abnormalities and as previously stated hysterosalpingography is advised in any patient with this type of history.

SM 4/continued

Three more cases of miscarriage in conjunction with uterine malformation were encountered but here they were first pregnancies and therefore not possible to say whether the malformation caused the miscarriage or not. One of these cases had a bicornuate uterus and the other two of the subseptate type; all were discovered during removal of retained products of conception.



23. Subseptate Uterus.

Retracted uterus following Lower Segment Caesarean Section, fundal depression not well shown - SM 5.

Repeated Breech Presentations and Retained Placentae in Association with a Uterus Bicornis Unicorpus Unicollis Subseptus.

SM 5

Mrs.H.P.S., aged 29 years. Married 8 years.

Type V

a - 4
b - 0
c - 3
d - 2
e - 0
f - 5
g - 2
h - 8
i - 6
j - 0
k - 3
l - 1
m - 3

Menstrual Type K13 Cycle 5/35 regular, normal loss, moderate pre-menstrual dysmenorrhoea.

Obstetric History

Gravida 4 Para 3.

All three infants were delivered as breech presentations, and the placentae were retained in each pregnancy requiring manual removal. The first child, a girl weighing 3 lb. 14 oz. was delivered at the 34th week and died from a cerebral haemorrhage 10 minutes after birth. No cause found for the cerebral haemorrhage.

A live male infant was delivered at term in the second confinement after a difficult breech extraction of a flexed breech. The child weighed 8 lb. 2 oz. and had bilateral talipes deformity.

A third pregnancy was complicated by an ante-partum haemorrhage at the 36th week of gestation,

SM 5/continued

and this was found to be due to a type 11 placenta praevia. The flexed breech presented, and although the haemorrhage was controlled by bringing down a leg, the child, a female weighing 8 lb. was stillborn. Again the placenta had to be removed manually.

Unfortunately no postmortem examination was performed on the two female infants so it was not possible to determine whether any uterine malformation was present.

Present Pregnancy

The patient first attended the anatenatal clinic at the 28th week of pregnancy. In view of the previous obstetric history the presence of a uterine malformation was suspected. On abdominal examination the fundus of the uterus was noted to be unusually broad, assymetrical with a marked cleft in the middle; the breech was presenting. On pelvic examination the pelvis appeared to be adequate and only one cervix was felt. At the thirty second and fourth weeks attempts to turn the breech were unsuccessful, version as far as transverse presentation only being possible.

SM 5/continued

In view of the bad obstetric history, although there was no apparent pelvic disproportion, elective Caesarean section was decided upon.

The patient went into labour spontaneously one week before term, and a Lower Segment Caesarean Section was then performed, and a live female infant delivered. The fundus of the uterus was noted to be deeply notched and with a median septum running downwards to within two inches of the internal os from the bottom of the notch. The septum was noted after the placenta, which was morbidly adherent, had been removed manually. After the uterine incision had been closed and the uterus had retracted firmly, the two cornua separated by a deep notch were much more apparent. Both kidneys felt normal on palpation.

Puerperium was uneventful and during this period the patient volunteered the information that she had two sisters, who between them had three children, and whose pregnancies and labours had been normal.

Similar Uterine Malformation in Two Sisters

With Android Features.

(Accepted for publication by the
British Medical Journal, May 1953)

SM 6

Mrs.E.M., aged 29 years.

Type 111 Menstrual Type K 18 Cycle 10-12/50-60 days.

a - 2
b - 0
c - 2
d - 0
e - 0
f - 2
g - 2
h - 2
i -
j - 0
k - 0
l - 0
m - 0

Married 5 years. Involuntary infertility.

Attended the gynaecological Out-Patients with a history of 4 months amenorrhoea and morning sickness.

On Examination Well marked hirsutism face and limbs, male distribution of pubic hair. Height 5' 2". Blood pressure 140/90 mm. Voice normal feminine timbre, moderate hypertrophy of clitoris. Breasts well developed with prominent Montgomeries tubercles. Vaginal examination revealed the uterus enlarged consistent with a ten weeks cyesis.

The patient was referred to the antenatal clinic where arrangements were made for her confinement in hospital.

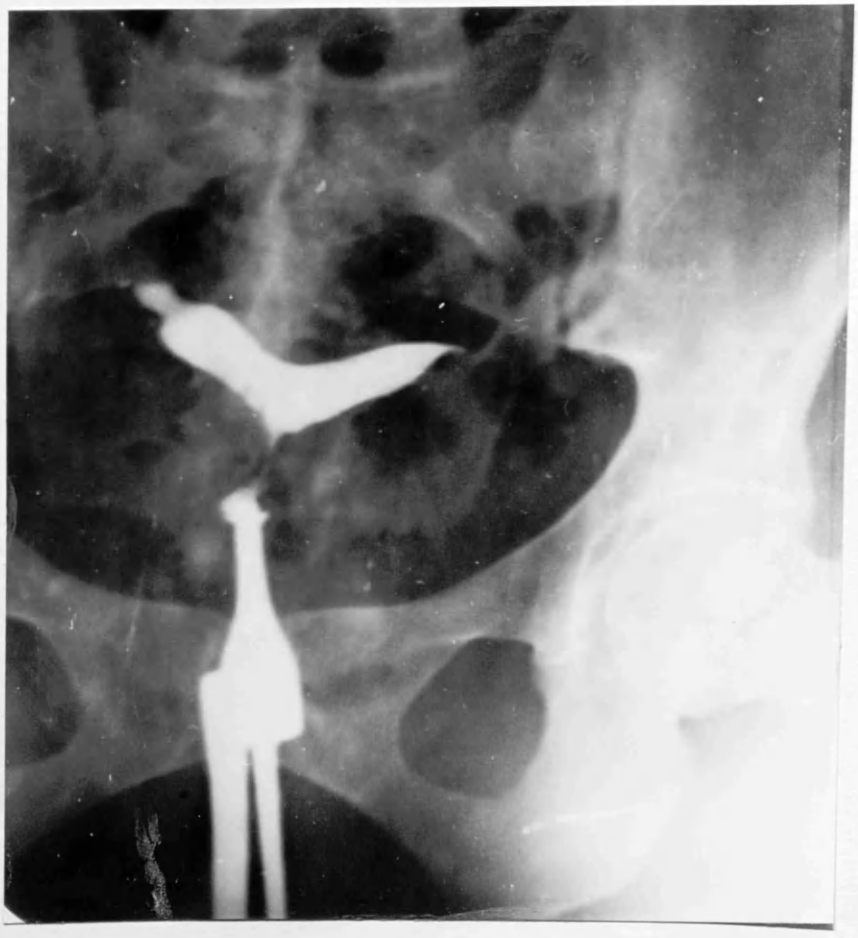
SM 6/continued

At subsequent antenatal visits the enlarging uterus was noted to be assymetrical with a diffuse swelling to the right which could not be felt apart from the body of the uterus.

At the 35th week of pregnancy the patient was admitted to hospital with signs of mild pre-eclamptic toxaemia and unstable presentation. As there was no cephalopelvic disproportion a uterine malformation was suspected.

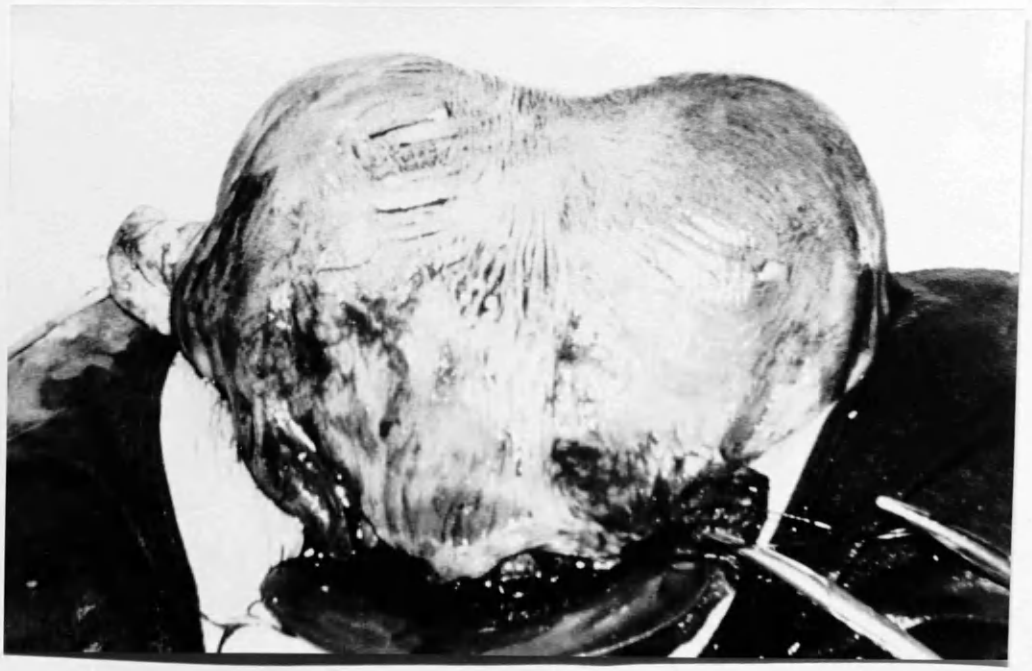
In spite of rest and sedation the toxaemia worsened and because of this and the unstable presentation it was decided to deliver the child by Caesarean section. At operation a bicornuate uterus was found with the pregnancy contained in the left horn. Both ovaries appeared normal. A healthy male child weighing 6 lb. 2 oz. was delivered and the patient made an uninterruped convalescence being discharged from hospital on the seventeenth day of the puerperium with the child fully breast fed.

A further investigation of the patient was undertaken three months after delivery.



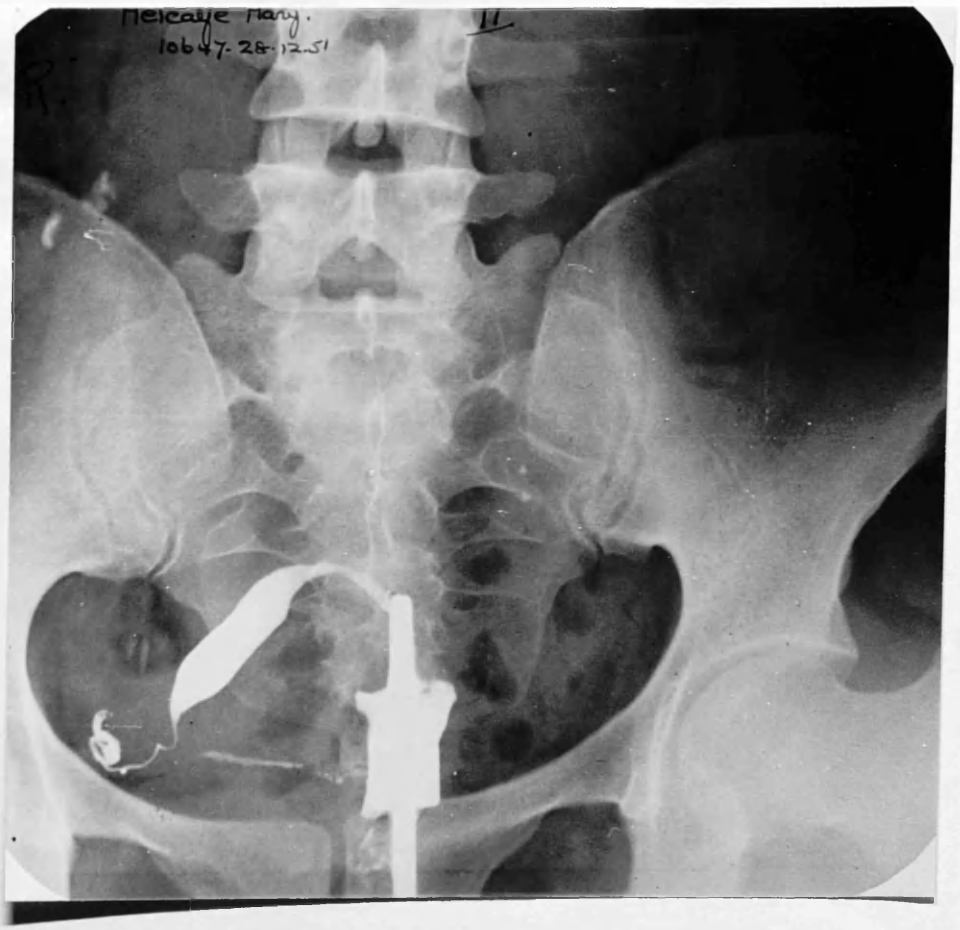
24. Bicornuate Uterus.

Appearance more suggestive of an arcuate uterus in this hysterogram. SM 6.



25. Bicornuate Uterus.

The uterus of the same patient SM 6 immediately following Lower Segment Caesarean Section.



26. Bicornuate Uterus

Hysterogram - SM 7

Only the right horn is outlined
simulating uterus unicornis.
This patient is the sister of SM 6.

SM 6/Continued

1954 Patient attended antenatal clinic with a history of 8 weeks amenorrhoea and on pelvic examination the right horn of the uterus was found to be enlarged consistent with the period of amenorrhoea, while small mass of harder consistency was felt to the left and probably represented the non-pregnant horn.

The presentation was persistently breech despite attempts to correct it. The blood pressure which had been 140/85 mm.Hg. at the first antenatal attendance commenced to rise in the third trimester, and at the 38th week was 155/105 mm.Hg. when she was admitted to hospital for rest. In view of her previous history and because the blood pressure remained high, delivery was effected by Lower Segment Caesarean Section. A healthy female infant weighing 7 lb. 5 oz. was delivered, and in whom no genital abnormality was noted.

Convalescence was uneventful.

SM 7

Mrs. Mary Metcalfe, aged 35.

Type

L.M.P. 9th March, 1954.
E.D.D. 16th December, 1954.

Progesterone mg.150 implanted left thigh on 2nd April, 1954. Admitted as threatened miscarriage on 27th May, 1954 with history of slight painless red loss for two days which continued intermittently until the 20th June when it ceased completely and patient was discharged.

At this time the uterus was enlarged consistent with an 18 week pregnancy although it was difficult to be certain of the fundal height, owing to the asymmetrical outline of the uterus but the patient was now aware of foetal movements.

Further antenatal attendance was normal apart from the presentation which was a breech, but in view of the patient's obstetric history it had been decided to effect delivery by Caesarean section and therefore no attempt was made to correct the malpresentation.

SM 7 /continued

At the 39th week the patient was admitted to hospital and delivery effected by Lower Segment Caesarean Section. It was noted that the cleft between the two horns had to a great extent disappeared and that the fundus showed only a slight depression at the junction of the two uteri, although the right horn in which the pregnancy was contained was much larger than the left non-pregnant honr. The placenta and membranes which were attached to the postero-lateral wall were easily expressed and no communication could be felt between the two horns. After closure of the uterine incision when the uterus was firmly retracted, it was observed that the bicornuate nature of the uterus was again more apparent. Both kidneys were palpated before closure of the abdomen, and felt normal. The infant, a female weighing 6 lb. 13 oz. cried at birth, and examination of the vulval area failed to reveal any genital abnormality. The puerperium was uneventful, mother and infant being discharged on the 17th day.

SM 7

Mrs.M.N., aged 31 years.

Type 111 Menstrual Type K 17 Cycle 6-8/30-35 days.

a - 1
b - 2
c - 1
d - 0
e - 0
f - 1
g - 1
h - 2
i - 0
j - 0
k - 0
l - 0
m - 2

Married 9 years. Gravida 2, both pregnancies having terminated spontaneously at the 10th week.

On examination - Hirsutism present. External genitalia normal.

17-ketosteroids 8 mg./24 hours.

Hysterosalpingogram showed a bicornuate uterus with non-patency of the left fallopian tube.

Shoulder Presentation in Association With a Uterus Cordiformis Subseptus

SM 8

Mrs.L.M., aged 27 years, married 2 years.

Type V

a - 1
b - 0
c - 1
d - 0
e - 0
f - 1
g - 0
h - 2
i - 1
j - 0
k -
l -
m - 1

Menstrual Type K 16 Cycle 6-7/32 days, regular, moderate loss, severe premenstrual dysmenorrhoea.

Obstetric History Gravida 2 para 0

Spontaneous miscarriage at ten weeks.

Present Pregnancy Since the 4th month of pregnancy the patient had been attended by the district midwife, who stated that the pregnancy had proceeded normally and that the vertex had become engaged in the pelvis at the 37th week.

The membranes had ruptured spontaneously at the 39th week and 20 hours later uterine contractions had commenced. The midwife noticed that the foetal heart had become irregular and on vaginal examination she felt a hand presenting; this was some 36 hours after the membranes had ruptured.

SM 8/continued

On admission to hospital strong uterine contractions present, foetal heart not heard. Vaginal examination revealed the cervix to be $\frac{3}{4}$ dilated, the shoulder presenting, and the right arm and cord prolapsed into the vagina; the cord was not pulsating.

Under general anaesthesia internal version was easily performed and a stillborn male child weighing 6 lb. 1 oz. delivered. During this operation a depression of the uterine fundus could be made out and in view of this the placenta was removed manually and a partial uterine septum was then felt, to which the placenta was morbidly adherent. The septum did not completely divide the uterine cavity but reached to within 2" of the internal os.

The patient made an uninterrupted convalescence and was discharged from hospital on the 12th day of the puerperium. Three months later the pelvis was shown to be normal on X-ray pelvimetry and hysterosalpingogram revealed a well marked cordiform uterus.

Commentary

Here again a minor uterine deformity was the cause of a major obstetrical abnormality. As the question of pelvic disproportion was ruled out by X-ray pelvimetry, it is certain that the oblique lie was due to the septate uterus. Doubt must be raised by the midwife's assertion that the vertex was engaged before the onset of labour.

Antepartum Haemorrhage in Association

With a Uterus Bicornis Bicorpus Unicollis.

SM 9

Mrs.R.G., aged 37 years, married 15 years.

Type 111

a - 1
b - 0
c - 1
d - 0
e - 0
g - 1
g - 0
h - 2
i - 1
j - 0
k -
l -
m - 1

Menstrual Type K 14 Cycle 3-5/28 days, regular. Loss always heavy, moderate premenstrual dysmenorrhoea.

Present Pregnancy - The patient was referred for admission to hospital on account of severe vomiting of 12 days duration. A history of 7 weeks amenorrhoea was given by the patient who stated that she could not be pregnant as she had been informed 12 years ago, following an infertility investigation, that pregnancy was impossible.

On examination there were signs of moderate dehydration with a marked ketonuria, but no bile or albumen in the urine. Vaginal examination revealed uterus enlarged consistent with a 6 weeks cyesis; later pregnancy was confirmed by a male toad test. The vomiting was treated by rest, sedation and fluids only by mouth, but it was not until fluids by mouth had been stopped and intravenous glucose saline given that the condition ameliorated and oral feeding recommenced. After 10 days in hospital the patient was discharged well with instructions to attend the antenatal clinic.

SM 9/continued

Information from the hospital where the patient had been investigated stated that she had had a D & C and insufflation of the tubes, when a bicornuate uterus had been found with non patency of the tubes. In view of this knowledge a further vaginal examination was made when a small mass could be discerned to the right of the pregnant uterus. Apart from saying that the mass was attached to the uterus, no definite diagnosis of a bicornuate uterus could be given.

Three weeks before term the patient was admitted to hospital with a complaint of sudden painless bleeding per vaginam, which had commenced whilst at her housework.

On examination the blood pressure and urine were normal and there was no uterine tenderness. The height of the fundus was 36 weeks and the vertex was entering the pelvis but not engaged, foetal heart heard. Three days after admission to hospital in spite of rest, a further slight haemorrhage occurred, and it was decided to examine the patient under anaesthesia. During this procedure it was found that the vertex could be made to engage in the pelvis. The cervix admitted two fingers and a thin rim of placenta could be felt high up but not reaching to the internal os. Low rupture of the membranes

SM 9/continued

was performed and labour commenced 12 hours later and lasted 15 hours with the spontaneous delivery of a female child weighing 5 lb. 10 oz.

Hysterosalpingography performed 12 weeks later showed a bicornuate uterus.

Commentary

It is probable that neither the hyperemesis gravidarum nor the placenta praevia were due to the uterine abnormality, although a higher than normal incidence is reported by several authorities. As one would expect placenta praevia frequently occurs with the septate form of uterus but no more commonly with the severe types of deformity.

It is interesting to note that the uterine malformation was not detected until attention had been drawn to its presence.

Manual Removal of an Abnormally Adherent Placenta in Association with a Uterus Cordiformis Subseptus.

SM 10

Mrs.J.R.,Married one year.

Type V

a - 1
b - 0
c - 1
d - 0
e - 1
f - 0
g - 0
h - 0
i - 1
j -
k -
l -
m - 1

Menstrual Type K13 Cycle 5-5/26 days, regular, normal loss, moderate dysmenorrhoea.

Obstetric History

The patient, a primipara, was an emergency admission from the district, with a retained placenta and post-partum haemorrhage following normal delivery at term.

On admission to hospital the patient was pale and shocked. The placenta had been retained two and a half hours and the blood loss had been estimated at 50 oz. by the district midwife. Following the setting up of an intravenous drip of dextran the patient's condition improved and manual removal of the placenta was decided upon. During this operation the placenta was found to be separated in its lower one third and separation of the attached portion of the placenta was commenced. The line of cleavage became increasingly difficult to discern as the uterine fundus was approached,

SM 10/continued

the placenta being morbidly adherent in this area, and one cotyledon had to be removed piecemeal, being firmly attached to a uterine septum. During these intra-uterine manipulations, a marked depression could be felt in the middle of the fundus, and running down from this was the fleshy uterine septum. These abnormalities were much more marked when the placenta had been removed and the uterus firmly retracted, when the septum could be felt reaching almost to the internal os.

Recovery was uneventful.

Commentary

According to most authorities it is this type of uterus most commonly associated with retention of the placenta and post-partum haemorrhage. It may be that the poorly developed uterine musculature, especially where a uterine septum is present, allow undue penetration of the chorionic villi into the decidua.

Bicornuate Uterus

SM 11

Bicornis Unicollis

Unstable Lie

Breech Delivery

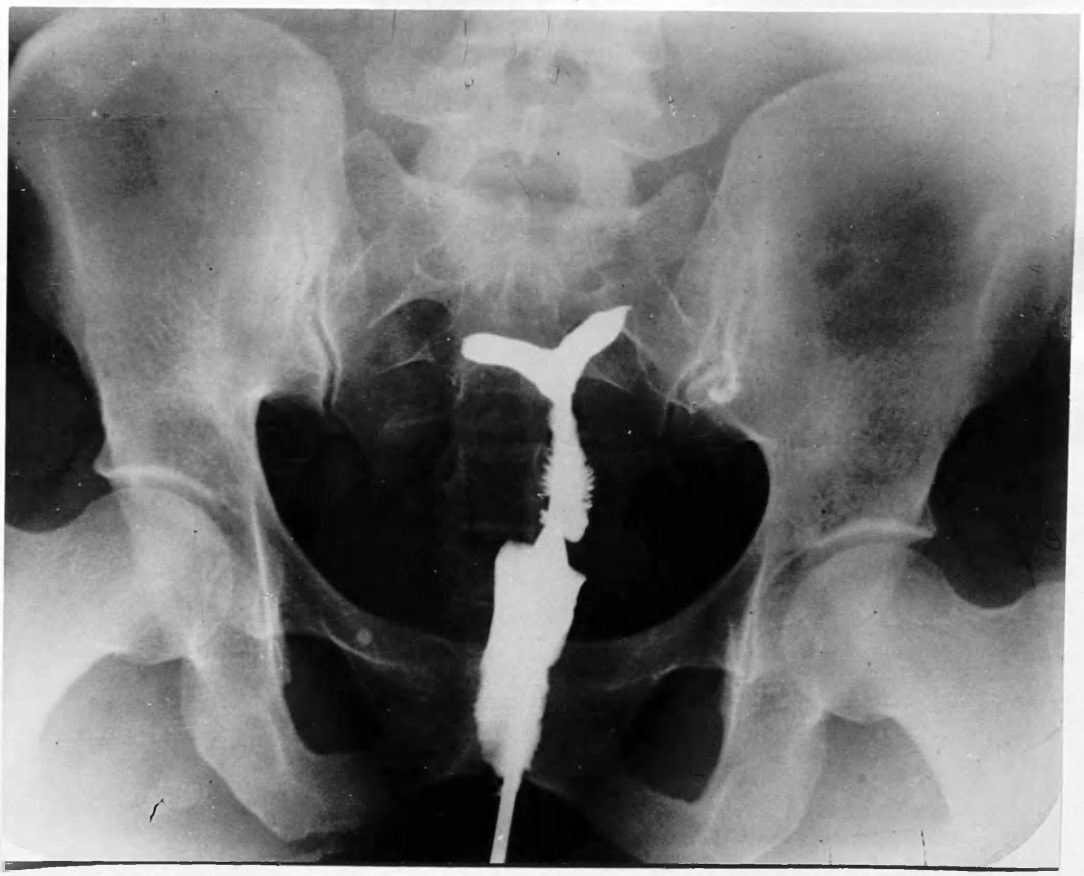
Mrs.Joan Harde, aged 27

Type 111

a - 1
b - 0
c - 1
d - 0
e - 0
f - 1
g - 0
h - 1
i - 1
j - 0
k - 0
l - 0
m - 1

Menstrual Type K 13 Cycle 5/28, normal loss, painless.

1953 - booked case attending antenatal clinic. Transverse lie at 30 weeks. Breech presenting and turned to a vertex at 32 weeks. Breech turned to vertex at 34 weeks. Broad asymmetrical saddle shaped fundus aroused suspicion of a congenitally malformed uterus especially with the unstable lie of the foetus. Admitted in labour at 35th week. Assisted breech delivery of a living female infant weighing 5 lb. 9 oz. Third stage normal. Hysterosalpingography demonstrated a bicornuate uterus.



27. Subseptate Uterus.
Hysterogram SM 12.

This patient was delivered of a stillborn female infant who at postmortem was found to have a congenital anomaly of the uterus.

See 28 and 29.

Uterus Bicornis Subseptus

SM 12 Unstable Presentation

Prolapse arm and cord

Internal podalic version and manual removal placenta.

Anne Norton, aged 20.

Type V Menstrual Type K 15 6/28 loss heavy, moderate dysmenorrhoea, unmarried.

a - 1
b - 0
c - 1
d - 1
e - 0
f - 1
g - 0
h - 1
i - 1
j - 0
k - 1
l - 1
m - 3

1954 - booked case. Attending antenatal clinic. 34th week transverse lie easily corrected to a vertex. Fundus felt notched. 36 weeks - admitted in labour with prolapsed arm and cord (not pulsating). Cervix $\frac{3}{4}$ dilated - internal podalic version - stillborn female infant weighing 6 lb. 2 oz. Placenta removed manually; thick median septum from fundus half way to internal os, not encroached upon by placenta. Puerperium morbid.

Postmortem on stillborn infant showed cause of death to be asphyxia. Congenital malformation of uterus and vagina. (photograph: uterus bicorpus bicollis. Vagina septate. No anomaly of urinary system noted.

28. Uterus bicornis bicollis with septate vagina from stillborn infant - size compared with a sixpence.

Uterus Bicornis Unicollis

SM 13

Unstable Lie

Caesarean Section

Mrs.Ann Hewitt, aged 20

Type 111

a - 1
b - 0
c - 1
d - 0
e - 0
f - 1
g - 1
h - 1
i - 1
j - 0
k - 0
l - 0
m - 1

Menstrual Type K 11 Cycle 4/27 regular, loss normal, painless.

1955 - booked case. Antenatal period normal until 34th week when a transverse lie was found and corrected to a vertex. At 39 weeks lie oblique with breech in left iliac fossa. Version under sedation unsuccessful. Pelvis adequate both clinically and on radiological examination but breech did not enter pelvis with moderate uterine contractions and membranes ruptured. Lower Segment Caesarean Section performed when a female infant weighing 7 lb. 8 oz. was delivered. Fundus of uterus markedly saddle shaped. Right horn with pregnancy. Left size 16 weeks pregnancy. No communication between two horns.

Uterus Subseptus

Normal Delivery

Retained Placenta

SM 14

Mrs.H.Brickley,aged 33

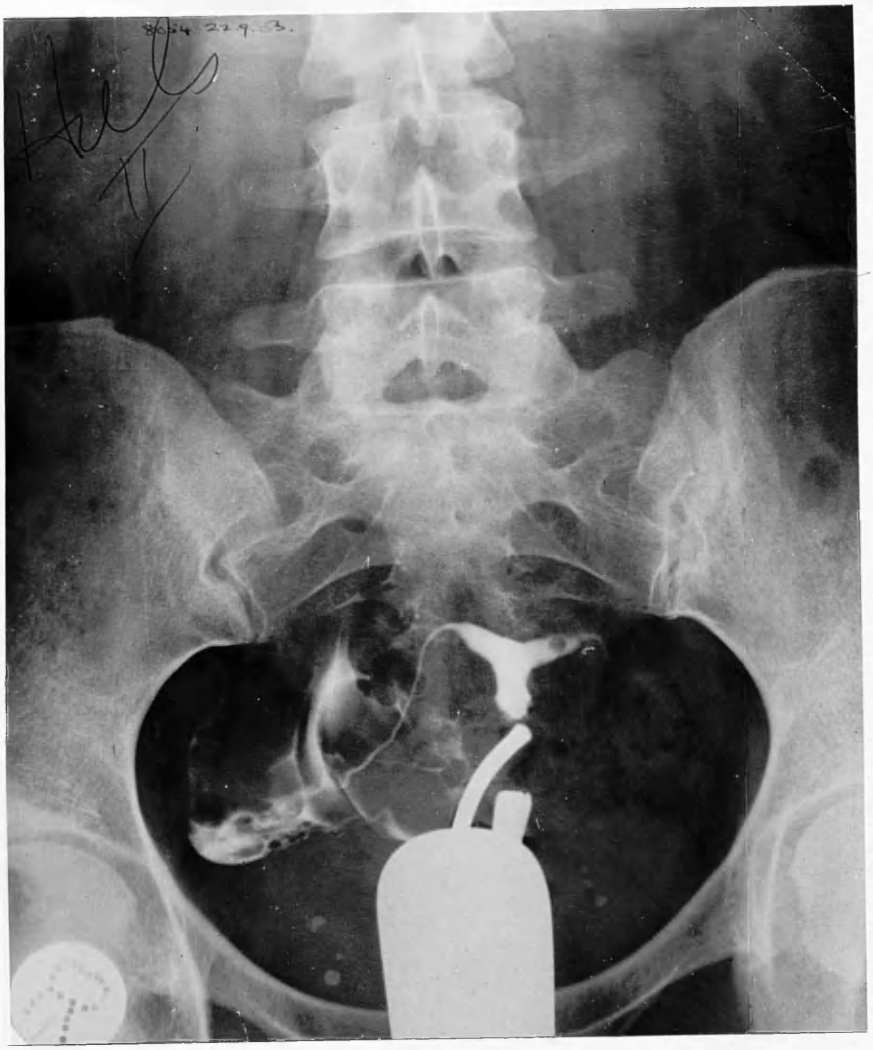
Type V

a - 1
b - 0
c - 1
d - 0
e - 1
f - 0
g - 0
h - 0
i - 1
j - 0
k - 0
l - 0
m - 1

Menstrual Type K 15 Cycle 7/28 heavy loss, regular always, dysmenorrhoea.

Voluntary Infertility. Married 6 years.

1954 - booked case, spontaneous delivery after normal pregnancy. Female infant weighing 6 lb. 2 oz. Placenta retained but no post-partum haemorrhage. On manually removing placenta the uterine fundus was found to be notched and the placental attachment encroached into a median septum of approximately 3" in length. Placenta appeared morbidly adherent in this area and was removed piecemeal.



30. Uterus Arcuatus.
Hysterogram.

Uterus Unicorpus Subseptus

Miscarriage

SM 15

J. Thomas, aged 19

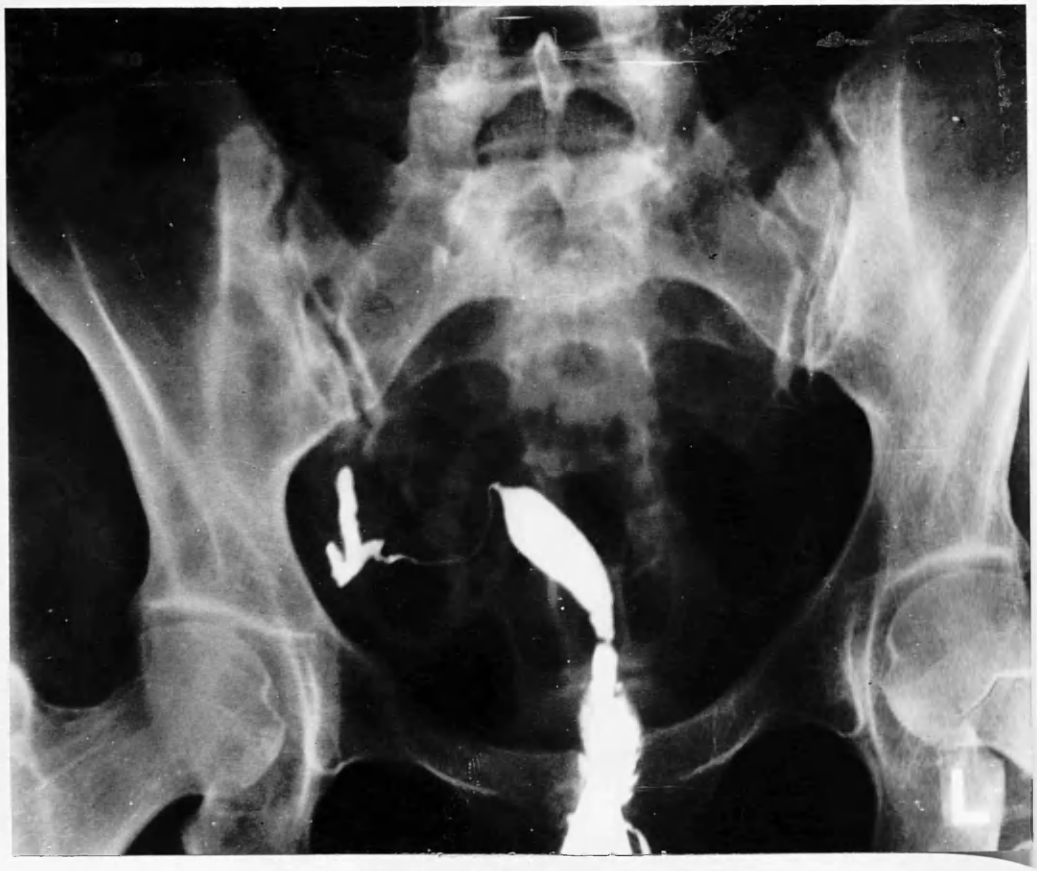
Type V

a - 0
b - 1
c - 0
d - 0
e - 0
f - 0
g - 0
h - 1
i - 0
j - 0
k - 0
l - 0
m - 1

Menstrual Type K12 4-5/30 regular, normal loss, painless.

Married 8 months.

1952 - Admitted to hospital as incomplete miscarriage of 4 months. Evacuation of uterus for retained products of conception. Piece placental tissue removed from fleshy central septum extending from fundus half way to internal os.



31. Uterus Bicornis.
Hysterogram simulating uterus unicornis as only one horn has filled with radio opaque media.

Uterus Bicornis Unicollis

Premature Labour

Stillborn Twins

SM 16

M.Davey, aged 31

Type 111

a - 2
b - 0
c - 0
d - 2
e - 2
f - 0
g - 0
h - 1
i - 1
j - 0
k - 0
l - 0
m - 1

Menstrual Type 14 Cycle 6/28 regular, normal loss, Moderate dysmenorrhoea.

Married 2 years.

1954 - Primigravida admitted in labour at 29 weeks gestation, membranes ruptured. Twin male foetus, stillborn. Binovular placenta. Evacuation uterus for retained products conception when anomaly discovered. Left horn much longer than right, piece of placental tissue removed. It appeared that both pregnancies had been in left horn.

Uterus Bicornis Unicollis

Repeated Breech Presentation

Cornual Pregnancy

SM 17

Mrs.Nadia Sekotina, aged 33

Type 111

a - 3
b - 3
c - 3
d - 2
e - 1
f - 2
g - 0
h - 6
i - 2
j - 0
k - 0
l - 0
m - 4

Menstrual Type K 12 Cycle 6/29 regular, normal loss, moderate dysmenorrhoea.

Gravida 6 Para 3

1951 - Attended antenatal clinic. This was her 5th pregnancy having had two stillbirths both delivered as breeches, one spontaneous miscarriage and one therapeutic abortion (indication on social grounds being performed abroad). Present pregnancy proceeded normally until 34th week when the foetus was found to be presenting by the breech. External cephalic version performed successfully without anaesthesia. Normal delivery at term of a male infant weighing 6 lb. 13 oz. Third stage normal.

1955 - Cornual Pregnancy.

Uterus Bicornis Unicollis

Repeated Breech Presentation

Cornual Pregnancy

SM 17

Mrs.Nadia Sekotina, aged 33

Type 111 Menstrual Type K 12 Cycle 6/29 regular, normal loss, moderate dysmenorrhoea.

a - 3
b - 3
c - 3
d - 2
e - 1
f - 2
g - 0
h - 6
i - 2
j - 0
k - 0
l - 0
m - 4

Gravida 6 Para 3

1951 - Attended antenatal clinic. This was her 5th pregnancy having had two stillbirths both delivered as breeches, one spontaneous miscarriage and one therapeutic abortion (indication on social grounds being performed abroad). Present pregnancy proceeded normally until 34th week when the foetus was found to be presenting by the breech. External cephalic version performed successfully without anaesthesia. Normal delivery at term of a male infant weighing 6 lb. 13 oz. Third stage normal.

1955 - Sacculation Pregnant Half of Bicornuate Uterus. Referred to hospital complaining of recurrent attacks of lower abdominal pain and slight vaginal bleeding of nine days duration. The pain which varied in severity preceeded the bleeding and was felt mainly just to the right of the midline in the hypogastric area.

SM 17/continued

The last normal menstrual period had been 12 weeks previously and the patient believed herself to be pregnant.

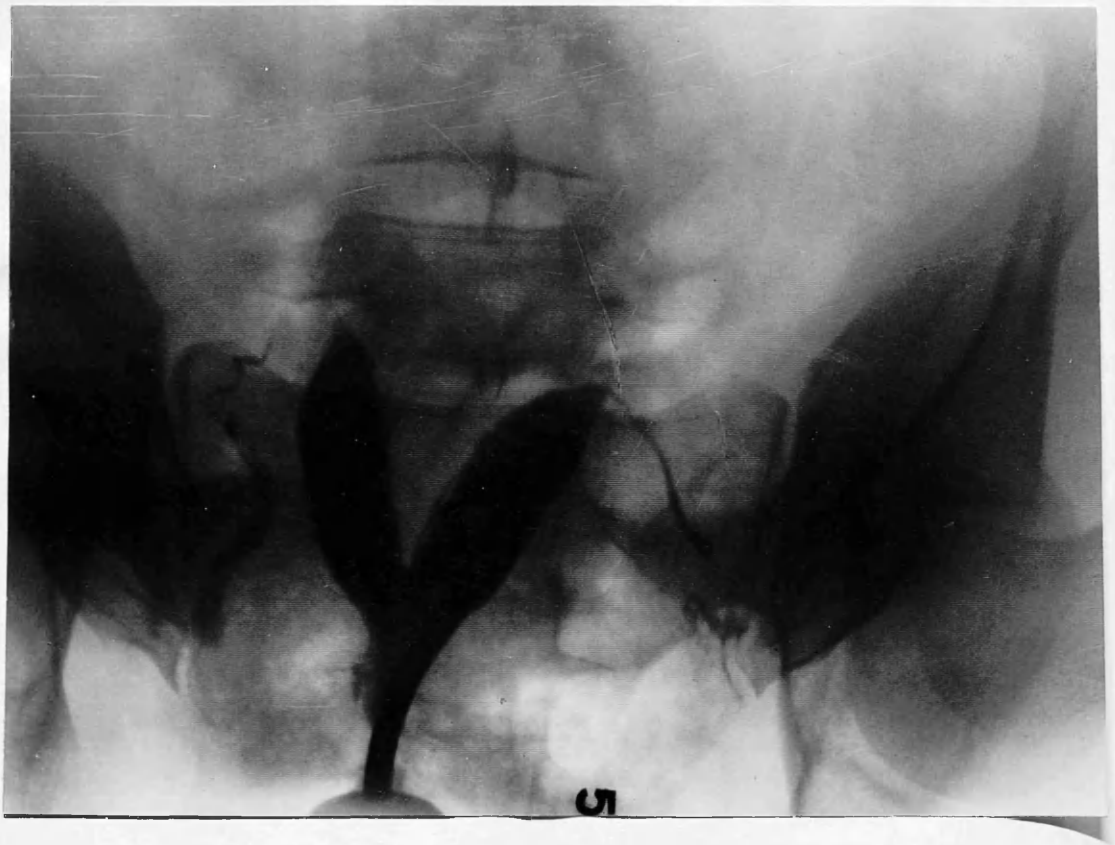
On abdominal examination a tender rounded semicystic mass, the size of a cricket ball could be felt in the right iliac fossa, and bimanually the mass was felt to the right of the uterus but not apart from it. The uterus was anteverted and considered to be slightly bulky but although the cervix was softened, to have the consistency of the non-pregnant organ.

A provisional diagnosis was made of advanced extra-uterine pregnancy or pregnancy in the rudimentary horn of a bicornuate uterus. During the next few days the patient's condition was observed without there being any amelioration of her symptoms and during this time the pregnancy test was reported as positive. Because of the uncertainty of the diagnosis and the danger of treating the provisionally diagnosed conditions conservatively, laparotomy was decided upon and just prior to operation a bimanual vaginal examination was made under anaesthetic and confirmed the previous findings.

SM 17/continued

Laparotomy - on opening the abdomen through a midline sub-umbilical incision the uterus was seen to be asymmetrically enlarged, the asymmetry being produced by a sacculation of the right half of the uterus, and represented the mass which had been felt in the right iliac fossa. The round ligament was noted to be lying lateral to the sacculation, the walls of the latter being so thin that no only was the foetus palpable through them but could actually be seen. The left half of the uterus was enlarged and of a firmer consistency than usual in pregnancy. In view of the extreme thinness of the walls of the aneurysmal-like dilatation and the apparent likelihood of rupture, the uterus was emptied by hysterotomy. The sacculation was then found to be separated from the left half by a thin partition extending from the fundus to the internal os. The foetus corresponded in size with a 12 week gestation and the placenta was attached in the region of the right cornu and was easily separated. After closing the uterus it became firmly retracted and the sacculation much less pronounced.

Three months later a hysterosalpingogram showed a bicornuate uterus the two horns of which were approximately equal in size.



32. Uterus Bicornis

Hysterogram from infertility investigation.

Uterus Bicornis Unicollis

Normal delivery.

SM 18

Mrs.Grace Whitcomb, aged 21

Type 111

a - 1
b - 0
c - 1
d - 0
e - 1
f - 0
g - 0
h - 1
i - 0
j - 0
k - 0
l - 0
m - 1

Menstrual Type K 14 cycle irregular 3-5/21-33.
loss heavy, painless.

Married 14 months.

Para 1 Gravida 1.

Vaginal examination at antenatal clinic revealed a uterus consistent in size with the period of gestation, namely 16 weeks. To the left of the uterus but seemingly attached to it was a firm mass the size of a plum.

Examination under anaesthetic confirmed the above findings and the mass was considered to be the non-pregnant horn of a bicornuate uterus.

Normal delivery of a male infant weighing 8 lb. 4 oz. at the 38th week. Third stage normal.

Duration labout 17 hours 40 minutes.

Four months post-partum hysterosalpingography confirmed the diagnosis of bicornuate uterus.

Queen Charlotte's Maternity Hospital

Type of Anomaly	1	111	V	V11	Total
	8	11	2	1	22
a	11	14	2	2	29
b	4	6	4	0	14
c	9	9	2	0	20
d	2	2	0	0	4
e	1	7	1	0	9
f	9	6	1	1	17
g	5	3	1	1	10
h	3	19	3	2	27
i	6	41	0	0	47
j	1	0	0	0	1
k	2	1	0	0	3
l	0	0	0	0	0
m	9	9	1	0	19

Uterus Bicornis Bicollis Septate Vagina

Breech Presentation

Classical Caesarean Section.

QC 1 1945
825

Type 1

a - 1
b - 1
c - 1
d - 0
e - 0
f - 1
g - 1
h - 2
i - 0
j - 0
k - 0
l - 0
m - 1

Aged 28

1943 - Spontaneous miscarriage at 18 weeks when anomaly detected

1945 - Persistent breech presentation external version unsuccessful presenting part remained high. Classical Caesarean Section at 37th week. Male infant weighing 6 lb. 2 oz. delivered.

Bicornuate Uterus
Bicornuate Uterus
Breech delivery Stillborn Infant.

QC 2 1946
1435

Type 111

a - 1
b - 1
c - 0
d - 1
e - 0
f - 1
g - 0
h - 2
i - 2
j - 2
k - 0
l - 0
m - 0

Aged 30

1945 - Spontaneous miscarriage at 12 weeks - anomaly detected during evacuation uterus for retained products conception.

1946 - Admitted in labour with infant's limbs and trunk delivered - head delivered by Mauriceau Smellie Veit manoeuvre. By dates 41 weeks but stillborn infant weighed 5 lb. 6 oz.

Third stage normal.

Uterus Bicornis Bicollis Septate Vagina

Normal Delivery

QC 3 1947
2543

Aged 27

Type 1

a - 1
b - 0
c - 1
d - 0
e - 1
f - 0
g - 0
h - 0
i - 0
j - 0
k - 0
l - 0
m - 0

Primigravida normal delivery at term of a male infant weighing 8 lb. 2 oz. Third stage normal.

Anomaly detected at antenatal clinic.

Uterus Bicornis Bicollis Vagina Septate

Unstable Lie

Caesarean Section

QC 4 1949
B8827

Type 1

a - 3
b - 1
c - 2
d - 2
e - 0
f - 2
g - 1
h - 4
i - 3
j - 0
k - 1
l - 0
m - 1

Aged 31

1939 - One Spontaneous Miscarriage.
Anomaly found at D & C.

1945 - Breech delivery at 39 weeks of
female infant weighing 6 lb. 12 oz.
died 3rd day cerebral haemorrhage.

1947 - Assisted breech delivery at 30 weeks
of a female infant weighing 2 lb. 4 oz. died
4th day - prematurity.

1949 - Unstable lie - persistently transverse.
Lower Segment Caesarean Section at
39 weeks female infant weighing 7 lb. 8 oz.
Puerperium normal.

Bicornuate Uterus

Dermoid cyst complicating pregnancy

Ovarian cystectomy

Normal delivery

QC 5
A2736
Type 111
a - 1
b - 0
c - 1
d - 0
e - 1
f - 0
g - 0
h - 1
i - 0
j - 0
k - 0
l - 0
m - 1

Aged 19

1950 - Primigravida - cystic mass size of a grape fruit felt through left fornix at 16 weeks pregnancy. Uterus found to be bicornuate at laparotomy pregnancy in left horn. Dermoid cyst arising from left ovary. Ovarian cystectomy performed. Normal delivery at term of a female infant weighing 6 lb. 9 oz. Third stage normal.

Uterus Subseptus

Normal delivery

Retained Placenta, Manual Removal.

QC 6
A2805
Type V
a - 1
b - 0
c - 1
d - 0
e - 1
f - 0
g - 0
h - 0
i - 1
j - 0
k - 1
l - 0
m - 1

1950 - Primigravida normal pregnancy.

Spontaneous delivery at 39 weeks, male infant weighing 8lb. 1 oz. Retained placenta. Uterine septum found during manual removal placenta. Placenta encroached upon septum which reached half way to internal os. Only 10 oz. haemorrhage.

QC 7
1952

7332

Type 1

a - 3
b - 0
c - 3
d - 0
e - 0
f - 3
g - 3
h - 1
i - 0
j - 0
k - 0
l - 0
m - 1

Uterus Bicornis Bicollis

Septate Vagina

Persistent Breech Presentation

Classical Caesarean Section.

Mrs.Winifred Mansford, aged 35.

Under the care of Mr.Bell.

In 1944, when aged 28, the patient was booked here when a primigravida. Attempted external version at 36 weeks was unsuccessful even under general anaesthesia and at this time the patient was found to have a septate vagina and double cervix. The pregnancy was apparently in the right horn. A classical Caesarean section was performed at the 37th week and a live male child weighing 6 lb. 2 oz. was delivered. The presence of a double uterus was confirmed at operation, the pregnancy being in the right horn. The uterine wall was very thin and there was some slight sub-peritoneal haemorrhage, in view of which Mr.Bell advised further sections should the patient again become pregnant.

QC 7/continued

In 1949, she had an elective Lower Segment Caesarean Section for a bicornuate uterus and in 1952 a repeat Lower Segment Caesarean Section for the same reason. On each occasion, she was delivered of live babies and all the family is doing very well.

The state of affairs in this patient appears to be a uterus bicornis bicollis and septate vagina.

Uterus Bicornis Unicollis
Antepartum Haemorrhage
Normal Delivery

QC 8 1951
A 3607

Type 111

Jennie Heaney, aged 31.

Under Mr.Briant Evans.

a - 1
b - 0
c - 1
d - 0
e - 1
f - 0
g - 0
h - 2
i - 1
j - 0
k - 0
l - 0
m - 1

No past obstetric history, but had been X-rayed at Hammersmith Hospital in 1948 when the presence of a double uterus was noted.

L.M.P. 15th June, 1950.
E.D.D. 22nd March, 1951.

Antenatal admission 12th September,1950, patient complained of lower abdominal pain. On examination a tender lump was found to the right side of the uterus. It was uncertain what was the nature of this lump (since the left side of the uterus enlarged rapidly while the lump on the right side appeared to disappear into the pelvis, I think it is a reasonable supposition that the lump on the right side was the non-pregnant horn of a bicornuate uterus).

QC 8/continued

She was re-admitted on the 6th January, 1951, with a small antepartum haemorrhage but there was no recurrence and no local cause found. She commenced in spontaneous labour and on 15th March, 1951 was delivered of a live female child spontaneously by the vertex after an episiotomy for foetal distress. The baby weighed 6 lb. 8 oz. at birth and both mother and baby did well.

There is no note of a septate vagina or presence of any pelvic anomaly found either at the time of delivery or subsequently so that the history of a double uterus in this case appears to depend on the Hammersmith Hospital X-ray in 1948 and there does not appear to be a letter from them confirming this.

Uterus Bicornis Bicollis Vagina Septate
Persistent Breech Presentation
Failed Version
Assisted Delivery

QC 9
1952

A 11411 Aileen Whitehead, aged 30. Primigravida.

Type 1 Booked under Mr.Winterton in 1952.

a - 1
b - 0
c - 0
d - 0
e - 0
f - 1
g - 0
h - 2
i - 1
j - 0
k - 1
l - 0
m - 3

Presence of double uterus and septate vagina noted on first attendance.

There was a complete vaginal septum and two cervices and a male distribution of pubic hair.

At the 30th week of pregnancy, the foetus presented by the breech and this presentation persisted. Attempted external cephalic version at 35th week failed.

She commenced in spontaneous labour at the 37th week and after a labour lasting 3 hours 45 minutes she was delivered per vaginam by the breech after division of the vaginal septum and episiotomy. Forceps were applied to the after coming head. A live male child resulted weighing 5 lb. $6\frac{1}{2}$ oz. and both mother and child did well.

QC 10
1952
A 8617

Emily Broughton, aged 35.

Type 111

a - 1
b - 1
c - 1
d - 0
e - 0
f - 0
g - 1
h - 1
i - 1
j - 0
k - 0
l - 1
m - 0

This patient was booked under the care of Mr.Gibberd in 1952.

Past History:

In 1947 the patient had been pregnant and at 3 months had had a sudden painless haemorrhage. Laparotomy was performed as there was some doubt whether or not the patient was suffering from an ectopic. At operation, a bicornuate uterus was found, the pregnancy being situated in the left horn. When seen in 1952, she appeared to be pregnant in the right horn.

Pregnancy continued satisfactorily until the 30th week but the presentation tended to be unstable and at the 30th week the lie was transverse and the patient had a small painless haemorrhage. A provisional diagnosis of placenta praevia was made and the patient admitted to hospital.

QC 10/continued

A week later all vaginal loss had ceased and on the 19th July, when 31 weeks pregnant, the patient commenced in spontaneous labour. A live healthy female child was delivered spontaneously face to pubes, after an episiotomy, labour having lasted 13 hours 25 minutes. The baby weighed 4 lb. 5 oz. at birth and was breast fed.

The puerperium was uneventful and on returning to the post-natal clinic on 27th August all was satisfactory. The uterus at that time was fairly well involuted and felt to be bi-lobed.

QC 11
1953

A 12383

Uterus Bicornis Bicollis Septate Vagina
Congenital absence right kidney and ureter
Ruptured Pyosalpinx.

Type 1

a - 0
b - 1
c - 0
d - 0
e - 0
f - 0
g - 0
h - 1
i - 0
j - 1
k - 1
l - 0
m - 0

Mrs.Mary Debenha, aged 35.

No previous pregnancy. She had attended a fertility clinic elsewhere, no clinical abnormality being found; she conceived before any investigations were performed. Appendicectomy had been carried out in 1937.

Attended ante-natal clinic at the 8th week of pregnancy when the uterus was found to be lying obliquely (? fibroid present).

She was admitted at the 24th week of the pregnancy, complaining of pain in the right iliac fossa for 5 days duration and a vaginal discharge. She had no urinary symptoms and the digestion was normal. On examination there was a tender area on the side of the uterus, this was thought to be a degenerating fibroid. There was no pyrexia, no leucocytosis, no pyuria, with slight constipation.

QC 11/continued

Five days after admission she had a shivering attack without sweating. A small enema was given, the patient collapsed becoming shocked very rapidly and died soon after.

At a post mortem examination, the cause of death was found to be a ruptured pyosalpinx(right) due to a streptococcal infection and septicaemia. There was a congenital absence of the right kidney and ureter, with a double uterine body and cervix, and a septate vagina. A normal pregnancy of about 24 weeks gestation occupied the left uterus, with normal tube and ovary on that side.

QC 12
1953

A 11760

Double Uterus and Vagina
Deep Transverse Arrest
Forceps Delivery

Type 1

Mrs.Doris Duff, aged 30.

Primipara. Booked 22.12.52.

a - 1
b - 0
c - 1
d - 0
e - 0
f - 1
g - 0
h - 3
i - 1
j - 0
k - 0
l - 0
m - 3

L.M.P. 16.10.52.
E.D.D. 23.7.53.

At her first attendance she brought a letter from her doctor saying that she attended the South London Hospital a year previously for investigation of infertility. There she had seen Miss Bloomfield and was found to have a double uterus.

On examination on 16th March, 1953, it was noted that there was a soft swelling to the right of the pregnant uterus, and on vaginal examination tags of a previous vaginal septum were noted though only one cervix was felt. Otherwise pregnancy continued uneventfully, external cephalic version without anaesthesia being performed at the 31st week.

QC 12/continued

She commenced in spontaneous labour at the 39th week and on admission the membranes were ruptured. First stage of labour lasted 14 hours and after 3 hours and 15 minutes in the second stage the head, having engaged as an occipito posterior was found to be lying with the saggital suture transversely in the pelvic cavity. Manual rotation and forceps extraction after episiotomy was performed and a live male child weighing 7 lb. 6½ oz. delivered. Both mother and baby made satisfactory progress in the puerperium. Breast feeding failed, the baby being artificially fed on leaving hospital. She was seen in the postnatal clinic on the 24th August and her only complaint was backache. Involution was satisfactory and the vagina and perineum had healed. There is a note to say the cervix was extensively scarred and lacerated but apparently only one cervix was visualised as no mention is made of the other.

Uterus Bicornis Unicollis

QC 13
1953

Repeated Miscarriages

A 13040

Type 111

Mrs. Ivy Cullen, aged 31.

Under the care of Mr. Gibberd.

a - 0
b - 3
c - 0
d - 3
e - 0
f - 0
g - 0
h - 3
i - 0
j - 0
k - 0
l - 0
m - 0

Past History: 2 previous miscarriages, one at $5\frac{1}{2}$ months in 1948; one at $2\frac{1}{2}$ months in 1950.

Attending Fertility Clinic at the Chelsea Hospital for Women since February, 1952.

L.M.P. 13.3.53.
Progesterone implant 6.5.53.

Booked here for confinement. Admitted as threatened abortion at 11 weeks on 5th June, 1953. Became a missed abortion. A D & C was performed at 24 weeks on 9th September, 1953, for evacuation of retained products. Uterus was the size of a 12 weeks cyesis and retroverted. The uterus was found to be bicornuate, the right horn larger than the left.

Uterus Bicornis Unicollis
Uterine Inertia
Lower Segment Caesarean Section
Repeat Section for Cephalo-pelvic Disproportion

QC 14
1953

A 11878

Type 111

a - 2
b - 0
c - 2
d - 0
e - 2
f - 2
g - 0
h - 0
i - 2
j - 0
k - 0
l - 0
m - 0

Mrs.Hacking, aged 22.

Previous Caesarean section in 1948 at Irvine Central Hospital for inertia associated with a bicornuate uterus. A live male child resulted weighing 7 lb. 8 oz.

When first seen with her second pregnancy in 1953, she was 34 weeks pregnant, probably in the right horn.

Pregnancy progressed uneventfully to term but the head was floating, displaced to the left and could not be made to engage. X-ray pelvimetry was performed, a Group B pelvis found but Dr.Rohan Williams suspected the presence of placenta praevia. Clinically, however, Mr.Gibberd did not think there was any evidence of this (the patient was under Mr.Gibberd's care).

Following the onset of labour, strong contractions did not result in descent of the head, and this coupled with the previous Caesarean section was considered adequate indication for a repeat.

QC 14/continued

At this operation the pregnancy was found in the right horn of a uterus bicornis unicollis. The left horn being directed posteriorly and laterally.

Mother and baby made satisfactory progress. A live female child weighing 8 lb. 4 oz. at birth having been delivered.

Uterus Bicornis Unicollis
Persistent Breech Presentation
Failed Version
Non-descent Breech in Labour, Lower Segment Caesarean Section

QC 15
1953

A 12164

Type 111

Daphne de Looper, aged 35.

Under the care of Mr.Gibberd.

Date of booking 4.2.53.
L.M.P. 26.9.52.
E.D.D. 3.7.53.

a - 1
b - 1
c - 0
d - 0
e - 0
f - 1
g - 1
h - 2
i - 1
j - 0
k - 0
l - 0
m - 2
n - 0

Past Obstetric History:

1 miscarriage at 2 months in February, 1952.
D & C at Chelsea Hospital for Women.

Pregnancy apparently continued normally and no abnormality was suspected until 35th week when the foetus presented by the breech and the uterus felt as if it was bicornuate.

Attempted version without anaesthesia failed.

She commenced in spontaneous labour at 41 weeks(by dates) and after 6 hours in the first stage of labour, the foetus was found to be lying obliquely and this was confirmed radiologically. Lower Segment Caesarean section was performed, the indication being (1) oblique lie (2) bicornuate uterus (3) elderly primigravida. At the time of operation, the uterine anomaly was confirmed but although in the notes it is labelled as a bicornuate uterus it would seem more likely that it was a bilobed fundus in an arcuate uterus as it would seem that the head of the baby had occupied the right cornu.

QC 15/continued

A live female child weighing 5 lb. 7¼ oz. was delivered and both mother and baby made good progress.

Uterus Bicornis Unicollis
Laparotomy for Abdominal Pain
External Cephalic Version Failed
Prolapse Cord in Labour
Assisted Breech Delivery

QC 16
1953

A 12436 Mary Hearn, aged 30.

Under Mr.Briant Evans

Type 111

Booked 9.3.53.
L.M.P. 22.11.52.
E.D.D. 29.8.53.

a - 1
b - 0
c - 1
d - 1
e - 0
f - 1
g - 0
h - 2
i - 2
j - 0
k - 1
l - 0
m - 1

During the pregnancy the patient had previously been in Battersea General Hospital where she had been operated upon for suspected appendicitis as at that time she complained of pain in the right iliac fossa. At the time of operation the appendix was normal but the uterus was asymmetrically enlarged and it was thought that the patient might have an angular pregnancy. She was transferred to Putney Hospital under the care of Mr.MacLeod and when seen by him in March the pregnancy appeared to be a normal 12 weeks intra-uterine cyesis.

On 29th May she was admitted with abdominal pain and on abdominal palpation a bicornuate uterus was felt. A diagnosis of cornual pregnancy was made and the patient treated conservatively.

Two attempts at external version under anaesthesia at 36 weeks both proved unsuccessful. The patient commenced in labour at term and was delivered by the breech after a labour lasting $7\frac{3}{4}$ hours.

QC 16/continued

The foetal heart was heard early in labour but when the cervix became fully dilated, the membranes ruptured and the cord prolapsed. No pulsation could be felt in the cord and the foetal heart could not be heard.

A stillborn female foetus, weighing 6 lb. 4 oz. was delivered by the breech (assisted breech delivery) and at the time of delivery the uterus felt bicornuate.

The patient developed a superficial thrombophlebitis during the puerperium but eventually recovered.

Uterus Subseptus
Instrumental Delivery
Morbid Puerperium
Repeated Miscarriages

QC 17
1952

A 12990 Claudia Wagstaff, aged 30.

Under the care of Mr.Arthure.

Type V

Past Obstetric History:

a - 1
b - 4
c - 1
d - 0
e - 0
f - 1
g - 0
h - 4
i - 3
j - 0
k - 2
l - 0
m - 1

Instrumental delivery at the Bearsted Memorial Hospital.

Live male child was delivered weighing 8 lb. 2 oz.

The patient had a 3rd degree tear repaired at the time of delivery and the puerperium was complicated by pulmonary embolism and pneumonia.

She had had three miscarriages all at $2\frac{1}{2}$ months in 1944, 1945 and 1946 respectively.

When seen at the first examination on 12th May 1953, she was not examined vaginally but on 2nd June 1953 she was admitted with an incomplete abortion. Evacuation of the uterus was performed at which time the uterus was found to be sub-septate, the septum extending from the fundus half way to the internal os, the pregnancy having been in the right side of the uterine cavity.

QC 18
1955

A 18278

Unicornuate Uterus

Multiple Pregnancy

Pre-eclamptic Toxaemia

Caesarean Section

Type V11

Aged 37 Gravida 1

a - 2
b - 0
c - 0
d - 0
e - 0
f - 1
g - 1
h - 2
i - 0
j - 0
k - 0
l - 0
m - 0

Multiple pregnancy admitted for pre-eclamptic toxaemia which in spite of bed rest deteriorated. Lower Segment Caesarean Section at 34th week. Male twins delivered weighing 3 lb. 4 oz. and 2 lb. 8 oz. respectively. Left ovary, tube and broad ligament noted to be absent at operation. Placentae binovular.

Intravenous pyelogram showed the right kidney and ureter to be absent.

QC 19
1955

A 18708

a - 1
b - 1
c - 1
d - 0
e - 0
f - 1
g - 0
h - 2
i - 1
j - 0
k - 0
l - 0
m - 1

Uterus Bicornis Bicollis Septate Vagina

Assisted Breech Delivery

Aged 23

1954 - Spontaneous miscarriage when anomaly detected

1955 - Persistent breech presentation - external cephalic version unsuccessful. Assisted breech delivery at 36th week of a female infant weighing 5 lb. 11 oz. Third stage normal.

QC 20

A 9043

Type 111

a - 3
b - 0
c - 1
d - 0
e - 1
f - 1
g - 0
h - 2
i - 2
j - 0
k - 0
l - 0
m - 0

Bicornuate Uterus

Twin Pregnancy

Aged 23

1952 - spontaneous delivery. Living male infant weighing 6 lb. 12 oz. after normal pregnancy and labour. Anomaly detected antenatally. Confirmed three months post-partum.

1955 - Twin pregnancy with delivery of a male infant weighing 5 lb. 2 oz. and female infant of 4 lb. 6 oz. The first normal vertex, the second as an assisted breech delivery. Placentae binovular. Third stage normal.

QC 21

A 19392

Type 111

a - 1
b - 0
c - 1
d - 0
e - 1
f - 0
g - 0
h - 0
i - 0
j - 0
k - 0
l - 0
m - 0

Bicornuate Uterus

Normal Delivery

Aged 28

Normal delivery of a female infant weighing 6 lb. 14 oz. at term. Third stage normal. Anomaly detected at infertility clinic.

QC 22
1955

16839

Uterus Bicornis Unicollis

Normal Delivery

Type 111

a - 2
b - 1
c - 2
d - 0
e - 1
f - 1
g - 0
h - 1
i - 1
j - 0
k - 0
l - 0
m - 0

Mrs.Marjorie Mascall, aged 30.

Menstrual Type K 15 cycle 5/21 regular, heavy loss, severe dysmenorrhoea.

Para 2 Gravida 3

1950 - miscarriage at 12 weeks pregnant. Anomaly detected during evacuation uterus for retained products conception.

1952 - Forceps Delivery for delay in second stage labour of male infant weighing 6 lb. 2 oz. Third stage normal.

1955 - Normal pregnancy and labour. Female infant weighing 7 lb. 5 oz.

URINARY INCONTINENCE FROM ECTOPIC URETER OPENING INTO THE VAGINAL VAULT WITH ASSOCIATED UTERINE MALFORMATION.

Miss A.M. - unmarried. Aged 17 years.

Menarche aged 15 years.

Menstrual Type 6/28 days, regular, normal loss, severe premenstrual dysmenorrhoea.

Patient attended the urological clinic complaining of urinary incontinence. She stated that she was constantly wet and required to wear a sanitary towel constantly. Since early childhood this complaint had been treated as a case of enuresis but had become worse in the past two years, causing her to seek fresh advice.

Urological Investigation

Intravenous pyelogram showed a double ureter on the right side, normal single ureter on the left.

Cystoscopy normal, but after intravenous injection of methylene blue, the dye appeared in the vagina as well as the bladder.

Examination under general anaesthetic

External genitalia appeared to be normal. On repeated injection of methylene blue, the dye was observed coming through a minute opening in the vaginal vault to the right of the cervix. There was no vaginal septum and the cervix was single but the uterine body was bicornuate.

Later the ectopic ureter was ligated and divided close to its entry into the vagina. Following this the patient's symptoms of incontinence disappeared.

Commentary

This case illustrates the association of congenital malformations of the urinary and genital tracts. Where malformations of either system are discovered, a routine investigation of the other system should be made to exclude congenital malformations.

Haematometra in a Rudimentary Uterine Horn

P.D. Aged 14 years

Admitted to hospital as an abdominal emergency, with a history of eight days lower abdominal pain. No vomiting but diarrhoea and tenesmus for the past four days. Frequency of micturition, both diurnal and nocturnal for a similar length of time. Last menstrual period was seven days overdue and commenced eight days before admission to hospital, that is on the same day as the onset of the abdominal pain.

Menarche, aged 13½ years.

Menstrual Type 4/28 days, regular. No premenstrual dysmenorrhoea but colicky lower abdominal pain for the past three months during the actual flow.

On Examination

Temperature 99°. Pulse rate 90. Flushed. Abdominal examination revealed tender mass in the right iliac fossa, which could also be felt rectally bulging through the anterior rectal wall.

White cell count: 23,000 c.mm. 85% polymorphs, leucocytes.

A provisional diagnosis of appendix abscess was made and the patient treated expectantly. The patient's symptoms lessened during the next four days and then returned with increasing severity. Rectal examination revealed a tender fluctuant swelling bulging the anterior rectal wall and extending into the right iliac fossa. The mass was aspirated through the anterior rectal wall and instead of pus a thick brownish fluid was withdrawn and found to be broken down red blood corpuscles.

The patient was now transferred to the gynaecological department and arrangements were made to examine the patient under anaesthetic.

Examination under Anaesthetic

Only one cervix could be felt and a large cystic mass bulging through the anterior vaginal wall appeared to be in continuity with the cervix; no definite uterine body could be felt.

Laparotomy

On opening the abdomen, the cystic mass which had been felt through the vaginal wall was found to be a rudimentary horn with attached fallopian tube greatly distended by blood. The left horn, tube and ovary appeared to be normal. A wide bore needle was inserted into the distended horn and 14 oz. thick chocolate coloured fluid withdrawn.

The right horn and tube were now ressected at the point where the rudimentary horn joined the supravaginal cervix; it was found that the rudimentary horn ended blindly just above the supravaginal cervix.

The abdomen was closed with drainage and apart from slight pyrexia for a few days immediately post-operatively the convalescence was normal.

Commentary

The case illustrates the occurrence of an acute abdominal emergency in association with a congenital uterine malformation. Where pregnancy occurs in a rudimentary horn which does not communicate with the main uterine body, a much more serious condition occurs, as the weakened uterine musculature is prone to rupture with profuse intraperitoneal haemorrhage.

BICORNUATE UTERUS FOUND AT HYSTERO-SALPINGOGRAPHY IN THE INVESTIGATION OF INFERTILITY

Mrs.D.H. - aged 31 years.

Menarche aged 13 years.

Menstrual Type 5-7/35-40 days, scanty loss, no dysmenorrhoea or dyspareunia.

Patient attended the gynaecological clinic complaining of infertility of one year's duration. Married four years, voluntary infertility for three years. History of appendix abscess aged twelve years, which was treated by drainage.

Vaginal examination revealed a single cervix normal vagina, uterus anteverted bulky and relatively fixed, no adnexal masses felt.

Seminalysis of husband normal.

Hysterosalpingogram one month later showed a bicornuate uterus with apparent non-patency of the left tube in the region of the cornua. Patency right tube and peritoneal spill present. Investigation of urinary tract normal.

Commentary

Whether the fertility rate is lowered or not in cases of uterine malformation is debatable but here the infertility is almost certainly due to the old pelvic peritonitis.

STATISTICAL ANALYSIS

Statistical Analysis and Case Records

The clinical records of five hospitals, three teaching and two non-teaching were examined for the purpose of obtaining sufficient material to evaluate the clinical significance of the congenital anomalies in pregnancy. This material consisting of 69 examples of congenital uterine anomaly with a total of 150 pregnancies was subjected to a careful analysis, on the model of Baker (1953) the results showing the incidence of abortion, foetal mortality, malpresentation, operative delivery, maternal morbidity, the complications of pregnancy and labour, foetal anomalies and other miscellaneous data, these being compared with data from another study and with values for the normal uterus. The clinical records of Guy's Hospital, Queen Charlotte's, St.George's, West Middlesex and St.Mary Abbot's Hospitals were used in the present survey.

Incidence Uterine Anomaly According to Hospital

		Incidence	Period of Survey
Guy's	10	1:538	1950-55 5 years
Queen Charlottes	22	1:1273	1945-55 10 years
St.George's	9	1:870	1940-55 15 years
West Middlesex	10	1:754	1951-53 3 years
St.Mary Abbot's	18	1:349	1949-55 5½ years

(Incidence calculated on the total number of cases booked for confinement).

Incidence of Uterine Anomaly in Relation to Type

1. Uterus bicornis bicollis (±) septate vagina.

   20 cases or 30% of total number.

III. Uterus bicornis unicollis.

   31 cases or 45%.

V. Uterus Septus, subseptus and arcuate.

   16 cases or 24%.

VII. Uterus unicornix.

   2 cases or 3%.

From the above table it will be seen that the incidence is significantly higher than any other Type III being the commonest form of anomaly being half again as common as Type 1, and twice as common as Type V.

If the relative incidence of the various types are compared, it will be found that Type 1 shows very little variation in the three teaching hospitals whose criteria for hospital confinement are approximately the same; similarly in Type III, Queen Charlotte's and Guy's Hospital show little difference, but at the same time the low incidence of this type at St.George's is difficult to explain.

A similar paradox is the very high incidence of Type 1 in the West Middlesex series, higher than in any of the teaching hospitals and very much higher than in the one other non-teaching hospital. As the teaching hospitals have a higher proportion of primigravidae it is to be expected that their figures would show a higher incidence of Type 1 for it is this anomaly which is most frequently diagnosed antenatally. On the other hand the West Middlesex is the 'parent' hospital for seven others and any cases of these anomalies are likely to be transferred to the 'parent' hospital and would not be shown as transferred.

Incidence of Uterine Anomalies in Relation to Type According to Hospital.

Hospital	Type of Uterine Anomaly		
	1	111	V
Queen Charlottes	36%	50%	9%
Guy's	30%	50%	20%
St.George's	33%	22%	33%
West Middlesex	40%	30%	30%
St.Mary Abbot's	12%	55%	33%

CONCOMITANT MALFORMATIONS

In the sixty-nine examples of congenital uterine malformation, there were seven with some concomitant malformation, an incidence of 10.1% but this figure is too low when one considers that only in 12 of the total 69 had any investigation of the renal tract been carried out yet three or 25% were found to have a renal anomaly.

Concomitant Malformation of	Uterine Anomaly			
	1	V11		
1. Renal Tract	2	1	3	Agenesis of renal tract on one side.
11. Gastro-intestinal Tract		111 1		Congenital Rectovaginal Fistula.
111. Minor Degrees Virilism		2	3	

Classification of Vital Statistics	Type of Uterine Anomaly				Total
	I	III	V	VII	
Total Cases Studied	20	31	16	2	69
Number Viable Births	28	40	17	3	88
Number of Abortions	11	29	22	0	62
Number of Infants	20	27	12	1	60
Number Foetal Deaths	5	8	6	1	20
Non-Operative Deliveries	14	14	8	0	36
Operative Deliveries	19	21	9	2	51
Caesarean Sections	7	8	3	1	19
Complications Pregnancy	17	55	29	3	104
Complications Labour	24	62	16	0	102
Maternal Morbidity	5	5	5	-	15
Maternal Mortality	1	0	0	0	1
Foetal Anomalies	0	0	2	1	3
Miscellaneous Operative Procedures	11	29	22	0	62

First Detected	Type Uterine Anomaly I	III	V	VII
Curettage	6	6	5	
Antenatal Clinic	5	4	1	
Laparotomy		7	0	1
Caesarean Section		4	2	1
Manual Removal Placenta	1	1	5	0
Hysterosalpingography	2	7	3	0
Examination under Anaesthetic	1	1		
Intra-partum	2			
Others				
Defloration Haemorrhage	1			
Haematocolpos	1			
Postmortem	1			
	20	31	16	2

Viability and Term Births

In a total of 150 pregnancies there were 88 viable births and of these 60 weighed more than $5\frac{1}{2}$ lb. giving a percentage viability of 59% and 70% term birth rate, both of which show a significant decrease from these associated with the normal uterus.

Prematurity

There were 28 infants whose birth weight was $5\frac{1}{2}$ lb. or less, an incidence of 31.8%.

Premature Labour in Relation to Type Anomaly

Type Anomaly	Number Premature Infants	Incidence per cent
1	8	33
111	13	35
V	5	17

From a total of twenty foetal deaths 11 were premature

Prematurity in seven cases was probably due to other factors than the anomaly per se, for six premature infants were twins and premature labour was induced for severe pre-eclamptic toxaemia at the 34th week in one other.

	1	111	V	V11
Multiple pregnancy		SM 16		
Spontaneous Labour		QC 20		
Caesarean Section and delivery twins where severe pre-eclamptic toxaemia present				QC 18
Surgical induction severe pre-eclamptic toxaemia	G 6			

Therefore corrected prematurity rate is 24%

and according to type

1 - 21% 111 - 25% V - 30% V11 - 0

Incidence of Abortion

In this series 62 pregnancies terminated prior to the 28th week of gestation, an incidence of 41.3% per total pregnancies (150). In seven of these termination of the pregnancy could not be attributed to the anomaly which gives a corrected abortion rate of 36%.

Termination of Pregnancy Prior to 28th Week, Not Attributable to the Anomaly

1 hysterectomy (SM 17)

1 therapeutic abortion (SM 17)

1 miscarriage following laparotomy for suspected rupture of uterine scar (G 4)

1 miscarriage following laparotomy for suspected ectopic pregnancy (QC 10)

2 hemihysterectomies for pregnancy in rudimentary horns (G 4 and WM 7)

1 mother died undelivered prior to the 28th week gestation (QC 11)

A total of seven pregnancies

Incidence of Abortion According to Type of Anomaly

Type Uterine Anomaly	Number Abortions	Incidence per cent	Corrected
1	11	28%	25%
111	29	43%	35%
V	22	55%	54%
V11	0	0	

Congenital Anomalies of the Foetus

There were three congenital anomalies among the 88 viable foetuses; an incidence of 3.4%. This is statistically significant yet a larger series would be desirable before drawing any conclusion.

Stillbirth and Neonatal Mortality

From 149 pregnancies, there resulted 88 infants which includes three sets of twins. Of the 87 infants delivered, there resulted 20 foetal deaths, an uncorrected incidence of 23%.

Foetal Mortality Related to Type of Uterine Anomaly.

Further analysis revealed that 5 infants, or 15% died in Group 1, 8 or 23% in Type 111 and 6 or 35% in Type V; there was one death in Type V11 but total deliveries for this Type was only three and the one death was unavoidable being an anencephalic foetus, so figures are not statistically significant.

Foetal Mortality Related to Type of Presentation.

When the total foetal mortality was analysed from the standpoint of presentation, it was noted that 8 deaths occurred in 53 vertex presentations, an incidence of 15%. In 8 forceps deliveries there was one death, no deaths in the 9 cases of Caesarean Section where the vertex presented, the remaining 7 deaths being associated with 36 spontaneous vertex deliveries, the major factor in these deaths being prematurity.

There were 10 deaths in 22 breech deliveries or an incidence of 45.4% but to this should be added the 10 breech presentations which were delivered by Caesarean Section without foetal loss, and gives an incidence of 31%. There were two deaths in four cases of oblique lie, and occurred in the two cases where the membranes were ruptured and the arm and cord had prolapsed, an incidence of 50%.

Method of Delivery Related to Type Anomaly and Presentation

If a further analysis is made of the foetal mortality in relation to the type of anomaly and method of delivery, the following information is obtained as shown in Table 111. There were 36 spontaneous deliveries, or 41.8% of all deliveries and these resulted in 7 deaths. Twenty-two breech deliveries or 25% of all deliveries, accounted for 10 deaths. Two internal podalic versions and extractions, or 2.2% of all deliveries accounted for two more deaths. There were 8 forceps deliveries of 9% of all deliveries and they accounted for one death. There were no deaths in 19 Caesarean Sections, of which three were of the classical type and the remaining 16 Lower Segment Caesarean Sections and accounted for 47.5 of all deliveries. The over-all incidence of operative delivery was 58.3% with a foetal mortality 25.5%. The operative delivery incidence is highest in Type 111 with an incidence of 60%, then Type 1 54%,

Method Delivery	Number	Incidence %	Type Anomaly				Infant Number	Mortality %
			1	111	V	V11		
Non-Operative (Spontaneous Vertex)	36	41.8%	14	14	8	0	7	19.3
Operative	51	58.2%	19	21	9	2	13	25.5
Breech 22			8	10	3	1	10	45.4
Forceps 8			4	3	1	0	1	12.5
Version & Extraction 2					2		2	100
Caesarean Section	19		7	8	3	1x	0	0
Classical 3								
L.S.C.S. 16								
Totals	87	100%	33	35	17	2	20	
No. Infant Deaths			5	8	6	1		
Incidence%			15	22.8	35%	33%		

x Twins

The incidence of Caesarean Section showed little difference between the various types being 23% for Type 1, 22.8 for Type 111 and 17.6 for Type V. The high incidence of Caesarean Section in Type 1 is at variance with the dictum 'the more complete the anomaly, the greater the chance of a normal delivery'.

Foetal Weight and Foetal Mortality Related to Anomaly

In a total of 20 foetal deaths 11 weighed $5\frac{1}{2}$ lb. or less and from the following table it can be seen that foetal weight has a significant bearing on foetal mortality

Type Anomaly				
1	111	V	V11	
5	8	6	1	20 Total foetal deaths
4	5	2	0	11 Number of foetal deaths where birth weight $5\frac{1}{2}$ lb. or less.

Caesarean Section

There were 19 Caesarean Sections of which 3 were of the Classical Type and the remaining 16 Lower Segment Sections from which twenty infants were delivered, including one set of twins. The incidence of Caesarean Sections to all form deliveries was 47.5 where the total operative delivery rate was 58.3%.

There were no maternal deaths and no foetal loss.

Incidence According to Type Anomaly

Type Uterine Anomaly	Number of Caesarean Sections
1	7
111	8
V	3
V11	1

INDICATIONS FOR CAESAREAN SECTION

WM 1 L.S.C.S. - previous unsuccessful pregnancy and hemi-hysterectomy.

SG 1 Classical - breech presentation only indication, elective section.

5 L.S.C.S. - breech presentation and minor degree pelvic contraction

7 L.S.C.S. - breech presentation, foetal distress early in labour

SM 5 L.S.C.S. - 2 previous stillborn breech deliveries.

6 L.S.C.S. - Unstable lie, pre-eclamptic toxaemia

6 L.S.C.S. - Repeat Section

7 L.S.C.S. - Repeated Miscarriages age

13 L.S.C.S. - Non descent of breech in labour

QC 1 Classical - breech presentation

4 L.S.C.S. - 2 previous stillbirths, unstable lie.

7 Classical - breech presentation, and 2 repeat lower segment.

7 L.S.C.S. - repeat

7 L.S.C.S. - repeat

14 L.S.C.S. - uterine inertia

14 L.S.C.S. - grade B pelvic contraction unsuccessful trial labour

15 L.S.C.S. - breech failure to descend in labour.

18 L.S.C.S. - Pre-eclamptic toxaemia, twin vertices.

COMPLICATIONS OF PREGNANCY

Malpresentation	Number	Incidence per cent	I	III	V	VII
Breech	33		14	13	5	1
Transverse	2				2	
Unstable	1		1			
Total	36		15	13	7	1

Premature Labour includes 3 sets of twins					
Figure of 23 obtained by subtracting 1 Caesarean Section for 1 surgical induction, 2 twins.	23	6	11	5	1 = 23
Pre-eclamptic Toxaemia	4	1	1	1	1 = 4
Mitral Stenosis	1				
Hyperemesis Gravidarum	1				
Pain Syndrome	5	0	5		
Antepartum Haemorrhage	6	1	2	3	
Pregnancy in Rudimentary Horn	2				

COMPLICATIONS OF PREGNANCY/contd

	1	111	V	V11
Laparotomy				
Septicaemia				
Peritonitis	1			
Multiple Pregnancy		2		1
Ovarian Cyst		1		
Bleeding in Early Months	1			
Threatened Miscarriage	1			
Pyelitis			1	
Postmature			1	
Decidual Cast		2		

COMPLICATIONS OF LABOUR

		1	111	V	V11
Delay Second Stage Labour	7				
a. from vaginal septum	5				
b. others	2				
Prolapse Arm	2				
Prolapse of the cord	5				
Oblique Lie	2				
Uterine Inertia	2				
Pelvic Contraction	3				
Trial of Labour	1				
Deep Transverse Arrest	1				
Foetal Distress	4				
Retained Placenta	8				
Postpartum Haemorrhage	4				
Third Degree Perineal Laceration	1				

MISCELLANEOUS OPERATIVE PROCEDURES

External Cephalic Version

Attempted 15

Successful 3

Digital Curettage

22

Surgical Induction

3 SM 9 G 4 G6

Manual Removal Placenta

10.

Laparotomies

G 9 suspected ectopic: QC 16 suspected appendicitis:

QC 10 suspected appendicitis: G 4: suspected rupture uterine scar:

SM 17 hysterotomy: QC 5 Ovarian cystectomy.

Division Vaginal Septum

QC 9 G2 WM 4 SM 2

Hemihysterectomy

G 5 WM 7

Metroplastic Operations

G 4: WM 10

Episiotomies

MATERNAL MORTALITY

There was one death in the 89 cases studied (QC 11) and was caused by streptococcal peritonitis following rupture of a pyosalpinx. Postmortem examination revealed a hitherto unsuspected congenital anomaly of the uterus and renal tract.

MATERNAL MORBIDITY

This is probably much higher than figures suggest for examination of case records showed many in which relevant details were absent or not fully documented.

No.		Postpartum Factors Causing Maternal Morbidity	Type Uterine Anomaly				Method of Delivery
			1	111	V	V11	
SG	6	Notifiable pyrexia following manual removal placenta	1				Forceps
	5	Postoperative	1				Caesarean section
	4	Postoperative	1				Caesarean section
G	5	Postoperative		1			Hemihysterectomy
	1	Placental cotyledon passed		1			Spontaneous breech. P.P.H.
	2	Urinary infection		1			Forceps
	6	? genital canal infection	1				Breech Extraction
QC.	16	Thrombophlebitis		1			Breech
	17	Pulmonary embolism			1		Forceps
	9	Unknown	1				Breech delivery
SM	3	Breast abscess		1			Spontaneous
	5				111		Breech with manual removal.
	12	Probable birth canal Infection			1		Internal version and manual removal placenta.

CONCLUSION

	Congenitally Anomalous Uterus			Norman Organ
	Present Study %	Baker %	Fenton and Singh %	American %
Abortion Rate(Uncorrected)	41.3	18.9	16.5	5.6
Viable Birth Rate	59	77.2	66.4	93.1
Term Birth Rate	70	57.5	56.1	85.0
Accidental Haemorrhage	4.5	0.9	5.5	1.2
Placenta Praevia	2.2	2.8	7.4	1.9
Toxaemias of Pregnancy	4.5	3.7	11.1	14.1
Malpresentation	38	28.7	17.5	3.9
Premature Labour	31.8	22.1	44.0	6.8
Uterine Inertia	2.2	7.3	22.8	4.0
Operative Delivery	58.2	63.0	50.0	20.0
Retained Placenta	11.4	0.9	9.0	1.9
Post-partum Haemorrhage	4.5	5.5	5.2	0.4
D & C for Retained Products of Conception	15	0.9	12.9	0.8
Foetal Mortality	23			0.9
Foetal Abnormalities	3.4	2.8		
Concomitant Malformations	10.1*			
Maternal Morbidity				

CONCLUSIONS

Congenital Uterine Anomalies would appear to be commoner than generally acknowledged.

The adoption of a universally accepted classification would resolve much of the difficulty at present experienced in clinical assessment of the anomalous uterus.

Although the association of congenital anomalies of the genital and urinary tracts is well recognised, the discovery of an abnormality in one system, is rarely followed by routine investigation of the other.

The nearer the anomaly approaches the normal organ the more likely it is to disturb pregnancy and conversely normal pregnancy and labour is most usual with the most marked forms of anomaly.

Recognition of the minor anomalies is dependent upon a familiarity with their clinical manifestations in pregnancy while the most marked forms will be detected on physical examination.

The common complications of pregnancy are abortion, malpresentation, premature labour and retained placenta and are commonly repeated in succeeding pregnancies.

Pain is a frequent concomitant of early pregnancy in the minor anomalies and the clinical appearance may closely simulate ectopic and angular pregnancies.

Dystocia from prolapse of the non-pregnant horn is a rare complication although formerly considered as common.

Statistics based on abstracts of the literature tends to give a distorted picture for only those cases judged of sufficient clinical interest are published and this will account for the high incidence of dystocia from prolapse of the non-pregnant horn so often reported yet in this and most contemporary studies it was found to be rare.

Further evidence is presented of the inheritance and familial incidence of these anomalies.

The anomalous uterus per se is not an indication for Caesarean section yet the high incidence of this form of delivery in the Type 1 anomaly suggests that frequently this is the only indication.