

CONGENITAL ABNORMALITIES OF GENITAL TRACT - UTERINE MALFORMATION

Pages with reference to book, From 261 To 266

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Abstract

Thirteen cases of congenital malformations of uterus were discovered during past ten years at Abbasi Shaheed Hospital, KMC, Karachi. They varied from uterus didelphys to uterus unicornis. Similar malformations presented different clinical pictures in different individuals. However, most of the congenital malformations encountered did not interfere with fertility and eight cases had conceived within a year of marriage though three ended in abortions. Intravenous pyelograms were also found to be normal in eight out of the nine cases, on whom this investigation was carried out. In conclusion it is pertinent to state that early diagnosis is important to state that immediate operative treatment is very effective in alleviating symptoms and preventing complications (JPMA 36: 261, 1986).

INTRODUCTION

Two müllerian ducts appear as buds in the outer part of each intermediate cell mass in the 5th and 6th weeks of intrauterine life. The ducts from each side fuse together and are canalised to form two fallopian tubes, the uterus and upper portion of vagina. Thus varying degrees of structural abnormalities of uterus and tubes can occur due to imperfect fusion and canalisation of müllerian ducts. The anomalies of müllerian ducts are genetically determined and are linked up with chromosomal make up of the individual, but they are also dependent on intra-uterine environment. Wilson and Harris¹ believed that two out of every thousand women have a sufficiently severe degree of uterine malformation to interfere with pregnancy.

Greiss and Mauzy² estimated the incidence to be 3.3% in their cases. Many a times the congenital malformations pass un-noticed as they do not always produce symptoms. Hence the incidence may even be still higher. In the present series, therefore, an attempt is made to discuss all the cases of congenital malformations encountered during a period of ten years.

MATERIAL AND METHOD

Thirteen cases with ages ranging from 16 to 55 years were detected to have congenital malformations of the uterus amongst 11,574 total operations performed (both major and minor) from 1974 to 1983 at Abbasi Shaheed Hospital, Karachi. Amongst these cases, eight were incidentally discovered at laparatomies carried out for various gynaecological indications and five were picked up at diagnostic curettages, through exploration of uterine cavities during evacuations and diagnostic curettage is routine practice in the department for detection of uterine abnormalities. Eight cases had symptoms related to abnormality while other five did not relate symptoms pertaining to malformation detected. All cases were questioned to establish the relationship of a particular anomaly with its relevant symptomatology.

Four cases (1, 2, 3 and 4) belonged to complete failure of fusion of müllerian ducts with didelphys uterus, 2 cases (5 and 6) had bifid uterus, 2 cases (7 and 8) were of sub septate uterus, 3 cases (9, 10 and 11) of bicornuate uterus, and case (12) of hypoplasia of one müllerian duct with undescended gonad of the same side and one case (13) was of unicornuate uterus.

Various operative techniques employed included dilatation and curettage, excision of vaginal septa, salpingectomy, utriculoplasty and enucleation and removal of ovarian cysts. All these cases are serially summarised in a table.

TABLE
Summary of Classification of 13 Cases of Uterine Malformation.

Type of Abnormality	No. of Cases	Case No	Presenting Complaint	Age	Marital Status	Management
COMPLETE FAILURE OF FUSION OF MULLERIAN DUCT						
1 Uterus Didelphys	4	1	Menorrhagia	16	S	Diagnostic D&C
		2	Post Coital bleeding P ₀₊₀	22	M	Excision of Vaginal Septum
		3	Post Coital bleeding	18	S	Not willing for excision of Septum
		4	Ectopic pregnancy Gravida - 4	30	M	Left Salpingectomy, Excision of Vaginal Septum
(Figure 1)						
INCOMPLETE FUSION, HYPOPLASIA, ABSENCE OF MULLERIAN DUCT						
2 Subseptate	2	5	Abortion P ₀₊₀ (Figure 2)	22	M	Dilatation & Curettage
		6	Miscarriages, Persistent Breech P ₁₊₂ (Figure 3)	26	M	Dilatation & Curettage Lower Segment Caesarean Section
3 Bifid Uterus	2	7	Tubo-Ovarian mass Pelvic Pain, Gravida 10 (Figure 4)	45	M	Total abdominal hysterectomy
		8	Asymptomatic Gravida 4 Post Menopausal (Figure 5)	55	M	Total abdominal hysterectomy Cyst adenoma left ovary
4 Bicornuate Uterus	3	9	Incomplete Abortion Gravida 4 Para Nil (Figure 6)	29	M	Dilatation & Curettage Utriculoplasty advised
		10	Haematocolpos from Vaginal atresia (Figure 7)	13	S	Utriculoplasty done
		11	Rupture of horn of bicornuate Uterus in pregnancy Primigravida (Figure 8)	22	M	Utriculoplasty done
5 Uterus Unicornuate	1	12	Ovarian cyst secondary amenorrhoea (persistent) P ₀₊₀ (Figure 9)	22	M	Enucleation of ovarian cyst
6 Hypoplasia of Right Mullerian Duct	1	13	Polypoidal cervical lip and mass in abdomen (Cyst of Undescended Gonad) P ₁₊₀ (Figure 10)	28	M	Dilatation & Curettage. Removal of cyst of undescended Gonad Follow up incomplete
* Case discussed in the other articles of the author on Congenital Malformations of genital tract (Vaginal defects)						

RESULT AND DISCUSSION

Different authors have reported varied frequency of malformations of genital tract anomalies. In our study, 13 cases of significant malformations were detected over a period of 10 years during which 11,574 total operations were performed in the department. A frequency of 0.13% was recorded in our cases.

Though eight of the cases were detected incidentally, a sound relationship with symptoms could be established in retrospect on taking careful obstetrics and family history.

The variation in the anomalies of mullerian ducts depends upon the level at which canalization is interfered with, so the problems of individual depends upon the type of malformation inherited. In case No. 1 of uterus didelphys, profuse menorrhagia was seen at menarche due to large bleeding area, and in case 4 ectopic pregnancy was due to abnormally long tube and in later pregnancies in-co-ordinate uterine action was diagnosed necessitating lower segment caesarean section. Non pregnant horn of uterus didelphys enlarged upto 12-14 weeks size of pregnancy under the effect of hormones and did not cause obstruction to the presenting part, although it may do so in some cases. Jones³ described uterus didelphys, obstetrically a bad uterus, but MonroKerr⁴ stated that more complete the malformation the lesser is the likelihood of dystocia.

Bifid uterus may have rudimentary horn and implantation of pregnancy sac in rudimentary horn of uterus may result in rupture and produce profound shock like that of rupture tubal pregnancy, some time rudimentary horn may be so small that it is only recognised when ruptured ectopic tissue is subjected to microscopic examination.

Buell⁵ and Perkin and Rubovitz⁶ have described diverticulum of uterus as a true sac connected to the uterine cavity by a small aperture. The diverticulum of uterus may be due to improper fusion of mullerian ducts in the mid-line and bulges out during pregnancy.

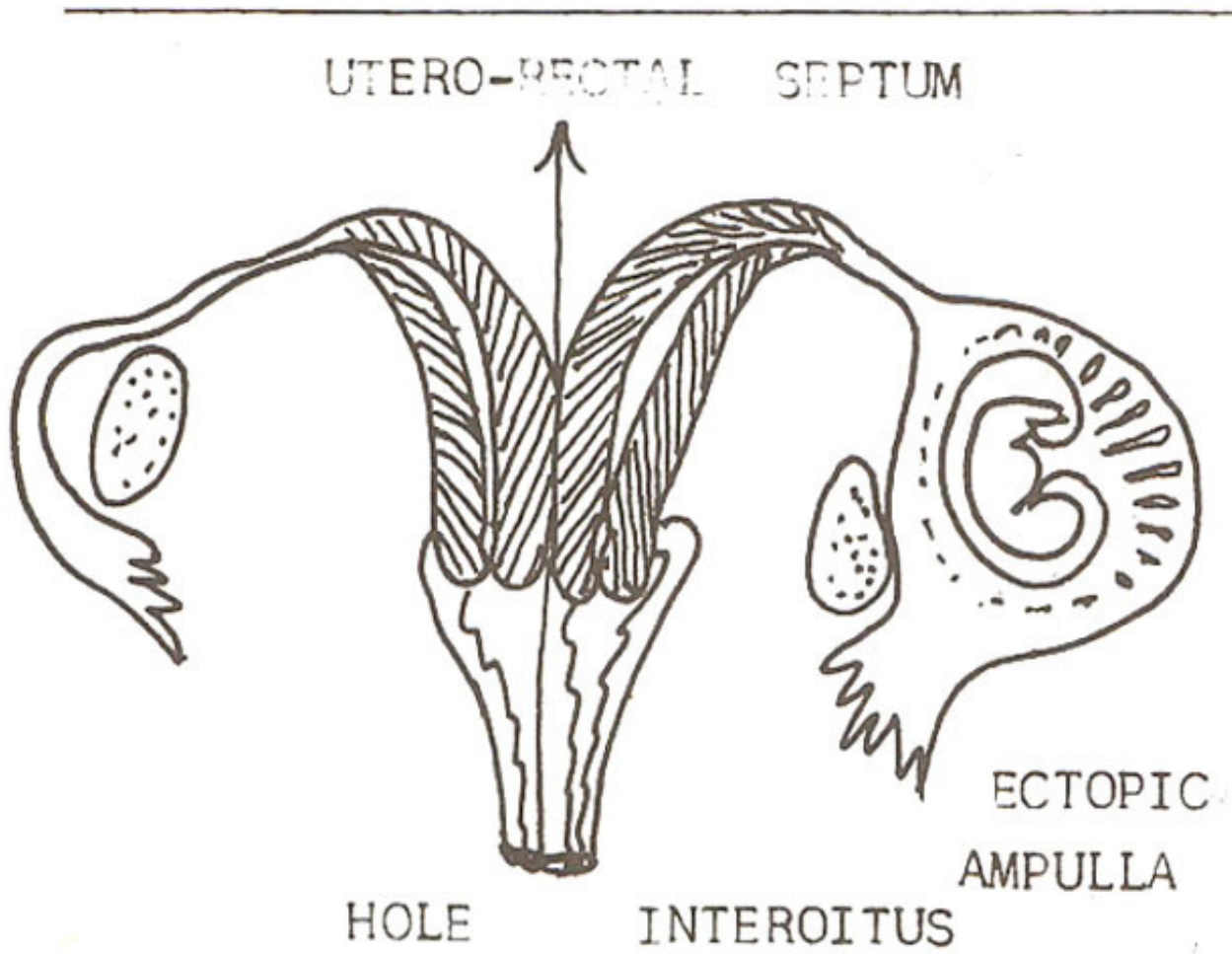


Figure 1. Case 4. Didelphys uterus with ectopic pregnancy.



Figure 2 (b). Case 5. Sub-septate uterus with double cervix.

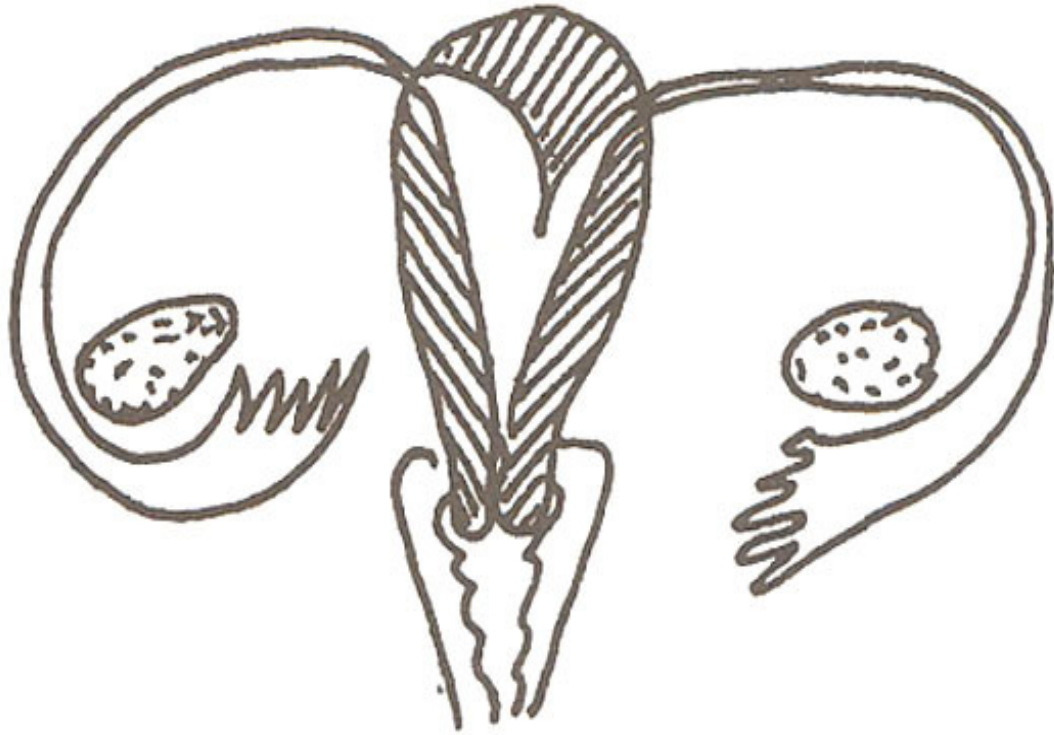


Figure 3. Case 6. Sub-septate uterus with two unequal halves with persistent breech-presentation and abortion.

RUDIMENTARY
RIGHT HORN OF UTERUS

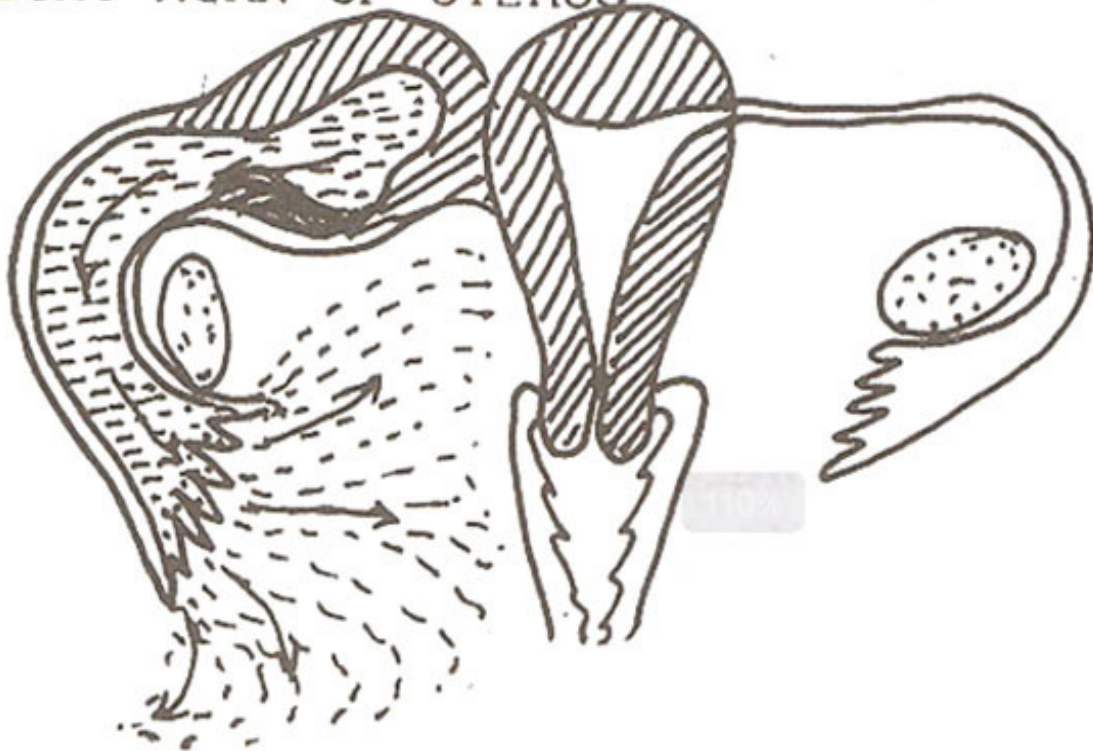


Figure 4(a). Case 7. Right rudimentary horn has no connection with normal left horn of uterus

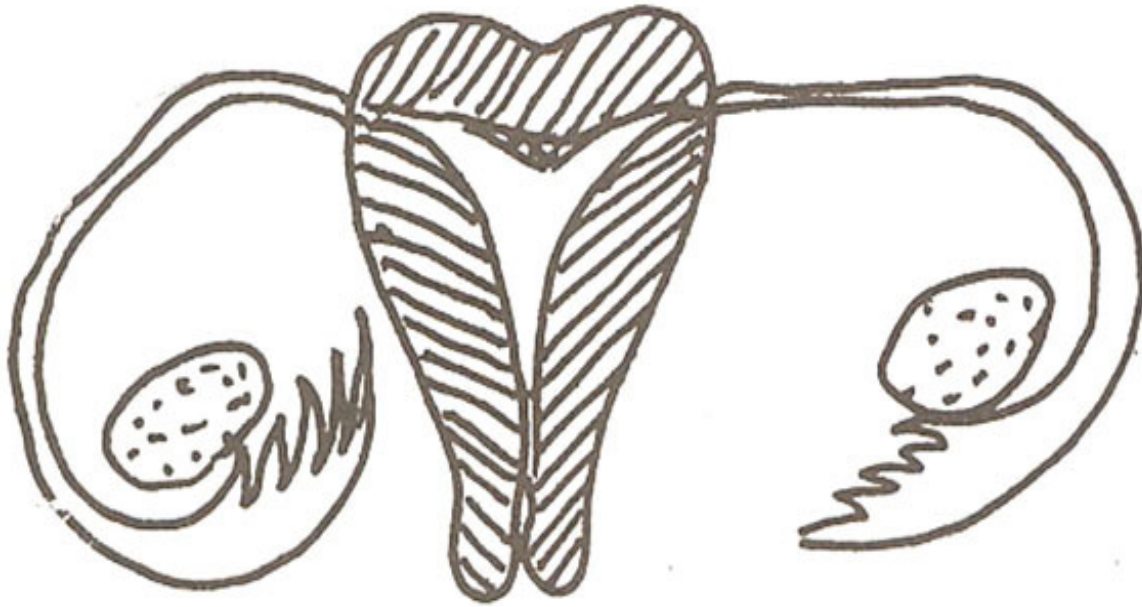


Figure 5. Case 8. Bifid uterus with single uterine cavity. 1 patient remained asymptomatic.

Bifid uterus has a depression on its fundus with a single cavity and cervix, such malformations often are asymptomatic and hence pass un-noticed (Case No 7 & 8).

In sub-septate uterus (Case No. 5 & 6) the fundus of uterus has a smooth curve as opposed to bidomuate uterus where two horn of uterus exist separately.

Uterus sub-septate (Case No. 6) should be strongly suspected if repeated attempts of external cephalic versions fail in transverse or breach presentation in primigravida. Hunter⁷ reckons that 12% of transverse lie are associated with this malformation and are more liable to cause inertia during labour. There is also an increased likelihood of placenta previa, as the uterus does not have enough space superiorly. Resection of septum is advised in cases of habitual abortions.

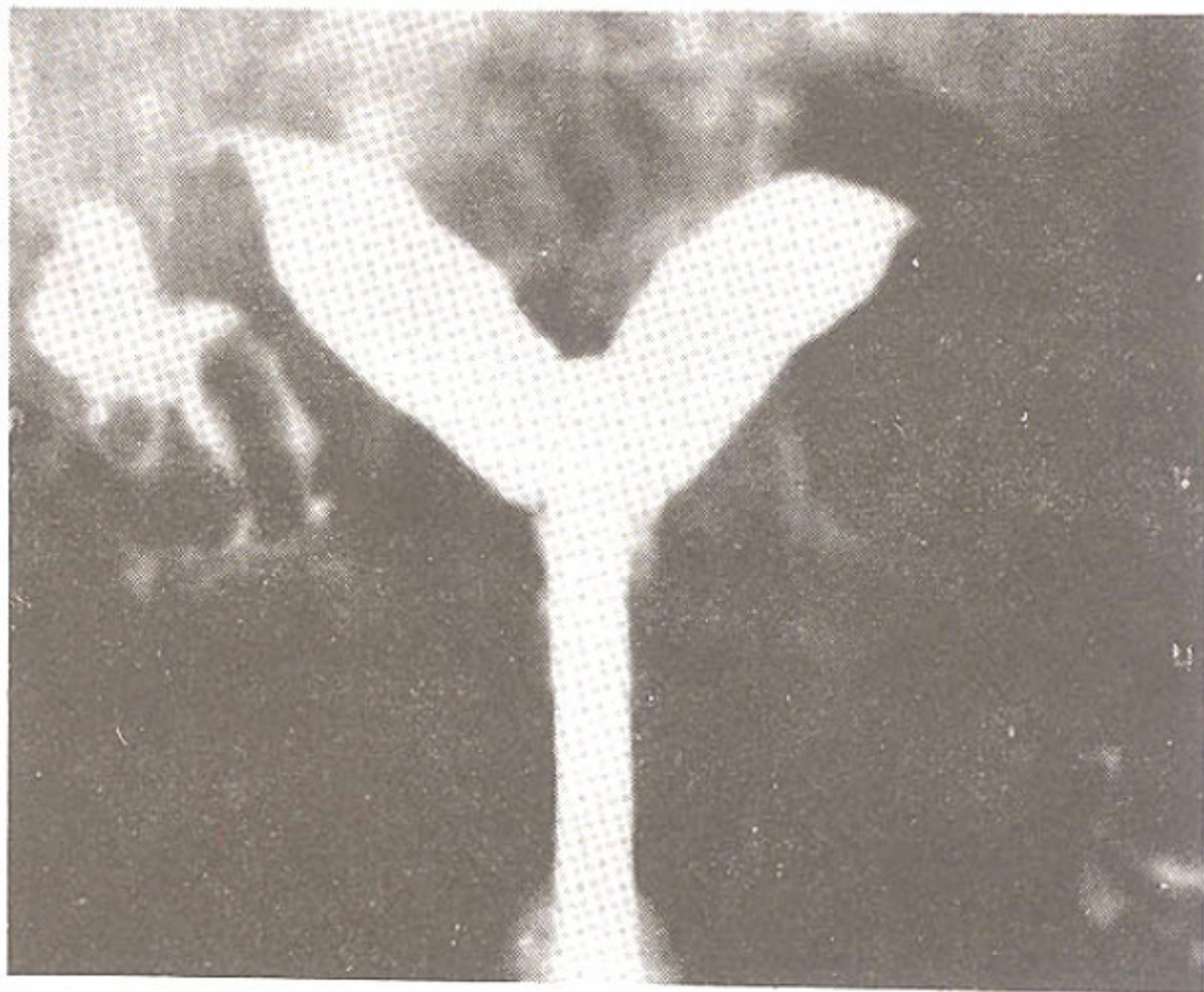


Figure 6. Case 9. Hystero-salpingogram showing uterus bicornis unicollis.

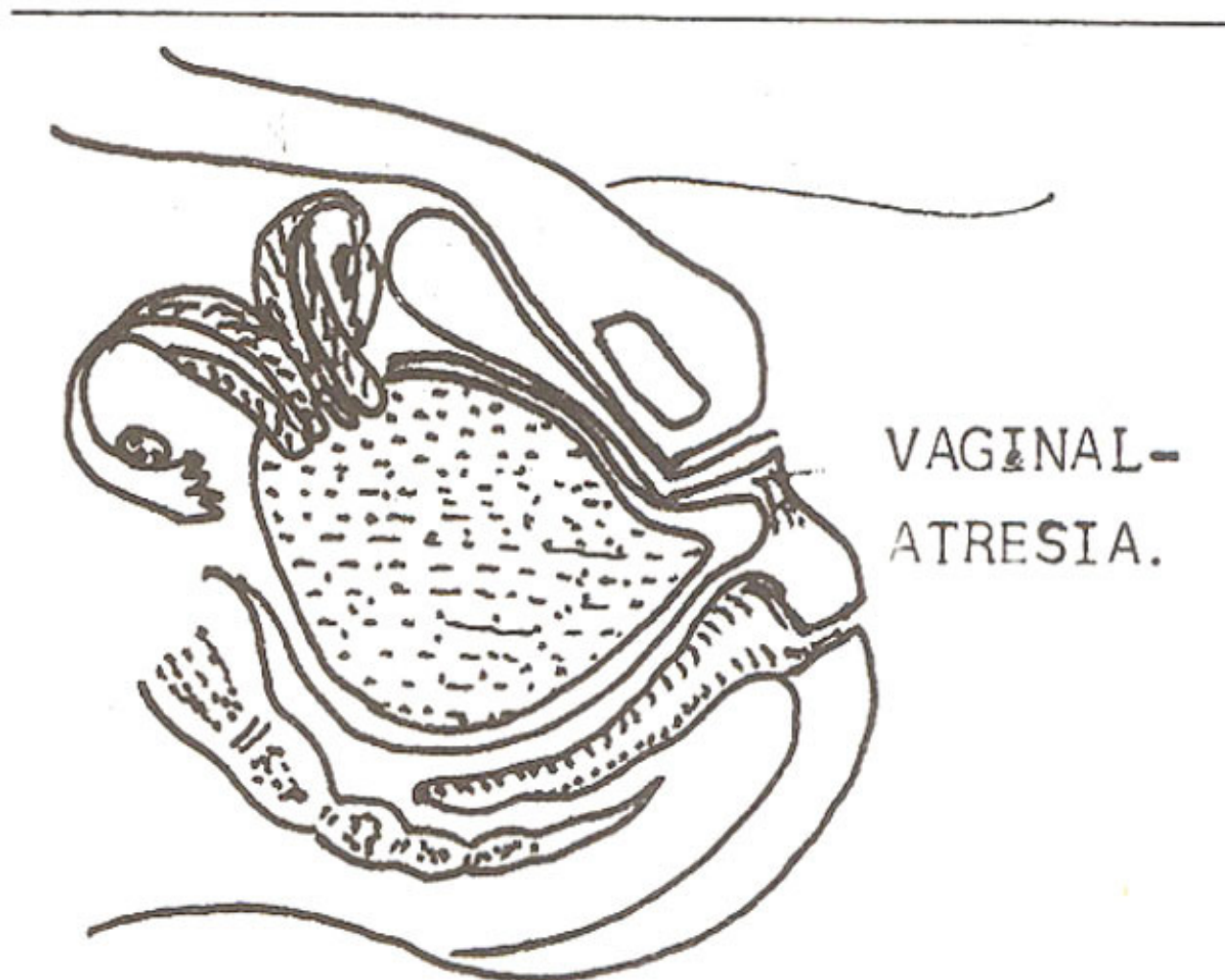


Figure 7(a). Case 10. Bicornuate uterus with haemato-
colpos and vaginal atrestia.

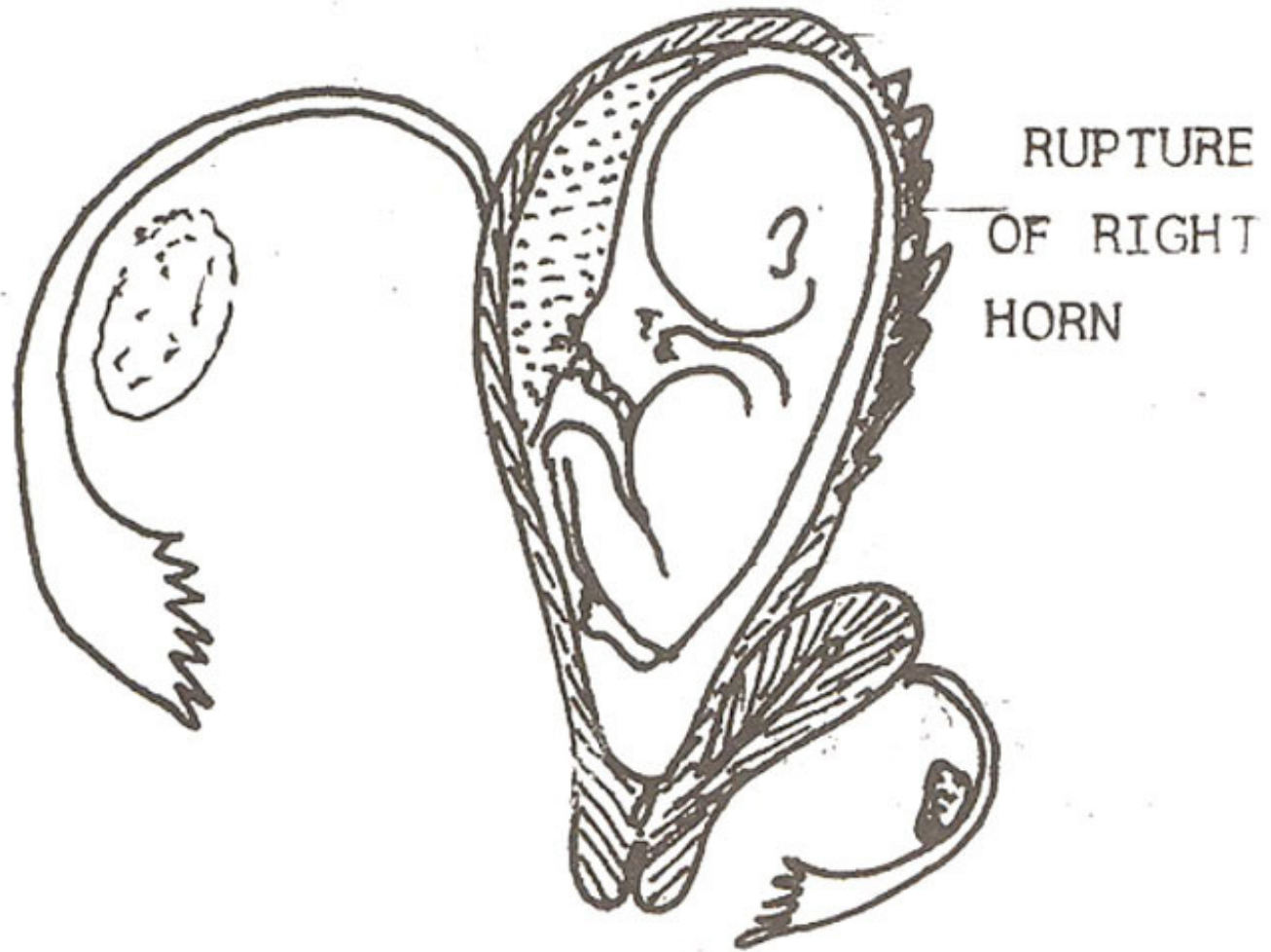


Figure 8. Case 11. Uterus bicornuas-unicollis.

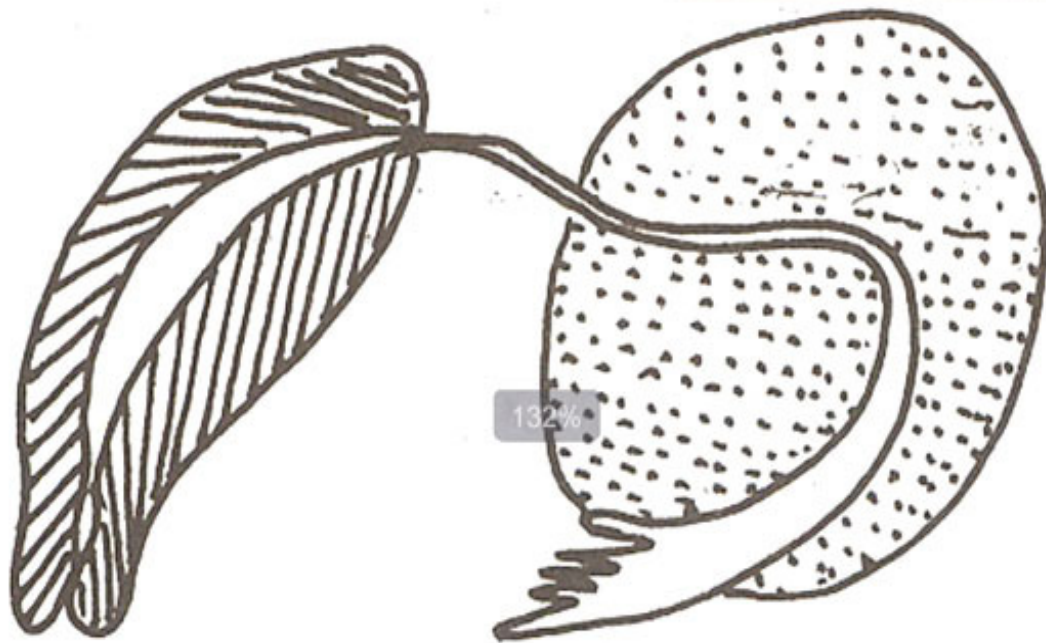


Figure 9. Case 12. Unicornuate uterus with ovarian cyst.

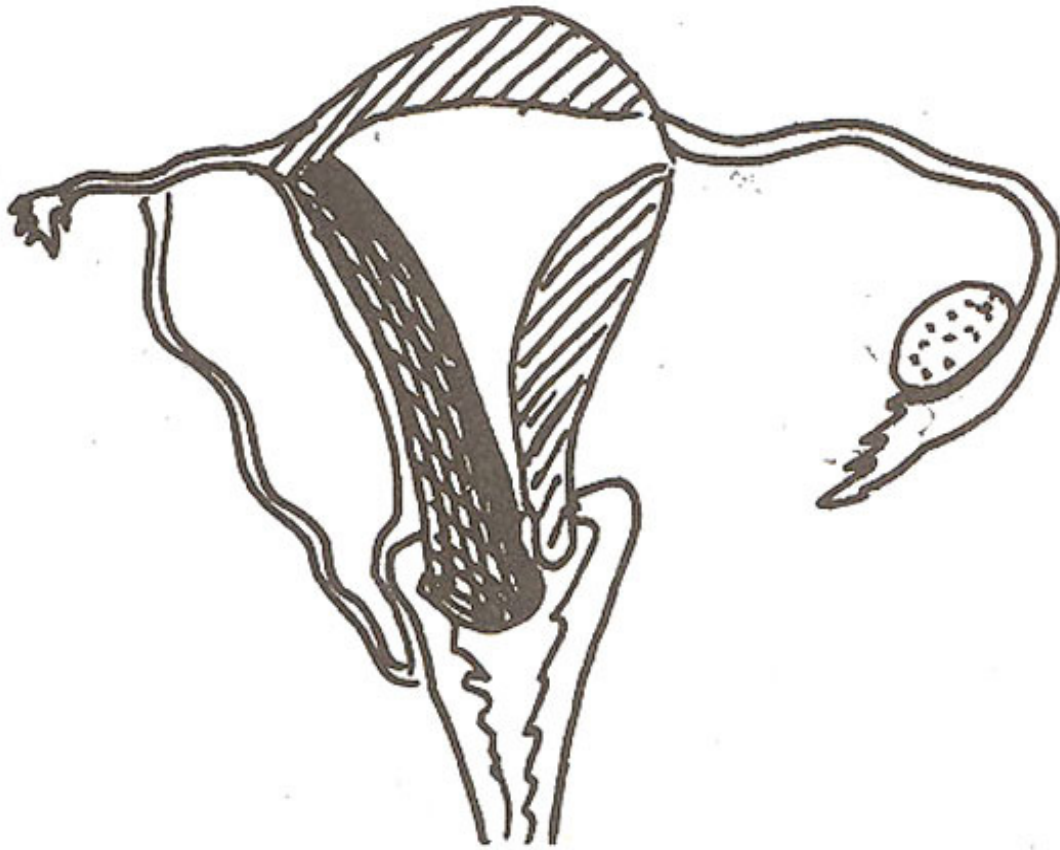


Figure 10. Case 13.

- Hypoplasia of right mullerian duct.
- Rudimentary right fallopin tube and round ligament.
- Undescended right gonad.

Bicornuate uterus may cause abortions, premature labour, dysmenorrhoea, dystocia, retention of gravid horn; utriculo-plasty was performed in case No. 10 and 11 which is standard technique and may have desired result. Patients should be kept under supervision in labour. Patients should be kept under supervision in labour.

Absence of one mullerian duct direct resulting in unicornuate uterus (Case No. 12) is an extremely rare condition. She had unicornuate uterus with benign cyst in the left ovary. She had brief history of menstruation earlier on, but was amenorrhoeic for past 12 years and did not respond to hormonal or clomid therapy by withdrawal bleeding. Her intravenous pyelogram was essentially normal. Her younger sister also suffers from a similar menstrual disorder thus suggesting a familial predisposition. Non-descent of gonad as in case 13, is a rare condition too, and undescended gonads either function for a short while or remain functionless. Hypoplasia of right mullerian duct resulted in poor development of right tube and probably non canalization of entire right wall of uterus and thickened posterior lip

cervix presenting as polyp. Non-descent of right ovary is explained partly by poorly development gubernaculum and eventually rudimentary right round ligament which is derived from it. Intravenous pyelogram showed hypoplasia of right kidney.

Abnormalities of mullerian ducts are closely associated with wolffian duct (Case No.13). Therefore all such cases should be scrutinised for congenital abnormalities of urinary tract.

CONCLUSION

It becomes apparent from this study that the recognition of congenital uterine malformations is not always possible unless a clue is provided by double cervix or vagina detected incidentally during minor or major gynaecological operations. This is because malformations of uterus may not always produce typical symptoms but their possibility has to be borne in mind in the cases of abortions, premature labours, cervical dystocia, unexplained severe dysmenorrhoea and shock in pregnant woman.

It is to be emphasised here that defects of external genitalia cannot provide a clue towards defects of genital organs above the hymen.

More recently ultrasound⁸ real time scanners with superior resolution have been found helpful for detection of congenital malformations of genital and renal tracts.

It is essential to realise that diagnosing female genital tract defects well in time can provide symptomatic relief, improve gestational performance of the affected individual and save lives in complicated cases.

Uterine malformations do not interfere with fertility of a woman.

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