CONGENITAL ABNORMALITIES OF GENITAL TRACT -
UTERINE MALFORMATION

Asif Zia Akhtar ( Department of Obstetric and Gynaecology, Abbasi Shaheed Hospital, Karachi.)

Abstract
Thirteen cases of congenital malformations of uterus were discovered during past ten years at Abbasi Shaheed Hospital, KMC, Karachi. They varied from uterus dideiphys 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 mullerian 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 mullerian ducts. The anomalies of mullerian ducts are genetically determined and are linked up with chromosomal make up of the individual, but they are also dependent on intra-uterine environment. Wilaon and Harris believed that two out of every thousand women have a sufficiently severe degree of uterine malformation to interfere with pregnancy. Greiss and Mauzy 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 mullerian ducts with didelphys uterus, 2 cases (5 and 6) had biffd 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 mullerian duct with undescended gonad of the same side and one case (13) was of unicorneuate 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>
<thead>
<tr>
<th>Type of Abnormality</th>
<th>No. of Cases</th>
<th>Case No</th>
<th>Presenting Complaint</th>
<th>Age</th>
<th>Marital Status</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>COMPLETE FAILURE OF FUSION OF MULLERIAN DUCT</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 Uterus Didelphys</td>
<td>4</td>
<td>1</td>
<td>Menorrhagia</td>
<td>16</td>
<td>S</td>
<td>Diagnostic D&amp;C</td>
</tr>
<tr>
<td>2 Post Coital bleeding P₀+₀</td>
<td>2</td>
<td>5</td>
<td>Abortion P₀+₀</td>
<td>22</td>
<td>M</td>
<td>Excision of Vaginal Septum</td>
</tr>
<tr>
<td>3 Post Coital bleeding</td>
<td>3</td>
<td>7</td>
<td>Tubo-Ovarian mass Pelvic Pain, Gravida 10</td>
<td>45</td>
<td>M</td>
<td>Total abdominal hysterectomy</td>
</tr>
<tr>
<td>4 Ectopic pregnancy Gravida – 4</td>
<td>4</td>
<td>9</td>
<td>Incomplete Abortion Gravida 4 Para Nil</td>
<td>29</td>
<td>M</td>
<td>Dilatation &amp; Curettage, Utriculoplasty advised</td>
</tr>
<tr>
<td>5 Uterus Unicomuate</td>
<td>1</td>
<td>12</td>
<td>Ovarian cyst secondary amenorrhoea (persistent) P₀+₀</td>
<td>22</td>
<td>M</td>
<td>Enucleation of ovarian cyst</td>
</tr>
<tr>
<td>6 Hypoplasia of Right Mullerian Duct</td>
<td>1</td>
<td>13</td>
<td>Polypoidal cervical lip and mass in abdomen (Cyst of Undescended Gonad) P₁+₀</td>
<td>28</td>
<td>M</td>
<td>Dilatation &amp; Curettage, Removal of cyst of undescended Gonad Follow up incomplete</td>
</tr>
</tbody>
</table>

* 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 dideiphys, 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 inco-ordinate uterine action was diagnosed necessitating lower segment caesarean section. Non pregnant horn of uterus dideiphys 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. Jones\textsuperscript{3} described uterus didelphys, obstetrically a bad uterus, but MonroKerr\textsuperscript{4} 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.

Buell\textsuperscript{5} and Perkin and Rubovitz\textsuperscript{6} 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.
Figure 4(a). Case 7. Right rudimentary horn has no connection with normal left horn of uterus.
Bifid uterus has a depression on its fundus with a single cavity and cervix, such malformations often are asymptomatic and hence pass unnoticed (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. Hunter 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 5. Case 8. Bifid uterus with single uterine cavity. 1 patient remained asymptomatic.
Figure 6. Case 9. Hystero-salpingogram showing uterus bicornis unicollis.
Figure 7(a). Case 10. Bicornuate uterus with haematocolpos and vaginal atresia.
Figure 8. Case 11. Uterus bicornuas-unicollis.

RUPTURE
OF RIGHT
HORN
Figure 9. Case 12. Unicornuate uterus with ovarian cyst.
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 amenorrhoic 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 ultrasound 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.

REFERENCES