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Congenital Abnormalities of Female Genital Tract

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Summary:

Objective: To observe the congenital abnormalities of female genital tract those are found in our population and their clinical implication.

Place of study: Faridpur Medical College Hospital, Faridpur.

Study period: January 2001 to December 2005.

Methods and Materials: During the study period all cases of congenital abnormalities of female genital tract diagnosed in Faridpur Medical College Hospital were studied.

Results: Total 74 women with different types of congenital abnormalities were detected. Though some cases were diagnosed incidentally, the various symptoms they produced were primary amenorrhea, apareunia, dyspareunia, hematocolpos and hematometra, infertility, prolapse uterus, abortions, ectopic pregnancy, premature labour and malpresentations. Among the 74 patients 35(47.3%) patients were treated at this centre, 24(32.4%) patients needed no treatment, 3(4%) patients were referred to higher centres for specialized treatment.

Conclusion: Different types of congenital abnormalities of female genital tract are found in our population which cause various symptoms and complications. Though not all abnormalities need treatment and some need very simple treatment, there are some cases which need specialized treatment and expertise.

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Introduction:

The incidence of congenital malformation of female genital tract is about 0.5% in female population.[1] 75% women with Müllerian abnormality will remain asymptomatic[2]. The remaining 25% will present with various symptoms. These are primary amenorrhea, hematocolpos and hematometra, dyspareunia, infertility, repeated miscarriage and obstetric complications.

The Müllerian abnormality can occur in isolation or in association with disorders of cloaca, urogenital sinus or anorectal areas because of their close embryological development.[3]

Proper assessment of these abnormalities needs a combination of clinical examination, examination under anesthesia, ultrasonography, hysterosalpingography and laparoscopy.[4] However magnetic resonance imaging should now replace hysterosalpingography and diagnostic laparoscopy as second line investigations.[5]

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The aim of this study is to observe the congenital abnormalities of the female genital tract those are found in our population and their clinical implications and also to share the experiences regarding their diagnosis, complications and management.

Methods and Materials:

This is a prospective hospital based observational study which was undertaken in Faridpur Medical College Hospital (FMCH) from January, 2001 to December, 2005.

All women with congenital abnormalities who: (a) attended in the outpatient department; (b) were admitted in the hospital with complication or for operative treatment; (c) were diagnosed during cesarean section or laparatomy were noted and studied. All women underwent clinical examination and some needed examination under anesthesia for assessment. Ulrtasonogram was done in every case to assess the uterine condition and to exclude associated renal abnormalities. Few patients presented here had laparoscopy or hysterosalpingography done in other centre of the country or abroad.

The patients with Müllerian anomalies were treated at this centre according to the facilities and expertise

available. Some patients were referred to higher centres. Some patients presented here already had treatment at other centres of home or abroad.

Limitation of the study: For assessment of the abnormalities, in addition to clinical examination and examination under anesthesia, ultrasonography is the only investigation done. Follow up study was not possible in all patients who were treated here as only few patients came for follow up.

Diagnosis and Results

During the study period, total 74 cases of congenital abnormalities of genital tract were detected.

The age of the women at presentation varied from 13 years to 45 years. None of the women presented before puberty.

Table-I shows the different abnormalities and their clinical presentations.

Table-I

Clinical Presentations of congenital anomalies of genital tract (n = 74)

Abnormalities	Number	Presentation
1. Müllerian agenesis with absent vagina	10	Primary amenorrhea apareunia, infertility.
2. Uterus Didelphys (a) with vaginal septum	(a) 2	Incidental diagnosis during laparotomy.
(b) without vaginal septum	(b) 1	Abortion.
3. (a) Arcuate	(a) 4	Incidental diagnosis,recurrent abortions,
(b) Septate	(b) 7	premature labor, malpresentations.
(c) Subseptate uterus	(c) 5	
4. Bicornuate uterus	2	Malpresentation, abortions, premature labour.
5. Unicornuate uterus	3	Incidental diagnosis, malpresentation
6. Unicornuate uterus with noncommunicated horn	5	Ectopic pregnancy.
7. Cervical aplasia	2	Hematomerta.
8. (a) Elongated cervix	(a) 9	Something coming down per vagina, infertility.
(b) Congenital uterovaginal prolapse	(b) 4	
10. Longitudinal Vaginal septum	4	Incidental diagnosis, postcoital bleeding, infertility,dyspareunia,
11. Transverse vaginal septum.	4	Hematocolpos, hematometra, dyspareunia.
12. Transverse vaginal band.	2	Incidental diagnosis during vaginal delivery.
13. Cribriform hymen.	1	Apareunia
14. Imperforated hymen	9	Hematometra & hematocolpos.
Total	74	

Müllerian agenesis with absent vagina was found in 10 women. All of them had normal secondary sexual characteristics. Among them 8 women were unmarried and presented with primary amenorrhea. The other 2 were married and presented with apareunia in addition to primary amenorrhea.

Uterus didelphys was found in 3 women, 2 of them had vaginal septum. One case was diagnosed at laparotomy which was done for twisted ovarian tumor. Two other cases were diagnosed during D&C for incomplete abortion.

Arcuate uterus was found in 4 women, all which were diagnosed incidentally during cesarean section.

Septate uterus was found in total 7 women.

Five cases of septate uterus were diagnosed during cesarean section, 2 with breech presentations, other 3 with cephalic presentations. Among them one woman had history of previous 3 consecutive abortions and one premature delivery. Her 5th pregnancy reached to term and had cesarean section but her 6th pregnancy again ended in late missed abortion. Her previous hysterosalpingography reported the uterus as bicornuate but during cesarean section the uterus was found septate. In 6th case, the woman was diagnosed as a case of ectopic pregnancy when she presented with early pregnancy, bleeding and ultrasonography reported the pregnancy sac was outside the uterine cavity. Laparotomy was done but there was no pregnancy sac outside the uterus and the uterus was found soft enlarged with wider fundus and depression at the centre indicating septate uterus. Repeat ultrasonography confirmed the intrauterine pregnancy which was ended in missed abortion. During D&C, presence of the septum in the uterus was confirmed.

The 7th case was diagnosed when a woman came for D & C as her ultrasonogram of uterus done for lower abdominal pain reported that she had a uterus with wider fundus and containing echogenic material in the middle indicating incomplete abortion. She had no clinical symptoms or signs of incomplete abortion. The woman had history of 3 vaginal deliveries at home, two by breech and one by cephalic presentation. All three children were alive. Clinical examination, analysis of her obstetrical history and careful repeat ultrasonography proved that the echogenicity in the middle of the uterus is nothing but the septum of a septate uterus.

Subseptate uterus was found in 5 women. One was found during manual removal of placenta. The other four were found during cesarean section, one with transverse lie, one with breech presentation and two with cephalic presentation.

Bicornuate uterus was found in two women. In one woman it was found during cesarean section for transverse lie. In another woman it was diagnosed by hysterosalpingography.

She had history of repeated pregnancy loss by abortions and premature labour.

Unicornuate uterus was found in 3 women during cesarean section, two done for breech presentations the other for foetal distress in cephalic presentation.

Unicornuate uterus with noncommunicating fused horn was found in 5 women. In one case the woman was admitted with missed abortion of about 22 week size. She was at first induced by intrauterine Foley's catheter but failed. Then she was given oxytocin drip, but the uterus ruptured. Immediate laparotomy was done and it was found to be a case of ruptured cornual pregnancy. Similarly the second case was diagnosed when laparotomy was done following rupture of cornual pregnancy which occurred when a woman with missed abortion of about 20 week size was induced with oxytocin drip. In the third case, D&C was attempted for early missed abortion when the uterus was found empty. Careful bimanual palpation revealed an adenexal mass. Laparotomy followed and the case was diagnosed as unruptured cornual pregnancy. Another case of unruptured cornual pregnancy was diagnosed when laparotomy was done in a woman with early pregnancy who had negative suction in attemted MR (Menstrual regulation) and ultrasonography showed live pregnancy sac outside the uterine cavity. The last case was diagnosed when laparotomy was done for ruptured ectopic pregnancy and finding it as a case of ruptured cornual pregnancy.

Cervical aplasia was found in 2 women. Both presented with amenorrhea and lower abdominal mass due to hematometra. In addition to cervical aplasia one had vaginal anus.

Elongated cervix was found in 9 women. They were diagnosed by finding the long vaginal cervix which is prolapsed and the deep vaginal fornices without having vaginal prolapse. Among them two were unmarried and seven were married. Among the married women two were nulliparous and five were parous. They had feeling of something coming down per vagina. One nulliparous married woman also complained of primary and the two parous women complained of secondary infertility. All parous women developed cervical prolapse after their first childbirth. Among them two developed secondary infertility but others had completed their family with prolapsed cervix.

Congenital 2nd degree uterovaginal prolapse was found in 4 women and they presented with infertility. One of them had ectopia vesicae.

Longitudinal vaginal septum was found in 4 women. One case was diagnosed incidentally during vaginal examination. The symptoms of other cases were dyspareunia, post coital bleeding and infertility.

Transverse vaginal septum was found in 4 women. Among them 3 women were unmarried and presented with amenorrhea and hematometra, hematocolpos. The other women was married and complained of dyspareunia. She had regular menses and examination under anaesthesia revealed a tiny opening in the septum.

Transverse vaginal band was found in two women incidentally during vaginal delivery.

Cribiform hymen was found in one woman who complained of apareunia. She had normal regular menses.

Imperforated hymen was found in 9 girls. All of them had amenorrhea with hematocolpos and hematometra.

Table-II shows the associated abnormalities of other system found along with Müllerian anomalies.

Fused pelvic kidney was found in one woman with Müllerian agenesis. One kidney was absent in two other women with Müllerian agenesis. Ectopia vesicae was found in one woman with 2nd degree uterovaginal prolapse. Vaginal anus was found in one woman with cervical aplasia.

Management and Discussion :

In Müllerian agenesis with absent vagina, creation of neovagina is indicated only when there is prospect of marriage and sexual activity. Before operation, proper counseling is needed regarding future menstrual function and fertility. In our society there is very little scope of this operation for these women. In this series, only two women were married and vaginoplasty was done in these two women. Habiba et al studied 45 cases of Müllerian agenesis from 1965 to 2002 at BIRDEM hospital Dhaka.[6] Among them only two patients were married with adequate vagina and needed no treatment. Plan was made for the remaining 43 patients to create a neovagina when they decide to go for a conjugal life.

Uterus didelphys was found incidentally in this study. In one woman uterus didelphys produced no symptoms and in other two women incomplete abortion occurred but whether it was due to uterus didelphys that could not be proved. No treatment was given for uterus didelphys. But the associated vaginal septa were excised.

Arcuate uteri were diagnosed incidentally producing no symptoms and needed no treatment.

Septate and subseptate uteri can be effectively treated by hysteroscopic resection. Septate and subseptate

Table-II

Associated other Anomalies

Associated Anomalies	Number	Müllerian Abnormalities
1. Fused pelvic kidney	1	Müllerian agenesis.
2. Absent one kidney	2	Müllerian agenesis.
3. Ectopia vesicae	1	2nd degree uterovaginal prolapse
4. Vaginal anus	1	Cervical aplasia.

uteri of this series which caused no symptoms or malpresentation were given no treatment. But the woman with septate uterus who had repeated pregnancy loss was referred to BIRDEM hospital, Dhaka.

Careful and transvaginal ultrasonography can avoid the mistake of finding the gestational sac outside the uterus by transabdominal ultrasonography (that occurred in this study) when actually it is in the one compartment of a septate uterus.

Metroplasty is considered in case of a bicornuated uterus when it causes repeated miscarriages. No treatment was given to women with bicornuate uterus of this study. The woman who presented with transverse lie and diagnosed during cesarean section was primigravida and had no previous history of pregnancy loss. She was properly counseled regarding her uterine abnormality and occurrence of repeated malpresentation and advised for proper antenatal check up during her next pregnancy. The other woman who had repeated abortions and premature labour was referred to Dhaka for confirmation of bicornuate uterus (because septate uterus appeared as bicornuate on hysterosalpingography), proper assessment and treatment.

Women with a small unicornuate uterus are unlikely to carry a pregnancy to term and have a high incidence of recurrent abortions.[7] In our study all 3 unicornuate uterus were found during cesarean section at term, two done for malpresentations and one for foetal distress. These unicornuate uteri causing malpresentation needed no treatment.

Unicornuate uterus with noncommunicating fused horn was found in five women. All of them presented with ectopic pregnancy but cornual pregnancy were evident only following laparotomy. Occurrences of ectopic pregnancy in these cases was by the process of external migration of ovum or sperm. In all cases the fused horns were excised.

There are very few reports of successful pregnancy in women with congenital absence of the cervix despite reconstructive surgery and hysterectomy is a more common outcome.[8]

Among the two cases in this series, one woman refused admission on the day of presentation at outpatient department and did not return back. For the other patient cervical reconstruction through abdomino-perineal route was planned for after proper counseling regarding prognosis and reproductive outcome. But the patient left the hospital by her own the day before operation.

In elongated cervix, amputation of the cervix was done in one unmarried woman who complained of severe symptoms, in one nulliparous married women who had infertility for 2 years and also in two parous women who had secondary infertility. No treatment was given to the other nulliparous woman who was recently married having no period of infertility. Vaginal hysterectomy was done in 3 other parous women who completed their family. Within 6 months of amputation of cervix, the unmarried girl and one of the parous women developed recurrent 2nd degree uterine prolapse. Manchester operation was done for congenital 2nd degree uterovaginal prolapse. The woman with ectopia vesicae and prolapse was operated in a private hospital at Dhaka along with reconstruction of the bladder. This woman had conceived and delivered by cesarean section. Following delivery she developed recurrent 2nd degree prolapse.

The two other cases, one operated in India and one operated here at this centre did not conceive but also developed recurrent prolapse. Several studies showed the complication of Manchester operation. Complications included dyspareunia, dysmenorrhea, recurrent uterine prolapse and enterocele.[9] Conception occurred in only 10% to 20% of patients following this operation.[10] Abdominal or laparoscopic sacrohysteropexy with synthetic mesh for uterovaginal prolapse in young women to preserve the uterus showed satisfactory results in different studies.[11,12,13,14] The last patient of congenital uterovaginal prolapse with infertility who presented recently and the women who developed recurrent prolapse following amputation of the cervix and Manchester operation were advised for sacro-hysteropexy with mesh and was referred as expertise are not available at this centre.

Septae were excised in all four cases of longitudinal vaginal septum.

Hematocolpos and hematometra produced by obstructive Müllerian anomalies if remain untreated, tubal damage and endometriosis may eventually occur.[15] Incidences of ectopic pregnancy will also increase.[16] In this series all cases of obstructive anomalies were presented 1 to 5 years following the age of menarche.

Vaginal reconstruction was done in all 4 cases of transverse vaginal septum by excising the septum. In two cases where the septum was thick advancement of vagina was done. During operation rectum was injured in one patient and two patients returned with recurrent vaginal stenosis and hematocolpos. Repeat reconstruction was done in both cases followed by amnion graft. A mould was prepared for each and given to them with advice to wear it for 6 months.

The vaginal bands found incidentally during delivery were excised.

The cribriform and imperforated hymens were excised.

Table-III shows the treatment summaries.

Table-III

Treatment Summaries of congenital anomalies of genital tract (n = 74)

	Number of patients	Percentage %
Treatment given at FMCH	35	47.3
Treatment given at other centre	2	2.7
Treatment not needed	24	32.5
Treatment refused	2	2.7
Patients referred	3	4
Treatment will be needed later	8	10.8
Total	74	100

Conclusion:

It is not very uncommon to find different types of congenital malformations of female genital tract in our practice. Various mistakes of diagnosis can be avoided by keeping in mind about these abnormalities during our practices. Early diagnosis and treatment of these conditions can prevent various obstetrical complications. Though not all abnormalities need treatment and some need very simple treatment, there are abnormalities which need proper assessment by various investigations and need specialized treatment and expertise which are not available everywhere in our country.

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