# CONGENITAL ABNORMALITIES OF GENITAL TRACT - VAGINAL DEFECTS

Pages with reference to book, From 256 To 261

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**Abstract**

Of 68,000 gynaecological cases seen during the past decade, only 25 had congenital malformations of the vaginal tract. The different types of congenital malformations, their presentation and management is presented (JPMA 36: 256, 1986).

## INTRODUCTION

Causes of most vaginal atresia are obscure. Three possible origins of such defects in XX females are familial transmission, teratogenic interference around 37th day and expression of recessive trait. Frequency of Congenital defects involving vaginal tract has been reported as ranging from 1 in 4,000 to 1 in 15,000 in the literature.[1]

Complete failure in the development of mullerian ducts causes absence of uterus, tubes and upper two-third and two-fifth of the vagina. [2] Lower one-third or one-fifth of the vagina is formed by canalization of sino-vaginal bulbs. in the former defect, the introitus is present and a depression exists in the lower part of the non-canalized vagina with normal development of vulva. Non-canalization of sino-vaginal bulbs results in complete absence of vaginal introitus.

Abnormalities of genital tract are reported to be closely associated with those of the woiffian ducts but developments of ovaries is embriologically different and hence is found to be normal in function and formation in spite of gross mullerian duct malformations.

In this study, an effort is made to discuss the frequency, symptoms and managements of various types of vaginal tract malformations.

## MATERIAL AND METHOD

Over 10 years period, from 1974 to 1984, 25 cases of Congenital Vaginal defects were encountered amongst 68,000 gynaecological cases seen in the out patients department of gynaecology at Abbasi Shaheed hospital.

Complete assessment of cases was carried out, including chromosomal analysis, an intravenous pyelography, when significant malformation were suspected. Surgical procedures consisting of simple operations on hymen, excision of transverse or longitudinal vaginal septa and vagino-plasty were performed as indicated.

All cases of primary amenorrhoea were subjected to examination under anaesthesia in order to assess the exact status of uterine development. Selection for vagino-plasty was based on the fact that the subjects were married or were contemplating marriage in the near future. Four cases under went vagino-plasty. One was done by simple dissection and dilation of the newly created vagina, but the results were not very encouraging. The other three vagino-plasties were carried out by using a loop of sigmoid colon, according to the method described by Goligher.[5] Major cases were followed for 6 months and the rest were followed for 6 weeks.

## RESULTS AND DISCUSSION

Congenital absence of the vagina was described by Realous Columbus in 1572. In 1817 Depuytren[3] attempted surgical correction of the defect. Since then different surgical methods have been used for example simple pressure technique[4], use of intestinal segment[5], full thickness and split thickness grafts[6], and creation of vulval space as described by Wffliam[7]. All procedures are useful with proper selection of patients.

Evan et a1[8] in 1981 published a series of 254 patients with vaginal malformations, of which 167 had total vaginal atresia. Nine percent cases had either absent or rudimentary uterii. Major urinary anomalies were demonstrated in 3.3% cases.

In the present study too, few cases of vaginal atresia (Table II)

TABLE-II
Cases of Vaginal Defects with associated Uterine Malformations .

| Case Number | Types of Vaginal Defects | Uterine Malformation | Gonads | Intravenous Pyelography Chromosomal Analysis | Further investigative procedures & Management |
|---|---|---|---|---|---|
| 1. | Upper 2/3rd of Vaginal atresia | Absent uterus and tubes | Palpable | 1. Normal IVP*<br>2. Barr bodies present | Examination under anaesthesia done |
| 2. | 2/3rd Vaginal atresia | Absent uterus and tubes | Palpable | Bifid pelvis of Kidney. Barr bodies present | Examination under anaesthesia done |
| 3. | Upper Vaginal atresia | Rudimentary Rt. horn of uterus and tubes | Normal with Corpus luteum | Normal IVP | Laparotomy done for Vagino-plasty |
| 4. | Upper Vaginal atresia | Non Canalized lower ends of mullerian ducts | Normal with Corpus luteum | Normal IVP | Laparotomy done for Vagino-plasty |
| 5. | Upper Vaginal atresia | Partly Noncanalized mullerian ducts | Normal with Corpus luteum | Normal IVP | Laparotomy done for Vagino-plasty |
| 6. | Upper Vaginal atresia | Rudimentary Unicornuate uterus | Normal ovary | Normal IVP | Laparoscopy |
| 9 to 12 | Longitudinal Vaginal Septa | All four cases had uterus didelphys | Normal Ovaries | Normal IVP | Examination under anaesthesia |
| 25 | Atresia of Lower 1/3rd of Vagina | Bicornuate uterus | Normal ovaries | Normal IVP | Laparotomy |

(* I.V.P. – Intravenous Pyelography)

were associated with absent or rudimentary uterus, and non-canalised mullerian ducts (Figures 1-3).

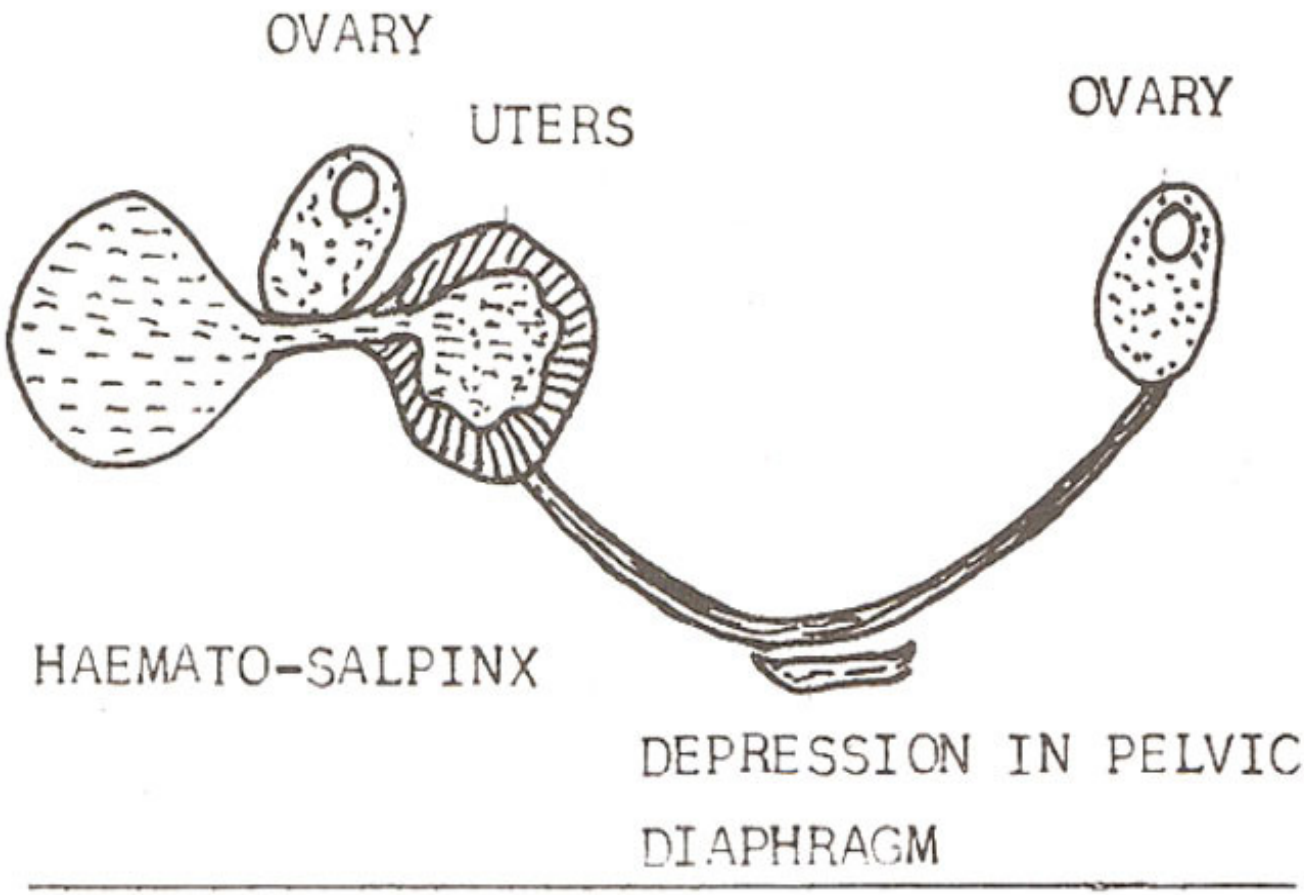


Figure 1. Case 3.

- Rudimentary right horn of uterus.
- Non-canalized left mullerian duct.
- Absent left tube.
- Both Ovaries showing corpus-luteum.

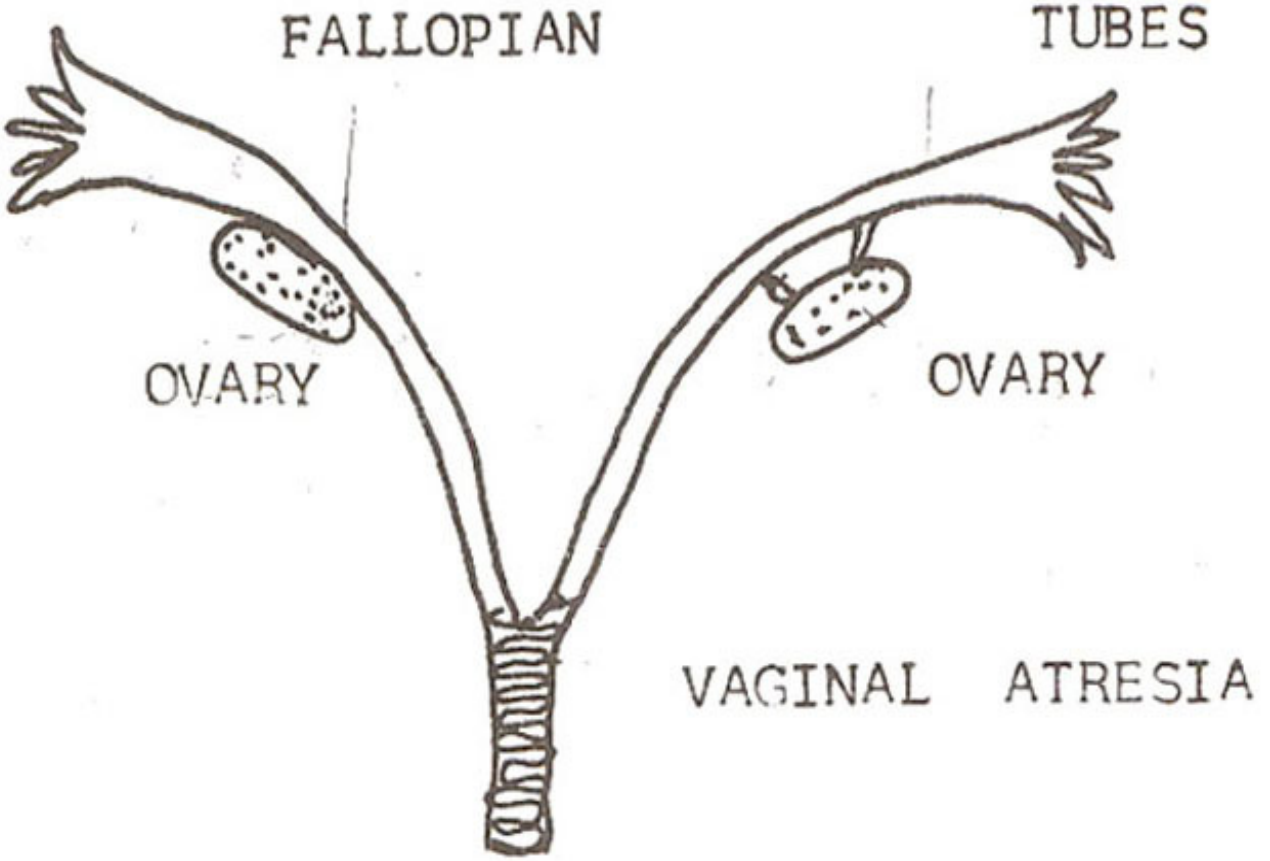


Figure 2.Case 4. Bilaterally Non-canalised and partly fused lower ends of mullerian ducts with normal ovaries.

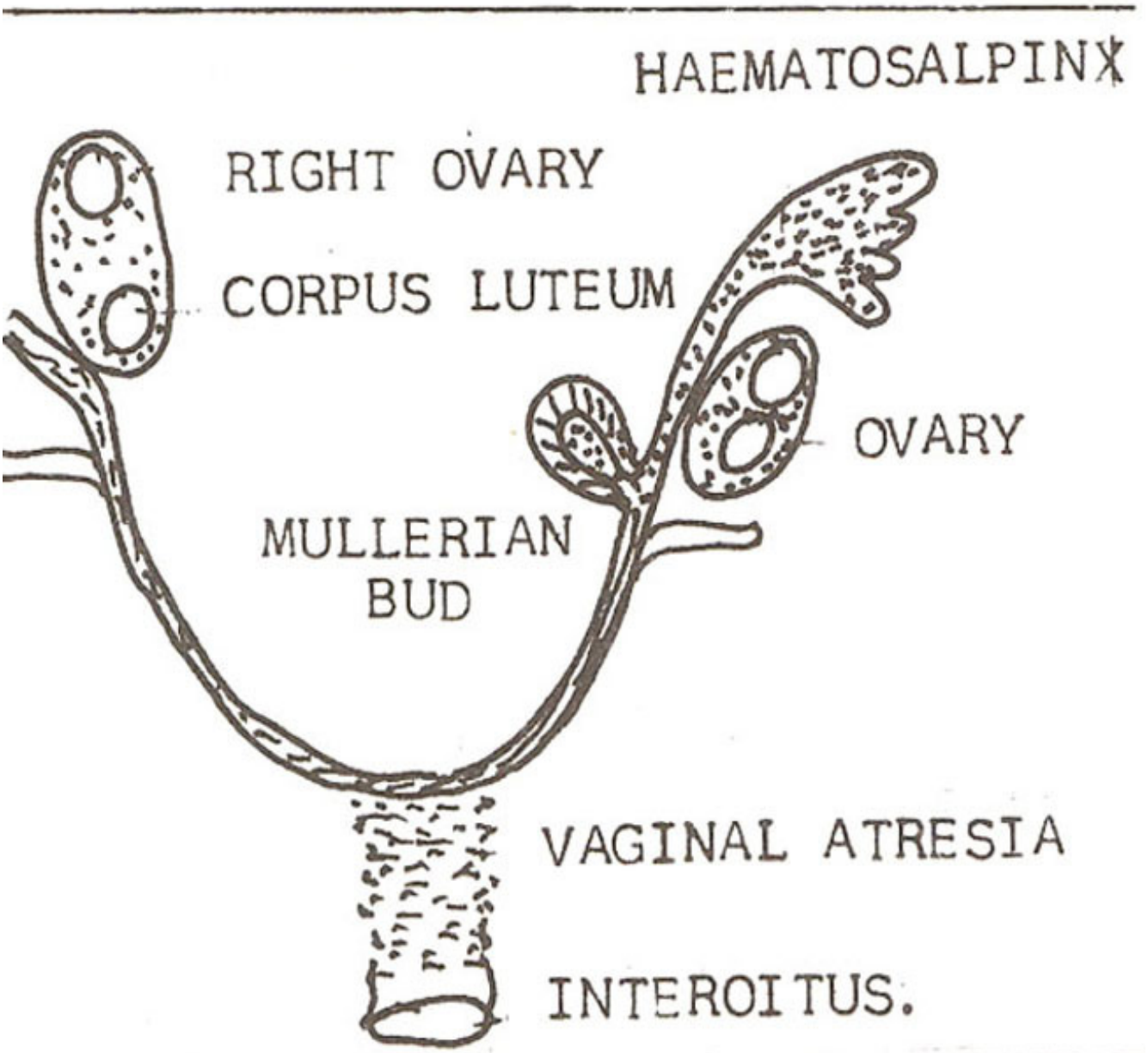


Figure 3.Case 5. Non-canalized right mullerian duct and left mullerian bud.

On intravenous pyelogram, only one case of bifid pelvis of kidney was detecied. Functioning uteri were usually present with lesser degree of vaginal malformations.

Isolated vaginal septum is probably the rarest of reproductive tract anomalies and is quite often a clue to the discovery of additional malformations[9]. Transverse septum may either be a focal manifestation or be associated with complete agenesis of mullerian duct.

Incomplete canalization of vaginal tract at different levels after the paried mullerian ducts have fused together, explain the conditions seen in case 71 (Figure 4).

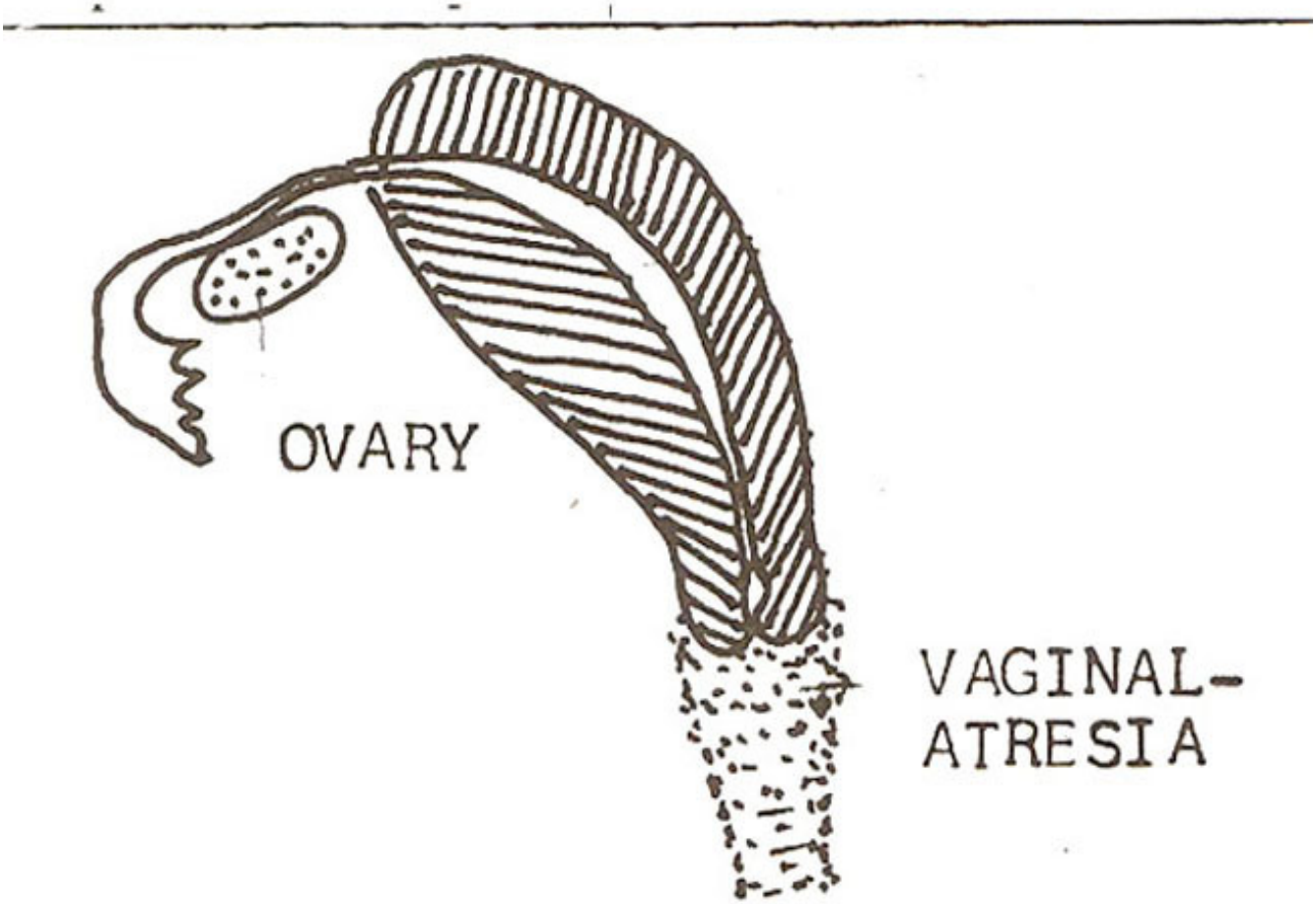


Figure 4. Case 6. Unicornuate uterus with vaginal atresia and a normal ovary.

In this case, excision of the septum was followed by drainage of chocolate coloured blood. The dilated uterine cavity was digitally explored and the cervical lips were not felt. The cervix was either not formed, or was fully taken up due to dilatation. It was probably a case of cervical atresia, which is rare[10] Patients get severe attacks of abdominal pain in cervical or proximal vaginal; atresia, whereas atresia of lower part of vagina can cause dilatation of upper vagina which can accommodate large amounts of blood without causing pain[11] (Figure 5).

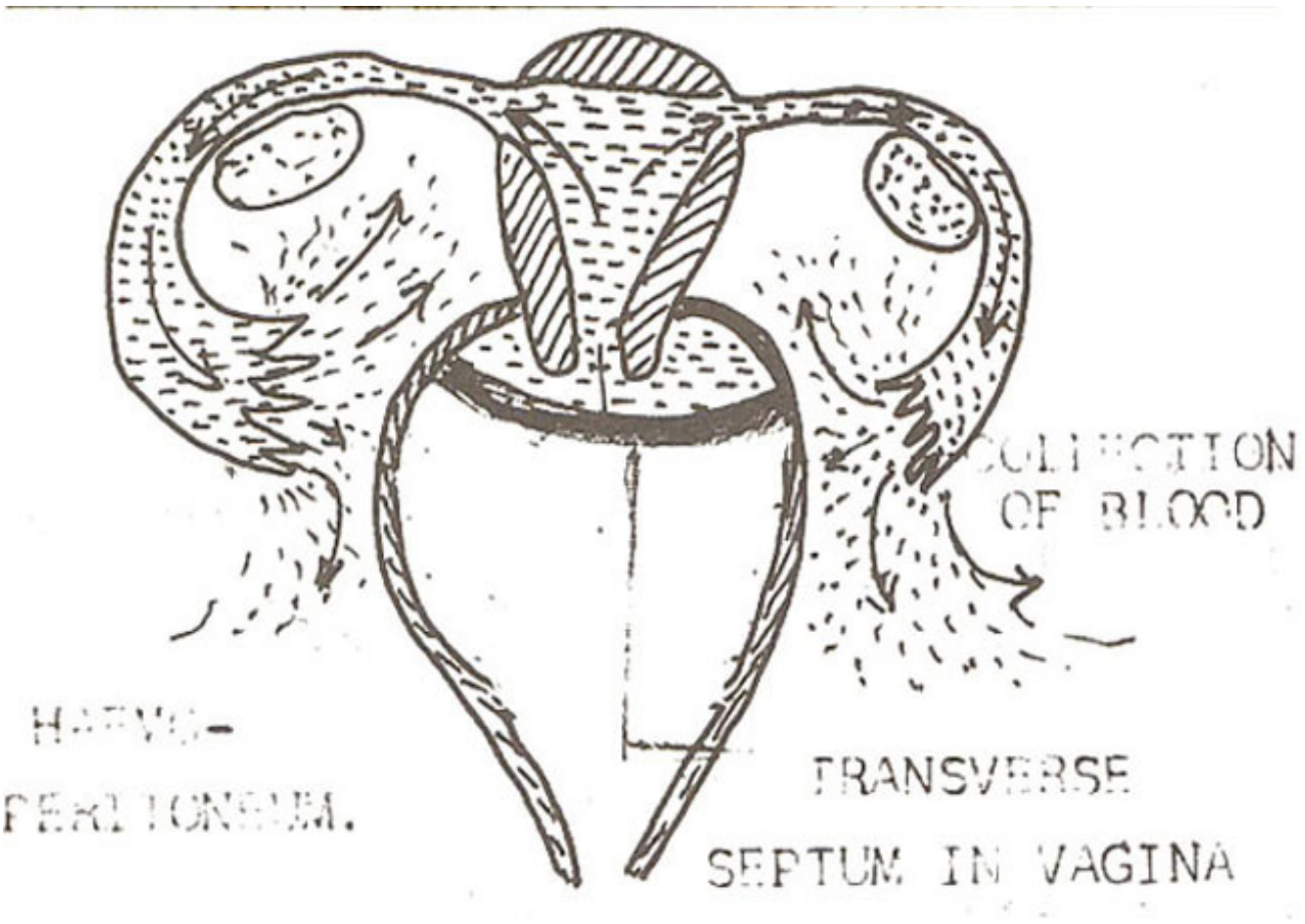


Figure 5.Case 7. Transverse septum in vagina causing obstruction to menstrual flow.

This fact was amply demonstrated in a young girl (Case 16) who presented at puberty with a painless cystic swelling rising upto the umblicus. She had marked difficulty in micturation. Needling was done elsewhere, but she again got filled up after 3 months. This time an operation for reconstruction of vagina was carried out, and the patient established normal menstrual blood flow but due to limited space between the urethra and rectum, the newly formed vaginal orifice was rather small and needed further surgical improvement before marriage.

Manorma[12] has quoted Rokitansky-Kuster Haus& syndrome which refers to a clinical entity comprising of normal female secondary sex characteristics, normal external genitals, congenital absence of vagina, rudimentary uterus in the form of bilateral non.canaliculated muscular buds, normal tubes and ovaries and normal endocrine cytogenetic disposition.[12] In some cases the two buds may be fused together in the mid line as a single bud. Case 4 (Figure 3) fits into this description.

Goligher[5] used a ioop of sigmoid colon as a pedicled graft in 7 cases of vaginal atresia. Similar technique was used in three of our cases with rudimentary uterus (Table I).

TABLE – I
Summary of Cases of Vaginal Defects.

| Type of Congenital Abnormalities | Serial Number of Cases | Presentation | Age | Marital Status | Management | Results |
|---|---|---|---|---|---|---|
| Atresia of upper 2/3rd Vagina | 1 | Amenorrhoea | 16 | S | Vagino-plasty advised later | |
| | 2 | Amenorrhoea | 28 | M | Not willing for operation | |
| | 3 | Amenorrhoea ( Fig. 1) | 18 | S | Vagino-plasty using loop of Sigmoid colon | Good |
| | 4 | Amenorrhoea ( Fig. 2) | 20 | M | Vagino-plasty done using loop of Sigmoid colon | Good |
| | 5 | Amenorrhoea (Fig. 3) | 32 | M | Vagino-plasty using loop of Sigmoid colon | Good |
| | 6 | Amenorrhoea ( Fig. 4) | 16 | S | Creation of Vaginal space by simple dis-section and dilation | Shortening of Vagina |
| Transverse Vaginal Septum | 7 | Acute Abdomen Haemoperitoneum and Haematosal-pinx Cervical atresia ( Fig. 5 ) | 12 | S | Excision of Septum | Good |
| | 8 | Obstruction to the head Presenting (Fig. 6) | 20 | M | Not willing for removal of septum L.S.C.S. done to deliver foetus | |
| Longitudinal Vaginal Septum | 9 | Asymptomatic (Fig. 7) | 22 | M | Excision of Septum | Good |
| | 10 | Menorrhagia | 13 | S | Excision advised before marriage | |
| | 11 | Post-coital haemorrhage | 20 | M | Excision of Septum | Good |
| | 12 | Post-coital haemorrhage | 22 | S | Not willing for operation | |
| Imperforate Hymen | 13, 14 15, 16 | Haemato colpos | 13 to 15 yrs | S | Cruciate incision of membrane | Good |
| | 17, 18 19, 20 | Nil | 11 to 12 yrs | S | Spontaneous development of Hymenal Opening | |
| | 21, 22 | | 6 mths to 1 yr | | Lost to follow up | |
| Fold of Hymenal membrane | 23 | Worried about Marital Life (Fig. 8) | 20 | S | Fenton Operation | Good |
| | 24 | Infertility | 28 | M | Fenton Operation | Conceived within a year of Operation |
| Atresia of lower 1/3rd of Vagina and introitus | 25 | Haematocolpos (Fig 9) | 13 | S | Reconstruction of lower part of Vagina and introitus | Fair |

Two of these patients were married and reported satisfactory marital adjustment, whereas the 3rd case was an unmarried girl and vaginoplasty was found to be satisfactory on six month follow up examination.

This method avoids formation of keloid and- scarring in the vagina. Lubricating secretions àf the

colonic graft had no disagreeable smell, though the procedure required ‘undergoing a laparotomy.

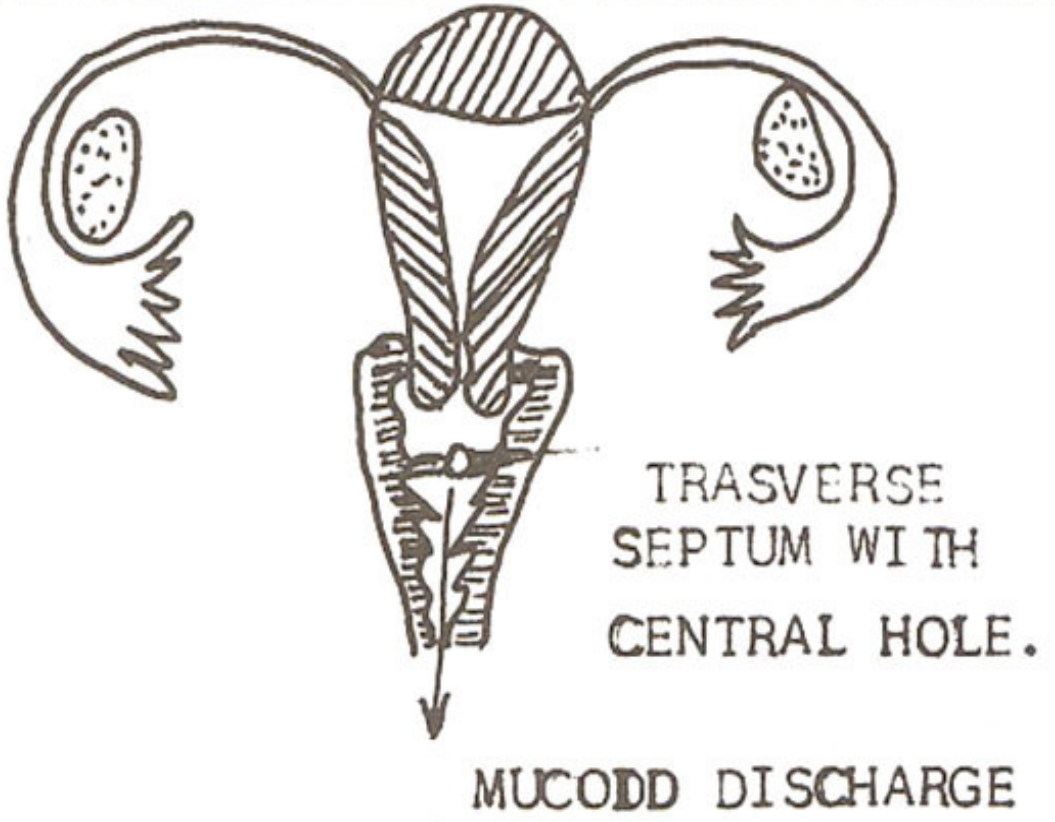


Figure 6. Case 8. Mucoid discharge trickling from a tiny hole in transverse septum in vagina.

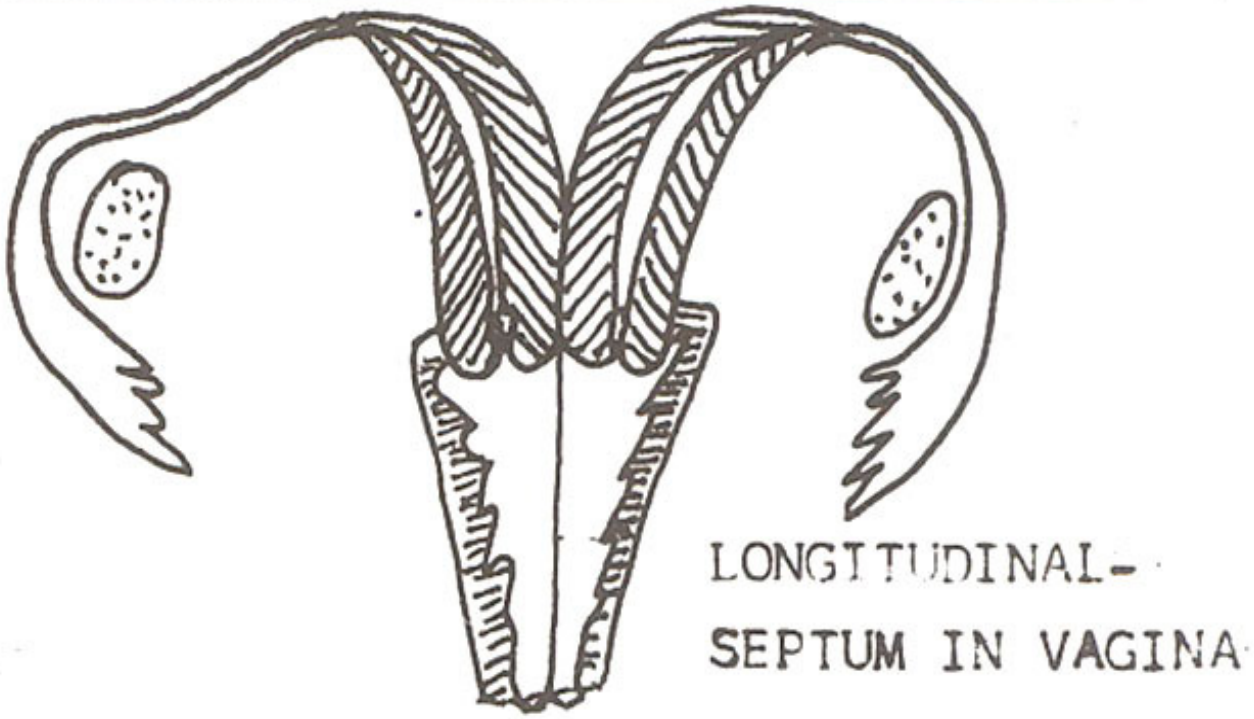


Figure 7. Case 9. Longitudinal vaginal septum with uterus didelphys.

Figure 8. Case 23. Folds of hymenal membrane covering vaginal orifice.

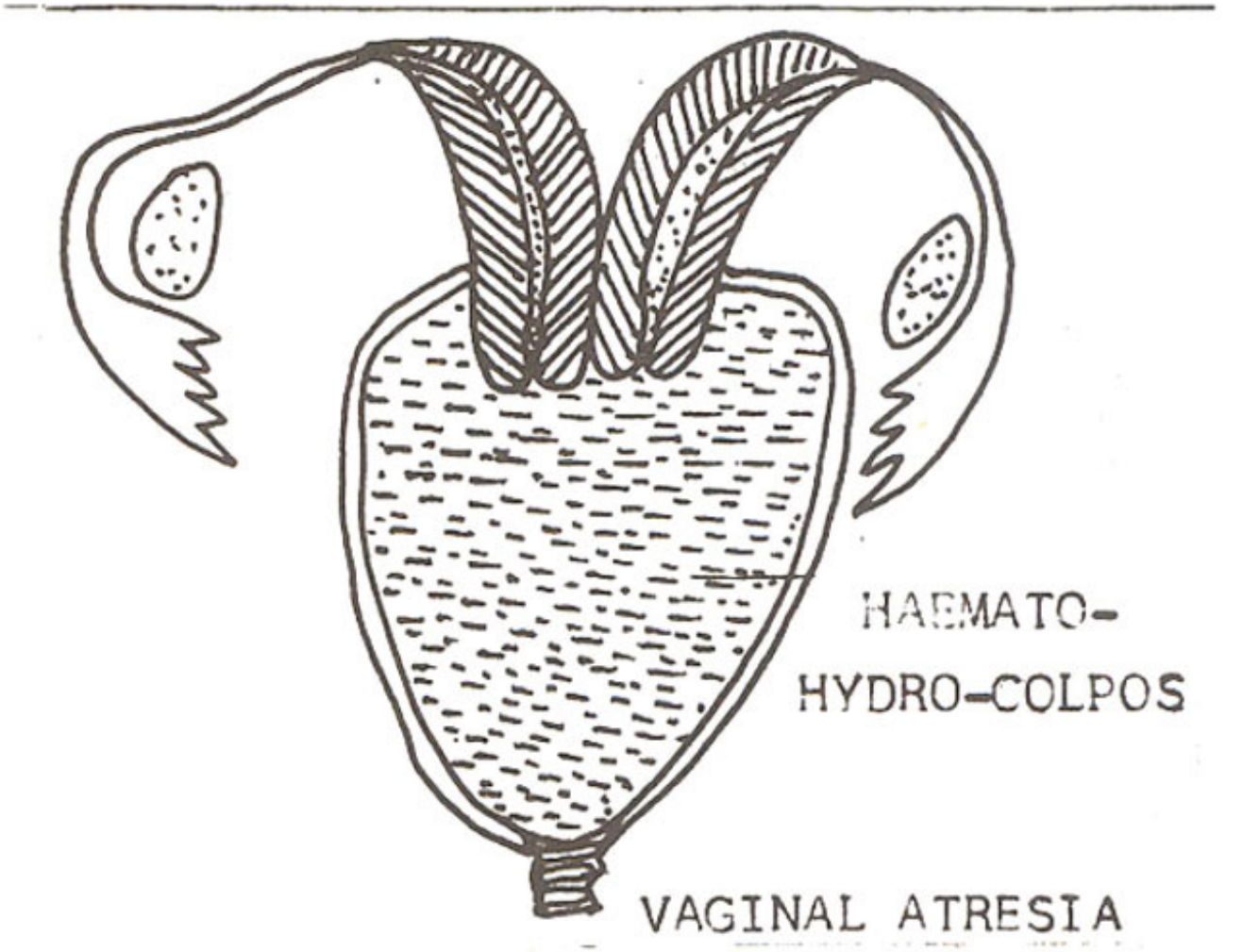


Figure 9. Case 25. Haemato-Hydro-Colpos causing distension of vagina presenting as tumour in abdomen.

Four cases of imperforate hymen noticed in childhood developed spontaneous opening of hymen (2 were sisters). The plane of cleveage developed from above downwards along the central line of fusion in the hymenal membrane.

It is concluded that cases with total vaginal atresia were mostly associated with absent or rudimenfary uterus. Clinically they present as cases of amenorrhoea. Development of external genitalia is of different origin. Thus abnormalities or external organs may give no clue of abnormalities above the hymenal ring. Similarly the formation and functioning of ovaries is inconsistent with a normal menstrual cycle in congenital absence of uterus.[13] Before selecting the type of operation for re-establishment of a functional vaginal tract, the uterine or gonadal status should be explored by laparoscopy. In cases of testicular feminization the gonads should be removed because of the risk of malignancy. The psychological and socio-sexual overtones to this problem should be considered. Proper operative procedures carried out in well selected cases can be very useful for successful marital relationships.

**REFERENCES**

1. Bryan, A. L., Nigro, J.A. and Councellor, V.S. One hundred cases of congenital absence of the vagina. Surg. Gynaecol. Obstet., 1949; 88: 99.
2. Smith-Wendall, C.P. and William, P.L: Development of Gonads and ducts. 3rded. London, Pitman, 1984; p. 97.
3. Whitacre, F.E. and Chen, C.Y. The surgical treatment of absence of Vagina. Report of two cases. Am.J. Obstet.Gynecol., 1945;49: 789.
4. Frank, R.T. The formation of an artificial vagina without operation. Am. J. Obstet. Gynecol., 1938; 35:105 3.
5. Goligher, J.C. The use of pedicled transplants of sigmoid or other parts of the intestinal tract for vaginal construction. Ann. R. Coil. Surg. Engl., 1983; 65:35 3.
6. Whitley, T.M., Parrott, M.H. and Rowland, W. Split thickness skin graft tedinique in the correction of congenital or acquired vaginal atresia. AmJ.Obstet.Gynecol., 1964;89: 377.
7. William, E.A. Congenital absence of vagina. A simple operation for its relief. J. Obstet. GynaecoL Brit. comm., 1964; 71: 511.
8. Evans, T.N., Poland, M.L. and Boving, R.L. Vaginal malformations. Am. J. Obstet.Gynecol., 1981; 141: 910.
9. Greiss, F.C.Jr. and Mauzy, C.H. Genital anomalies in women . An evaluation of diagnosis, incidence, and obstetric performance. Am. J. Obstet. Gynecol., 1961; 82: 330.
10. William Bryan: Congenital Cervical and Vaginal atresia. J. Obstet. Gynaecol. Brit. Comm., 1963; 19: 301.
11. Jeffcoate, T.N.A. Principles of gynaecology. 3rd ed. London, Butterworths, 1967; P. 171.
12. Purwar, M.B., Deshpande, A.S. and Jawade, S.B. Rokitansky-Kuster-Hauser syndrome with fibromyoma arising in rudimentary buds. Asia Oceania J. Obstet. Gynaecol., 19$3;9: 55.
13. Macrae Mohammadally: Ovarian functions in congenital absence of the uterus. J. Obstet. Gynaecol. Brit. Comm., 1969; 76: 114.