ABSTRACT: INTRODUCTION: Bicornuate uterus is a congenital malformation caused by dysgenesis and fusion defects having two uteri and one cervix [bicorns, unicollis]. **METHOD:** Among 446 hysterectomy specimens received between April 2010 to April 2013, there were four specimen of bicornuate uterus. **OBSERVATION:** In this study, along with bicornuate uterus, we observed other associated pathological conditions as follows, 1) Fibroids and adenomyosis, 2) Tubercular endometritis and bilateral tubercular salpingitis. 3) Non-communicating rudimentary horn with hematometra and 4) Endometriosis. **CONCLUSION:** As there is lot of debate on the symptomatology, in malformed uteri, associated other pathological conditions have to be kept in mind, which are treatable and can reduce the incidence of infertility and other complications. **KEYWORDS:** Bicornuate uterus, fibroid, adenomyosis, tuberculosis.

INTRODUCTION: Uterus develops from mullerian [paramesonephric] duct. Mullerian duct anomalies are congenital, which will be either due to 1] Aplasias and dysgenesis. 2] Fusion defects. 3] Failure of septum dissolution. 4] Failure of canalization. Some of these anomalies will have combined pathogenetic pathways.\(^1\) Over all the incidence of uterine malformations varies from 0.16 - 10%.\(^2\)

Bicornuate uterus is also known as bicorns and unicollis is characterized by a single cervix and single vagina. This anomaly is due to combined pathogenetic pathway of both dysgenesis and fusion defects. Majority of the women with the malformations of the genital tract are asymptomatic, a small minority will present with symptoms directly related to the presence of malformation, while others may suffer reproductive problems which are consequence of uterine anomaly.

However there is lot of debate and conflicting studies as for the symptoms and reproductive capabilities are concerned. The present study is intended to stress on the other pathological conditions which will contribute to the symptoms and fertility of women with bicornuate uterus.

**MATERIALS AND METHODS:** Among 446 hysterectomy specimens received between April 2010 and April 2013, there were four bicornuate uterus. All the clinical, radiological and pre-operative details were obtained. Clinical details were analyzed with histopathological findings.

**Case 1:** A 40 year old female presented with menorrhagia, metrorrhagia and underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy. Ultrasonography showed bicornuate uterus with fibroids. Histopathological examination revealed two intramural fibroids, the larger one measuring 3x2 cms, thick myometrium with trabeculations. [Figure 1]. Microscopy confirmed adenomyosis [Figure 2a] and leiomyoma [Figure 2b]. Both ovaries showed follicular cysts.
**Case 2:** A 60 year old post-menopausal female presented with lower abdominal pain, weight loss, underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy. Ultrasonography and gross appearance showed thick endometrium measuring 0.9 cms, suggested suspicion of endometrial hyperplasia or carcinoma [Figure 3]. Microscopic examination revealed multiple epitheloid cell granulomas, Langhan’s type of giant cells and areas of necrosis [Figure 4]. These granulomas were extended into myometrium and were also seen in the both fallopian tubes. Both ovaries were unremarkable.

**Case 3:** A 30 years female presented with dysmenorrhea, underwent abdominal hysterectomy with unilateral salpingo-oophorectomy. Ultrasonography showed bicornuate bulky uterus. Histopathological examination revealed uniformly thick myometrium with trabeculations [Figure 5]. Microscopy confirmed adenomyosis. [Figure 6]. Ovary was unremarkable.

**Case 4:** A 35 years female presented with severe lower abdominal pain during cyclical menstrual period and oligomenorrhea, underwent abdominal hysterectomy. Ultrasonography showed bicornuate uterus with one horn showing dilated endometrial canal. Histopathological examination revealed one non-communicating horn filled with blood [Figure 7].

**DISCUSSION:** The mullerian or paramesonephric ducts arise as longitudinal invaginations of the coelomic epithelium along the anterolateral surfaces of the nephrogenital ridges in the posterior abdominal wall. These develop into the main genital ducts of the female. Initially three parts can be recognized in each duct (a) a cranial vertical portion that opens into the abdominal cavity (b) a horizontal part that crosses the mesonephric duct (c) a caudal vertical part that fuses with its partner from the opposite side with the decent of the ovary. The first two parts fuse to form the uterine tube and the caudal parts fuse to form the uterine canal.³

By convention the term ‘malformation’, when applied to female genital tracts, refers to the anatomical abnormalities of the tract in individuals who have no phenotypic, sex chromosomal, gonadal or endocrinological abnormalities.¹ There have been number of attempts to classify the malformation of female genital tract, these usually being based upon the known or presumed mechanism of their origin.⁴ ⁵ ⁶

In general, female genital tract female malformations can be considered under four headings:

1. Aplasia and dysgenesis
2. Fusion defects
3. Failure of septum dislocation
4. Failure of canalization.

Genetic factors may play a role in the development of some anomalies. There have been a number of reports like familial mullerian duct fusion defects and familial Mayer-Rokitansky-Kuster-Hauser syndrome.⁷ ⁸ ⁹

Among the overall incidence of uterine malformation which varies from 0.16%-10%, the incidence of bicornuate uterus noted in Stompe-Sorensen (1988) study is 1.2% of 167 women
undergoing laparoscopic steralisation. In our study incidence accounts to 0.9% of all hysterectomy specimens.

As far as clinical presentation of non-obstructive fusion defects are considered, many will be asymptomatic. Patients with a functioning non-communicating rudimentary horn can develop a haematosalpinx and suffer severe dysmenorrhoea. One of our patients presented with non-communicating horn, hematometra and severe dysmenorrhoea.

There is no doubt that many, probably most women with such uterine malformations have normal reproductive history and the literature abounds with reports of normal full term pregnancies in unicornuate, bicornuate, non-fused and septate uteri.

Despite this documented reproductive success in women with malformed uteri, there is still considerable debate about infertility, early pregnancy loss, premature labor and intrauterine growth retardation and pre-eclampsia. This association of uterine malformations with endometriosis is well known entity, but is in fact only true for those cases in which there is obstruction to the menstrual out flow. This is one of the strongest arguments for the menstrual reflex hypothesis of the pathogenesis of endometriosis. But in the present study there were two non-obstructive cases, one associated with fibroids showed endometriosis.

CONCLUSION: As there is lot of debate on the symptomatology, reproductive capability, infertility, abortion, premature labor and intrauterine growth retardation in malformed uteri, associated pathological conditions like endometriosis, fibroids and tuberculosis have to be kept in mind, which are treatable and the incidence of infertility and complications can be reduced.

REFERENCES:

Figure 1: Gross appearance of bicornuate uterus two fibroids and thick trabeculated myometrium
Figure 2:
  a. Shows endometrial glands and stroma [arrow] amidst the myometrium [adenomyosis]. (H & E stain. 40X)
  b. Benign spindle cells arranged in interlacing bundles and fascicles [Leiomyoma] (H & E stain. 40x)

Figure 3: Bicornuate uterus showing irregular thickening of endometrial cavity with gray white granular and cheesy material

Figure 4: Microphotograph showing epitheloid cell granulomas with Langhan’s type of giant cells [thin arrow] and caseous necrosis [thick arrow]. (H & E stain.10x)
Figure 5: Bicornuate uterus showing hypertrophied myometrium with trabeculations suggestive of extensive endometriosis [arrow]

Figure 6: Endometrial gland and stroma with areas of hemorrhage within the myometrium. [adenomyosis] (H & E stain. 40x)

Figure 7: Endometrial gland and stroma with areas of hemorrhage within the myometrium. [adenomyosis] (H & E stain. 40x)