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CASE REPORT

# Complex Müllerian malformation: report of a case with a hypoplastic non-cavitated uterus and two rudimentary horns

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**Various classifications have been used for congenital anomalies of the Müllerian system. We report a case of a previously unknown anomaly of the uterus, and propose its possible embryological causes. The patient presented with primary amenorrhoea and infertility, and during laparoscopy three distinct uterine horns were observed. The tubes were connected to the two most lateral horns, each juxtaposed to a normal ovary. The middle horn had a seemingly normal attachment to the right uterosacral ligament, whereas its attachment to the left uterosacral ligament appeared attenuated and less normal. Furthermore, the right horn was immediately attached to the middle horn, whereas the left horn was, like its ipsilateral uterosacral ligament, attached to the middle horn by a more attenuated, stretched fibrous bridge. Only the middle horn, with its uterosacral ligaments, had an attached, although obstructed, cervix. Ultrasonographic examination revealed no endometrium echogeneity in any of these uterine bulbs. No etiologic factors were noted in the patient's history; her mother denied known ingestion of estrogens or other drugs while carrying her daughter. The pathogenesis of this anomaly cannot be clearly defined, but may involve sequential embryological errors of duplication of the Müllerian tracts, failure of fusion of each set of the Müllerian tracts with expected failed canalization of each tract and, finally, agenesis of the medial horn of the left duplicated tracts.**

*Key words:* Müllerian defects/rudimentary horn/uterine anomaly

## Introduction

Congenital anomalies of the Müllerian system are common defects, reported in up to 3.2% of all women (Simon, 1991). Many are asymptomatic and therefore unrecognized. Diagnosis is usually made with clinical presentations of infertility, miscarriage, premature birth, abnormal fetal presentation, cyclic pain, or as incidental findings during pelvic or monographic examinations. They occur due to partial development of the Müllerian ducts. Increasing degrees of abnormal lateral fusion of the ducts can form the arcuate uterus, bicornuate uterus and uterus didelphys. Such anomalies can be symmetric or asymmetric, and the uterine horns obstructive or non-obstructive. Failure of fusion may lead to failure to cannulate, with no endometrium developing within the uterine horns. Any portion or segment of a duct may experience agenesis.

Various classifications have been proposed for congenital uterine anomalies based on the understood embryology of the Müllerian system (Buttram and Gibbons, 1979; American Fertility Society, 1988). However, our review of the literature has failed to produce a 'uterine triplication'; we herein report the first known case of an amenorrhoeic, infertile woman with three uterine horns, the 'tricornuate uterus'.

## Case report

A 26-year-old woman presented for evaluation of primary amenorrhoea and infertility. Physical examination revealed that breast development, axillary and pubic hair were normal. The vagina was normal, but the sole cervix was small in size and its lumen not patent. Bimanual examination revealed a hypoplastic uterus and bilateral adnexal masses. Karyotype was 46 XX. FSH, LH, estradiol and free testosterone levels were all suggestive of the female reproductive pattern. Ultrasonography revealed the 'uterus' was 16×22×37 mm and no endometrium was observed. The lateral masses measured 17×42 mm and 16×28 mm and no endometrial echogenity

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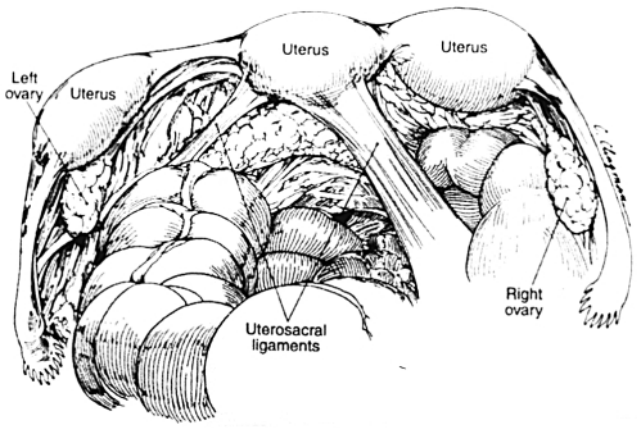



**Figure 1.** Schematic drawing of the complex Müllerian anomaly reported in a 26-year-old woman.

was observed in either mass; the ovaries, being displaced laterally, were not imaged on initial sonography.

At subsequent laparoscopy, three small uterine horns were observed in the pelvis (Figure 1). The middle horn was slightly deflected to the right side of the pelvis and had two uterosacral ligaments—the right one being thicker and much more normal in appearance. The right lateral horn was attached by a short fibrous bridge, as seen in uterus didelphys, with a normal Fallopian tube and ovary attached to it. The left lateral horn was displaced more laterally and connected to the dominant mid-horn via a more attenuated bridge, with its normal ovary and tube attached. There was no endometrium seen on sonography, and no haematometra or endometriosis at laparoscopy despite normal estrogen levels, suggesting a complete absence of endometrium. Hysteroscopy could not be performed through the impatent cervix. With no indication for hysterectomy, the procedure was concluded. The woman is followed on an outpatient basis, and is considering the use of a surrogate mother. Intravenous pyelography demonstrated no concomitant urinary system malformation.

## Discussion

During fetal development, by week 5 of gestation, the urogenital ridges develop from mesoderm on the dorsal of the coelemic cavity. Paramesonephric (Müllerian) ducts begin to appear lateral to the mesonephric ducts at 7 weeks gestation. While each duct forms a solid bud, the caudal end of each Müllerian duct crosses the Wolffian duct. Eventually Müllerian ducts fuse in the midline and the uterus is formed at about week 10, with the fusion beginning in the middle of the uterus and proceeding cephalad and caudad. The unfused cranial parts of the ducts differentiate into Fallopian tubes. The fused Müllerian ducts descend to the urogenital sinus and join, and the resorption of the septum between ducts then begins in the isthmus of the uterus and extends cranially and caudally (Muller *et al*., 1967). It has been reported that canalization occurs following fusion (Davis *et al*., 1992).

In the literature, only one case has been reported that is similar to ours, which was about a patient having a unicornuate uterus with two cavitated, non-communicating rudimentary uterine horns (Nezhat and Smith, 1999). However, the described horns are stacked one on top of the other. In our case there were three non-caviated, non-communicating rudimentary horns laterally placed. Our best theory of the embryogenesis of this anomaly is as follows: each Müllerian duct duplicates, and the medial duplicated tract of each pair descends towards the urogenital sinus. The one tract that reaches the sinus forms the rudimentary cervix and its right-sided normal uterosacral ligament. The medial tract of the left pair fails to reach the urogenital sinus and undergoes atresia, producing far more attenuated ligamentous attachments than the right pair of tracts. Failure of fusion follows, then failure of canalization. Without the agenesis of one horn, the probable result would have been uterus didelphys with each horn having an attached rudimentary horn as well. Obviously, since we do not have the histological diagnosis one cannot rule out the type of anomaly that may also favour a rudimentary horn on the left side and adenomyotic nodule on the right. However, attached tubes and ovaries bilateral to the rudimentary horns support the failure of fusion and canalization, rather than adenomyotic nodule.

In conclusion, pathogenesis of this anomaly is unclear, but its discussion helps demonstrate the embryology of the Müllerian tracts. Treatment continues to be unsatisfactory in these cases, and would probably require in-vivo intervention in the first weeks of the embryonic period.

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