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Case Report

# Unclassified Mullerian Anomaly: Retroperitoneal Uterus

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## Abstract

Anomalies of uterus might be encountered at Caesarean Section or routine investigation. In this article, we present a rare case of double horns of uterus completely separated. One of them is situated retroperitoneally. Therefore, MRI or CT scans is recommend for unclassified Mullerian anomalies, to confirm pelvic or other anomalies and to reduce the likelihood of complications arising during the operation or delivery.

**Keywords:** retroperitoneal space; uterine retro flexion; uterine retroversion, uterus

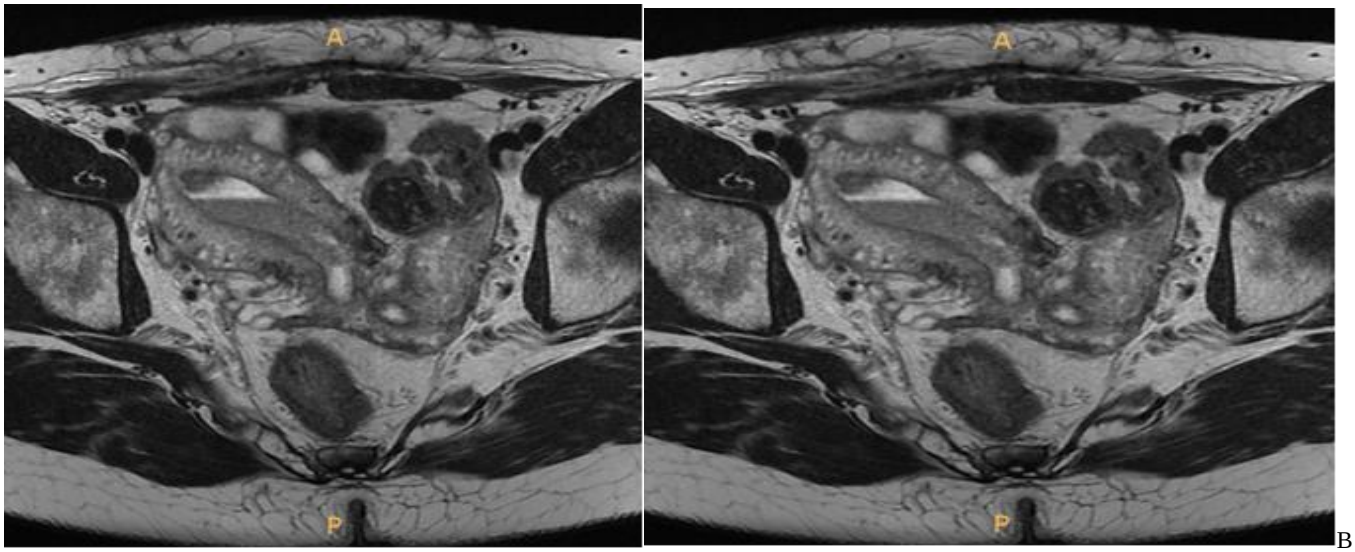
## Introduction

Classification of Mullerian duct anomalies is described according to a fusion pattern. Unclassified Mullerian duct anomalies or uncommon complications were discovered incidentally. We have also added our data pertaining to OHVIRA syndrome [1], with vaginal calcified masses such as stones, to the report by Živkovic et al. [2]. Unclassified Mullerian anomalies should be considered for classification to reduce complications during delivery or operation.

We have encountered, albeit very infrequently, unclassified Mullerian anomalies, such as a retroperitoneal uterus, which was misdiagnosed as unicornuate uterus with a double cervix. We searched the PubMed and EM-DAT databases, but uncovered no additional data pertaining to this case.

## Case History

A 29 year-old female, at 39 gestational weeks, was transferred to our institution with a unicornuate uterus with double cervix, and complete longitudinal vaginal septum. The woman with a gravid uterus underwent cesarean section for a breech presentation. The patient had undergone no previous surgeries and had a normal medical history. Following delivery of a 2,830 g female infant, the patient's uterus was checked. A right unicornuate uterus, right ovary and right cervix were observed, but there was no left uterus in the cul-de-sac or other pelvic cavity. Following the operation, we checked the right and left cervixes for vaginal septum. We were unable to dilate the left cervix or left uterine cavity, but detected a hard mass at the upper site of the left cervix. At the 4-month follow-up, we recommend MRI to confirm any unclassified Mullerian anomalies i.e., unicornuate uterus or double cervix. MRI findings revealed that both uterine horns were widely separated (i.e., the angle between the endometrial canals was > 180 degrees). We found that another uterine body was located in retroperitoneal space: MRI revealed a widely-separated uterus, and no other uterus was found in the pelvic cavity during cesarean section (Figure1).We did not uncover any similar cases in our literature search, but unclassified Mullerian anomalies have been reported previously, including double cervix and double vagina with a normal uterus. Mullerian anomalies were classified according to morphologic pattern rather than the location of the anomaly site.




A B

**Figure 1 (a-b):** *T2-weighted axial image demonstrating uterus didelphys. Both uterine horns are widely-separated (i.e., the angle between the endometrial canals is >180 degrees).*

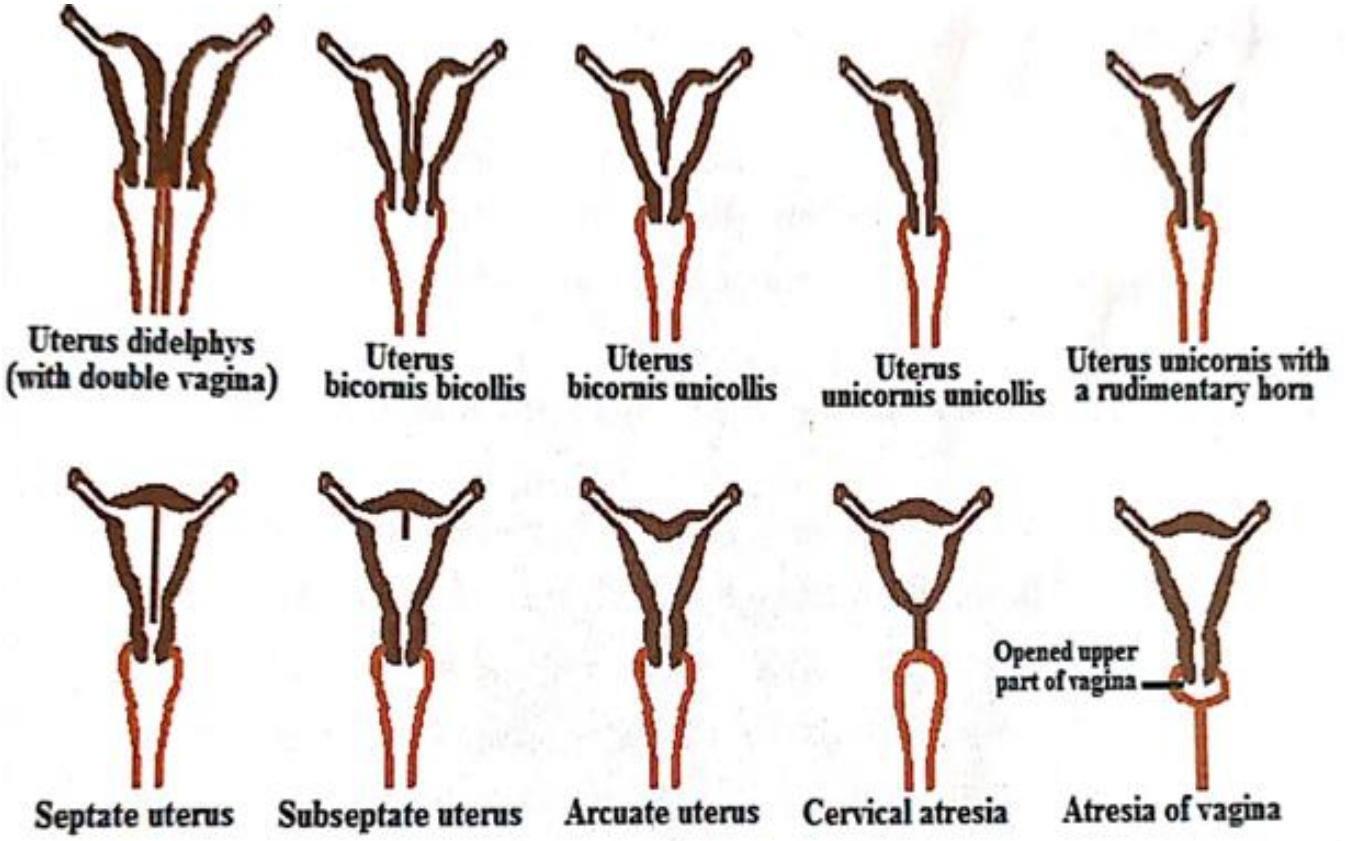



**Figure 2:** *Diagrams showing congenital anomalies of uterus and vagina (about Hegazy, 2014)*

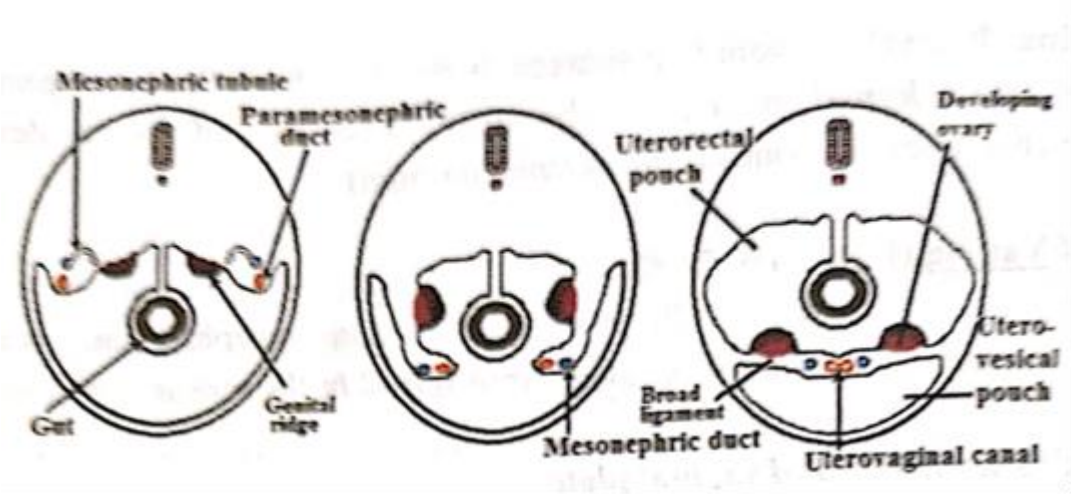



**Figure 3:** *Diagram showing transverse section in body of fetus with invagination of Mullerian ducts ANF formation of broad ligaament (about Hegazy, 2014)*

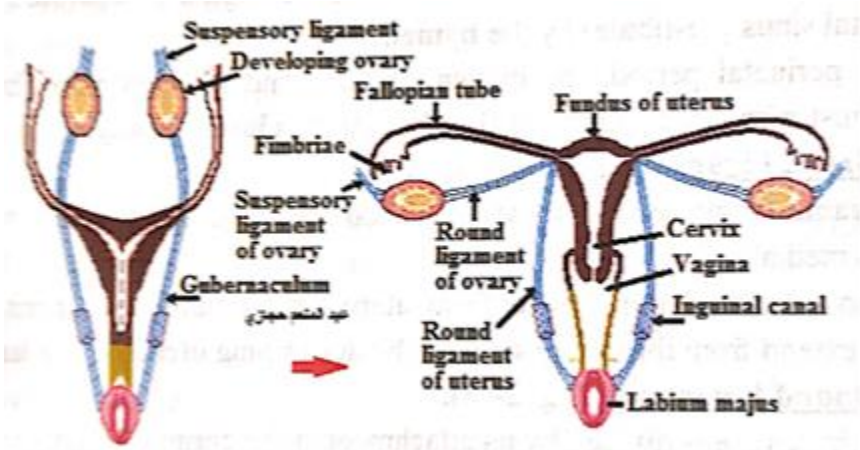



**Figure 4:** *Diagram showing derivatives of Mullerian ducts in females (about Hegazy, 2014)*

## Discussion

These rare cases should be pooled and classified with other anatomically-unusual variants. However, Hegazy [3] stated that this case matches a congenital anomaly called uterus didelphys where there are 2 uterine horns, 2 cervices and septate vagina. It could result from imperfect fusion of the parts of 2 Mullerian ducts forming uterus. These embryonic ducts give rise to the 2 Fallopian tubes and unite together to form the uterus and upper portion of vagina. The ducts originate in the intrauterine life behind the coelomic epithelium; then invaginate ventrally and caudally to join in their middle and lower parts forming what is called uterovaginal canal that differentiates into uterus and upper portion of vagina. In the current case, one Mullerian duct fails to invaginate ventrally and remains separated from the other that reached the abdominal cavity.

We recommend MRI or CT scans for unclassified Mullerian anomalies, to confirm pelvic or other anomalies and to reduce the likelihood of complications arising during the operation or delivery. Our case report of Mullerian anomalies should prove informative for obstetricians.

## Conflict of interest statement

The authors declare no competing interests.

## Ethical Issues

Informed consent was obtained from the patient.

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## Acknowledgement

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