The Mullerian ducts are the human anlage of the female reproductive tract. They differentiate to form fallopian tubes, uterus, uterine cervix and the superior aspect of the vagina. Congenital uterine anomalies, also known as Mullerian duct malformations, are quite common in women. Approximately 0.001 to 10% of women are diagnosed with a congenital uterine anomaly depending on the studied population and the method used. Most identified anomalies of the female reproductive tract are found within the uterus. Isolated cervical, vaginal or fallopian tube defects are much rarer compared to uterine anomalies.

Introduction to Imaging of Mullerian Duct Anomalies

The most common risk factors for uterine anomalies are the maternal use of thalidomide or diethylstilbestrol during pregnancy. Fortunately, awareness about the teratogenicity of these two medications has contributed significantly to lower the incidence of Mullerian duct anomalies.
Embryogenesis and Abnormal Development of the Female Reproductive Tract

At six weeks of embryological development the male and female fetus have two pairs of genital ducts known as the Mullerian and Wolffian ducts. These come from a common embryonic mesoderm that gives rise to the genital system in females and males, respectively.

At this stage, the genitourinary systems are identical and cannot be differentiated. The Wolffian duct system initially grows faster than the Mullerian ducts, forming a pathway for growth of the later. Moreover, the Wolffian ducts initially drain to the primitive mesonephric kidney thus the close association of Mullerian duct anomalies and urinary system anomalies.

Regression of the Wolffian duct system is based on the absence of the testosterone induced SRY gene found on chromosome Y. The ducts elongate in week 9 to form three distinct regions, i.e.,

- The cranial vertical that forms the fimbriae and fallopian tubes.
- The horizontal that develops into the remaining structures of the fallopian tubes.
- The caudal vertical segment of each side that develops to fuse with each other at the midline of the future pelvis and form the uterovaginal primordium that later becomes the uterine and upper vagina region.

Abnormal development at any stage leads to abnormalities as follows:

- Bilateral deficient Mullerian duct development leads to absent vagina, cervix and uterus in addition to absent fallopian tubes.
- **Failure of Mullerian ductal fusion is responsible for the anomalies uterine didelphys, bicornuate uterus and arcuate uterus.**
- Failure of resorption of the medial walls of the vagina and uterus is the mechanism of formation of a complete septate or partial septate uterus.
- Abnormal hormone balance which is linked to the maternal use of diethylstilbestrol can cause a T-shaped uterus. The most common type of uterine anomalies is arcuate or hypoplastic uteri. Unicornuate or bicornuate uterus are two common examples of an arcuate uterus.

Up to one tenth of the cases of congenital uterine malformations also have associated anomalies of the urinary tract, gastrointestinal tract, musculoskeletal system or the heart. **Endometriosis is more common in malformed uteri compared to normally developed ones.** Renal agenesis is very common in patients with uterus didelphys and unicorneute uterus.

Despite the association with other anomalies, the risk of neoplasms in patients with congenital uterine malformations appears to be equal to that of the general population.

Diagnostic Workup and Imaging Studies of the Malformed Uterus

Most patients with congenital uterine malformations present to the clinic because of a history of recurrent spontaneous abortions or amenorrhea.

**Note:** Spontaneous abortions are most common with T-shaped uteri, septate uteri and
unicornuate uterus.
Ultrasound and MRI are the most common imaging modalities used in the diagnosis of Mullerian duct anomalies as they give the type of the anomaly and the presence/absence of concomitant renal malformations with high accuracy compared to other modalities. The diagnosis of a malformed uterus can be delayed due to the operator dependency of ultrasonography, the first imaging modality used in the evaluation of structural uterine anomalies in a patient with recurrent abortions. The increased incidence of spontaneous abortions and infertility might be related to the direct effect of the abnormal uterus on the implantation of the uterus or the co-presence of cervical incompetence. Cervical incompetence is found in up to one-third of the women with congenital uterine malformations.

Ultrasonography and Congenital Uterine Malformations

The accuracy of transvaginal ultrasound in the identification of congenital uterine malformations is excellent, 82 to 92%. Vaginal and cervical atresia in addition to uterus didelphys are reliably identified on ultrasound. On the other hand, septate uterus, unicornuate uterus and other less common uterine malformations are poorly visualized and recognized with ultrasonography.

Another drawback of ultrasonography in the identification of congenital uterine malformations is the operator dependency. Additionally, obese patients usually have ultrasound images of lower quality and the identification of uterine anomalies might be problematic.

Magnetic Resonance Imaging and Congenital Uterine Malformations

Magnetic resonance imaging studies have an estimated accuracy of 100% in identifying almost any type of uterine anomalies. It can identify vaginal, cervical and uterine anomalies reliably. Septate, unicornuate and bicornuate uterine malformations can be easily identified on a magnetic resonance imaging study. Additionally, MRI of the fallopian tubes, ovaries and other pelvic structures makes it easy for the radiologist to identify endometriosis even in initial stages. The standard pelvic MRI protocols employed include T1-weighted, T2-weighted and contrast enhanced images.

Note: The main disadvantage of ultrasonography and magnetic resonance imaging studies is the fact that both can be only diagnostic but not therapeutic.

Laparoscopy and Congenital Uterine Malformations

The direct visualization of the external contour of the uterus can be helpful in the differentiation between bicornuate and septate uterus. Hysteroscopy is helpful in the identification of intrauterine adhesions and septa. These two techniques can be also used for adhesolysis or other treatments.
Examples of Uterine Malformations and the Imaging of Malformations

In the following table, we show some common congenital uterine malformations, the imaging modality of choice for the visualization of that malformation, and the main characteristics one can see on that specific imaging modality for that specific anomaly.

<table>
<thead>
<tr>
<th>Uterine Anomaly</th>
<th>Imaging Modality of Choice</th>
<th>Main Imaging Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypoplastic Uterus</td>
<td>Magnetic Resonance Imaging</td>
<td>A small uterus with regular morphology and normal relationship to the uterus. Hysterosalpingography can be also used with good results.</td>
</tr>
<tr>
<td>Unicorunate Uterus</td>
<td>Magnetic Resonance Imaging</td>
<td>Decreased uterine volume with asymmetry. The presence of a uterine horn that is rudimentary. Ultrasonography can be confusing and some cases were missed by conventional transvaginal ultrasound.</td>
</tr>
<tr>
<td>Uterine didelphys</td>
<td>Ultrasound or Magnetic Resonance Imaging</td>
<td>Uterine didelphys can be easily diagnosed with either transvaginal ultrasound or magnetic resonance imaging. Divergent uterine horns, fundal cleft, two services and a vaginal septum are the most common findings on both imaging modalities.</td>
</tr>
<tr>
<td>Bicornuate Uterus</td>
<td>Ultrasound, Magnetic Resonance Imaging</td>
<td>This uterine anomaly can be easily identified by any of the three mentioned imaging modalities, therefore, the most cost-effective method is advisable, i.e., ultrasound. T2-weighted MRI images are best at the visualization of the two uterine horns.</td>
</tr>
<tr>
<td>Septate Uterus</td>
<td>Magnetic Resonance Imaging or hysterosalpingography</td>
<td>T2-weighted images on magnetic resonance imaging are very accurate in the identification of the uterine septum. Hysterosalpingography is helpful as it can show a septum that is associated with uterine contraction. The septum appears hypoechoic on ultrasonography but it can be missed in a significant number of cases.</td>
</tr>
<tr>
<td>T-shaped uterus</td>
<td>Hysterosalpingography</td>
<td>In contrast to all the previously mentioned anomalies, a T-shaped uterus is best visualized with a hysterosalpingography. Magnetic resonance imaging and ultrasound are inferior in the identification of this anomaly. Suspicton of this anomaly is based on the maternal history of using diethylstilbestrol during pregnancy.</td>
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References

[Imaging diagnosis of congenital uterine malformation](via sciencedirect.com)

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Notes