UTERINE MALFORMATIONS

A type of female genital malformation resulting from an abnormal development of the Müllerian duct(s) during embryogenesis.

Related Diagnoses:
Endometriosis | Repeated implantation failure | Uterus septus | Uterus duplex | Uterus subseptus | Amenorrhoea | Unicornuate uterus | Bicornuate uterus

About Uterine malformations

In the female, Müllerian ducts will develop to form the uterine tubes, uterus, cervix, and the upper one-third of the vagina; in the male, they are lost. This abnormal development can lead to uterine malformations resulting from aplasia of a part of Müllerian duct. Morphological abnormalities can vary from insufficient fission of tissues causing bicornuate uterus, double uterus or preserved uterine septum or in case of Müllerian aplasia to absent uterus.

The prevalence of uterine malformation is estimated to be 6.7% in the general population, slightly higher (7.3%) in the infertility population, and significantly higher in a population of women with a history of recurrent miscarriages.

Judged by the extent of morphological pathology the American Fertility Society (now American Society of Reproductive Medicine) classification distinguishes several types of malformation, as described below.

Class I: Absent uterus (Müllerian agenesis)

The condition is also called Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome, which is characterized by congenital aplasia of the uterus and the upper part (2/3) of the vagina in women showing normal development of secondary sexual characteristics and a normal 46, XX karyotype.

Class II: Unicornuate uterus (a one-sided uterus)

The uterus is formed from one only of the paired Müllerian ducts while the other Müllerian duct does not develop or only in a rudimentary fashion. The sometimes called hemi-uterus has a single horn linked to the ipsilateral fallopian tube that faces its ovary.

Class III: Double uterus (uterus didelphys, also uterus didelphis)

One of the least common amongst Mullerian duct anomalies (MDAs). Both Müllerian ducts develop but fail to fuse, thus the patient has a "double uterus". This may be a condition with a double cervix and a vaginal partition, or the lower Müllerian system fused into its unpaired condition.

Class IV: Bicornuate uterus (uterus with two horns)

Only the upper part of that part of the Müllerian system that forms the uterus fails to fuse, thus the caudal part of the uterus is normal, the cranial part is bifurcated. The uterus is "heart-shaped". It is a uterus composed of two "horns" separated by a septum. In humans, a bicornuate uterus is a type of uterine malformation, but in some other mammalian species, including rodents and pigs, it is normal.
Class V: Septated uterus (uterine septum or partition)

The two Müllerian ducts have fused, but the partition between them is still present, splitting the system into two parts. With a complete septum the vagina, cervix and the uterus can be partitioned. Usually the septum affects only the cranial part of the uterus.

A form of septate uterus, i.e. an incompletely septated uterus or uterus subseptus, is one of the most common forms of congenital uterine malformations.

Class VI: DES uterus

The uterine cavity has a "T-shape". The t-shaped malformation is commonly associated with in-utero exposure (the so-called "DES-daughters").

A rudimentary uterus

An uterine remnant not connected to cervix and vagina and may be found on the other side of unicornean uterus.

Diagnosis

For diagnosing uterine malformation, the physician will need imaging techniques besides a physical examination to determine the character of the malformation: gynecologic ultrasonography, pelvic MRI, or hysterosalpingography. A hysterosalpingogram is not considered as useful due to the inability of the technique to evaluate the exterior contour of the uterus and distinguish between a bicornuate and septate uterus. In addition, laparoscopy and/or hysteroscopy may be indicated.

Associated diseases

Patients with uterine abnormalities may have associated renal abnormalities including unilateral renal agenesis, vertebral, and, to a lesser extent, auditory and cardiac defects, associated gynecological and obstetric complications such as infertility, endometriosis, hematometra, urinary tract anomalies, abortions, preterm deliveries and malpresentation, double vagina or genital neoplasms. In the female, Müllerian ducts will develop to form the uterine tubes, uterus, cervix, and the upper one-third of the vagina; in the male, they are lost. This abnormal development can lead to uterine malformations resulting from aplasia of a part of Müllerian duct. Morphological abnormalities can vary from insufficient fission of tissues causing bicornean uterus, double uterus or preserved uterine septum or in case of Müllerian aplasia to absent uterus.

Complications

Depending on the nature of the handicap, mostly associated with gynecological and obstetric complications such as infertility, endometriosis, hemometra, urinary tract anomalies, abortions, preterm deliveries, fetal malformations, malpresentation.

Risk factors

Uterine malformations are associated with genetic and teratogenic factors. Using uncertified contraceptives (e.g. diethylstilbestrol), and any other substance with teratogenic potential, can be seen as problematic. Fetus is the most vulnerable in the period of 2 to 12 weeks.

Impact on fertility

In case of Müllerian agenesis for example, when uterus is absent, the conception and in some cases, sexual intercourse is physically impossible. In other types of inflection, fertility can be unaffected, however mostly, the inception ratio is lowered. Because the most problematic is gravidity and delivery itself, the patient should be carefully monitored by obstetrician.
Prevention

None possible, except for avoiding using diethylstilbestrol – teratogenic drug, which used to be prescribed for example for: gonorrheal vaginitis, atrophic vaginitis, menopausal symptoms, and postpartum lactation suppression to prevent breast engorgement, later it was used like a postcoital contraceptive. Oral postcoital contraceptive. Avoiding other teratogens.

Symptoms

Depending on the malformation type, symptoms range from amenorrhea, infertility, recurrent pregnancy loss, and cramps or pain, to normal functioning depending on the nature of the defect. In some patients the vaginal development may be affected.

Therapies

Self therapy

Because of the morphological nature of the diagnosis, self or alternative therapy is not applicable. Still, as some sort of an alternative therapy, mental care could be recognized. Psychological distress is very important in young women with Müllerian duct anomalies. It is essential for the patients and their families to attend counseling before and throughout treatment.

Conventional medicine

The most important thing is proper diagnose, the therapy based on clinical symptoms differs from careful monitoring by obstetrician to surgical solution.

Pharmacotherapy

Mostly additional treatment to the surgical solution. Drugs to help epithelization processes and anti-adhesion drugs are administered after the surgery. Antibiotic therapy should be part of the pharmacotherapy. Hormonal therapy is used to help women with conception.

Surgical therapy

There are several surgical techniques depending on the type of malformation. Laparoscopic or hysteroscopic methods are used:

Laparoscopy

It is an operation performed in the abdomen or pelvis through small incisions (usually 0.5–1.5 cm) with the aid of a camera. The laparoscope allows doctors to perform both minor and complex surgeries with a few small cuts in the abdomen. There are a number of advantages to the patient with laparoscopic surgery versus an open procedure. These include reduced pain due to smaller incisions and hemorrhaging, and shorter recovery time.

Hysteroscopy

The inspection of the uterine cavity by endoscopy with access through the cervix. It allows for the diagnosis of intrauterine pathology and serves as a method for surgical intervention (operative hysteroscopy). A hysteroscope is an endoscope that carries optical and light channels or fibers. It is introduced in a sheath that provides an inflow and outflow channel for insufflation of the uterine cavity. In addition, an operative channel may be present to introduce scissors, graspers or biopsy instruments. A hysteroscopic resectoscope is similar to a transurethral resectoscope and allows entry of an electric
loop to shave off tissue. A contact hysteroscope is a hysteroscope that does not use distention media.

Assisted reproduction

The most often method is IVF-ICSI in combination with some surgical solution and pharmacotherapy. Own oocytes or donated ones may be used, depending on woman age and quality of her eggs. Multiple pregnancies may have a negative impact on the likelihood of a full term pregnancy; therefore, a singleton pregnancy should be ensured by eSET (elective single embryo transfer) in these patients. If the uterus is absent, surrogate mother is needed to carry a baby.

Find more about related issues

Diagnoses

Endometriosis
A state in which pieces of the tissue alike to the lining of the uterus (endometrium) grow in other parts of the body.
Learn more at: www.fertilitypedia.org/therapy/diag/endometriosis

Repeated implantation failure
The absence of implantation after three or more transfers of high quality embryos or after placement of 10 or more embryos in multiple transfers.
Learn more at: www.fertilitypedia.org/therapy/diag/repeated-implantation-failure

Uterus septus
A form of a congenital malformation where the uterine cavity is partitioned by a longitudinal septum. It is one of Müllerian duct anomalies.
Learn more at: www.fertilitypedia.org/therapy/diag/uterus-septus

Uterus duplex
Congenital uterine malformation where both Müllerian ducts develop but fail to fuse, thus the woman has a "double uterus".
Learn more at: www.fertilitypedia.org/therapy/diag/uterus-duplex

Uterus subseptus
A form of a congenital malformation where the uterus is partially divided by a longitudinal septum. It is one of Müllerian duct anomalies.
Learn more at: www.fertilitypedia.org/therapy/diag/uterus-subseptus

Amenorrhoea
The absence of a menstrual period in women of reproductive age.
Learn more at: www.fertilitypedia.org/therapy/diag/amenorrhoea

Unicornuate uterus
Congenital uterine anomaly (one of the Müllerian duct anomalies) usually associated with communicating or non-communicating rudimentary horn.
Learn more at: www.fertilitypedia.org/therapy/diag/unicornuate-uterus

Bicornuate uterus
Inborn morphological deviation of the uterus - one of the Müllerian duct anomalies where the uterine cavity is divided in the upper part.
Learn more at: www.fertilitypedia.org/therapy/diag/bicornuate-uterus

Organs

Cervix
The narrow inferior portion of the uterus that projects into the vagina.
Learn more at: www.fertilitypedia.org/edu/organs/cervix
Fallopian tubes
Two very fine tubes that transport sperm toward the egg, and allow passage of the fertilized egg back to the uterus for implantation.
Learn more at: www.fertilitypedia.org/edu/organs/fallopian-tubes

Uterus
The uterus is the largest and major organ of the female reproductive tract that is the site of fetal growth and is hormonally responsive
Learn more at: www.fertilitypedia.org/edu/organs/uterus

Vagina
Sex organ that is a part of the female genital tract having two primary functions: sexual intercourse and childbirth.
Learn more at: www.fertilitypedia.org/edu/organs/vagina

Reproductive cells

Endometrial cell
Cells composing an inner layer of the uterine lining.
Learn more at: www.fertilitypedia.org/edu/reproductive-cells/endometrial-cell

Endometrium
The innermost layer of uterus forming the uterine lumen where the implantation of an oocyte happens.
Learn more at: www.fertilitypedia.org/edu/reproductive-cells/endometrium

Oocyte
A female germ cell involved in reproduction.
Learn more at: www.fertilitypedia.org/edu/reproductive-cells/oocyte

Reproductive functions

Fertilization
The fusion of an ovum with a sperm to initiate the development of a new individual organism.
Learn more at: www.fertilitypedia.org/edu/reproductive-functions/fertilization

Fetal development
The process in which a human embryo or fetus gestates during pregnancy, from fertilization until birth.
Learn more at: www.fertilitypedia.org/edu/reproductive-functions/fetal-development

Implantation
The very early stage of pregnancy at which the embryo adheres to the wall of the uterus.
Learn more at: www.fertilitypedia.org/edu/reproductive-functions/implantation

Risk factors

Abortion
The ending of pregnancy by removing a fetus or embryo before it can survive outside the uterus.
Learn more at: www.fertilitypedia.org/edu/therapy/rf/abortion

Symptoms

Absence of menstrual periods
The absence of a menstrual period in a woman of reproductive age.
Learn more at: www.fertilitypedia.org/edu/symptoms/absence-of-menstrual-periods-1
Absent vagina
A rare symptom that occurs when the vagina doesn’t develop.
Learn more at: www.fertilitypedia.org/edu/symptoms/absent-vagina

Double vagina
Longitudinal division of the vagina into two separate organs.
Learn more at: www.fertilitypedia.org/edu/symptoms/double-vagina

Impossible sexual intercourse
A condition that affects a woman’s ability to engage in vaginal penetration.
Learn more at: www.fertilitypedia.org/edu/symptoms/impossible-sexual-intercourse

Infertility
The failure to achieve a clinical pregnancy after 12 months or more of regular unprotected sexual intercourse.
Learn more at: www.fertilitypedia.org/edu/symptoms/infertility

Preterm birth
A birth of the baby before 37 completed weeks of gestational age.
Learn more at: www.fertilitypedia.org/edu/symptoms/preterm-birth

Recurrent miscarriage
A disease distinct from infertility, defined by two or more failed pregnancies.
Learn more at: www.fertilitypedia.org/edu/symptoms/recurrent-miscarriage

Therapies

Egg donation
Process by which a woman donates eggs for purposes of assisted reproduction or biomedical research.
Learn more at: www.fertilitypedia.org/edu/therapies/egg-donation

Elective single embryo transfer
The procedure of transfer one single good quality embryo in cleavage stage or in stage of blastocyst that was selected as the most appropriate.
Learn more at: www.fertilitypedia.org/edu/therapies/elective-single-embryo-transfer-1

Hormone replacement therapy
Learn more at: www.fertilitypedia.org/edu/therapies/hormone-replacement-therapy

ICSI
A micromanipulative fertilization technique in which a single sperm is injected directly into an egg.
Learn more at: www.fertilitypedia.org/edu/therapies/icsi

Sperm donation
The procedure in which a man (sperm donor) provides his sperm for fertility treatment.
Learn more at: www.fertilitypedia.org/edu/therapies/sperm-donation

Standard IVF
A process in which an egg is fertilised by sperm outside the body; in vitro. Own or donated gametes may be used.
Learn more at: www.fertilitypedia.org/edu/therapies/standard-ivf

Surrogacy
The embryo is gestated in a third party’s (surrogate) uterus.
Learn more at: www.fertilitypedia.org/edu/therapies/surrogacy
A types of female genital malformation resulting from an abnormal development of the Müllerian duct(s) during embryogenesis.

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