GYNATRESIA

Labial Fusion, Labial Adhesion, Labial Synechiae, Labial Agglutination, Labial Adherence, Gynatresia, Vulvar Fusion, Vulvar Synechiae, Cervical Atresia, Vaginal Atresia, Vaginal Aplasia, Atresia Of Hymen

Occlusion of some part of the female genital tract, especially occlusion of the vagina by a thick membrane.

診断: 女性

About Gynatresia

Gynatresia means the narrowing of the female genital tract, which often leads to occlusion or obstruction. In most cases it is caused by a thick membrane in any part of the tract.

Gynatresia can be congenital or acquired. Some congenital conditions which cause the obstruction include cervical atresia, vaginal atresia and aplasia, atresia of hymen and labial fusion.

In very rare situations, the obstruction can be caused by long-term infection or injury.

Cervical atresia

The situation when the cervix is present but deformed and nonfunctional, are known as cervical atresia (Pic. 1). Patients with cervical atresia typically present in early adolescence, around the time of menarche, with amenorrhea and cyclic pelvic pain caused by the obstruction of menstrual flow from the uterus. If untreated, the accumulation of menstrual fluid in the uterus caused by cervical agenesis can lead to hematocolpos (a medical condition in which the vagina fills with menstrual blood), hematosalpinx (a medical condition involving bleeding into the fallopian tubes), endometriosis (a disease in which
tissue that normally grows inside the uterus grows outside it.), and pelvic adhesions.

**Vaginal atresia**

Vaginal atresia is a condition in which the vagina is abnormally closed or absent. The main causes can either be complete vaginal hypoplasia (underdevelopment or incomplete development of the vagina), or a vaginal obstruction, often caused by an imperforate hymen or, less commonly, a transverse vaginal septum.

**Vaginal aplasia**

Vaginal aplasia is one of the most significant congenital anomalies of the female reproductive tract from physical and psychological perspective. The primary goals of surgical intervention in patients with vaginal aplasia are to relieve obstruction and pain, to restore a normal sexual functioning, and to preserve the patient’s reproductive potential. The timing of surgery depends on the patient’s anatomic configuration and on the presence or absence of functional endometrial tissue. Neovagina creation needs to reconstruct a vagina of adequate length and diameter and need a stable lining. Vaginal aplasia is associated with Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome which is characterized by congenital aplasia of the uterus and the upper part of the vagina in women showing normal development of secondary sexual characteristics. The aplasia is frequently associated with extragenital anomalies. These other manifestations are renal, skeletal and heart malformations.

**Atresia of hymen**

Atresia of hymen or an imperforate hymen is a congenital disorder where a hymen without an opening completely obstructs the vagina. It is caused by a failure of the hymen to perforate during fetal development. It is most often diagnosed in adolescent girls when menstrual blood accumulates in the vagina and sometimes also in the uterus. It is treated by surgical incision of the hymen.

**Labial fusion**

Labial fusion (Pic. 2) is a common gynecological disorder in the pediatric population and is defined as complete or partial adherence of labia majora or minora. It is commonly seen at the age of 13-23 months with an incidence of 1.8%. Labial fusion is never present at birth, but rather acquired later in infancy, since it is caused by insufficient estrogen exposure and newborns have been exposed to maternal estrogen in utero.
Labial fusion are usually caused by a combination of local inflammation, chronic infection. Inflammation and irritation secondary to vaginitis may have caused superficial epithelium of labia to heal subsequently with fibrous adhesions. Chronic and untreated inflammation creates an environment that causes a tendency to fibrosis and eventual scarring despite normal estrogen levels. The reason why not all of the cases with untreated severe vaginitis develop with labial adhesion is another point of discussion. On the other hand the rapid development of the labial adhesion in a period of only 3 weeks, without any mechanical and traumatic etiology, hormonal abnormality, or underlying dermatological disease in this case, is also dramatic.

Most presentations are asymptomatic and are discovered by a parent or during routine medical examination. The condition can be diagnosed based on inspection of the vulva. In patients with labial fusion, a flat plane of tissue with a dense central line of tissue is usually seen when the labia majora are retracted, while an anterior opening is usually present below the clitoris.

Labial fusion in adults is a rare clinical entity with only few described cases, in particular in post-menopausal women. The major symptom in those reported cases is urination anomalies and most commonly infections of the urinary tract. The pathophysiological mechanism is believed to be chronic inflammation and/or irritation of the vulvar skin arising from various conditions such as local inflammation, lichen sclerosus (a disease of unknown cause that results in white patches on the skin, which may cause scarring on and around genital or sometimes other skin), recurrent urinary tract infections as well as lack of sexual activity, lack of estrogen and local trauma. The major presenting symptom is urinary incontinence and voiding dysfunction whereas the mechanism by which this occurs is most likely due to physical obstruction of the urinary flow, directly related to the labial fusion. In addition, the mechanical obstruction of urine flow commonly results in a retrograde flow inside the vaginal cavity.

In severe cases where the labia minora are entirely fused, causing urinary outflow obstruction or vaginal obstruction, the labia should be separated surgically. Recurrence after treatment is common but is thought to be prevented by good hygiene practices.

**Associated diseases**

Labial adhesions have been found secondary to female circumcision, herpes simplex, dermatological conditions, caustic vaginitis, local trauma, and vaginal laceration following childbirth. Vaginal aplasia is also associated with Mayer-Rokitansky-Küster-Hauser syndrome (absence of the upper two-thirds of the vagina and the uterus is absent or undeveloped). Vaginal or cervical atresia can in severe cases caused the hematocolpos (a medical condition in which
the vagina fills with menstrual blood) and hematosalpinx. Some cases of gynatresia are associated with endometritis, which is inflammation of inner layer of uterus caused by stagnant blood and increased amount of bacteria.

**Complications**

Labial fusion can lead to urinary tract infection, vulvar vestibulitis (an inflammation of the vulva and the vulvar vestibule) and inflammation caused by chronic urine exposure. In severe cases, labial adhesions can cause complete obstruction of the urethra, leading to anuria (passage of less than 100 milliliters of urine in a day) and urinary retention. Retention of blood in uterus or Fallopian tubes before the obstruction can lead to hematocolpos or hematosalpinx.

**Risk factors**

- inflammation
- low hygiene
- local trauma

**Impact on fertility**

Very often most labial adhesion will separate naturally over time. In cases where they not the surgery is necessary, because the penis is not be able to penetrate into the vagina through the adhesion. After the surgery is performed, woman is capable of normal sexual life and she can conceive child naturally.

**Prevention**

The level of knowledge about the rules of girl intimate hygiene is one of the most important factors in the prevention of this disorder.

**Symptoms**

Patients may present with associated symptoms of dysuria (painful urination), urinary frequency, refusal to urinate, or post-void dribbling. Some patients present with vaginal discharge due to pooling of urine in the vulval vestibule or vagina. In severe cases gynatresia can lead to hematocolpo and
Therapies

Self therapy

Good hygiene practice may prevent the inflammation of vulva.

Conventional medicine

Topical estrogen is used as pharmacotherapy in children and in adults in less severe cases of labial adhesions. More often it is necessary to undergo a surgery when the adhesions are divided mechanically.

In cases of cervical atresia or vaginal aplasia the reconstructive surgeries are the option.

Pharmacotherapy

With labial adhesion in children favorable results were usually obtained with topical estrogen. This topical treatment can go up to 3 to 4 weeks until recovery is achieved.

Surgical therapy

Labial adhesion

For adult patients, estrogen therapy is not always successful as a first-line treatment and surgery may be necessary. The usual solution for cases of adult labial adhesion is surgical adhesiolysis (Pic. 3, Pic. 4). Adhesiolysis is the technique of lysing adhesions using either microscissors (recommended) or thermal energy modalities.

Cervical atresia

Surgical interventions for congenital cervical atresia range from complete hysterectomy with canalization to conservative options, such as uterine cavity catheterization. As a high level of surgical expertise is required to conduct complex reconstructive procedures (Pic. 5), many
Clinicians choose hysterectomy as the optimal primary surgical treatment for the malformation.

**Vaginal aplasia**

Optimum therapeutic approach in vaginal aplasia has always been an area of extensive controversies. Although surgical management gained priority due to the evolution of techniques, there is currently no consensus in the literature regarding the best type of surgical approach. The most commonly preferred surgical procedure among gynecologists is McIndoe operation which involves the creation of a space between bladder and rectum, insertion of a mold covered with split-thickness skin graft into that neovaginal space, and use of postoperative vaginal dilation to avoid stenosis.

**Assisted reproduction**

About 5% of women with absent vagina (vaginal aplasia) have a functioning uterus. Spontaneous conceptions are rare after correction surgery but there are techniques of assisted reproduction which can be used to help women became pregnant. There is possibility to transfer embryo through fallopian tubes or through myometrium (the middle layer of the uterine wall, consisting mainly of uterine smooth muscle cells) which is called transtubal or transmyometrial embryo transfer. The following delivery of child is by caesarean section.

**Find more about related issues**

**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

**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](http://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](http://www.fertilitypedia.org/edu/therapies/standard-ivf)

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**Gallery**

**Pic**
*Diagrammatic anatomical comparison between the traditional cervix classification for abnormal development and the proposed cervical atresia typing.*

**Pic**
The labia majora are completely fused, leaving a very small opening to allow urination.
**First surgical intervention.**

**Postoperative 15th day view on patient.**

**Cervical atresia before, during, and after reconstructive procedure.**

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