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Mini Review

Congenital and Acquired Malformations in Vulva and Vagina - Pediatric Age

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Abstract

Vulva and vagina malformations are significant because of their effect on future reproduction. They may be asymptomatic and discovered during clinical check up examinations. They may be symptomatic especially at puberty. For the diagnosis, clinical examination detects vulva and vagina abnormalities. The use of sonogram will be helpful for the diagnosis of Mullerian duct anomalies.

Keywords: Vulva; Vagina; Pediatric

Introduction

The pediatric gynecologist is frequently consulted because of anomalies detected by the pediatrician during the evaluation of symptomatology as pain or discharge affecting the child. The development of female genital tract starts at 5-6 weeks gestation by the paramesonephric ducts or mullerian ducts [1]. These ducts grow distally as the embryo grows. The medial walls of these ducts grow closer together and then start to degenerate between week 13 and 20 – at this stage, the genital tract has proximal separated paramesonephric ducts and this will be the fallopian tubes. The rest of the fused ducts will be the uterus, cervix, and upper ¾ of the vagina . The lower ¼ of the vagina develops from the urogenital sinus. Abnormalities in longitudinal fusion of mullerian ducts results into double uterus, septate uterus, actuate uterus, and longitudinal vaginal septum. Abnormalities in fusion of mullerian bulb and urogenital sinus may lead to development of transverse vaginal septum, and imperforate hymen [2].

Imperforate hymen

This anomaly is usually discovered at puberty – patients present with amenorrhea and cyclic pain. Examination of vulva reveals normal labia majora and minora. The hymen is intact and completely obstructing the vaginal introitus. Menstrual blood collects above the hymen and distends the vagina. The treatment is surgical incision of the hymen. The collected menstral blood in the vaginais now drained out and the patient is relieved of her pain and will have regular cycles [3].

Labial agglutination

These children present to the pediatric gynecologist between 3 months and 6 years. The reason for the presentation is absent introitus - the condition results from adhesions of the labia minora thus obscuring the hymen and uretheral orifice. The condition results from mild infection and hypoestrogenism. One of the symptoms is dribbling of urine that collects behind the labial adhesions. The treatment of this condition is taking proper care of hygiene and using warm water baths every day. In addition, to apply estrogen cream to the labia minora twice per day. Usually, this management leads to resolution of these labial adhesions between 2-4 weeks. Sometimes, there is no response to this medical management. The labial adhesions are separated surgically under anesthesia, followed by estrogen cream application [4,5].

Transverse vaginal septum

This usually occurs in the lower part of the vagina as a result of failure of canalization of the vagina at the level of mullerian tubercle and urogenital sinus. The patient will have amenorrhea, cyclic pain at puberty. The treatment is surgical excision [6].

Mullerian agenesis [7,8]

This anomaly is the result of failure of development of the paramesonephric ducts. The evaluation reveals a rudimentary uterine horn and absent vagina. The anomaly is discovered at puberty because of primary amenorrhea. The general examination reveals normal thelarche (breast development), normal adrenarche

(axillary and pubic hair). The treatment is to develop a vagina either by using dilators that is successful in 90-96% of the cases; or surgically by using partial thickness skin graft as in McIndoe procedure - Chromosomal karyotyping of these patients is 46XX [7,8].

Androgen insensitivity syndrome

This is a condition resulting from absent androgen receptors. The karyotyping is 46XY. The gonads are testes and serum testosterone level is in the male category. However, they have no hair growth at puberty. They have breast development. They are raised as females with mullerian agenesis. The management is removal of the gonads, and vaginal reconstruction surgery. They will be on hormone replacement in the form of estrogen treatment [9,10].

Tumors of the vulva

In pediatric age, patients sometimes present because of swelling in the labia. By examination several conditions are diagnosed:

- Hypertropied labia. This is usually unilateral and is congenital. Sometimes, it may need surgery for Labioplasty [11,12].
- Sebaceous cyst usually it is small less than one centimeter. If there is pain or discomfort, then excision is the treatment.
- Bartholin duct cyst. The results from obstruction of the Bartholin duct due to infection. The treatment is marsupialization of the duct [13].

In conclusion, patients of pediatric age are referred to a gynecologist because of abnormality of vulva, pain, and or discharge - pelvic examination of the external genitalia usually leads to diagnosis of vulva abnormality. Also abnormalities of vagina and uterus are evaluated by the use of sonogram and or CT scan.

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