*Van Med J 31 (3):179-185, 2024*
*DOI: 10.5505/vmj.2024. 63935*

# Obstructive Uterovaginal Anomalies In Children

**Burhan Beger [1], Cihan Etgül [1], Bulent Sonmez [2], Metin Simsek [3], Baran Serdar Kızılyıldız [2]**

*[1]Van Yüzüncü Yıl University, Pediatric Surgery Department, Van, Türkiye*
*[2]Van Training And Research Hospital, Pediatry Department, Van, Türkiye*
*[3]Van Training And Research Hospital, Pediatric Surgery Department, Van, Türkiye*

**Abstract**

**Introduction:** The congenital vaginal obstructions, which may be associated with labial synechia, imperforate hymen, complete vaginal transverse septum, partial vaginal agenesis or atresia are extremely rare reproductive system anomalies in children and adolescents. A complete consensus has not been achieved in the diagnosis and management of uterovaginal canal obstructions in children and adolescents. In this study, our uterovaginal canal obstruction experiences were evaluated with the literature.

**Materials and Methods:** Data of patients with uterovaginal obstruction were evaluated, between 2015 and 2018. Values were expressed as counts and percentages. The files containing age, symptoms, diagnosis, radiological modalities and surgical management of the patients diagnosed with reproductive tract obstruction, between 1st April 2015 and 1st August 2018 were evaluated retrospectively. In addition to the general physical examination, a careful urogenital system examination was performed in the initial evaluation of the patients

**Results:** Twenty-one female patients with uterovaginal obstruction were detected. The mean age was. The most common anomalies detected were labial synechia and imperforate hymen. Moreover, the labial synechia was determined in 9 patients, the imperforate hymen in 7 patients , the distal vaginal atresia in 2 patients, the OHVIRA in 2 patients.

**Conclusion:** Most of the uterovaginal obstructions can be treated with simple surgical intervention when differential diagnosis is kept in mind. However, complex cases require a multidisciplinary approach.

**Key words:** Child; uterovaginal canal; obstruction; pelvic mass.

## Introduction

The congenital vaginal obstructions, which may be associated with labial synechia, imperforate hymen, complete vaginal transverse septum, partial vaginal agenesis or atresia are extremely rare reproductive system anomalies in children and adolescents (1-3). As a result of the defect during the development of the urogenital sinus in the caudal direction, the obstruction may be at different levels embryologically (1). Due to the fact that vaginal and uterine fluids cannot drain from the opening depending on the obstruction, fluid accumulation occurs. If the amount of fluid increases, hydrometra, hematometra or pyometra (i.e. infection presence) may occur depending on the character of the liquid (2,3). In the literature, a consensus has not been achieved in terms of optimal surgical method, postoperative management, and conditions that cause long-term complications. This situation is frequently related to the rarity of the procedure, complex anatomy and patient diversity (4). Preoperative identification of vaginal obstruction can help to make appropriate consultancy and appropriate surgical decision (2,5). In the light of this information, we aimed to evaluate the results of uterovaginal obstruction cases in our own clinic

## Material and Methods

Ethics Committee approval was obtained from Van Training and Research Hospital Ethics Committee with decision number 2019/15 (01.08.2019). The files containing age, symptoms, diagnosis, radiological modalities and surgical management of the patients diagnosed with reproductive tract obstruction, between 1st April 2015 and 1st August 2018 were evaluated retrospectively. In addition to the general physical examination, a careful urogenital system examination was performed in the initial evaluation of the patients. The patients diagnosed with reproductive tract obstruction were examined for differential diagnosis and accompanying diseases. The first radiological imaging method was ultrasonography. Preoperative definitive diagnosis was performed with MRI((magnetic resonance imaging) in all patients thought to have complex uterovaginal obstruction. Topical estrogen (Premarin vaginal cream, 0.625 mg conjugated estrogen, Wyth) was administered twice daily for 14 days to the patients with asymptomatic labial synechia. Labial separation under topical anesthesia (5% lidocaine-EMLAR) was performed in the patients, of whose

*Corresponding Author: Van Yüzüncü Yıl University, Pediatric Surgery department, Tuşba, Van/Türkiye E.mail: burhanbeger@hotmail.com,
**Orcid:** Burhan Beger 0000-0002-1565-8062, Cihan Etgül 0009-0004-7823-3493 , Bulent Sonmez 0000-0002-4810-7072 , Metin Simsek 0000-0003-4736-7160 , Baran Serdar Kızılyıldız 0000-0001-7252-3650

**Received:** 20.03.2024, **Accepted:** 23.05.2024

conservative treatment was unsuccessful; therefore, medical treatment was continued for 5 days in order to prevent recurrence. Under general anesthesia, hymenectomy was performed with " + " incision to the patients with imperforate hymen. In patients with distal vaginal atresia, after the catheterization of the bladder and rectum, the vagina mucosa was found with an incision between the rectum and bladder in the lithotomy position and then vaginal pull-through and perineum anastomosis were performed. The transverse vaginal membrane was excised cystoscopically with cautery. On the other hand, if non-combinated part opening could not be achieved, the transverse vaginal membrane was excised with laparotomy in patients with upper vaginal obstruction. If vaginal opening could be achieved, we expected for 1 week to epithelialize of the lumen placed a foley or pezer catheter. Due to sepsis and complex surgical procedure, the purulent fluid was drained with a drainage from abdominal or introitus guided by ultrasonography in neonatal pyocolpos cases. After the septic period regressed and surgically optimal conditions were met, definitive surgery was planned.

**Ethical approval:** Ethics Committee approval was obtained from Van Training and research Hospital Ethics Committee with decision number 2019/15 dated 01.08.2019

**Statistics analysis:** Analytical statistics were not used in this study, but only descriptive statistics (number and percentage) were used.

## Results

Twenty-one girls with reproductive tract obstruction (at mean age: 13 years, range: 3 days-17 years) were included in this study. The symptoms detected during the first examination (e.g. primary amenorrhea, periodic abdominalgia, abdominal pain, menstrual irregularity, and pelvic mass) were given in Figure 1.

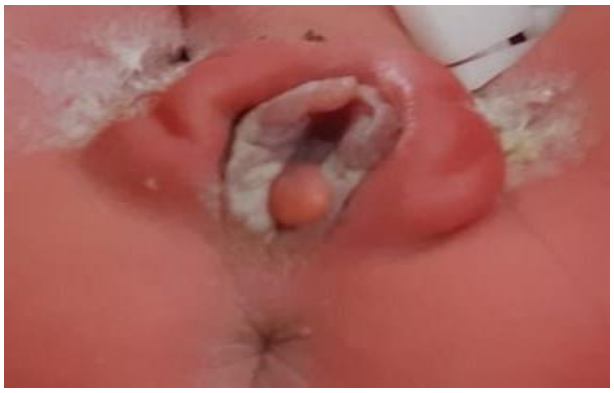

**Figure 1:** Vaginal polyp evaluated as uterovaginal obstruction

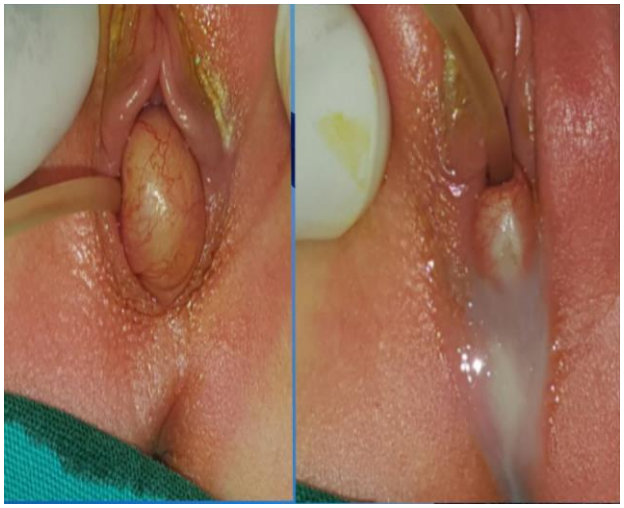

**Figure 2:** Opening of the imperforate hymen and drainage

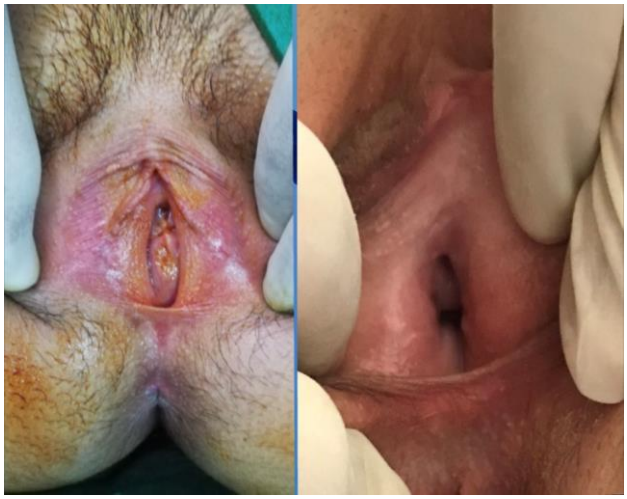

**Figure 3:** Distal Vaginal Atresi and MRKH syndrome

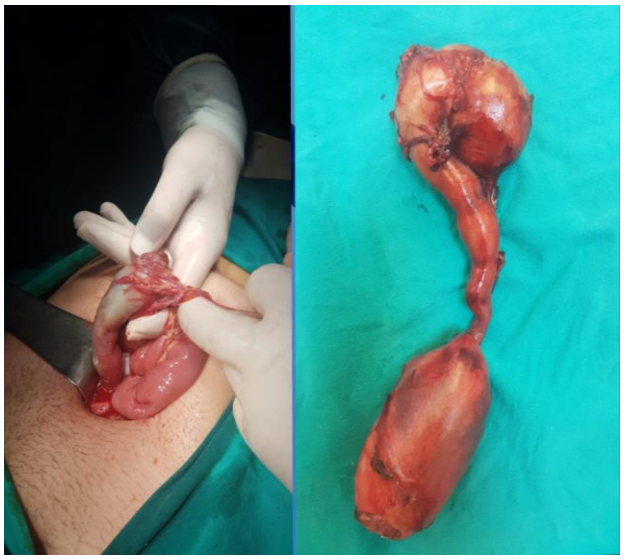

**Figure 4:** Obstructed Hemivagina

**Table 1:** Clinical manifestations for 21 patients

| | Patients (n) | Percentage of total(%) |
|---|---|---|
| Primary amenorrhea | 7 | 17 |
| Cyclic pelvic pain | 9 | 22 |
| Abdominal pain | 12 | 30 |
| Pelvic mass | 6 | 14 |
| Menstrual disorder | 6 | 14 |
| Periodic fever | 1 | 3 |

**Table 2:** Associated anomalies and Syndromes for 21 patients

| | Patients (n) | Percentage of total (%) |
|---|---|---|
| Spinal anomaly | 1 | 5 |
| Pyokolpos | 2 | 10 |
| Renal anomaly | 3 | 15 |
| Situs Inversus Totalis | 1 | 5 |
| Pelvic Mass | 6 | 55 |
| Mc-Cusic Cauffman Syndrome | 1 | 5 |
| Bardet-Biedle Syndrome | 1 | 5 |

Moreover, the labial synechia was determined in 9 patients, the imperforate hymen in 7 patients (figure 2), the distal vaginal atresia in 2 patients (figure 3), the OHVIRA(Obstructed hemivagina with uterus didelphys and ipsilateral renal anomaly) in 2 patients (figure 4), and the longitudianal vaginal septum in 1 patient. The most common complaint was found as abdominal pain (Table 1). The most common accompanying anomaly was determined as pelvic mass and renal anomaly (Table 2). The most commonly used modality radiologically was abdominal ultrasonography. However, MRI was used most frequently in treatment planning, especially in complex cases (figure 5).

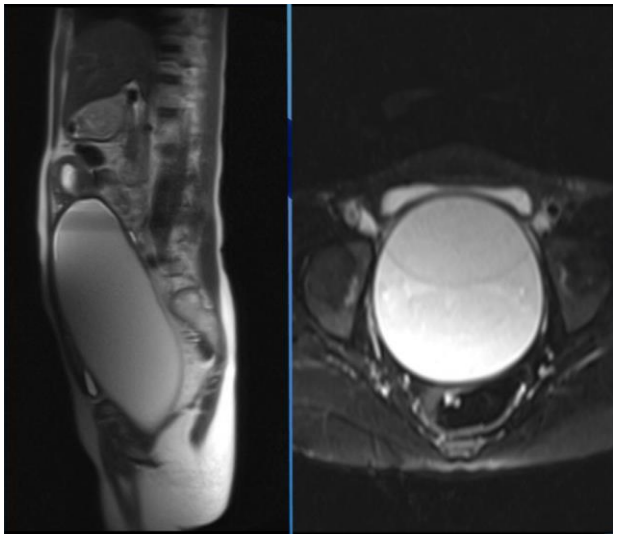

**Figure 5:** MRI of 14 years girl with distal vaginal atresia

In 2 of 9 patients with labial synechia, relapse was detected despite prophylactic medical treatment and sitting bath. The mean follow-up was 2 months.No stenosis was detected in any of the impregnated hymen cases. The mean follow-up was 6 months. One of the 2 cases with distal vaginal atresia was followed up for 9 months. Stenosis was not detected and dilatation was not needed. The septic view disappeared after abdominal percutaneous drainage due to pyocolpos in the other 3-month-old patient. Due to the low birth weight, it was decided to wait until the patient was 10 kg. In 2 cases with OHVIRA, noncombinated uterine cystic mass was excised by laparotomy. In a 3-days-old patient with longitudinal vaginal septum and pyocolpos, the abdominal percutaneous abscess was drained, and then the patient was expected to be 10 kg for definitive surgery. The mean follow-up was 9 months (2 months - 2 years).

## Discussion

Genetic and environmental factors are believed to play a role, although the cause of most Müllerian canal anomalies is unknown. Ionizing radiation, infection, drugs (e.g. diethylstilbestrol and thalidomide) exposed during the development of the genital system may cause some Mullerian canal anomalies (6,7). For the diagnosis and effective treatment of congenital anomalies, embryology should be well understood (8). In embryo aged with 5-6 weeks of gestation, the genital system is named as the indifferent stage due to the association of both pairs of canals. Due to the

absence of the mullerian inhibitory factor (MIF), the Müllerian canals develop towards caudal and joins mutually in the midline. They form the uterovaginal canal with a single lumen that turns into the uterus and upper vagina in the future periods. The non-merged upper parts of the Müllerian canals differentiate into the fallopian tubes. The lower 1/3 of the vagina develops from the urogenital sinus. After the 20th gestational week, the vaginal plaque degenerates and the vaginal lumen is formed; therefore, the connection between the inner and outer parts of the genital canal occurs (6,9). After elongation and fusion, the cervical mesodermal and endodermal tissues change simultaneously (8). The defect that occurs during elongation of the Müllerian canal causes congenital vaginal obstructions. Although some classifications are reported, none of them cover all anomalies (10). The frequently used Society for Reproductive Medicine classification divides the Müllerian anomalies into 6 groups; as hypoplasia/agenesis, unicornuate, didelphus, bicornuate, septate, arcuate. Gynecological abnormalities are reported to be approximately 5.5-7% (10). In the classical presentation, the adolescent girls show the increased pelvic pain and dysmenorrhea. The cycle is irregular in the first 3 years. Often it does not produce any signs immediately after menarche. These patients may be detected incidentally due to urinary infection. In addition, they may be determined with acute abdominal pain, fever and vomiting due to pelvic mass (10). With the effect of mass after menarche, peritoneal escape with retrograde menstruation causes hematometra and hematosalpinx. Due to maternal estrogen stimulation in the newborn, vaginal mass with hydrocolpos and mucocolpos in adolescent patients can cause primary amoanorrhea and cyclic abdominal pain (1,10). Laparoscopy and laparotomy were used to define the anatomy in the past; however, these methods are abandoned due to the development of imaging modalities (10). Pelvic MRI is the gold standard radiological modality. The presence, thickness, location (low, mid, or high) of the transverse vaginal septum with the MRI can be defined for preoperative surgical planning. With the MRI, the high transverse vaginal septum can be separated from the cervical agenesis (10). Preoperative diagnostic accuracy of the MRI is reported to be approximately 100% due to its ability to detail the Müllerian canal anatomy and to identify potential urinary anomalies (1). Moreover, the MRI can determine the volume of the hematocolpos causing vaginal enlargement. It helps to plan operation stages by estimating the distance of other tissues from the vaginal swelling (5,10). Labial adhesion, an acquired child disease, is a problem that labia minora fuses over the vestibule. Although its cause is unknown, the labial adhesion is associated with low estrogen levels in the prepubertal period (11,12). Only observation is sufficient in asymptomatic patients, but topical treatment with conjugated estrogens is the basis of conservative treatment in symptomatic patients. If the labial adhesion is severe or associated with pain and infection, surgical intervention should be performed by manual separation or lysis (12). The labial adhesion prevents the excretion of vaginal secretions and urine. Microperforated hymen is that the formation of urinary retention depending on accumulated vaginal secretion since hymen does not have sufficient opening. In addition, girls with micro-perforated hymen are at risk for pelvic infections and recurrent vulvovaginitis due to ascending infections (13). Imperforate hymen is the most common cause of congenital distal vaginal obstruction (10). Asymptomatic imperforate hymen may not give symptoms until adolescence, but manifested by recurrent lower abdominal pain with menarche. Patients with this anomaly can be detected incidentally in childhood. In the literature, spontaneous opening has been reported with microperforations. Symptoms are related to the size of the hymenal opening. Girls with micropeforation often have a normal cycle, but they see irregular post-menstrual spotting due to incomplete drainage. This abnormality occurs in one of 1000 healthy born girls (13,14). Vulvar swelling is not seen in all cases and is frequently missed (15). In imperforate hymen, cruciate or circular incision is sufficient to provide menstrual flow. Stenosis is rare and dilatation is not required. After the association of obstructed hemivagina and uterus didelfis was defined in 1922, the triad was reported in 1950 with the same side renal anomaly (16,17). This triad was called Herlyn-Werner-Wunderlich syndrome in 1980 and abbreviated as OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) in 2007 to provide a common basis for the syndrome. In classical presentation, the diagnosis of OHVIRA is diagnosed with the presence of vaginal and pelvic mass and often requires additional analysis (18). Its prevalence is estimated to be between 0.1-3.8% (19). Resection of the vaginal septum is the optimal operation for patients (17). Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome seen as a result of disruption in the embryonic development of the Müllerian canals is the most serious developmental congenital anomaly of the female genital system. In this syndrome, the upper

2/3 of the vagina and uterus develop to different degrees, including not fully or partially developing. Its incidence is estimated to be 1/4500 in newborn girls (10). There is pelvic pain associated with rudiment uteri. If non-communicating, laparoscopic supracervical excision of these residues is performed (6). Vertical junction disorders include transverse vaginal septum and cervical agenesis or atresia. This situation stems from the fusion of the urogenital sinus with the Müllerian canal (6). On examination, normal external genitalia and perforated hymen are detected. Vaginal length can be measured through the hymenal opening. Septum is called low type when the length is less than 3 cm, intermediate type when between 3-6 cm, and high type when 6 cm larger. Treatment is excision of the septum. The thin septa (<1 cm) is completely excised. The thick septa (> 1 cm) is also completely excised; however, the vaginal epithelium should be revised and the upper vaginal vaginal epithelium should be pulled down and anastomosed with the lower vaginal epithelium (10). Isolated distal vaginal atresia, a rare Müllerian anomaly, is seen in one living girl between 4000 and 1/10000 (22,23). It can be detected as distal or total vaginal atresia. Except for rare cases, it always includes an anatomically normal corpus and cervical aplasia (20). Distal vaginal atresia can be effectively treated with pull-through vaginoplasty. In this method, vaginal opening is found via an incision performed between the bladder and the rectum, and an anastomosis of the vagina mucosa and perineum is made (21). Cervical atresia, a rare Müllerian anomaly, occurs 1 in 80,000 to 100,000 births. Its treatment and management is very difficult and hysterectomy is recommended if canal formation cannot be achieved (22). If vaginal obstruction is not treated, it may cause complications such as peritonitis and endometriosis due to retrograde menstruation depending on fluid accumulation (2). In the reproductive tract, an accumulation of fluid produced by vaginal and cervical glands is seen on the closed side due to the mechanical obstructions. This condition is determined clinically as pelvic mass and dysmenorrhea. Depending on the liquid and the area it accumulates, hydro-metro-colpos, muco-metro-colpos and hemato-metro-colpos may occur (10,23,24). Pyocolpos occurs if the fluid is infected. Pyocolpos is an emergency surgical condition and may cause mortality via vesicovaginal fistula, destruction and sepsis in case of delay (23,24). It is surgically important to make a differential diagnosis of this pelvic mass with conditions such as sacrococcygeal teratoma, ovarian cyst, old ovarian torsion, megacystis, anterior sacral meningocele, anal atresia, gastrointestinal duplication cyst. In addition, hydronephrosis, urinary tract infection, sepsis, constipation and vomiting may be seen depending on the ureteral compression causing by mass in the lower abdomen (15). Fluid accumulation associated with maternal hormonal stimulation in non-communicating area can be detected in girls with vaginal obstruction in the neonatal period. The hydrometrocolpos approximately with 6: 10000 and pyocolpos approximately with 1: 30000 are rarely seen (24). The vaginal obstruction may be associated with Bardet-biedle syndrome (18), Ellis van Creveld syndrome, VACTERL association (vertebral, anal, cardiovascular, tracheooesophageal, renal and limb anomalies), MURCS association (Mullerian duct aplasia / hypoplasia, renal agenesis / ectopia / hypoplasia cervicothoracic somite dysgenesis such as Klippel Feil abnormality, anomalous ribs or Sprengel deformity) (25), Peter plus syndrome (23), Fraser syndrome, Winter syndrome (2), postaxial polydactyly, retinal dystrophy or retinitis pigmentosa, obesity, nephropathy, and mental redation (26). The treatment is planned according to the location of the obstruction. However, in the case of pyocolpos and sepsis, complex surgical procedures can be planned after abscess drainage with a catheter inserted from percutaneous abdominal or intoroitus under the guidance of ultrasonography (27).

**Study limitations:** Due to the rare anomalies, sufficient number of cases could not be provided and also the study was designed retrospectively.

## Conclusion

Genital examination should be performed carefully in every child and uterovaginal obstructions should be kept in mind in the differential diagnosis of pelvic masses. Most of the uterovaginal obstructions can be treated with simple surgical intervention when preoperative diagnostic modalities are used. However, complex cases require a multidisciplinary approach.

**Ethical approval:** Ethics Committee approval was obtained from Van Training and research Hospital Ethics Committee with decision number 2019/15 dated 01.08.2019

**Conflict of interest:** The authors have no conflict of interest related to this study.

**Financial support:** No financial support was received for this study.

**Author contributions:** Concept (BB),Design (BB,CE,BS,MS), Data Collection and/or Processing (BSK), Analysis and/or Interpretation(BB,CE,BS,MS,BSK)

## References


1. Zhang M, Zhang Mx, Li Gl, Xu Cj. Congenital Vaginal Atresia: A Report Of 39 Cases İn A Regional Obstetrics And Gynecology Hospital. Journal Of Huazhong University Of Science And Technology.Med Sci 2017; 37(6): 928-932.
2. Awad Ee, El-Agwany As. Distal Vaginal Atresia Misdiagnosed As İmperforate Hymen: A Case Managed By Transperineal Vaginal Pull Through (Distal Colpoplasty). The Egyp J Rad Nuc Med2015; 46(4): 1155-1158.
3. Shaked O, Tepper R, Klein Z, Beyth Y. Hydrometrocolpos-Diagnostic And Therapeutic Dilemmas. J Ped And Adol Gyn 2008;21(6): 317-321.
4. Mansouri R, Dietrich Je. Postoperative Course And Complications After Pull-Through Vaginoplasty For Distal Vaginal Atresia. J Ped And Adol Gyn 2015; 28(6): 433-436.
5. Pandya Ka, Koga H, Okawada M, Coran Ag, Yamataka A, Teitelbaum Dh. Vaginal Anomalies And Atresia Associated With İmperforate Anus: Diagnosis And Surgical Management. J Ped Surg 2015; 50(3): 431-437.
6. Tokmak A, Sarıkaya E. Mülleryan Kanal Anomalileri. Jin-Obst Neon Tıp Derg 2015; 12(2):83-88.
7. Orofino A, Lanzillotto Mp, Savino Cl, Woldemicael A, Gentile O, Paradies G. (2018). Acute Abdominal Pain İn An Adolescent Female With Herlyn-Werner-Wunderlich Syndrome And Hemicervicovaginal Atresia. J Ped Surg Case Rep 2018; 31: 17-19.
8. Kimble R, Molloy G, Sutton B. Partial Cervical Agenesis And Complete Vaginal Atresia. J Ped And Adol Gyn 2016; 29(3):43-47.
9. Moraloğlu Ö, Sucak A, Tetikk. Konjenital Vajen Anomalilerinin Tanı Ve Tedavisi. Jin-Obst Neon Tıp Derg 2012; 8(33).
10. Skinner B, Quint Eh. Obstructive Reproductive Tract Anomalies: A Review Of Surgical Management. J Min İnv Gyn 2017; 24(6): 901-908.
11. Soyer T. Topical Estrogen Therapy İn Labial Adhesions İn Children: Therapeutic Or Prophylactic?. J Ped And Adol Gyn 2007; 20(4): 241-244.
12. Omar Ha. Management Of Labial Adhesions İn Prepubertal Girls. J Ped And Adol Gyn 2000; 13(4): 183-185.
13. Tardieu Sc, Appelbaum H. Microperforate Hymen And Pyocolpos: A Case Report And Review Of The Literature. J Ped And Adol Gyn. 2018; 31(2):140-142.
14. Ramphul M, Perry L, Bhatia C. Neonatal İmperforate Hymen With Hydrocolpos. Bmj Case Rep 2016;2016.
15. Chang, My. Congenital Hydrocolpos Mimicking A Mature Cystic Teratoma İn The Pelvis. Neon Med 2016; 23(2): 127-130.
16. Reis M, Vicente Ap, Cominho J, Gomes As, Martins L, Nunes F. Pyometra And Pregnancy With Herlyn-Werner-Wunderlich Syndrome. Rev Brasil Gin Obst 2016; 38(12): 623-628.
17. Zhu L, Chen N, Tong Jl, Wang W, Zhang L, Lang Jh. New Classification Of Herlyn-Werner-Wunderlich Syndrome Chin Med J 2015; 128(2): 222.
18. Santos Xm, Dietrich Je. Obstructed Hemivagina With İpsilateral Renal Anomaly. J Ped And Adol Gyn 2016; 29(1): 7-10.
19. Rathod S, Samal Sk. Secondary Vaginal Atresia Treated With Vaginoplasty Using Amnion Graft: A Case Report. J Clin Diag Res 2014; 8(11):5.
20. Han Tt, Chen J, Wang S, Zhu L. Vaginal Atresia And Cervical Agenesis Combined With Asymmetric Septate Uterus: A Case Report Of A New Genital Malformation And Literature Review Med 2018;97(3).
21. Mansouri R, Dietrich Je. Postoperative Course And Complications After Pull-Through Vaginoplasty For Distal Vaginal Atresia. J Ped And Adol Gyn 2015;28(6): 433-436.
22. Singh Rr. A Case Report Of Cervıco Vagınal Atresıa. Yuva JMed Sci 2016; 2(3): 75-76.
23. Ramareddy Rs, Kumar A, Alladi A. Imperforate Hymen: Varied Presentation, New Associations, And Management. J Ind Ass Ped Surg 2017;22(4): 207.
24. Agarwal N, Singh S, Daya, R, Pathak A. Pyocolpos İn A 4 Year Old: A Rare Scenario. Int J Rep, Cont Obst And Gyn 2016; 2(3): 488-490.

25. Nagaraj Br, Basavalingu D, Paramesh Vm, Nagendra Pdk. (2016). Radiological Diagnosis Of Neonatal Hydrometrocolpos-A Case Report. J Clin And Diag Res 2016;10(3):18.
26. Bodduluri Vl, Srinivasan L. Hydrometrocolpos: A Lower Mesodermal Defects Sequence. J Fet Med 2015; 2(4): 191-196.
27. Kim Ik, Vellody R, Pohl Hg, Sharma K, Yadav B. (2018). Ultrasound-Guided İntroital Drainage Of Pyometrocolpos. J Ped Surg Case Rep 2018;33:4-6.