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Case Series of Reproductive Outcomes after Surgical Correction of Obstructed Hemivagina in OHVIRA

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Abstract

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) is a rare congenital developmental syndrome manifested by uterine duplication, lower genital tract obstruction, and unilateral renal anomaly. Literature on reproductive outcomes in this patient population is limited. The aim of this study is to describe obstetric outcomes after surgical correction of obstructed hemivagina in a longitudinal cohort of patients with a diagnosis of OHVIRA. All cases of OHVIRA presenting to a single tertiary care children's hospital from 1990 to 2021 were retrospectively reviewed. Three cases demonstrating a variety of clinically important reproductive outcomes are described in detail including risks such as retained products, endometritis, preterm labor, and malpresentation. Understanding the reproductive outcomes associated with this diagnosis is important for practitioners seeking to counsel and care for patients with this diagnosis. This case series demonstrates a wide array of potential gynecologic and obstetric risks, though ultimately with successful term and near-term pregnancies.

Keywords
- hemivagina
- renal agenesis
- infertility
- Mullerian anomaly

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) is a rare congenital developmental syndrome manifested by uterine duplication, lower genital tract obstruction, and unilateral renal anomaly.[1] The exact incidence is not known but is estimated to occur in 0.1 to 3.8% of the general population.[2] Prior reports have detailed surgical outcomes for patients with OHVIRA, though literature on reproductive outcomes in this patient population is limited. Among the studies reporting on obstetric outcomes in patients with OHVIRA, the most common complications include recurrent miscarriage, malpresentation, postpartum hemorrhage, retained placenta, fetal growth restriction, preterm birth, and premature rupture of membranes.[2,3] The aim of this study is to describe obstetric outcomes after surgical correction of obstructed hemivagina in a longitudinal cohort of patients with a diagnosis of OHVIRA.

Methods

All cases of OHVIRA presenting to a single tertiary care children's hospital from 1990 to 2021 were retrospectively reviewed. Among 111 individuals diagnosed and surgically treated for OHVIRA, 69 had corresponding medical records at an affiliated academic institution that provides adult obstetric care. Three individuals were identified to have a record of pregnancy at this affiliated obstetric hospital. Chart review was performed to gather data on reproductive care, including infertility treatment, pregnancy, prenatal care, labor and

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delivery. This study was approved by the Institutional Review Boards of both the adult and children's hospitals.

Cases

Patient A

Patient A was known from infancy to have a solitary kidney and duplicated ureteral system. At the age of 9 years, she began menstruating with associated severe dysmenorrhea. She was noted by her pediatrician to have a vaginal duplication. Ultrasound imaging demonstrated a right obstructed hemivagina and ipsilateral renal agenesis. At the age of 9 years, she had a vaginoplasty to create a single vagina, and intraoperatively, she was noted to have two cervices. At the age of 12 years, she underwent a diagnostic laparoscopy in the setting of chronic pelvic pain, which revealed two hemiuteri and stage I endometriosis based on the American Society for Reproductive Medicine staging system[4] for which she was initiated on depo medroxyprogesterone acetate.

During a lapse in contraception use, she became spontaneously pregnant at the age of 21 years. She was diagnosed with an ectopic pregnancy that was treated successfully with methotrexate at an outside institution. She was followed up annually by gynecology at our institution and had a term cesarean section at the age of 23 years (2013), but records from delivery at an outside hospital were not available. Following this delivery, she had a bilateral tubal ligation and was transitioned back to depo medroxyprogesterone acetate with later addition of norethindrone acetate for control of dysmenorrhea.

Patient B

Patient B first presented to gynecology care at the age of 30 years after an abnormal pap smear with her primary care provider. She underwent a colposcopy with benign findings and was followed up closely with serial pap smears. On initial pelvic examination, her uterus was notably deviated to the right. She returned to gynecologic care at the age of 33 years for infertility evaluation. During hysteroscopic evaluation, she was noted to have an apparently unicornuate uterus. However, a follow-up magnetic resonance imaging confirmed uterine didelphys with a left obstructed hemivagina and ipsilateral renal agenesis.

At the age of 33 years, she underwent an operative laparoscopy with lysis of adhesions and destruction of stage II endometriosis. A left vaginoplasty was also performed to relieve the obstructed left hemivagina. She was intraoperatively noted to have two cervices and two uteruses.

She underwent successful intrauterine insemination (IUI) with donor sperm due to male factor infertility. Implantation occurred in the right uterine horn. Her pregnancy was uncomplicated until preterm rupture of membranes at $36^{5/7}$ weeks' gestation. In 2009, she underwent an uncomplicated primary cesarean section per maternal request for a vertex presenting, 2,700 g newborn. She continues to be followed up by our institution for persistent abnormal pap smears (atypical squamous cells of uncertain significance, cannot rule out high grade) and a recent loop excision procedure of both cervices without malignancy. To date, there have been no further attempts of pregnancy.

Patient C

Patient C first presented to our institution's emergency department at the age of 15 years for evaluation of abdominal pain and fever. Pelvic ultrasound revealed a solitary right kidney and a single uterus with hematocolpos. A computed tomography scan was performed which revealed appendiceal inflammation concerning for appendicitis in addition to uterine didelphys with dilated left vagina. The patient was taken to the operating room for laparoscopic appendectomy, with intraoperative findings notable for a normal appearing appendix, though with inflamed pelvic structures and confirmed uterine didelphys. Given concern for infection secondary to the hemivaginal obstruction, she underwent a vaginoplasty with release of purulent material from the obstructed left hemivagina.

At the age of 17 years, she underwent laparoscopic destruction of stage I endometriosis in the setting of severe dysmenorrhea refractory to combined oral contraception. Depo medroxyprogesterone acetate was administered for postoperative menstrual suppression. During a lapse in contraceptive use, she became spontaneously pregnant and subsequently underwent a pregnancy termination via dilation and curettage. Her chronic pelvic pain persisted despite depo medroxyprogesterone acetate and norethindrone acetate. At the age of 21 years, she underwent diagnostic laparoscopy, which revealed no residual endometriosis. She also underwent hysteroscopy and chromopertubation for infertility workup, which confirmed tubal patency and normal appearing endometrial cavities.

Following an unrevealing infertility workup, she attempted spontaneous conception over 3 years with two chemical pregnancies and eventually, at the age of 24 years (2020), conceived in the right uterine cavity with the first cycle of IUI. She had an uncomplicated prenatal course followed by a late-term induction of labor. She underwent cervical ripening with misoprostol and a Cook balloon and then transitioned to oxytocin. She received an epidural, and artificial rupture of membranes was performed. She pushed for 60 minutes for an uncomplicated vaginal delivery of a 3,090 g newborn in the occiput anterior position. She had an uncomplicated placental delivery. Her postdelivery course was complicated by chorioamnionitis treated with IV antibiotics.

She reinitiated menstrual management with depo medroxyprogesterone acetate. However, she experienced persistent daily bleeding both with this method and with transition to the etonogestrel implant. She had an endometrial biopsy of the right uterine cavity, which revealed chronic endometritis, but could not tolerate biopsy of the left uterine cavity. She continued to have abnormal bleeding despite completion of empiric antibiotics. At the age of 25 years, she underwent hysteroscopy revealing the right uterine cavity to have a small focus of calcified material along the medial wall. This lesion was resected and pathology revealed chronic endomyometritis with rare degenerating

trophoblast cells suggestive of an early placental site nodule. Hysteroscopic evaluation of the left uterine cavity noted a 2-mm blood-filled cyst in the endometrium that was intraoperatively lysed.

Following this procedure, she became spontaneously pregnant in the right hemiuterus at the age of 26 years (2022). Her pregnancy was complicated by polyhydramnios with a maximum vertical pocket of 11 cm and was subsequently induced at 39 weeks' gestation. Her induction began with a Cook balloon, oxytocin, and early epidural. The balloon fell out after 2 hours and followed by artificial rupture of membranes. She progressed in active labor to complete over 6 hours. She pushed for 20 minutes followed by an uncomplicated vaginal delivery of a 3,400 g male newborn in the occiput anterior position. She intends to have a bilateral interval salpingectomy for permanent sterilization.

Discussion

The cases presented here illustrate the complexity that accompanies reproductive and obstetric counseling in individuals with a diagnosis of OHVIRA after surgical repair. Our findings are similar to other international studies investigating reproductive outcomes in patients with OHVIRA or other uterine anomalies. Haddad et al reported on a survey of the reproductive outcomes of 42 patients after surgical repair of obstructed hemivagina, 9 of whom reported a total of 20 pregnancies.[3] The live birth rate was 69%, with nine reported as vaginal deliveries and four cesarean sections.[5] Heinonen found a high rate of preterm birth (24%), malpresentation (51%), and cesarean section (84%) among patient with uterine didelphys and longitudinal vaginal septum.[6] Among the nine patients with a repaired obstructed hemivagina, six had successful deliveries.[4] Our case series adds to this international data by providing background on the initial clinical presentation and surgical management of the obstructive Mullerian anomalies and other gynecologic history prior to achieving pregnancy. To our knowledge, this is the first case series to describe the surgical management of OHVIRA in detail with long-term follow-up of corresponding obstetric outcomes in the United States. The details and outcomes of which are summarized in ►**Table 1**.

By the seventh week of embryogenesis, the Mullerian ducts have elongated and crossed the metanephric ducts to meet in the midline. At 12 weeks, the caudal Mullerian ducts fuse, forming a uterovaginal canal. Following this, canalization of each duct occurs and the resulted septum is reabsorbed by 20 weeks.[3] Multiple studies have demonstrated increased rates of preterm birth in the setting of canalization defects.[7] However, limited trials have suggested no benefit of septum resection or metroplasty on reproductive outcomes.[8] Though routine cervical length monitoring is performed in all patients with uterine anomalies at our institution, there is no evidence that routine cerclage placement improves obstetric outcomes in this population. The elevated risk of preterm delivery is thought to be a consequence of uterine size and shape rather than cervical insufficiency.[5] Further research is needed to ascertain if interventional procedures or alterations in prenatal

Table 1 Clinical and surgical characteristics of patients with OHVIRA and their associated obstetric outcomes

Patient	Presenting complaint (age, y)	Mullerian anomaly	Surgical repair (age)	Additional gynecologic morbidities	Pregnancy outcomes/obstetric complications
A	Dysmenorrhea (9)	Uterine didelphis and right obstructed hemivagina with ipsilateral renal agenesis	1. Vaginoplasty to create single vagina (9) 2. dx laparoscopy with ablation and excision of endometriosis (12)	Stage I endometriosis	G1—ectopic (methotrexate) G2—term cesarean section
B	Infertility (33)	Uterine didelphys with left obstructed hemivagina and ipsilateral renal agenesis	1. Diagnostic laparoscopy with ablation of stage II endometriosis and concurrent vaginoplasty for obstructed left hemivagina (33)	Stage II endometriosis	G1—termination G2—pPROM at 36 wk, elective cesarean section for vertex presenting infant
C	Pelvic pain (15)	Uterine didelphys with dilated left vagina	1. Vaginoplasty with release of purulent material from obstructed left vaginal vault (12) 2. Laparoscopic ablation/excision of endometriosis stage I (17) 3. Diagnostic laparoscopy (no endometriosis) and hysteroscopy with chromopertubation with patent's fallopian tubes (21)	Stage I endometriosis, infertility	G1—SAB G2—SAB G3—IUI pregnancy in right horn, 39 wk IOL and uncomplicated SVD G4—spontaneous pregnancy in right hemiuterus, induced at 39 wk for polyhydramnios

Abbreviations: IOL, induction of labor; IUI, intrauterine insemination; OHVIRA, obstructed hemivagina and ipsilateral renal anomaly; pPROM, preterm premature rupture of the membranes; SAB, spontaneous abortion; SVD, spontaneous vaginal delivery.

surveillance may improve pregnancy outcomes in patients with Mullerian anomalies.

Endometriosis frequently coincides with a diagnosis of obstructive reproductive tract anomaly. The majority of cases of even advanced stage endometriosis will resolve with relief of the vaginal obstruction, though Silveira and Laufer described a case series of patients in whom endometriosis persisted despite surgical correction of the outflow anomaly.[9] Two of the three described cases required continued menstrual suppression for relief of endometriosis symptoms even after vaginoplasty, highlighting the importance of maintaining endometriosis on the differential of abdominal pain in patients with corrected outflow obstruction. Furthermore, endometriosis is known to impact fertility outcomes, perhaps contributing to the high rate of assisted reproductive technology utilized by our cohort.[10]

Our case series also supports prior literature on the potential obstetric delivery considerations associated with OHVIRA syndrome or other Mullerian anomalies. Two patients underwent cesarean section, though the indications for cesarean delivery were not known for Patient A. Prior literature demonstrates an elevated cesarean section rate in patients with Mullerian duplication anomalies, often due to malpresentation and labor dystocia.[5] Risk of uterine rupture is known to be associated with various Mullerian anomalies, though most often occurring in women with a unicornuate uterus with pregnancy in a rudimentary horn. A single 25-patient study looking at rates of rupture in the setting of trial of labor after cesarean section in women with Mullerian duct anomalies found a high (8%) risk of uterine rupture in that population, as compared to a 1% risk in matched controls.[11] Patient C had a successful vaginal delivery of two term infants, further supporting the safety and feasibility of vaginal delivery after vaginoplasty correction of an obstructed hemivagina.

Patient C experienced retained products of conception leading to abnormal uterine bleeding and chronic endometritis. We hypothesize that the anatomic variation and often widely divergent angles of the uterine horns may lead to difficulty of outflow of products of conception after miscarriage, termination, or placental delivery and techniques to ensure complete evacuation of the uterine cavity should be further investigated in patients with Mullerian anomalies. Patient B continues to undergo surveillance for abnormal pap smears of both cervices, highlighting the importance of identification and sampling of both cervices for cervical cancer screening in patients with cervical duplication.[12,13] This anatomic variation also extends to challenges in access to both endometrial canals for evaluation of abnormal uterine bleeding as experienced with Patient C.

Of note, all of the patients in this cohort experienced known pregnancies in the uterine horn contralateral to the prior vaginal obstruction. Though pregnancies most often occur in the contralateral hemiuterus, studies have demonstrated that pregnancies can occur in the hemiuterus ipsilateral to the repaired vaginal obstruction, supporting the role of vaginoplasty repair rather than removal of the obstructed uterine horn to preserve fertility potential on the obstructed side.[3,4] The lack of ipsilateral pregnancies is likely secondary to the small sample size, limiting generalizability. Though our institution is a referral center for congenital Mullerian anomalies, the report of obstetric outcomes in our study sample was limited as many patients either relocated or sought obstetric care at another nonaffiliated institution if they became pregnant.

Conclusion

Long-term follow-up of patients with OHVIRA after surgical correction of outflow obstruction is limited. The reproductive and obstetric outcomes in this uncommon Mullerian anomaly are varied and warrant continued investigation to optimize preconception counseling. In an era of increased access to imaging, the diagnosis of OHVIRA is likely to become more common in the preconception population. Understanding the reproductive outcomes associated with this diagnosis is important for practitioners seeking to counsel and care for patients with this diagnosis. This case series demonstrates a wide array of potential gynecologic and obstetric risks, though ultimately with successful term and near-term pregnancies.

Attestation

- Data regarding any of the subjects in the study has not been previously published unless specified.
- Data will be made available to the editors of the journal for review or query upon request.

Capsule

Reproductive outcomes after OHVIRA surgery are generally favorable but include a number of clinical risks that are important for patient–provider counseling.

Funding

None.

Conflict of Interest

None declared.

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