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HERLYN-WERNER-WUNDERLICH SYNDROME IN LATE PREGNANCY: A CASE REPORT ON EMERGENCY CESAREAN DELIVERY FOR LEAKING PER VAGINUM IN THE PRESENCE OF A CERVICAL STITCH

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ABSTRACT

Background: Herlyn-Werner-Wunderlich syndrome is a rare Müllerian anomaly characterized by uterine didelphys, obstructed hemivagina and ipsilateral renal agenesis, often diagnosed in adolescence or early adulthood. Pregnancy in such anomalous uteri carries high risks of miscarriage, preterm birth, malpresentation and cervical incompetence, frequently necessitating prophylactic cerclage and operative delivery. **Methodology:** This case report was conducted in the Department of Obstetrics and Gynecology, CPR Hospital, Kolhapur, to document the presentation and management of a pregnant woman with Herlyn-Werner-Wunderlich syndrome presenting with leaking per vaginum and a cervical stitch in situ. Data were extracted from records, examination notes, ultrasound reports, operative findings and postnatal follow-up, then arranged chronologically to highlight diagnostic dilemmas, therapeutic decisions and outcomes, with informed consent and confidentiality maintained. **Case Details:** A 27-year-old G3P1D1A1 at 35.3 weeks, with known Herlyn-Werner-Wunderlich syndrome and prior excision of an oblique vaginal septum with pyocolpos drainage, presented with two hours' history of lower abdominal pain and leaking per vaginum. Her obstetric history included one preterm neonatal death due to cardiac anomaly and one early abortion; in the current pregnancy, a prophylactic modified Shirodkar cerclage had been placed on the right cervix in a didelphys uterus. Examination revealed a laterally localized gravid uterus, stable vitals and findings suggestive of prelabour rupture of membranes in one uterine horn with stitch in situ. Serial ultrasounds had shown a single live fetus in a separated cavity with adequate liquor and normal growth. With PROM, uterine anomaly and cerclage, an emergency lower-segment cesarean section was performed, confirming uterine didelphys, allowing stitch removal and safe delivery of a healthy neonate without intraoperative or postoperative complications. **Conclusion:** This case demonstrates successful late-pregnancy management of Herlyn-Werner-Wunderlich syndrome complicated by preterm premature rupture of membranes and cervical cerclage through timely emergency cesarean delivery. Individualized planning at a tertiary centre preserved maternal reproductive potential while ensuring favorable perinatal outcome.

KEYWORDS : Herlyn-Werner-Wunderlich Syndrome; Uterine Didelphys; Obstructed Hemivagina; Cervical Cerclage; Emergency Cesarean Section

INTRODUCTION

Herlyn-Werner-Wunderlich (HWW) syndrome, also known as Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) syndrome, represents a rare congenital anomaly of the female genitourinary tract characterized by the classic triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis[1]. This condition arises from abnormal embryological development during the eighth week of gestation, where improper fusion of the Müllerian ducts and defective development of the Wolffian ducts lead to concurrent reproductive and urinary tract malformations[2]. The estimated incidence ranges from 0.1% to 3.8% among women with Müllerian duct anomalies, though the true prevalence remains underestimated[3,4]. Delayed diagnosis poses significant risks for long-term complications such as endometriosis, pelvic inflammatory disease, tubal adhesions, and infertility[5].

Pregnancy in women with uterine didelphys presents substantial obstetric challenges, with spontaneous abortion rates reported between 32% and 52% and preterm birth rates ranging from 20% to 45%[6]. The anatomical configuration of duplicated uterine cavities, each possessing its own cervix, predisposes to cervical incompetence due to congenital weakness of the internal os and accommodation difficulties[7]. Cervical cerclage emerges as a critical intervention in these high-risk pregnancies, with both therapeutic and prophylactic applications demonstrated to improve obstetric outcomes[8]. Transabdominal cerclage offers optimal results when the cervix is hypoplastic or lacks adequate portio vaginalis for vaginal approaches[9]. However, the presence of a cervical stitch introduces potential complications, including suture displacement, cervical laceration, and ascending infection, particularly when accompanied by premature rupture of membranes or chorioamnionitis[10].

The occurrence of leaking per vaginum in the presence of an in-situ cervical stitch represents a particularly complex clinical scenario in HWW syndrome patients[11]. Furthermore, the obstructed hemivagina component of HWW syndrome may mask or complicate the source of vaginal leakage, necessitating meticulous differentiation between amniotic fluid from the gravid uterus and inflammatory or hematocolpos discharge from the obstructed system[12]. The management requires multidisciplinary coordination between maternal-fetal medicine specialists, urogynecologists, and neonatologists to optimize both maternal and fetal outcomes while preserving future fertility potential[13].

The present case report is aimed to describe the clinical presentation, diagnostic evaluation and obstetric management of a pregnant woman with HWW syndrome presenting with leaking per vaginum with a cervical stitch in situ at a tertiary care facility. The rarity of this condition duplicated cervical anatomy introduces unprecedented complexity in diagnosing rupture of membranes versus alternative sources of vaginal discharge.

METHODOLOGY

This case report was done in the Department of Obstetrics and Gynecology, CPR Hospital, Kolhapur. The purpose of the report was to describe the clinical presentation, diagnostic evaluation and obstetric management of a pregnant woman with Herlyn-Werner-Wunderlich syndrome presenting with leaking per vaginum with a cervical stitch in situ. Data were collected from the patient's medical records, clinical

examination notes, antenatal ultrasound reports, operative findings and postoperative follow-up documentation. All relevant observations were analyzed in chronological order to highlight diagnostic challenges and management decisions. Patient identity was kept confidential and informed consent was obtained for academic publication.

Case History

A 27-year-old woman, gravida 3 para 1 live 1 abortion 1, presented at 35.3 weeks of gestation with sudden onset lower abdominal pain and leaking per vaginum for two hours. The present pregnancy was conceived spontaneously and had been uneventful until late third trimester. She was on routine antenatal supplements and had undergone all scheduled scans. The pregnancy had been monitored closely due to a known congenital Müllerian anomaly suggestive of Herlyn-Werner-Wunderlich syndrome. Earlier in the pregnancy, she underwent a cervical encirclage using a modified Shirodkar technique, which was placed on the right cervix as the patient had two cervices. Her obstetric history included a preterm delivery at seven months resulting in neonatal death due to cardiac anomaly and one early spontaneous abortion. She also had a significant gynecological history of excision of a left oblique vaginal septum with drainage of pyocolpos in 2018. Patients had taken routine ANC supplements, progesterone support and prophylactic uterine relaxants.

Clinical Examination

On admission she was alert, oriented and vitally stable. Abdominal inspection showed a gravid uterus with visible striae and healed laparoscopic scars. Fundal height corresponded to approximately 34–36 weeks. Palpation revealed a lateralized uterine contour, with a uniformly convex area on the left suggestive of fetal back and irregular prominences on the right consistent with fetal limbs. First pelvic grip indicated the presenting part was not engaged. The uterus felt deviated toward the right. Cardiovascular and respiratory examinations were normal, with no edema or signs of systemic illness. Per abdomen findings and the complaint of leaking per vaginum in presence of in situ cervical stitch strongly suggested prelabor rupture of membranes with early labor pains in a pregnancy occurring in one horn of a duplicated uterus. No evidence of hernia or abdominal tenderness beyond contractions was noted.

Investigations

Serial antenatal ultrasounds confirmed a single live fetus located in a laterally separated uterine cavity rather than a single midline cavity. Early viability scans correlated with gestational age, and subsequent anomaly and growth scans showed normal Dopplers and appropriate interval growth. Fetal heart rates throughout pregnancy ranged between 148–173 beats per minute. The placenta was described in scans as left lateral and liquor was adequate. Imaging also repeatedly identified the altered uterine configuration and documentation of two cervices, supporting the suspicion of a uterine duplication anomaly. A cervical length of 3.0 cm and a closed os were recorded prior to cerclage insertion.

On admission laboratory investigations were within normal limits and did not indicate infection. Clinical diagnosis of membrane rupture with the cervical stitch in situ was made primarily based on examination findings supported by imaging history.

Final Diagnosis

A 27-year-old G3P1D1A1 (Gravida 3 Para1 Death 1 Abortion 1) at 35.3 weeks with Herlyn-Werner-Wunderlich syndrome and cervical cerclage in situ, presenting with prelabor rupture of membranes and labor pains, with pregnancy located in one horn of a didelphys uterus.

Management

Given the leaking per vaginum at 35.3 weeks with uterine contractions in the presence of a cervical stitch and congenital uterine malformation, an emergency lower segment cesarean section was planned. Intraoperatively, a didelphys uterus was confirmed with pregnancy in one horn. The cervical stitch was identified and removed during the procedure. The fetus was delivered safely through a lower segment uterine incision, and the placenta was removed intact. Hemostasis was secured, and the uterus was closed in layers with careful attention to the anomalous anatomy. There were no intraoperative complications and no excessive bleeding.

Postoperative management included analgesia, prophylactic antibiotics and monitoring of vitals, lochia and wound condition. The patient remained stable with good postoperative recovery and no signs of infection or sepsis. Breastfeeding support and routine postnatal counseling were provided. She was discharged on postoperative day four with advice for follow-up after ten days and documentation of the didelphys uterus and stitch removal for future pregnancies.

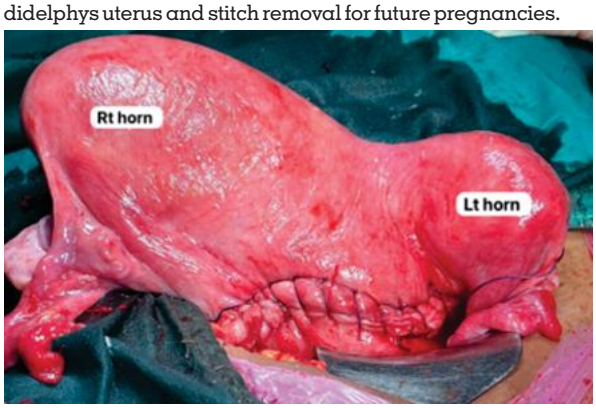

Image 1. Didelphys Uterus With Non-communicating Horns

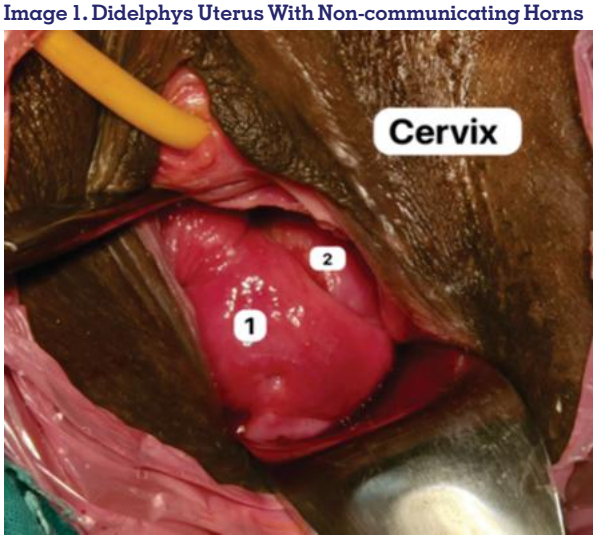

Image 2. Two Separate Cervix

DISCUSSION

The present case highlights the obstetric complexity of Herlyn-Werner-Wunderlich syndrome presenting with premature rupture of membranes at 35.3 weeks gestation in the presence of a cervical stitch, necessitating emergency cesarean delivery. The successful maternal and fetal outcomes underscore the critical importance of tertiary care management for this rare Müllerian anomaly, particularly when complicated by cervical incompetence requiring prophylactic cerclage.

Comparison with recent literature reveals both convergent and divergent management approaches. Tuqan et al.[1] reported a case of HWW syndrome managed with elective cesarean section at 38 weeks without preoperative cervical cerclage. This difference likely stems from our patient's history

of prior preterm delivery and documented cervical incompetence, factors that necessitated prophylactic cerclage placement early in gestation. Oliveira et al.[14] described a 40-year-old primigravida with HWW syndrome who experienced frequent contractions from 15 weeks yet achieved term delivery via emergency cesarean after failed forceps extraction. While their case lacked cervical cerclage, it similarly demonstrated the high rates of malpresentation and operative intervention characteristic of this population, with 80-84% of HWW pregnancies requiring cesarean delivery due to abnormal fetal lies.

The gestational age at delivery in our case (35.3 weeks) reflects the inherent risks associated with cervical stitch and premature rupture of membranes, differing from reports of term deliveries in surgically optimized patients. Bunnell et al.[15] documented a case series where patients achieved term pregnancies after preconception surgical correction of obstructed hemivagina. The earlier delivery in our patient may be attributed to persistent cervical weakness despite septum excision in 2018 and cerclage placement, suggesting that prior pelvic surgery does not fully mitigate the risk of preterm birth in HWW syndrome. This contrasts with Alqntash et al's[16] (2025) case report of term delivery following early diagnosis and surgical management, where the patient conceived spontaneously within 11 months postoperatively. The variation in outcomes likely reflects differences in baseline cervical integrity, timing of surgical intervention, and the presence of concurrent risk factors such as our patient's history of neonatal loss from cardiac anomaly.

The decision to perform emergency cesarean rather than expectant management or attempted vaginal delivery aligns with current evidence regarding PROM in Müllerian anomalies. Maiti et al.[6] previously described a case of uterine didelphys with cervical incompetence managed by cesarean at 37 weeks while leaving the merselene suture undisturbed. However, our approach of stitch removal was justified by the duplicated cervical anatomy and risk of suture-related complications during uterine involution. This individualized management reflects the absence of standardized protocols for HWW syndrome complicated by cerclage and PROM, necessitating case-by-case decision-making at experienced tertiary centers.

CONCLUSION

This case illustrates the successful management of a 27-year-old G3P1D1A1 at 35.3 weeks gestation with Herlyn-Werner-Wunderlich syndrome, presenting with preterm premature rupture of membranes and abdominal pain despite cervical cerclage. Emergency cesarean delivery following rupture of membranes ensures maternal and fetal safety while preserving reproductive potential.

Multidisciplinary evaluation, appropriate imaging, and individualized labor planning are proving essential in preventing complications and optimizing outcomes in late-pregnancy presentations of Müllerian anomalies. Long-term follow-up should include preconception counseling and multidisciplinary planning to improve obstetric outcomes and minimize recurrence of complications.

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