

Rudimentary Horn Pregnancy: Diagnosis and Management

Ahmed Samy Yassin^{1*}

¹Sidra Medical and Research Centre/Weill Cornell Medical College, Qatar

*Corresponding author: Dr. Ahmed Samy Yassin, Sidra Medical and Research Center, 13th Floor, Room 18 A – Al Nasr Tower, PO Box 26999, Doha, Qatar, Tel: 00974 30311505; Email: asay5960@aol.com, ayassin@sidra.org

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Abstract

Rudimentary Horn Pregnancy (RHP) is a rare form of pregnancy in a uterine rudimentary horn with a reported incidence of 1:76,000 and an estimated maternal mortality rate of 0.5%. Around 40% of women with RHP are asymptomatic when present during routine ultrasound scan especially in the first trimester and many women often have a history of previous normal pregnancies and vaginal delivery. Despite the frequent use of ultrasonography during pregnancy, up to 50% of RHP cases are diagnosed only following rupture. Advanced RHP may rupture with little warning at any time, with significant risk to maternal health; hence early diagnosis can help in planning management before its rupture. This review describe the Müllerian duct anomaly resulting in the formation of a rudimentary horn, the available methods for early diagnosis of RHP and the possible differential diagnosis of this rare type of pregnancy and the different treatment options available including modern laparoscopic treatment

Keywords: Rudimentary Horn Pregnancy; Ultrasound; Magnetic Resonance Imaging; Laparoscopic Excision

Introduction

Rudimentary horn pregnancy (RHP) (figure 1) is a rare form of pregnancy in a rudimentary uterine horn with a reported incidence of 1 in 76,000–1 in 150,000 [1, 2]. RHP results in rupture in 80–90 % of cases during the second or third trimester due to poorly developed musculature that cannot stretch and the increased reported presence of placenta percreta in cases of RHP due to the above mentioned poor musculature and the presence of thin or nonfunctional endometrium and scant decidualization resulting in life-threatening hemorrhage [2-6].

The maternal mortality rate associated with RHP has reduced from 18 % in the nineteenth century to 0.5% now with very few reported cases of fetal survival [7-11]. Delayed diagnosis

of RHP until it ruptures in second or third trimester is the main factor contributing to the high maternal mortality rate in comparison to that for tubal ectopic pregnancies.

Embryology

The female reproductive tract develops from a pair of Müllerian ducts that form the following structures: fallopian tube, uterus, cervix and the upper two-thirds of the vagina. Normal development of the Müllerian ducts depends on the completion of three phases: organogenesis, fusion and septal resorption [12].

Unicornuate uterus results from complete or near-complete arrest of organogenesis of one of the Müllerian ducts and in-

complete fusion of the two Müllerian ducts. The overall incidence of unicornuate uterus in the general population is 1/4020 [13].

The American Society of Reproductive Medicine developed one of the most commonly used classification system for Müllerian duct anomalies (MDA) [14].



Figure 1. Laparoscopy showing right unicornuate uterus and left rudimentary horn ectopic pregnancy.

According to this classification unicornuate uterus is classified as Class II MDA with four possible subtypes (figure 2): (Class II a) Unicornuate uterus with cavitory communicating rudimentary horn, (Class II b) Unicornuate uterus with cavitory non-communicating rudimentary horn, (Class II c) Unicornuate uterus with non-cavitory (non-functional) rudimentary horn and (Class II d) Unicornuate uterus without rudimentary horn [12,14].

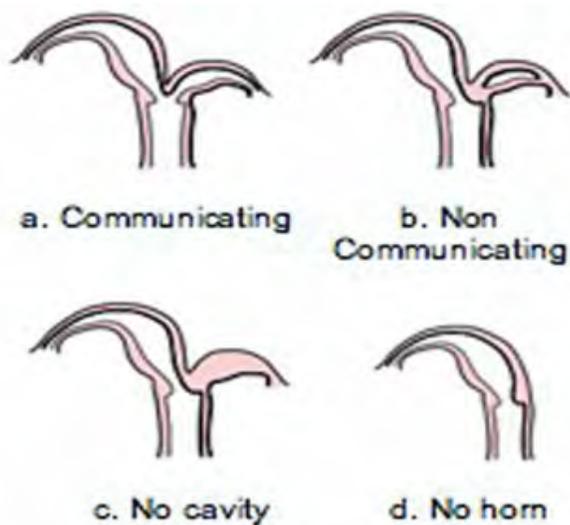


Figure 2. Unicornuate uterus (Class II) Subtypes ^{12,14}.

In more than 75 % of cases of unicornuate uterus, a contralateral rudimentary horn is present. A review of 366 cases (between 1966–2003) showed that the rudimentary horn had cavity in 82%, endometrium in 67%, and no communication with uterus in 92% of cases, reinforcing the theory of peritoneal transmigration of sperm or fertilised ovum to the non-communicating rudimentary horn in cases of RHP [7,8].

Renal anomalies reported to be present in up to 40% of unicornuate uterus patients and are ipsilateral to the rudimentary horn with renal agenesis is the commonly reported anomaly. Other anomalies include ectopic kidney, horseshoe kidney, renal dysplasia and duplicated collecting systems [12,15].

Clinical Presentation

Despite improvements in the diagnosis and treatment of RHP, 50% are diagnosed following uterine rupture in the second or third trimester and the morbidity associated with this condition remains very high [2]. That is why every effort should be made to diagnose RHP at an early stage during the pregnancy.

One of the reasons for the late diagnosis is that around 40% of women with RHP are asymptomatic when present during routine ultrasound scan especially in the first trimester and many women often have a history of previous normal pregnancy and vaginal delivery [8, 6, 16]. Because of that and the catastrophic life-threatening hemorrhage which may occur if RHP ruptured, high index of clinical suspicion should be exercised in pregnant women with or without abdominal pain and any of the following; the presence of adnexal mass, mild tenderness in the abdomen, free fluid, reduced fundal height, malpresentations, anhydramnios, intrauterine fetal demise, failed induction or termination of pregnancy, and an empty lower pole of the uterus. Pregnant women with the above should be carefully investigated, especially if they have previous history of menorrhagia, dysmenorrhea, endometriosis, infertility, recurrent miscarriages, and suspected malformation as these conditions are common in women with MDA [2,4,5,8].

Symptomatic women with RHP tend to complain of abdominal pain, which occurs at an average gestational age of 21 weeks [2].

A 10 years world literature review showed a total of seventy-seven cases of RHP reported over this period. Abdomen pain was the commonest symptom seen in 47 % of cases, followed by shock/ unstable vitals in 26 %, failed induction of labor in 16.85 %, bleeding per vaginal and intrauterine death in 13 % each, and anhydramnios in 9 % of cases. Signs of rupture, peritonitis, acute abdomen with increasing abdominal fluid and decreasing hematocrit are seen in cases of rupture or impending rupture of RHP. The location of the pain varies from the lower abdomen to the epigastrium [4].

Diagnosis

Early, prerupture diagnosis is of major importance and can provide the option of safe and effective treatment. However, early diagnosis of a RHP can be difficult. Many cases of late and false diagnosis leading to uterine rupture have been repeatedly reported in literatures, with some cases diagnosed only after failed attempt to evacuate the uterus for termination of pregnancy [5,16,17].

Ultrasound reported to be successful in prerupture diagnose of RHP and differentiating it from other types of pregnancy when done during the first trimester but it becomes more difficult as the pregnancy progresses [10].

The following criteria can be used to diagnose RHP on ultrasound examination (figure 3) [10]:

1) a single interstitial portion of fallopian tube in the empty unicornuate uterine body; 2) a gestational sac, mobile, separate from the empty uterus and surrounded by myometrium; 3) a vascular pedicle adjoining the gestational sac to the unicornuate uterus [10]. Other suggested findings including the presence of thin myometrium ring all around the gestational sac, absent continuity between the cervical canal and the lumen of the pregnant horn and hypervascularisation typical of placenta accreta may support the diagnosis [6,18].

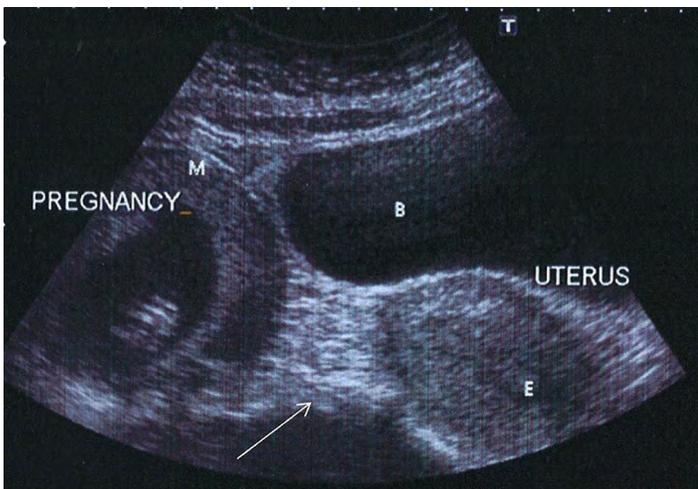


Figure 3. Ultrasound findings of right Rudimentary horn pregnancy with fetal pole surrounded all around by myometrium (M) beside an empty uterus with prominent endometrium (E), Bladder (B) and a band of tissue connecting the rudimentary horn to the displaced empty unicornuate uterus is seen (arrow).

The differential diagnosis of an ultrasonographically suspected RHP is tubal pregnancy, interstitial pregnancy and a pregnancy in a bicornuate or septate uterus (Table 1). A tubal pregnancy will not show a ring of myometrium surrounding the gestational sac, but the differentiation between the 2 latter conditions

and RHP may be difficult [6]. Interstitial pregnancy cannot be separated from the uterus (immobile gestational sac) and can be diagnosed by visualization of the interstitial line (echogenic line) adjoining the gestational sac and the lateral aspect of the uterine cavity and demonstrating the ipsilateral interstitial tube adjoining the uterine cavity and the gestational sac (figure 4) [10,19]. Variation in thickness of the myometrium in 2 horns and a marked distance between them favor the diagnosis of a RHP over that of pregnancy in a bicornuate uterus. In contrast to RHP, continuity between the endometrium lining the gestational sac and the other uterine horn is typical for a pregnancy in a bicornuate uterus [6].

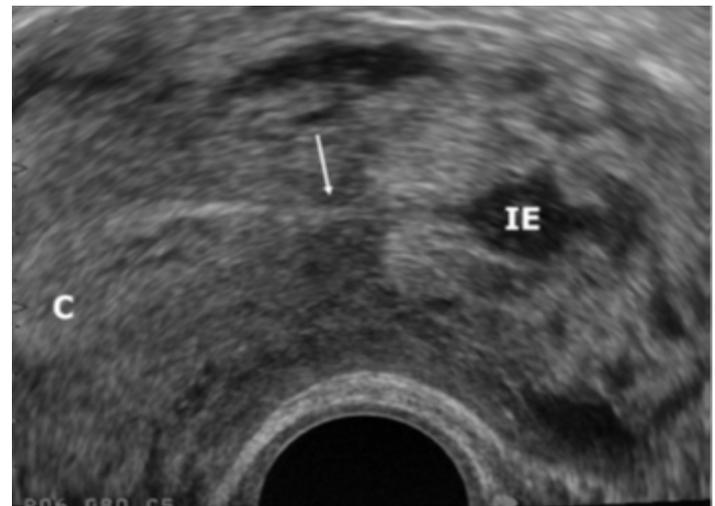


Figure 4. Ultrasound findings of Interstitial ectopic pregnancy (IE), Interstitial line sign (echogenic line) is seen (arrow) joining the endometrial cavity (c) ¹⁹.

In spite of the above, ultrasound sensitivity for diagnosis of rudimentary RHP ranges from 29 to 33 % only, that is why Magnetic Resonance Imaging (MRI) is recommended as a problem solving tool in cases of dilemma on ultrasonographic imaging for confirmation of RHP [8,16]. The main advantage of MRI as compared with ultrasound is the ability to visualize the whole uterus in one image plan, and thus identify the exact site of implantation. MRI provide images with high soft-tissue contrast, and multiplanar images which reveals both internal and external uterine structure, thus it is a very useful noninvasive tool for diagnosis of MDA, identifying communication between the uterine cavity and horn, and help to plan surgery by localizing the position of the horn attachment to the unicornuate uterus and identifying whether this attachment is broad attachment or just a small fibrous band [20-23]. The contralateral dominant, or nonrudimentary unicornuate horn will be laterally displaced and has been described as a “banana-shaped uterus” [22,24]. In cases of RHP, coronal sagittal and axial planes are used for accurate assessment of uterine connection to the horn and either confirm or rule out communication with endometrial cavity or endocervical canal [22].

Table 1. Differential diagnosis of rudimentary horn pregnancy.

Site of pregnancy	Rudimentary horn pregnancy	Interstitial pregnancy	Bicornuate uterus Pregnancy
Helpful scan findings	(1) Pseudopattern of an asymmetrical bicornuate uterus with variation in thickness of the myometrium surrounding the 2 horns and marked distance between the 2 horns (2) A gestational sac mobile and separate from the empty uterus (3) Absence of visual communication between tissue surrounding the gestational sac and the cervical endometrium	- In early pregnancy: (1) An empty uterus (2) The gestational sac cannot be separated from the uterus and at least 1 cm from the lateral edge of the uterine cavity (3) Thin myometrial layer ($\leq 5\text{mm}$) surrounding the supero-lateral portion of the gestational sac (4) The interstitial line sign is present (5) Narrow communication between the sac and the uterine cavity - in advanced pregnancy US is equivocal	(1) No variation in thickness of the myometrium around the 2 horns (2) Visual continuity between the endometrium lining the gestational sac and that of the other uterine cornua (3) Wide communication between gestational sac and the rest of the endometrial cavity
Helpful MRI findings	(1) The gestational sac is clearly located in a rudimentary horn with a band of tissue connecting the rudimentary horn to the uterus (2) the gestational sac is surrounded by a wall that should match the signal intensity of normal myometrium with all pulse sequences (3) no communication between the pregnant horn and endometrial cavity or endocervical canal (4) The contralateral unicornuate uterus will be laterally displaced and described as a “banana-shaped uterus” (5) Identification of myometrial discontinuity and hemoperitoneum in ruptured advanced cases	(1) A junctional zone is present between the gestational sac and the uterine cavity in early gestation (2) The gestational sac is located eccentrically in the uterine fundus and is contiguous with the uterine myometrium, (3) Is surrounded by asymmetric thin myometrium (4) Clear visualization of the decidua adjacent to the gestational sac in second trimester (5) Identification of myometrial discontinuity and hemoperitoneum in ruptured advanced cases	(1) Two separate uterine horns with continuity between the endometrium lining the gestational sac and the other uterine horn
Laparoscopy findings	(1) The gestational sac in a separate uterine horn with a fallopian tube (2) The horn is attached to the empty unicornuate uterus by a (narrow or wide) band (3) The round ligament of the rudimentary horn is attached lateral to the gestational sac	(1) The gestational sac (the uterine bulge) located lateral to the round ligament	(1) The gestational sac in one of the two uterine horns which is bigger and continuous with the other horn (2) The round ligament of the pregnancy horn is attached lateral to the gestation sac

It is also important to try to identify any renal tract malformations when RHP is diagnosed and before planning for surgery because of the high incidence of renal anomalies with MDA [15,21].

Treatment

Surgery is the main line of management. Advanced RHP may rupture with little warning at any time, with significant risk to maternal health, hence early diagnosis can help in planning elective surgery before rupture [4,10, 22].

One paper reported that when the ipsilateral tube was preserved in two cases after elective removal of the non-gravid rudimentary horn, one of the patients subsequently developed tubal pregnancy in the preserved tube [5]. In another report conservative treatment of RHP without removal of the pregnant rudimentary horn was followed by recurrence of RHP in the subsequent pregnancy [10]. In view of the above surgery for RHP should involve excision of the rudimentary horn and ipsilateral salpingectomy with conservation of the ipsilateral ovary [5].

The excision of a RHP is not technically difficult in early pregnancy and the risk of complications is low if through evaluation of the rudimentary horn and any associated urological anomalies was done before embarking on the surgical excision [15,17, 21]. Laparoscopic excision increasingly becomes a better choice during the first trimester [10,17]. If a thick fibromuscular tissue connecting the RHP and the unicornuate uterus is present, laparoscopic removal is technically more challenging, requires a longer operation time, and closing the myometrium with laparoscopic sutures is recommended to avoid potential future uterine rupture. However, if only a thin fibrous band was present between the RHP and the unicornuate uterus no need to close the myometrium [25].

In advanced cases it is best to perform open surgery, although recently few case reports of laparoscopic management of RHP in second trimester have been reported. These cases undergone radiological evaluation of the rudimentary horn to determine the feasibility of laparoscopic excision and received preoperative potassium chloride to induce feticide and reduce the size of the pregnancy which facilitated the removal of the excised pregnant horn with the help of morcellator and endoscopic bag through small abdominal incision [21,26].

Medical management of a viable RHP with methotrexate and intracardiac potassium chloride followed by gonadotropin-releasing hormone injections has also been used pre-operatively to reduce both size of pregnancy and the blood loss from associated placenta accreta before interval excision of the rudimentary horn 6 weeks later [10,21, 27].

Missed miscarriage in a rudimentary horn has been managed

expectantly with serial β -hCG measurement until resolution of the pregnancy. However, recurrence of RHP can occur again if the rudimentary horn is not removed electively after the conservative or medical treatment [10].

Prognosis and Future Pregnancy

After excision of RHP woman should be investigated to check of any associated renal tract anomalies if this was not done before surgery.

Women with a unicornuate uterus with or without rudimentary horn are at a higher risk of obstetric complications, such as first and second trimester miscarriage, preterm delivery, breech presentation, pregnancy induced hypertension (in women with associated unilateral renal agenesis), intrauterine growth restriction, intrauterine fetal demise and increased caesarean section rate [5, 28]. This risk will obviously persist after excision of the RHP and women should be informed about this risk.

Consideration for prophylactic resection of a non-communicating rudimentary horn in women with MDA has been advocated by some authors to prevent endometriosis and possible future RHP [29].

A case series of the reproductive and obstetric outcome after laparoscopic excision of non-pregnant functional, non-communicating broadly attached rudimentary horn showed no negative effect from the surgery on subsequent fertility and no reported case of uterine rupture during subsequent pregnancy in the remaining unicornuate uterus after the rudimentary horn excision [30]. However, all cases in this series delivery by caesarean sections five for obstetrics reasons and two elective sections.

Women should be informed that there is lack of evidence-based data from well-designed studies addressing the issue of route of delivery in uncomplicated subsequent pregnancy after excision of RHP.

Conclusion

RHP is a rare form of pregnancy in a rudimentary uterine horn, if not diagnosed 80–90 % of the cases may rupture during the second or third trimester causing significant maternal morbidity and mortality. Early diagnosis during the first or early second trimester allows the safe planning for surgical excision through the most appropriate route before rupture of the pregnant horn.

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