# Unicornuate uterus with a non-communicating functioning rudimentary horn, associated with ipsilateral renal agenesis

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The incidence of Müllerian defects varies currently from 7% to 8% of the normal fertile population and more than 25% among women with recurrent miscarriage. However, clear uterine malformations are observed in 5% of the general population, in 2% to 3% of fertile women, in 3% of infertile women and in 5% to 10% of patients with recurrent miscarriage.[1]

Unicornuate uterus with a rudimentary horn is a rare Müllerian anomaly, with an incidence of 0.06%.[2] A partial or complete lack of development of one Müllerian duct between the 7th and 8th week of gestation may result in the formation of a unicornuate uterus.

The functioning unicornuate uterus may or may not be associated with a rudimentary uterine horn. Seventy-five to ninety percent of unicornuate uteri with rudimentary horns are non-communicating.[3] Non-communicating cavitated rudimentary horns are the most clinically significant as they are more likely to be associated with pelvic pain from haematometra or from endometriosis due to retrograde menstruation. Also, pregnancies in these rudimentary horns may be diagnosed late, with a 70% risk of uterine rupture occurring before 20 weeks of gestation accompanied by potentially massive and life-threatening intraperitoneal haemorrhage.[4]

## Case

In December 1999 a 16-year-old single girl was admitted to the King Khalid University Hospital (KKUH) with severe dysmenorrhea. She had menarche at the age of 12 years with regular periods every 28 days. Menses lasted for 6 days and was associated with increasing severe dysmenorrhea not relived by nonsteroidal anti-inflammatory medications. The cyclical pain caused repeated absenteeism from the school and later discontinuation of her education.

In 1996 at the age of 13 years she had an apendicectomy in a private hospital for suspected appendicitis. The appendix was not inflamed. Two years later she had laparotomy in the same private hospital for persistent cyclical lower abdominal pain and a suspected right adnexal mass on ultrasound. Intravenous pyelography (IVP) revealed a single left ureter and kidney. The right kidney and ureter were not visualized. At laparotomy, there was a left unicornuate uterus with a normal cervix and a non-communicating right rudimentary horn, which was distended, cystic and attached to the unicornuate uterus by a thick pedicle-like tissue. There was a right haematosalpinx. The left ovary and tube looked normal. The non-communicating right horn was surgically connected to drain into the cavity of the left communicating left unicornuate uterus. She was discharged on danazol 200 mg twice a day for three months. However, the cyclic pain returned and became worse with time.

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Six months after her laparotomy she presented to King Khalid University Hospital with the same complaints and a tender cystic mass was felt in the right iliac fossa. Ultrasound scan showed a left unicornuate uterus and a right, non-communicating rudimentary horn distended with blood and a right haematosalpinx. The consent of the parents was obtained for a hysterectomy of the right non-communicating horn. A ureteric stent was inserted by the urologist for identification of the only ureter to avoid ureteric injury at hysterectomy. The round ligaments were identified and the uterovesical pouch was opened and the bladder was pushed down. The attaching pedicle of the non-communicating horn was doubly clamped, transfixed ligated and excised together with the right haematosalpinx. On the right ovary, there were endometriotic spots that were diathermised.

## Discussion

A precise classification of the unicornuate uterine did not occur until Buttram and Gibbons presented a classification scheme in 1979.[5] To improve on this classification, the American Fertility Society (AFS) produced a standard classification scheme in 1988, which included a scoring system that helps the surgeon not only to document the severity of the disease but also to formulate a prognosis.[6] Using the AFS scoring system, our patient would be IIb with an excellent (>75%) prognosis for conception and subsequent viable infant.

The basic objectives of surgical resection of a rudimentary non-communicating horn of unicornuate uterus are the pain relief and the maintenance of reproductive capacity. Therefore, consideration of prophylactic resection of a non-communicating uterine horn with a cavity should be considered in an asymptomatic, reproductive age patient once the diagnosis is made. Laparoscopic removal of a

rudimentary horn can be used to decrease the incidence of adhesions.

In patients in one study who presented with endometriosis and a unicornuate uterus with a cavity, rudimentary horn and tubal lumen, 50 had a communication between the endometrial cavity in the rudimentary horn and tubal lumen.[7] In our case endometriotic spots were seen on the right ovary and were diathermised. The first attempt in the private hospital to drain this non-communicating right horn to the communicating left horn was unconventional; we would have expected it to fail as this artificial opening would heal by fibrosis and close again.

We feel that an experienced gynecologist who is familiar with such abnormalities and the appropriate management should, ideally, manage such cases. He would also be in a position to understand the gynecological sequelae of such procedures and their possible impact on the woman's future obstetric performance.

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