We present a case of a patient with Uterus bicornis bicollis, imperforate hemivagina and ipsilateral renal agenesis. This group of congenital malformations is often asymptomatic until puberty, when it presents as cyclic dysmenorrhea, leucorrhoea or a pelvic mass. Magnetic resonance imaging is becoming the preferred modality for delineation of uterine malformations. When congenital abnormalities of the reproductive tract are encountered, a search should also be made for renal anomalies. Patients with Uterus bicornis bicollis and unilateral imperforate vagina are often seen with pain severe enough to mimic an acute abdomen. It is important to keep this unusual congenital malformation in mind in the differential diagnosis of vaginal discharge, pelvic mass and/or abdominopelvic pain in young women so as to avoid inappropriate surgical procedures.

INTRODUCTION
The true incidence of obstructive Müllerian anomalies is unknown but is believed to be between 0.1% and 3.8% overall (1). Double uterus is associated with a longitudinal vaginal septum and with a blind hemivagina in 10 – 30% (2, 3) of Müllerian anomalies. The incidence of imperforate hymen is estimated to be 0.1% (1) and transverse vaginal septum 1 in 70 000 females, making these some of the rarest anomalies of the female genital tract (4). Awareness of these rare malformations should lead to early diagnosis and early surgical interventions resulting in reduced long-term morbidity (5, 6).

The longitudinal vaginal septum is produced by incomplete disappearance of the partition between the fused Müllerian ducts (7). The presentation of a complete obstructing hemivagina caused by a longitudinal vaginal septum may be associated with uterine malformation and ipsilateral renal agenesis (8–10). Obstructive anomalies prevent normal menstruation, allow for collection of blood in the uterus and the vagina and may increase the incidence of retrograde
menstruation. A high index of suspicion is necessary to diagnose these disorders and an adequate workup is essential. This report will present a case of uterus bicornis bicollis, unilateral imperforate vagina and ipsilateral renal agenesis. The incidence, classification, workup and management will be reviewed.

CASE REPORT
A 17-year old nulliparous female presented with abnormal vaginal bleeding and painful periods. Menarche was at age 12 years with normal pubertal events. Her menses were normal until about a year before presentation when she developed heavy menses and approximately 10 days of postmenstrual spotting. She also developed worsening dysmenorrhoea and an abnormal vaginal discharge. She became sexually active at age 14 years but did not give a history of dyspareunia, postcoital bleeding or any sexually transmitted infections. Her past medical/surgical history was not significant.

Physical examination was unremarkable except for the pelvic examination which revealed normal external genitalia, normal introitus and vagina except that the right lateral wall was bulging into the vaginal cavity and the cervix was deviated to the left. Bimanual pelvic examination revealed a large right cystic adnexal mass.

Pelvic ultrasound scan revealed what was reported as a “bicornuate uterus and a large 8 x 8 cm diameter mass beneath the bladder and to the right of the cervix” (Fig. 1: a, b). The left ovary was normal but the right ovary was not identified. Based on the above findings a diagnosis of a bicornuate uterus with a large right endometrioma was made.

After appropriate counselling and consent, the patient had a diagnostic laparoscopy which revealed two uterine horns, normal Fallopian tubes and ovaries; no endometriotic deposits nor sign of a pelvic mass was noted (Fig. 2). Intravenous urogram and abdominal ultrasound scan revealed a solitary normal left kidney. A repeat transvaginal ultrasound scan confirmed uterus didelphys with an imperforate hemivagina. A final diagnosis of uterus bicornis bicollis, imperforate hemivagina and ipsilateral renal agenesis was made. The patient consented to excision of the vaginal septum.

SURGICAL PROCEDURE
At the time of admission for surgery, copious quantities of foul smelling, cream-coloured discharge per introitus was noted; a high vaginal swab was taken and she was started on co-amoxicillin-clavulanic acid twice daily.

Examination under anaesthesia revealed a complete longitudinal vaginal septum with a right haematocolpos. The septum was excised; a pyocolpos, an erythematous and inflamed vaginal mucosa and a second cervix were noted. The obstructed cervix appeared hyperaemic and flush with the vaginal fornices. After resection of the vaginal septum, an acroflavin pack was left in the vagina overnight. Swabs taken at admission and during the operation grew group B Streptococcus sensitive to co-amoxicillin-clavulanic acid.

Her postoperative course was uneventful and when reviewed three months later, speculum examination revealed healed vaginal mucosa and two normal-looking cervices and there were no further complaints of dysmenorrhoea.

DISCUSSION
Uterus bicornis bicollis with unilateral imperforate vagina and ipsilateral renal agenesis is an uncommon congenital anomaly in which the uterus and vagina are duplicated. The earliest case of “Double” uterus with unilateral haematocolpos was reported in 1925 (11).

Embryologically, the female genital tract develops almost entirely from the Müllerian ducts that appear in the first 6 weeks of intrauterine life. The Müllerian ducts begin as paired symmetrical structures that then fuse in the midline to form the uterus, cervix and upper part of the vagina. If each Müllerian duct develops separately without fusion at any point, the result will be two distinct hemiuteri, each with its own Fallopian tube, ovary, cervix and vagina. The exact cause of the vaginal septum is unknown, but may be caused by an embryonic arrest at 8 weeks gestation that simultaneously affects the Müllerian and metanephric ducts.

There is a tissue-specific factor called the hepatocyte nuclear factor 1 Beta (HNF1B) that plays an essential role in early development. Heterozygous mutations (SI51P, Q243,
and R137-K161) with the HNFIB gene are associated with juvenile diabetes, a variety of renal and genital tract malfunctions (13, 14). This type of gene mutation may be responsible for the anomalies seen in our patient and if that is so she may be at risk for early onset diabetes.

Renal agenesis commonly accompanies Müllerian duct malformations (Fig. 3), because they both generate from the same ureteric bud (7). Vercellini et al (12) reported on the presence of a uterus didelphys, obstructed hemivagina and renal agenesis on the right side as in this case.

In 1989, the American Society for Reproductive Medicine developed a classification system that allows organization according to major uterine anatomical types. Table 1 classifies Müllerian anomalies, whereas Table 2 lists the classification of vaginal anomalies. The case presented herein has a Class III uterine anomaly and a Class IIa vaginal anomaly.

The presentation of obstructive longitudinal vaginal septum is variable and ranges from no symptoms to cyclic pelvic pain and abnormal bleeding. They may also present on rare occasions with fever, peritonitis, purulent vaginal discharge and leukocytosis, leading to a presumptive diagnosis of pelvic inflammatory disease.

It is common for these patients to develop haematometra, haematosalphinx and endometriosis. In uterus didelphys with unilateral imperforate vagina, the most common symptoms are cyclic dysmenorrhoea and abdominopelvic pain. If the vagina or uteri communicate at any point, abundant and often foul-smelling leucorrhoea results (15).

### Table 1. ASRM classification of Müllerian anomalies

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<th>Classification</th>
<th>Anomaly</th>
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| Class I (agenesis/hypoplasia) | a. Vaginal  
                          | b. Cervical  
                          | c. Fundal  
                          | d. Tubal  
                          | e. Combined anomalies |
| Class II (unicornuate) | a. Communicating  
                          | b. Noncommunicating  
                          | c. No cavity  
                          | d. No horn |
| Class III (didelphys) | Didelphys |
| Class IV (bicornuate) | a. Complete  
                          | b. Partial |
| Class V (septate) | a. Complete  
                          | b. Partial |
| Class VI (Arcuate) | Arcuate |
| Class VII (DES related) | DES related |

ASRM, American Society of Reproductive Medicine; DES, diethylstilbestrol.

### Table 2. Vaginal classification

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<th>Classification</th>
<th>Anomaly</th>
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| Class I        | Transverse  
                          | a. Obstructing  
                          | b. Nonobstructing |
| Class II       | Longitudinal  
                          | a. Obstructing  
                          | b. Nonobstructing |
| Class III      | Stenosis/iatrogenic |

Magnetic resonance imaging (MRI) may be an important adjunct for diagnosis because of its ability to clarify the anatomy of the uterus and vagina (16, 17). Magnetic resonance imaging has been found to be 100% sensitive and 83–100% specific in the diagnosis of this anomaly (15, 16) and is considered to be more sensitive for imaging soft tissue pelvic anatomy than CT. However, both of these modalities are expensive. Ultrasound can confirm the absence of one kidney in these patients however, which in the presence of an obstructed genital tract will lead to the correct diagnosis 100% of the time (16).

Resection of the vaginal septum was effected easily as this patient was sexually active. In a series described by Altintas (8) transvaginal resection of the vaginal septum was done in six of seven cases; for the other patient, the blind vagina with haematocolpos and attending uterus were extirpated by an abdominal approach. Kim et al (9) described the use of an operative hysteroscope to excise the septum through the small aperture of the hymen in a patient who was virginal so as to preserve the anatomy of the hymen.
Obstetric concerns included an increased risk of miscarriage, prematurity, intrauterine growth restriction (IUGR), abnormal fetal position and increased Caesarean section rate. Due to the absence of intrauterine septae, pregnancies in didelphic uteri are not as prone to miscarriage or prematurity as other uterine anomalies. Ludmir et al (18) in 1990 reviewed the obstetric outcomes in 10 patients with didelphic uteri; 80% of the fetuses survived. This study also incorporated reproductive outcomes in unicornuate, bicornuate and septate uteri. A term pregnancy was achieved in 60% of the didelphic group, the highest rate of all the different types of uterine anomalies studied.

The prompt and accurate diagnosis of disorders of development of the female reproductive tract is essential to prevent complications from acute illness and preserve future fertility. Many cases are improperly diagnosed; we therefore emphasize the need for paediatricians, radiologists, gynaecologists and paediatric surgeons to become familiar with these disorders to avoid complications and diagnostic delay.

**REFERENCES**