UNILATERAL RENAL AGENESIS

Ura

The complete absence of development of one kidney accompanied by an absent urethra.

⚠️ Risk factor ⚫ Male & Female

About Unilateral renal agenesis

Unilateral renal agenesis (URA) is the absence of one kidney (Pic. 1). Unilateral renal agenesis is a frequent renal malformation with incidence of 1 per 1000 live born children. Usually asymptomatic, it can be early detected by prenatal ultrasound, allowing opportune detection and adequate follow up.

The exact cause of renal agenesis in newborns is not known. Etiology of unilateral renal agenesis (URA) is heterogeneous (diverse) with environmental and genetic influences. Most cases of renal agenesis are not inherited from the parents, nor do they result from any behavior by the mother.

The diagnosis is usually incidental, during imaging evaluation of patients. The diagnosis of unilateral renal agenesis must alert the physician to look for associated genitai abnormalities, ideally during neonatal period or at peri-pubertal age, avoiding complications or even unnecessary emergent surgery in patients with abdominal or pelvic pain that might be confused with other causes of acute pelvic pain. Associated gonadal and genital abnormalities occur in females as well as in males. In females, such abnormalities include ovarian agenesis, ovarian cysts, duplicate, unicornuate, bicornuate or rudimentary uterus, absent or double vagina, absent fallopian tube, abnormal external genitalia and others.

Currently, there is no established protocol to study and follow up single-kidney children and there is lack of uniformity in post-natal management of these patients.

Symptoms

URA is usually asymptomatic, but through prenatal ultrasound it is being increasingly recognized.

Associated diseases

Unilateral renal agenesis is frequently associated with other abnormalities, including isolated or syndromatic anomalies of the cardiovascular, skeletal, central nervous and urogenital systems. These include Müllerian duct anomalies (MDA) resulting in uterine and/or vaginal defects at the same side of the absent kidney.

MDA duct anomalies are rarely diagnosed prenatally. The neonatal period is an excellent opportunity to detect gynecologic malformations due to the physiologic enlargement of the uterus influenced by maternal and placental hormones. Thereafter, uterine malformations can go undetectable until puberty. Clinical manifestations often start during adolescence, with the onset of menarche, presenting with cyclic pelvic pain caused by an obstructed structure, with or without menstrual abnormalities. Additional complications include recurrent infections, pelvic adhesions and endometriosis (inner lining of the uterus is found in other parts of the body).

Complications

The diminished functioning renal mass due to URA may lead to compensatory hypertrophy (enlargement) and hyperfiltration nephropathy (situation where the filtration elements in the kidneys called glomeruli produce excessive amounts of urine). As hyperfiltration occurs as a physiologic response in normal pregnancies, it is important to consider whether a woman with unilateral renal agenesis may be at increased risk for adverse outcomes during pregnancy.

Risk factors

Literature mentions URA as most frequent on the left side and predominating in males with a ratio of 1:2:1. Prenatal factors associated to renal agenesis are diabetes mellitus, alcohol exposure, black race, and young maternal age (<18 years).

Prevention

Since the exact cause of unilateral renal agenesis is not known, so genetic factors cannot be avoided but maternal factors leading to unilateral renal agenesis like maternal smoking and binge drinking can be avoided or modified accordingly to lower the risk for
How it can affect fertility

Female fertility

Up to 40% of women with a urogenital tract anomaly also have an associated renal tract anomaly. This occurs due to similar embryologic origins of both the renal and the genital systems. Unilateral renal agenesis may be associated with an increased incidence of Müllerian duct (Pic. 2) abnormalities, which are abnormalities of the development of the female reproductive tract and can be a cause of infertility, blocked menstrual flow (hematocolpos), increased need for Caesarean sections, or other problems. Herlyn-Werner-Wunderlich syndrome is one such syndrome in which unilateral renal agenesis is combined with a blind hemivagina (defective lateral fusion of the caudal portions of the müllerian ducts during weeks 8 to 12 of pregnancy) and uterus didelphys (uterus present as a paired organ).

The incidence of uterine malformation leads to impaired fertility and a number of obstetric complications such as threatening preterm delivery pelvic and transverse presentation, premature departure of amniotic fluid, intrauterine growth restriction, threatening rupture of the uterus, caesarean section. Nevertheless, some defects may be asymptomatic and diagnosed in later in life, during investigation of infertility or recurrent miscarriage.

Male fertility

Unilateral renal agenesis may be associated with a rare condition called congenital absence of the vas deference (CAVD). Congenital absence of the vas deferens means that the vasa deferentia which connect the sperm-producing testicles to penis (Pic. 3) are not formed at birth. Therefore, those who are missing both vas deferens are typically able to create sperm but are unable to transport them appropriately. Their semen does not contain sperm, a condition known as azoospermia. It may either be unilateral (CUAVD; missing one vas deferens) or bilateral (CBAVD; missing both vas deferens).

Prognosis

Usually, prognosis is good for patients with unilateral renal agenesis. Some cases are caused by genetic mutations that are passed on from parents who either have the disorder or are carriers of the mutated gene. Prenatal testing can often help determine if these mutations are present.

Genital anomalies occur in 37–60% of females and 12% of males with congenital unilateral renal agenesis. Many investigators believe that fertility problems in affected individuals usually involve maintenance of pregnancy and not a decreased ability to conceive. These patients experience a high frequency of spontaneous abortion (SAB) in the late 1st or 2nd trimester, premature birth, and abnormal fetal presentations.

About 30–40% of cases with congenital absence of the vas deference is associated with unilateral renal agenesis. Whereas the condition is often associated with bilateral vas absence and azoospermia and most men with CUAVD are fertile, men with a single vas may present with mild oligoospermia (low sperm concentration) as testicular sperm output is cut in half.

Gallery

Pic In unilateral renal agenesis, complete absence of development of one kidney accompanied by an absent ureter.

Pic In the absence of SRY, ovaries differentiate, Wolfian ducts degenerate, and Müllerian ducts develop into a simple columnar epithelial tube that will differentiate into the oviducts, uterus, cervix, and upper portion of the vagina.
Vas deferens transport sperm from the epididymis to the ejaculatory ducts in anticipation of ejaculation.

Sources

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* Unilateral renal agenesis associated with ovarian cysts in a 19 year old woman in Orulu, South-East Nigeria [https://www.researchgate.net/publication/283729147_Undilateral_renal_agenesis_associated_with_ovarian_cysts_in_a_19_year_old_woman_in_Orulu_South-East_Nigeria]—by Anyabolu et al. licensed under CC BY 4.0

* Congenital absence of the vas deferens [https://fertilitypedia.edu/diagnoses/congenital-absence-of-the-vas-deferens]—sourced from Fertilitypedia licensed under CC BY-SA 4.0

* Figure 41.03.01 [https://commons.wikimedia.org/wiki/File:Figure_41.03.01.jpg]—by CNX OpenStax licensed under CC BY 4.0

* Route of vas deferens from testis to the penis [https://commons.wikimedia.org/wiki/File:Route_of_vas_deferens_from_testis_to_the_penis.png]—by Andrew Z. Colvin licensed under CC BY-SA 3.0

* The uterus differentiates from the fetal Müllerian ducts [https://commons.wikimedia.org/wiki/File:The_uterus_differentiates_from_the_fetal_M%C3%BCllerian_ducts.jpg]—by Teixeira et al. licensed under CC BY 3.0