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Congenital urinary tract obstruction

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Congenital urinary tract obstruction is a heterogenous condition with a varying natural history. Accurate diagnosis within the late-first and second trimesters allows for counselling of the parents and planning of multi-disciplinary care for the pregnancy and newborn. Antenatal investigations to predict postnatal renal function are of varying accuracy. However, some factors have been shown to be predictive of poor outcome in terms of renal function at birth and infancy. There is the possibility of in-utero intervention in these fetuses.

Key words: congenital urinary tract obstruction; hydronephrosis; posterior urethral valves; vesico-amniotic shunting.

INTRODUCTION

Congenital abnormalities of the genitourinary tract are one of the most common sonographically identified anomalies, with a rate of 1 in 250 to 1 in 1000.[1] Obstructive uropathies account for the majority of cases: approximately 1 in 2000 pregnancies.[2] Obstruction can be at the level of the ureteropelvic or uretovesical junction, or it might be urethral. It can be either unilateral or bilateral; although, by definition, obstruction at the level of the urethra must be bilateral and it might involve some or all of the urinary tract.

The majority of genitourinary abnormalities are diagnosed during the detailed second-trimester scan (usually performed at 18–20 weeks). However, with the increasing use of first-trimester screening, more severe renal anomalies are being noted between 11 and 14 weeks using ultrasound. The findings on antenatal ultrasound will depend on

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the level and severity of the obstruction and, in the presence of a minor obstruction with normal liquor volume and in which dilatation of the renal tract might not occur until the third trimester, the diagnosis can be missed. Antenatal diagnosis allows for the planning of appropriate prenatal and postnatal care, consideration of prenatal intervention and psychological adjustment of the parents; however, minor anomalies can cause undue parental anxiety. For some anomalies there is also uncertainty regarding best management.

The prognosis in upper urinary tract obstruction in the fetus is generally relatively good unless other co-existent congenital anomalies are present. However, the situation is very different for lower urinary tract obstruction (LUTO) because of its association with cystic renal dysplasia, oligohydramnios, pulmonary hypoplasia, mild dysmorphia and limb contractures. For this reason, prenatal in-utero therapy has been considered in selected cases of LUTO in an attempt to bypass the congenital urinary tract obstruction and attenuate the secondary developmental complications. Prenatal counselling in this situation is difficult because the prognostic value of the modalities used to assess fetal renal function are uncertain and the effectiveness of therapy remains to be established.

In this chapter we discuss the epidemiology and aetiology of congenital urinary tract obstruction and the pathophysiology behind these conditions. Current options for investigation and management of the fetus are presented, with discussion of future research developments.

EPIDEMIOLOGY

Upper urinary tract obstruction can be detected consistently at the second-trimester ultrasound scan. The largest European database is from the EUROSCAN Group, which involves 20 registries from 12 European countries. Data have been acquired from 709,030 cases, with a mean prevalence of all types of renal malformation of 1.6 per 1000 births. The most frequent diagnosis reported was upper urinary tract dilatation in 309 (27%) of patients, with 259 (84%) being detected prenatally.[3] Unilateral hydronephrosis is more common than bilateral[4] and most cases – 88% of bilateral and 69% of unilateral – can be described as mild.[5] Pelviureteric obstruction is the most common cause of hydronephrosis (reported incidences of 39–64%) with reflux accounting for a third (33%) and vesico-ureteric obstruction 9–14%.[6–9]

Only one study giving population-based information on LUTO and data from a regional congenital anomaly register (Northern Region) has been published in the medical literature. This study identified 113 registered cases in the 14-year period between 1984 and 1997 and noted that the registry had a high notification rate with an ascertainment level of 95%. The incidence of LUTO was calculated as 2.2 per 10,000 births (based on total birth denominator data) with posterior urethral valves (PUV; determined by postnatal investigations and autopsies) being the most common at 64% (1.4/10,000 births), followed by urethral atresia at 39% (0.7/10,000 births) and prune belly syndrome 4%.[10]

AETIOLOGY

Upper urinary tract obstruction

Ureteropelvic junction obstruction

Ureteropelvic junction (UPJ) obstruction is the most common lesion of the fetal urinary tract. Its aetiology is unclear. Causes include abnormal recanalisation of the