Congenital pelviureteric junction obstruction

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Congenital PUJO is the most common cause of unilateral hydronephrosis with an incidence of 1 in 500 live births. It may result from intrinsic obstruction secondary to an aperistaltic segment at the PUJ due to muscular hypoplasia. Other causes include a high insertion of the ureter into the pelvis and the presence of crossing aberrant vessels at the PUJ. It is more

Ureter draining lower moiety inserts laterally and superiorly into the bladder Ureter draining the upper moiety inserts medially and inferiorly into the bladder – ureterocele and upstream hydroureteronephrosis Sphincter Figure 82.4 Retrocaval ureter with a classic ‘reverse J’ sign seen on intravenous urogram (courtesy of Department of Urology, Christian Medical College, Vellore, India).

common in males and on the left side. Bilateral obstruction occurs in 10% of cases. Historically , PUJO presented as a palpable flank mass in an infant or a child, but most are now detected before birth with antenatal US. Older children may present with intermittent flank pain, UTI or a flank mass. Adults present with back or flank pain or recurrent pyelonephritis. Rarely , a patient may present with a history of severe flank pain following ingestion of large amounts of fluid, which is relieved after passing a large amount of urine (Dietl’s crisis). US may show symmetrical hydronephrosis and a dilated renal pelvis and can provide information on the severity of obstruction by measuring the degree of dilatation, parenchy mal thickness and cortical echogenicity . Isotope diuretic renog raphy is the current investigation of choice. Isotope uptake and washout of the isotope can be followed with time to produce a renogram curve. Usually , half of the peak isotope activity is cleared within 10–15 minutes (T). A rising curve following 1/2max administration of furosemide, a T of greater than 20 min 1/2max utes and a di ff erential function of less than 40% on the a ff ected side is suggestive of significant obstruction and is an indication for surgical intervention (Figure 82.5). CTU or MRU may also be used in the evaluation of PUJO. T he Anderson–Hynes dismembered pyeloplasty is the pro cedure of choice with a wide funnelled, dependent anastomo sis, maintaining good vascularity of the upper ureter and pelvis and excision of the redundant pelvis (Figure 82.6). The indica tions for pyeloplasty are persistent pain, hypertension, haema turia, secondary renal calculi and recurrent UTIs. Endoscopic management in the form of endopyelotomy is reserved for post-py eloplasty strictures. Dietl’s crisis, first reported by Josef Dietl , 1804–1878, in 1864, an Austrian doctor and pathologist known for his work on floating kidneys. James Christie Anderson , 1899–1984, urologist, Royal Hallamshire Hospital, She ffi eld, UK. Wilfred Hynes , 1903–1991, plastic surgeon, The Plastic and Jaw Department, The Royal Hospital, She ffi eld, UK. Anderson and Hynes devised the operation in 1949. Carl Weigert , 1845–1904, German pathologist and anatomist known for work on cellular staining. Robert Meyer ,

1864–1947, German pathologist and gynaecologist in Berlin, removed from his position for being Jewish, emigrated in 1939 to Minneapolis, MN, /uni00A0 USA. Morton A Bosniak , 1929–2016, Professor of Radiology , New Y ork University (NYU) Langone School of Medicine, New Y ork, NY , USA. Congenital anomalies /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF

Figure 82.5 Isotope renal scan using diethylenetriaminepenta-acetate showing an obstructive pattern on the time–activity graph with hold-up of contrast of up to 2 hours. This /f_i nding is consistent with pelviureteric junction obstruction (courtesy of Department of Urology, Christian Medical College, Vellore, India). Congenital anomalies are usually detected incidentally and often only manifest when effected by pathology such as stone disease or malignancy Ectopic ureter should be suspected in a female child who presents with continuous incontinence of urine and also voids normally Weigert–Meyer rule The ureter that drains the upper moiety is at a more inferior and medial position and is prone to obstruction and dysplasia The ureter that drains the lower moiety is at a more superior and lateral position and is prone to VUR Most cases of ANH are transient and resolve after birth

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