Ureteropelvic Junction Obstruction

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Pediatric ureteropelvic junction (UPJ) obstruction can occur in all age groups; increased detection in neonates due to prenatal hydronephrosis can be seen on ultrasonography. It is more common in boys than girls and more common on the left side.

1. Pathophysiology:
   (a) Causes of UPJ obstruction:
      (i) Intrinsic: interruption in development of circular musculature of the UPJ causing fibers to be widely separated and leading to functional discontinuity of muscular contractions, poor emptying; other causes include valvular mucosal folds or upper ureteral polyps.
      (ii) Extrinsic: aberrant, accessory, or early-branching lower pole vessel passing anteriorly to the UPJ.
      (iii) Secondary UPJ obstruction: severe VUR with tortuous course causing proximal kinking of ureter due to fixation causing obstruction.
   (b) Associated anomalies:
      (i) Renal dysplasia.
      (ii) Multicystic kidney.
      (iii) Duplicated collecting system: more typically lower segment.
      (iv) Vesicoureteral reflux.
      (v) Horseshoe kidney or ectopic kidney.
      (vi) VACTERL constellation of anomalies.

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2. Clinical presentation:
   (a) Infants: Asymptomatic; previously found due to palpable masses but now found on prenatal ultrasonography; occasionally due to failure to thrive, feeding difficulties, urosepsis, presence of nephrolithiasis.
   (b) Older children: Episodic flank pain, cyclic vomiting; occasionally present with hypertension due to reduced blood flow leading to renin-mediated hypertension.
3. Diagnosis:
   (a) VCUG: rule out VUR.
   (b) Renal scan: degree of obstruction and renal function.
   (c) Consider operative repair if decline in ipsilateral decline in renal function > 10 % or symptomatic.
   (d) If <10 % relative renal function of affected side, may consider nephrectomy.
   (e) If no intervention indicated, surveillance with renal ultrasound or renal scan every 3–6 months.
4. Treatment: surgical repair by open, endoscopic, laparoscopic, and robotic-assisted approaches with the three main groups being flap type, incisional-intubated type, and dismembered type.
   (a) Open approaches (open pyeloplasty):
      (i) Typically dismembered pyeloplasty: preserves lower pole or crossing vessels, excision of pathologic UPJ with repositioning, and successful reduction.
      (ii) Approach can be subcostal, flank or posterior lumbotomy.
      (iii) Problematic if inadequate ureteral length: can use spiral flap if sufficient extra-renal pelvis to accommodate this gap.
      (iv) Most common is Anderson-Hynes pyeloplasty: identify the UPJ, traction sutures placed, transect the UPJ, ureter spatulated on lateral side, pelvis opened medially or excision of redundant pelvis, align the pelvis and spatulated ureter to allow for dependent drainage, close with anastomotic suture; can be done with or without a stent.
      (v) Other options:
         1. Foley Y-V plasty: good for high insertion of the ureter.
         2. Spiral flap: long gap with adequate extrarenal pelvis.
         3. Vertical flap: dependent junction with large box-shaped extrarenal pelvis.
      (vi) Outcomes: dependent on minimal handling of ureter at time of repair, use of internal stenting or nephrostomy tube drainage.
         1. Early complications: uncommon, prolonged urinary leak from drain which can usually be managed conservatively or may require ureteral stenting.

(b) Endoscopic approaches (endopyelotomy):

(i) Antegrade or retrograde.
(ii) Balloon dilation, laser incision, or Acucise ureteral cutting balloon device (Applied Medical, Rancho Santa Margarita, CA).
(iii) Requires stenting for at least six weeks typically.
(iv) Higher rates of recurrence.

(c) Laparoscopic approaches (including robotic-assisted pyeloplasty):

(i) Typically the dismembered approach as discussed previously: allows for excision of stenotic segment and preservation of vessels.
(ii) Non-dismembered pyeloplasty technically easier as allows for easier tension free suturing.
(iii) Robotics allows for improved anastomotic suturing.
(iv) Transabdominal approach as compared to retroperitoneal open approaches.
(v) Minimally invasive, smaller scars.
(vi) Patient needs to be large enough to allow for adequate pneumoperitoneum and work space to complete repair: typically older patients.
(vii) Intraoperative complications: bleeding, trocar damage to other structures including bowel or vessels, conversion to open.
(viii) Postoperative complications: wound infection, persistent urine leak, UTI.