UPJ syndrome in adults

Definition
- Functional failure of the transport of urine from the renal pelvis to the ureter resulting in pyelocalyceal dilation of the affected kidney.

Pathogeny
- **Intrinsic congenital causes** (the most common):
  - *Aperistaltic segment of the ureter*: absence of spiral muscle bundles in the stenotic ureteral segment resulting in the absence of a peristaltic wave.
  - *True ureteral stenosis* (less common): a congenital stenosis due to excessive collagen deposits in the stenotic area. May also be caused by torsion or folding of the ureteral mucosa and musculature.
  - *High insertion of the ureter*: produces a true urine drainage failure.
- **Extrinsic congenital causes**:
  - *Extrinsic compression by crossing vessels*: this is controversial, as the true etiology is probably an intrinsic ureteral lesion.
- **Acquired causes**:
  - *Ureteral fibroepithelial polyps* or ureteral/extraureteral malignancies.
  - *Lithiasis*.
  - Postoperative or postinflammatory *fibrosis*.

Clinical presentation
- Intermittent *flank pain* related to fluid intake.
- *Nausea* and *vomiting*.
- Occasional *self-limiting hematuria* or recurrent urinary tract infections.
- *Hypertension* (uncommon as initial symptom).

Diagnosis
- **The goal of imaging studies** is to determine the location and functional importance of the obstruction. To arrive at a diagnosis, a combination of methods is used.
- **Ultrasound**: the first test performed if there is clinical suspicion. Can differentiate acquired causes such as lithiasis and determine the level of obstruction at the UPJ. The determination of resistance through *Doppler* ultrasound can help distinguish an obstructive from a non-obstructive dilation. Also differentiates UPJ syndrome from polycystic kidney disease.
- **IVP**: first-line option in cases of clinical suspicion supported by ultrasound results. Shows elimination delays in the pyelocalyceal system, which appears dilated, with a ureter of normal diameter, if visible. Diuretic stimulus may be used in this technique.
- **Isotopic renogram (DTPA)**: allows quantification (%) of renal function and differentiates obstructive from non-obstructive dilation after diuretic stimulus. Predicts the capacity for recovery of renal function in patients in whom IVP shows a loss of function.
- **Helical CT or uro-MRI with vascular sequence**: second-line exploration. Allows differentiation of obstructive causes (extrinsic or intrinsic). Vascular studies detect the presence of crossing vessels.
- **Whitaker test**: urodynamics study of the UUT that requires the placement of a nephrostomy tube. A pressure difference >15-22 cm of H2O between the renal pelvis and the bladder suggests UPJ syndrome. Disadvantages: an invasive method yielding imprecise results.
Treatment indications

- **Surgical treatment:**
  - Presence of symptoms associated with obstruction.
  - Impaired overall renal function or impaired homolateral function.
  - Presence of associated lithiasis or recurrent urinary tract infections.
  - Hypertension of renal origin.

- **Nephrectomy:**
  - Symptomatic patient in whom scintigraphy indicates affected kidney function <15% with a normal contralateral kidney.
  - Loss of function with extensive lithiasis and signs of chronic infection in the affected kidney with a normal contralateral kidney.

Surgical treatment

- **Open surgery techniques:** the traditional procedure, currently relegated to second-line status by laparoscopic and endourological techniques. Ureteral catheterization prior to surgical treatment is only indicated in cases of infection secondary to obstruction, renal failure, or severe and incessant pain. Technical variants include:
  - **Dismembered or Anderson-Hynes pyeloplasty:** versatile technique applicable to any anatomic abnormality except for intrasinus pelvis and in cases of long or multiple proximal ureteral stenoses. The success rate is 95%.
  - **Foley Y-V plasty:** indicated in cases of high ureteral insertion. Contraindicated when the pelvis must be reduced or when a transposition of anterior crossing vessels must be performed.
  - **Culp-De-Ward spiral flap:** indicated in cases of large, easily accessible, extrasinus pelvis in which the ureter is in a sloped oblique position. An ideal technique for long proximal ureteral stenosis.
  - **Scardino-Prince vertical flap:** similar indications to those above.
  - **Davis intubated ureterotomy:** indicated in long stenotic segments of the proximal ureter and other associated ureteral lesions. Combined with a vertical flap. Requires placement of a nephrostomy tube.
  - **Ureterocalicostomy:** indicated as a salvage technique if the aforementioned techniques fail or as a primary procedure in very small pelvises or kidneys with anatomical abnormalities.

- **Endourological techniques:** the goal is a full-thickness incision in the obstructive ureteral segment, from the lumen to the periureteral fat, with placement of a double J catheter. Require shorter hospital stays and recovery times than open surgery techniques.
  - **Percutaneous antegrade endopyelotomy:** indicated in patients with associated pyelic lithiasis. Should not be performed in long stenotic segments (>2 cm) or in cases of active infection or hemorrhagic diathesis. The incision should be made laterally to avoid crossing vessels. The success rate is 56-100%.
  - **Endopyelotomy with electrocautery balloon catheter:** contraindicated in patients with lithiasis, large hydronephrosis, and stenotic segments >2 cm. The success rate is 67-75%.
  - **Ureteroscopic endopyelotomy:** allows direct visualization of the pelvis. Contraindicated in large dilations and stenotic segments >2 cm. A posterolateral incision should be performed. May be associated with balloon dilation. The success rate is 70-97%.

- **Laparoscopic techniques:** they lead to lower morbidity, shorter hospital stays, and faster recovery times. Their indications and success rates are similar to those of open surgery. The main contraindications are clotting disorders, urinary tract infections, and cardiovascular disorders. The transperitoneal approach is the most commonly used approach.