Renal and ureteral involvement in Erdheim-Chester disease

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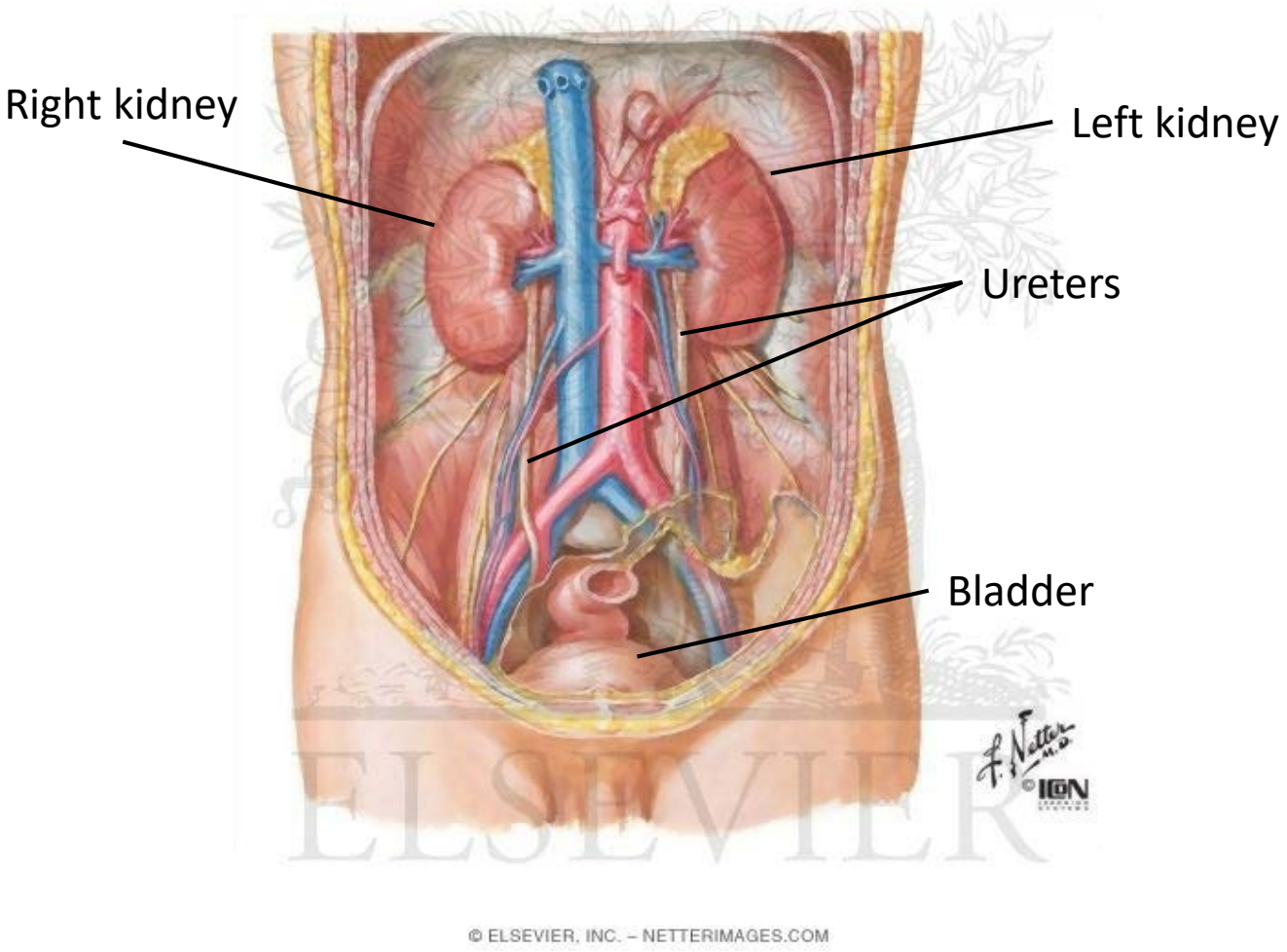
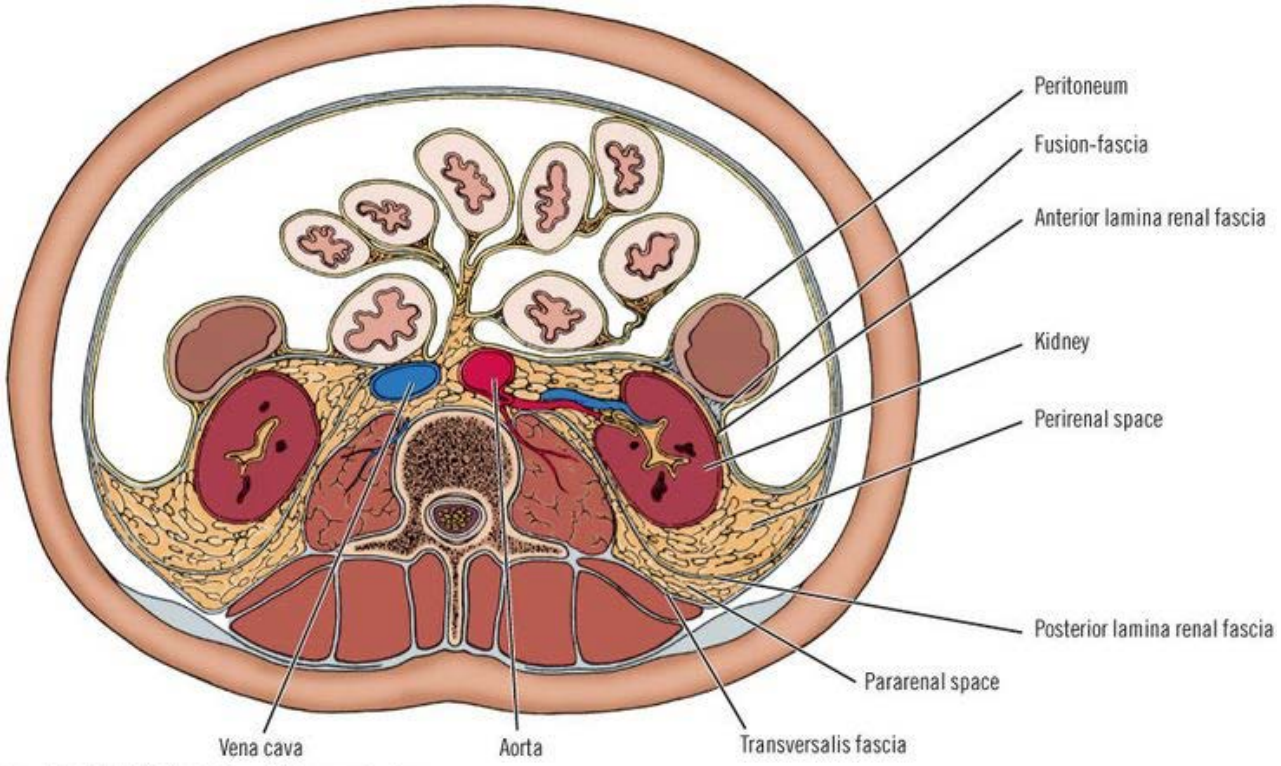
The kidneys and the urinary excretory system



The kidneys and the retroperitoneum



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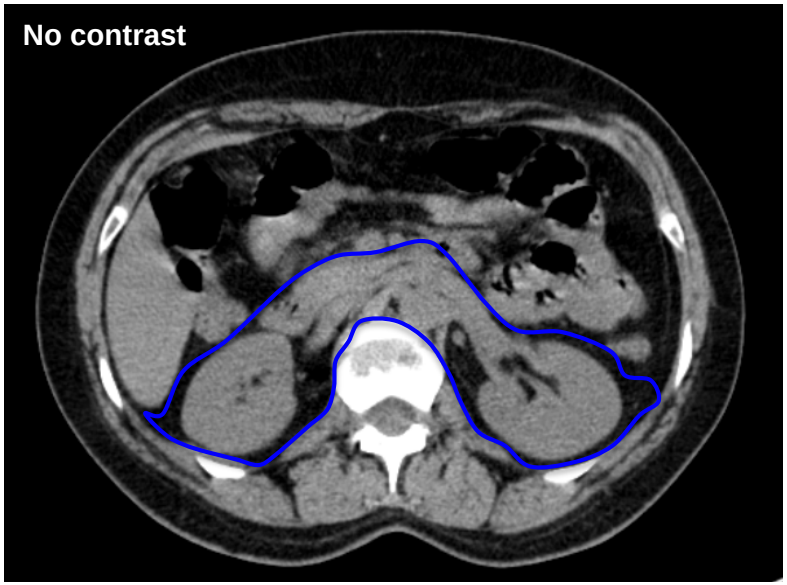
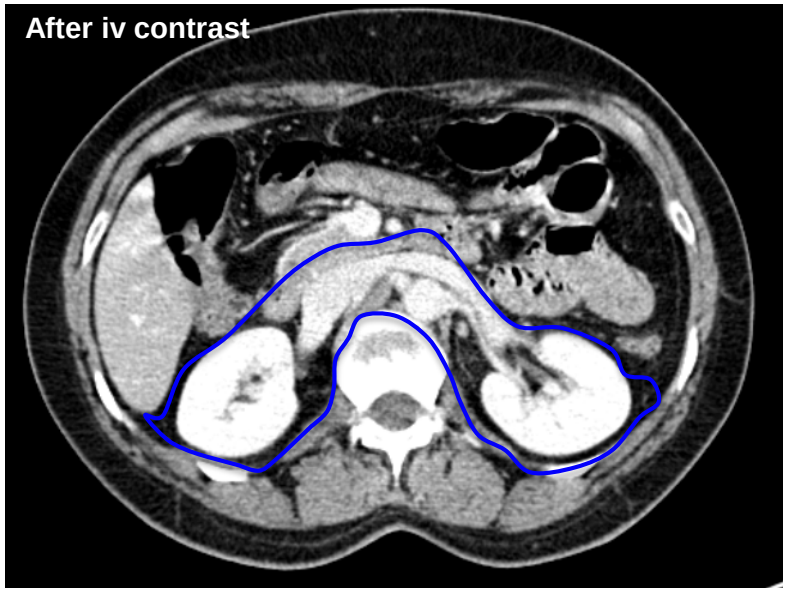
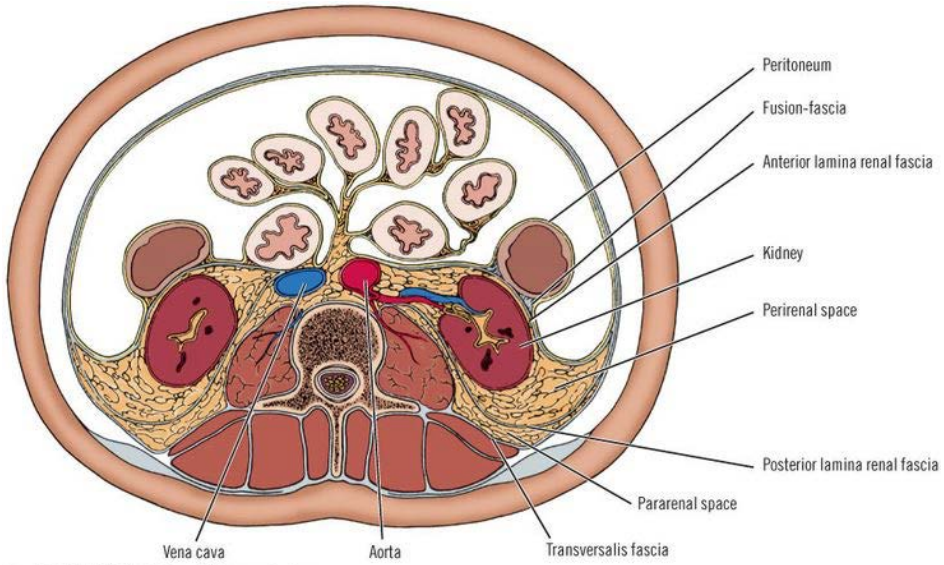
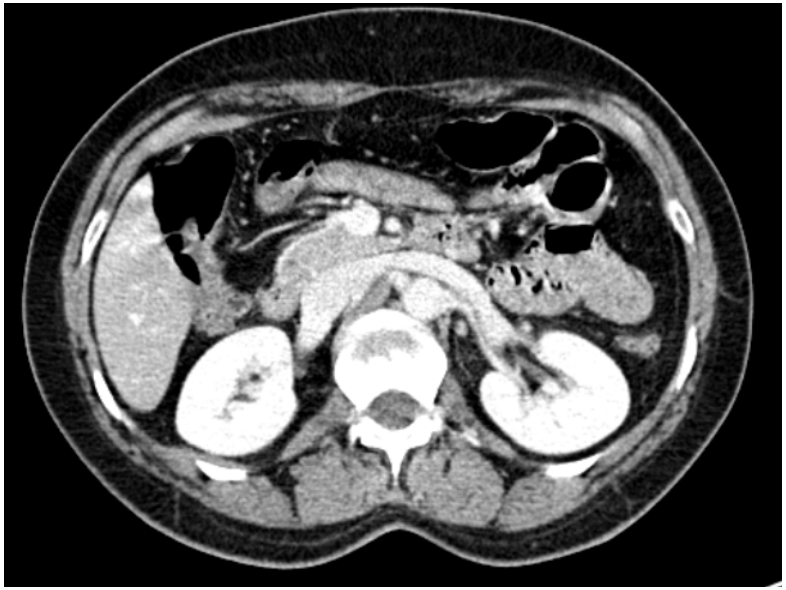


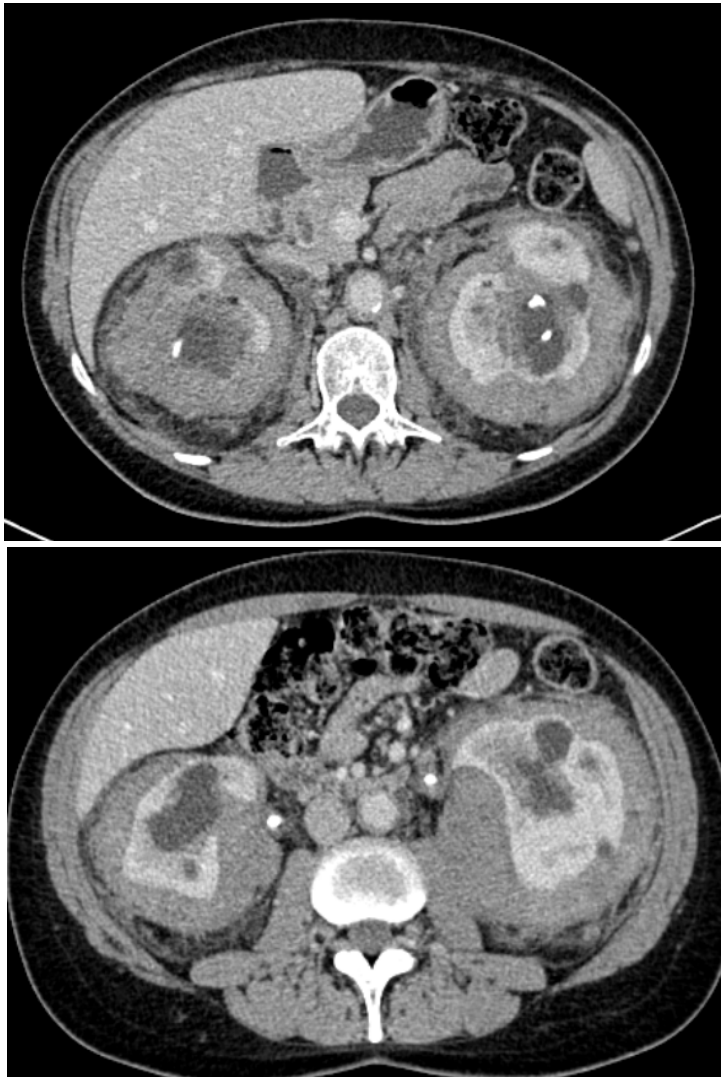




Perirenal infiltration in ECD (*hairy* kidneys)



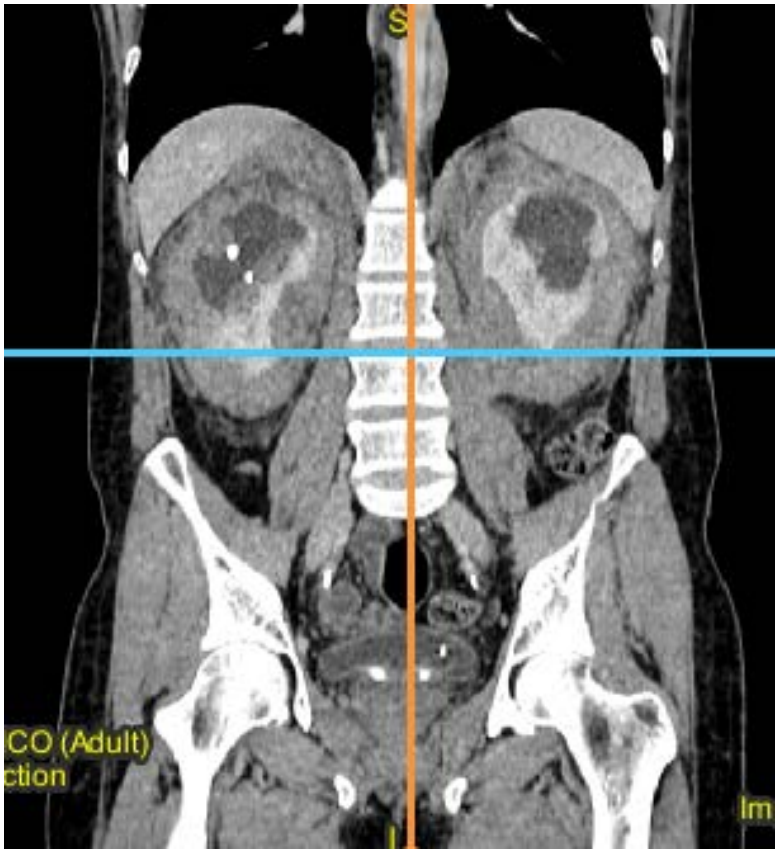
Abdominal CT scan in a healthy 38 yo lady



Abdominal CT scan in a 50-yo ECD man

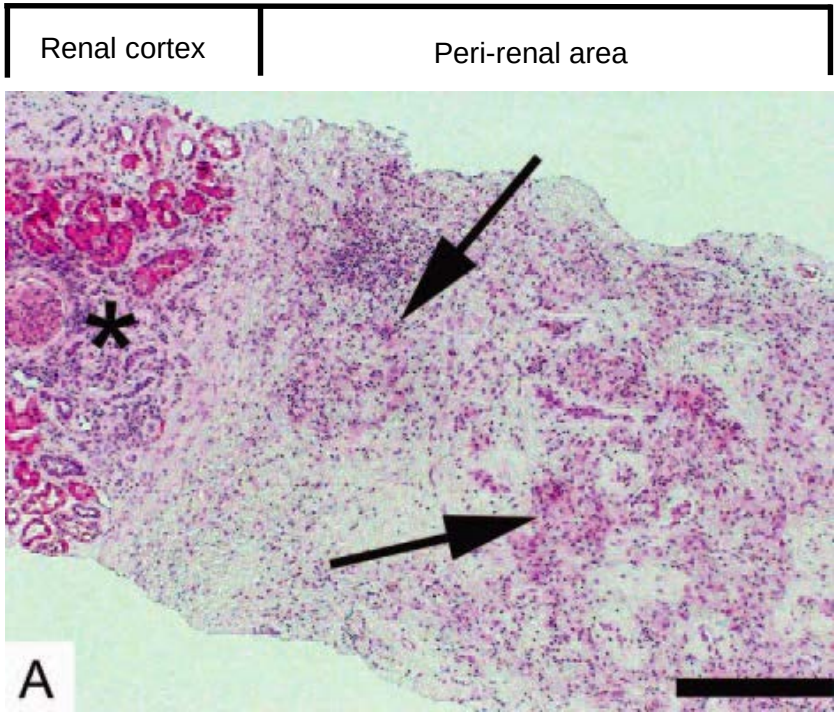
Perirenal infiltration in ECD (*hairy* kidneys)

Abdominal CT scan in a healthy 38 yo lady



Abdominal CT scan in a 50-yo ECD man

Perirenal infiltration in ECD (*hairy* kidneys)



- Approximately 50-70% of the cases
- The infiltration is often limited to the peri-renal space
- It usually extends to the renal hilum (renal artery and vein), the renal pelvis and the proximal ureter
- Peri-renal disease may limit the ability of the kidney to dilate when the ureters are compressed
- Good site for diagnostic biopsy

Imaging studies for retroperitoneal/perirenal ECD

Imaging technique	**PROs**	**CONs**
Sonography (ultrasound)	Allows visualisation of hydronephrosis; non-invasive, no radiation	Poor visualization of peri-renal or peri-ureteral tissue; limited usefulness for peri-renal tissue follow-up; operator-dependent
Computed tomography (CT)	Optimal visualization of all renal and ureteral complications of ECD; non-operator dependent	Radiation dose; potential nephrotoxicity; contraindicated if allergy to iodinated contrast medium
Magnetic resonance imaging (MRI)	Optimal visualization of all renal and ureteral complications of ECD; non-operator dependent; no radiation	Contraindicated in patients with severe renal failure, allergy to metals, bearing pacemakers or metal prosthesis
Positron emission tomography (PET)-CT	Allows evaluation of metabolic disease activity (active vs inactive) of peri-renal tissue; no significant contraindications	Radiation; does not reliably assess dimension of the lesion; high cost; does not provide renal functional evaluation
Renal Scintigraphy	Functional study; no significant radiation; can be repeated	Does not visualise anatomical structures

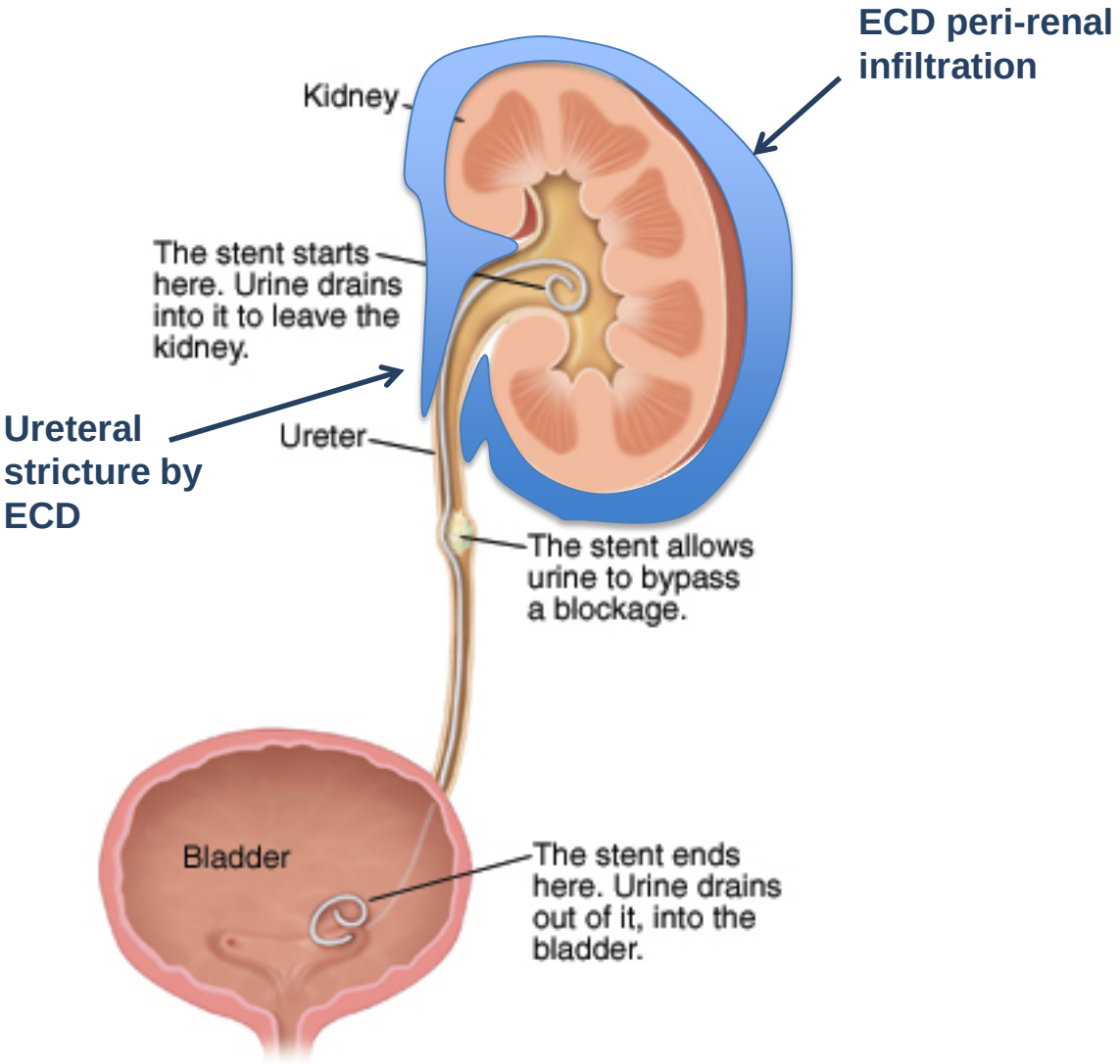
Ureteral obstruction in ECD

- Approximately 20-50% of the cases
- May be unilateral (often bilateral)
- Usually insidious onset, often asymptomatic or with dull back pain
- ECD infiltration usually causes stenosis of the proximal(upper) third of the ureter (unlike idiopathic retroperitoneal fibrosis)

How to relieve ureteral obstruction

Double- J ureteral stents

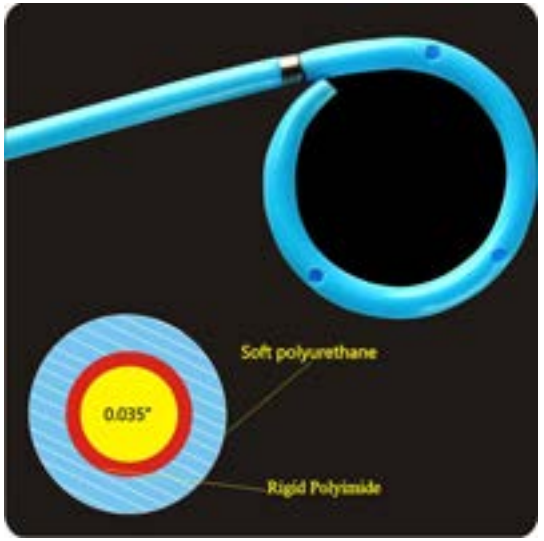
- Placed via cystoscopy
- Duration: 6-12 mths
- Complications: irritation, lower urinary tract symptoms, infections, bleeding
- Allow “internal” renal drainage avoiding external nephrostomy tubes



How to relieve ureteral obstruction

Double-J “tumor” stents

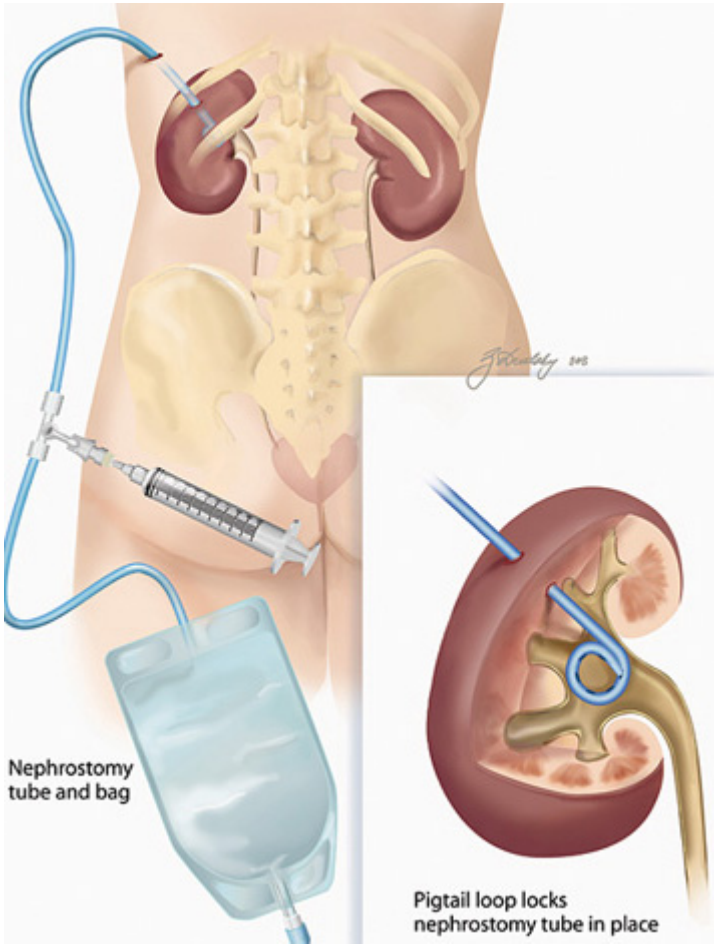
- Placed via cystoscopy
- Duration: 6-12 mths
- Reinforced internal layer for resistance to compression
- Different reinforced segments depending on the site of ureteral obstruction



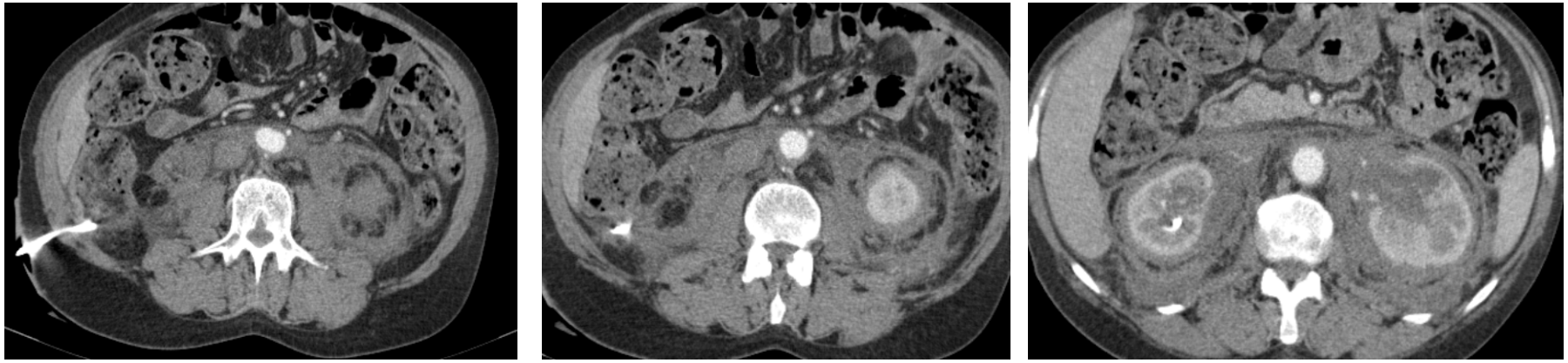
How to relieve ureteral obstruction

Nephrostomy

- Placed percutaneously
- Complications: infections, bleeding, poor quality of life
- Allows efficacious drainage of the obstructed kidney(s)



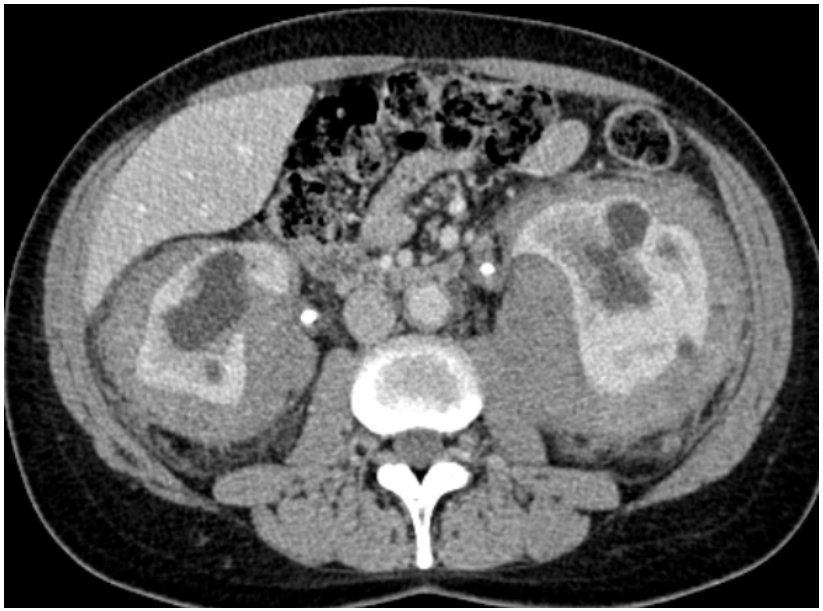
How to relieve ureteral obstruction



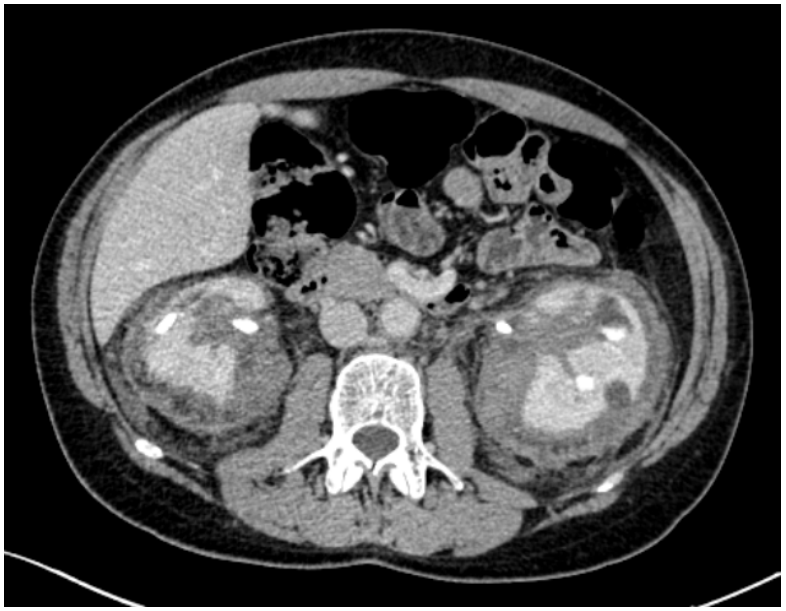
Abdominal CT scans show a nephrostomy tube in the right kidney of a 47-yo ECD patient

How to relieve ureteral obstruction

Ureteral obstruction must be treated with stents or nephrostomies AND medical therapy.
Although effective, this approach may not completely resolve obstruction and surgical ureterolysis can be performed (andecdotal reports, one in our series of 40 cases)



With stents, before therapy



With stents, month 6 of Everolimus therapy

Functional consequences of ureteral obstruction in ECD

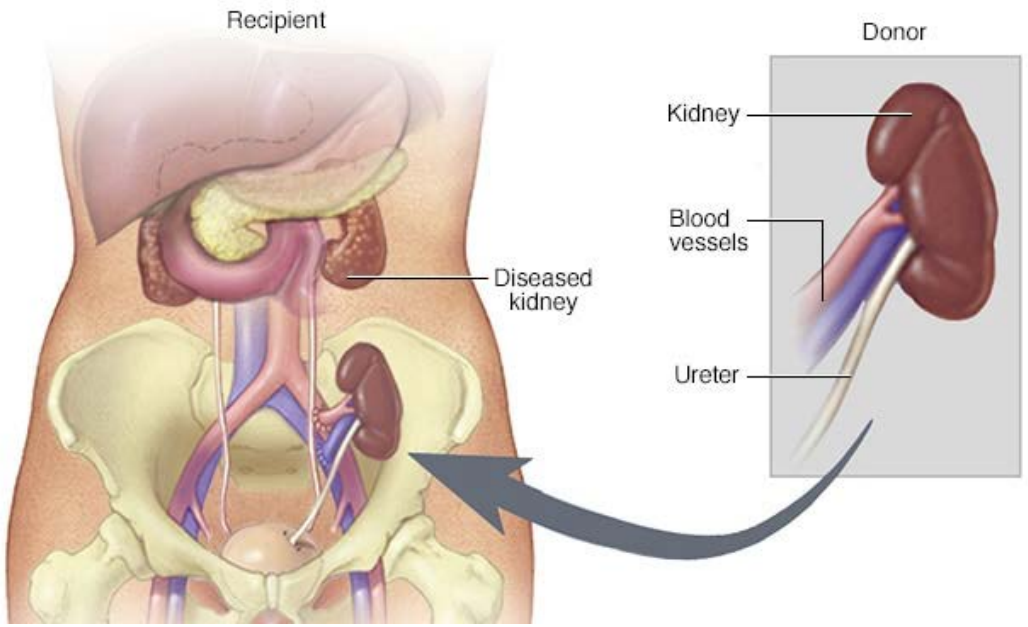
- Acute renal failure (uncommon)
- Chronic renal failure (chronic kidney disease, CKD) of varying degrees → possible progression to *end-stage renal disease*
- Renal atrophy
- Infection (pyelonephritis)

End stage renal disease

- Hemodialysis preferred over peritoneal dialysis
- Renal transplantation: no contraindications

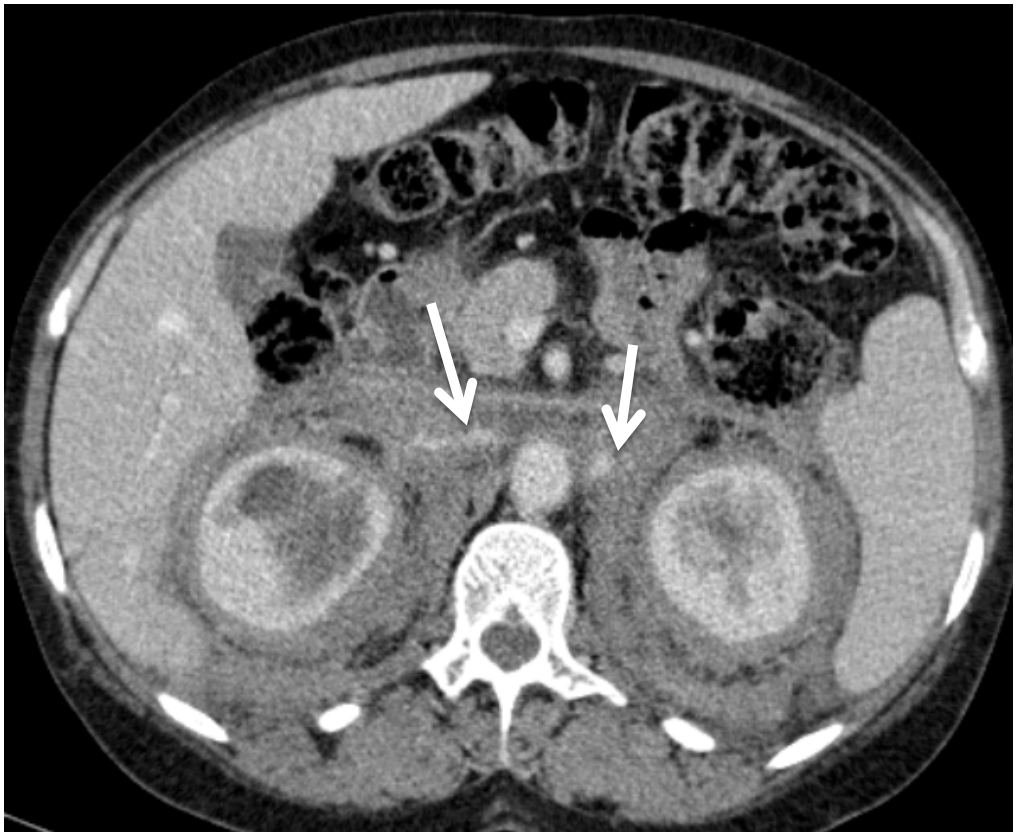
47-yo man with ECD and (moderate) chronic renal failure; CT scan shows hydronephrosis and renal hypotrophy/atrophy (right > left)

Renal transplantation in ECD



- No apparent technical contraindication if the iliac vessels of the patient are spared
- Contraindicated drugs after Tx: interferon alpha
- No contraindications to the use of Vemurafenib or anti-cytokine therapies
- mTORi are part of post-tx immunosuppressive regimens
- Risk of ECD relapse on the graft?

Renal artery (and vein) stenosis in ECD

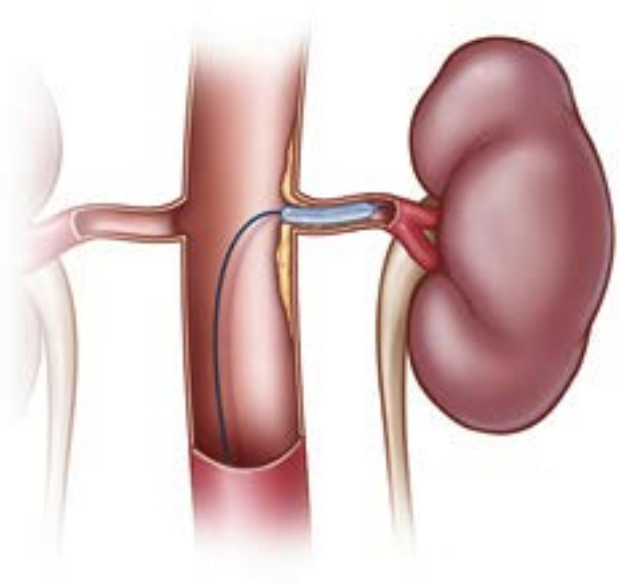


- Compression of the renal arteries causes *reno-vascular hypertension* , a particularly severe form of arterial hypertension
- It may also cause renal atrophy
- When bilateral, it can progressively lead to chronic renal failure
- Diagnosed by angio-CT, angio-MRI and traditional angiography

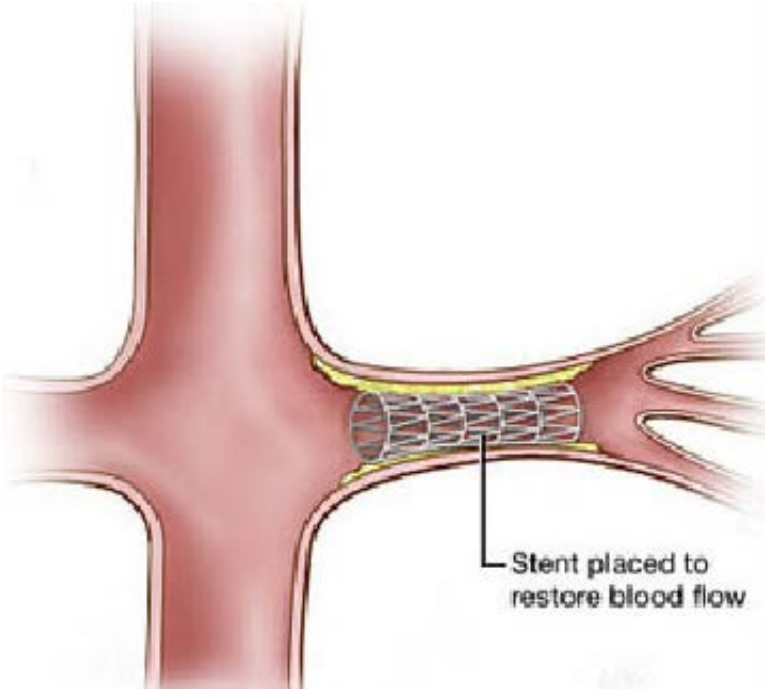
When to suspect renal artery stenosis in ECD

- Worsening hypertension
- Hypertension requiring multiple anti-hypertensive drugs
- Renal asymmetry on imaging studies
- Serological abnormalities
    - Worsening renal function (creatinine increase)
    - Hypokalemia (low K, below 3.5 mEq/L)
    - Increase in renin activity and aldosterone levels
    - Metabolic alkalosis (increase in pH and bicarbonate levels)

How to treat artery stenosis in ECD



Renal artery ANGIOPLASTY



Renal artery STENTING

Thank you