

Polycystic Kidney Disease

Characteristic Findings:

Commonest sign of PKD



- Polyuria
- Polydipsia
- **Pain** (dull, in the renal angle)
- Hematuria (in ~30%)
- Hypertension
- Abdominal discomfort
- **Stroke or subarachnoid haemorrhage may be the first sign!**

Decrease in urine-concentrating ability = early manifestation of the disease

Most patients WILL NOT BE NEPHROTIC
Most patients WILL BE PROTEINURIC

Renal failure happens in their 40s to 60s

CYSTS OCCUR IN OTHER ORGANS:
Liver, pancreas, spleen

On physical exam: Bilateral loin masses will be tender and palpable! This is the only other time you can ballot a kidney except for massive hydronephrosis.

Differentials to rule out:

- **Bilateral calculi** – but... they would be in more pain, it would be colicky, there would be frank red hematuria, etc...
- **Hydronephrosis** – any historical evidence of obstruction? Oliguria?
- **Pyelonephritis**- are they febrile? History of recurrent UTI? Any leucocytes in the urine?

main aim is to prove that the patient has big cysty kidneys, and that the cause of this is an autosomal dominant genetic defect

INVESTIGATIONS:

LOOKING FOR:
2 cysts in 1 kidney
or 1 cyst in each
 - in an at-risk patient younger than 30 years
OR
 At least 2 cysts in each kidney in an at-risk patient aged 30-59 years
OR
 At least 4 cysts in each kidney for an at-risk patient aged over 60

- FBC** -Elevated hematocrit may result from INCREASED EPO PRODUCTION from the cysts
-There may be normocytic normochromic anaemia if they are in end stage renal failure
- EUC** - electrolyte abnormalities are not usually seen- if they are, its either a late presentation or another problem underlying the presentation, like PKD as well as urinary obstruction
- Urinalysis** - Of course there will be BLOOD; but will there be LEUCOCYTES?
- if yes, it may be pyelonephritis or UTI instead of PKD
- Urine Microscopy** -where is the blood coming from? Dysmorphic RBCs?
- Abdo Ultrasound** { THIS IS "THE SHIT": will show you the characteristic kidney cysts, cysts in spleen and liver, etc... THE IMAGING OF CHOICE

CT scan is more sensitive for smaller cysts, but involves radiation.

HEAD MRI SCAN is necessary to investigate a possible ANEURYSM

Genetics: DNA linkage analysis for family members

MANAGEMENT

Relieve pain – NSAIDs, opiates

Control hypertension – these people get berry aneurysms:

Plus blood pressure control improves renal outcome

ACE INHIBITORS are the drug of choice;

Beta Blockers can be added for bonus effect

ALSO: lose weight, stop smoking, start exercising, etc...

Monitor renal function: eventually, will need dialysis and transplant

Genetic Councelling: risk to existing and future offspring

And **AVOID CONTACT SPORTS**, lest you burst a cyst!