SURGERY, TRANSPLANTATION AND POLYCYSTIC DISEASE

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What are polycystic kidneys and livers?

- **Cystic degenerative condition**
  - Multiple cystic (thin walled fluid filled cavity)

- **Genetic condition**
  - Autosomal dominant
  - (Autosomal recessive rare)
  - Approx 10% of renal disease
    - (1 to 2/1,000 births)

Big abdominal lumps that cause problems
In reality...

- Kidneys get big
- Kidney function gets worse
  - Treatment needed if end stage renal failure
- Renal replacement therapy (RRT) is dialysis or transplant
What is renal “failure”

- Stage 1: GFR above 90 mL/min
- Stage 2: GFR 60-90 mL/min
- Stage 3: GFR 30-60 mL/min
- Stage 4: GFR 15-30 mL/min
- Stage 5: GFR below 15 mL/min

GFR is roughly = % kidney function
## Symptoms and signs

### History
- **Pain**
  - Back/loin/abdomen
  - Enlargement of cyst(s)
  - Bleeding inside/outside cyst
  - Infection
- **Infection**
  - Pain
  - Fever
- **Kidney failure**
- **Incidental / Family history**

### Examination
- **Examination**
  - Big kidneys/liver
  - Abdominal distension
  - “Malnutrition”
  - Hypertension (blood pressure)
Malnutrition
How do we investigate?

- **Investigations**
  - **Blood tests** - FBC, U+E, Bone, LFT
  - **Urine tests** - protein, ACR, MSU
  - **X-rays** - pretty useless

- **Scans**
  - **Ultrasound** - good test - but no pics
  - **CT scan** - really good – nice pics…
Ultrasound

- **Diagnostic criteria:**
  - At least 2 cysts in 1 kidney or 1 cyst in each kidney in an at-risk patient younger than 30 years
  - At least 2 cysts in each kidney in an at-risk patient aged 30-59 years
  - At least 4 cysts in each kidney for an at-risk patient aged 60 years or older

- **Ultrasonography diagnostic criteria for ADPKD in patients with a family history but unknown genotype:**
  - Three or more (unilateral or bilateral) renal cysts in patients aged 15-39 years
  - Two or more cysts in each kidney in patients aged 30-59 years
  - Fewer than 2 renal cysts in the findings provides a negative predictive value of 100% and can be considered sufficient for ruling out disease in at-risk individuals older than 40 years.
CT - Polycystic kidneys
CT - Polycystic kidneys

![Normal kidneys CT scan]

![Diseased kidneys CT scan]

![Schematic diagram of normal and diseased kidneys]
Why would surgery be necessary?

- **Unrelated**
  - Dialyisis
  - Hernia

- **Directly related**
  - Symptomatic
    - Recurrent infection
    - Uncontrollable hypertension
    - Intolerable pain
  - Space
    - For transplant
Peritoneal dialysis

- Often possible
- Volume considerations
- Infections
Hernia

- Common in PCKD
- Recurrence high
Kidney surgery

- Nephrectomy
  - Means removal of kidney

- Bilateral nephrectomy
  - Removal both kidneys

- (Partial nephrectomy – part of a kidney)

- Laparoscopic
  - Aka keyhole
  - Using cameras and minimally invasive techniques

- (Embolisation)
How can we take them out?

Open

vs

Laparoscopic (keyhole)

vs

hand assisted

vs

(Embolisation)
In all operations...

- Must get to the kidneys
- Must mobilise the kidneys
- Must safely secure the blood supply
- Must divide the ureter
- Must get the kidney out
Open Nephrectomy
Open Nephrectomy

- Can approach both kidneys and vasculature
- Need to mobilise the peritoneal structures
  - Colon
  - Spleen
  - Liver
Squeamish...?

- Look away now.
Do you take out my own kidneys Doc?

- If causing problems
  - Infection
  - Hypertension
  - (nephrotic/protein loss)

- If too big!

- Infection
After bilateral (both) nephrectomy

- No kidney function at all
- Fluid restriction
- Blood pressure often low
Are there options?

- What about one side if it's just for space?

- Unilateral nephrectomy
  - Minimally invasive

- “Sandwich” technique

- Nephrectomy at time of transplant
Unilateral nephrectomy

- Do symptomatic kidney
- Largest kidney
- (Right sided transplant favoured)

- Timing…?
  - Pre dialysis
    - May push into end stage
    - Leaves a potential source of symptoms and problems
Fully Laparoscopic

- Dunn MD Am J Kid Dis 2000
  - 11 cases
  - Morcellation in 8
  - 7-11cm incision in rest
  - 6.3 hours
  - Size not mentioned

- Desai MR BJU 2008
  - N=13
  - Decreased blood loss and quicker recovery
Hand assisted

- Developed from donor nephrectomy
- Small series published

- Rehman J Urol 2001
  - 3 cases – 5.5 hours

- Lee and Clayman J Endourol 2004
  - 7 cases - 4.8-5.5 hours
Minimally invasive
Technique — retroperitoneal vs transperitoneal

- TP and RP laparoscopic nephrectomies provide good outcomes in patients with ADPKD. The choice of a TP route could decrease the length of hospital stay.

- Complication rates (25.6 vs 33.3 %, p = 0.44), transfusion rates (11.6 vs 20.5 %, p = 0.27) and conversion to open surgery (4.6 vs 7.7 %, p = 0.56) were similar between the TP and RP groups, respectively. Operative time was shorter for TP procedures (171.6 vs 210.5 min, p = 0.002), but there was no difference between the two approaches after 20 surgeries (p = 0.06). Patients in TP group had a shorter length of hospital stay (5.3 ± 1.9 vs 7.2 ± 2.5 days, p = 0.002). However, there was a trend towards shorter return of bowel function in the RP group (2.1 ± 0.9 vs 2.4 ± 0.8 days, p = 0.09).

World J Urol. 2015 Dec 10. [Epub ahead of print]

Laparoscopic nephrectomy for polycystic kidney: comparison of the transperitoneal and retroperitoneal approaches.

Benoit T1
Technique

- Depends on:
  - Surgeon
  - The patient
  - The kidneys
  - The indications

- One size doesn’t fit all
- Safety is the over riding priority
When?

- Take a kidney out when:
  - GFR ~15
  - Needs dialysis
  - !!! - If no LD (not uncommon in PCKD!)
  - Wait for a kidney in UK ~1000d
Timing

- Very difficult with little evidence
  - Opinion is only one pre-Tx / peri-ESRF
  - Pre-tx:
    - One kidney
    - Technique is dependent on surgeon and your kidneys
  - Concomitant (same time as transplant):
    - Has been done
    - Increased blood transfusions and wound problems
    - Generally avoided in UK
- Post transplant:
  - If problems. If not leave alone (16% required nephrectomy in 10yrs)
Simultaneous transplant and nephrectomy

- Ishmail H  2005
  - Concomitant (n=11) vs sequential laparoscopic (n=7)
  - Concomitant
    - Complications in 7 (Bowel injury, transplant urine leak, necrotic pancreatitis,)
    - 1 kidney lost
    - 68% required a second surgical procedure
  - Sequential laparoscopic
    - No major complications


Abstract

BACKGROUND: Many strategies regarding timing of native nephrectomies exist for patients with symptomatic polycystic kidney disease (PCKD).

METHODS: This is a single-center, retrospective study of 594 adults with PCKD who had renal transplants from 1994 to 2014. Three groups were analyzed: renal transplant-only recipients (tx alone), recipients of simultaneous bilateral nephrectomies and transplant (simultaneous), and recipients with pretransplant bilateral nephrectomies (pre). The primary outcome was graft survival. Secondary outcomes included postoperative complications.

RESULTS: Five hundred sixty-five adults with PCKD received kidney transplants (303 tx alone, 161 simultaneous, 27 pre). Ten-year posttransplant graft survival was 68.5%, 63.6%, and 65.7% for tx alone, simultaneous, and precohorts (P = 0.86). No statistically significant differences were observed in rates of postoperative ileus, deep vein thrombosis, small bowel obstruction, urinary stricture, urine leak, hernia formation, and delayed graft function. More wound complications were seen in prepatients (25.9% vs 11.1% tx alone, 5.1% simultaneous; P = 0.03), whereas simultaneous patients had a lower incidence of lymphocele (1.3% vs 11.1% pre, 10.2% tx-alone; P = 0.002). Importantly, simultaneous patients had more renal vascular thromboses (4.4% vs 1.3% tx alone, 0% pre; P = 0.04). 16.3% of renal transplant alone patients required nephrectomy at 10 years follow-up. Twenty-nine patients were referred for transplant having had nephrectomies and were ultimately not transplanted. In 4 of these patients who had data available for analysis, the mean panel-reactive antibody significantly increased after nephrectomy was performed.

CONCLUSIONS: Simultaneous bilateral nephrectomy can be safely performed at the time of renal transplantation, however, carries a significantly increased risk of renal vascular thrombosis.

Simultaneous had >3x risk of immediate Tx failure
Radiology

- Percutaneous drainage of cysts
Embolisation

- Instead or as well as surgery

- The artery is blocked off with via a catheter through the blood vessels

- The kidney shrivels up

- May be promising

- May be dangerous!
Partial “nephrectomy”

- What does it mean?
  - Cyst de-roofing
  - Cyst excision
  - “True partial nephrectomy”
  - Described for cancers
  - Nephron sparing
Transplantation

- Best form of renal replacement therapy (RRT)

- Outcome

  - Graft Survival in Patients With Polycystic Kidney Disease With Nephrectomy of Native Kidney Pre-transplant.

  - Graft survival after the first year was of 98% for group 1 and 95% for group 2.

  - The 5-year implant survival was 95% and 80% respectively.

  - (García-Rubio JH, 2015)
Recipient details – cause of ESRF

- PKD: 14%
- FSGS: 5%
- IgA: 11%
- vasculitis: 3%
- Urol: 8%
- other: 26%
- DM: 7%
- GN: 4%
- Small: 4%
- BP: 3%
- CNI: 1%
- ???: 14%
how do we transplant?

- **Donor**
  - Cadaveric
    - DCD
    - DBD
  - Living
    - Related
    - Unrelated
    - ABOi
    - HLA
    - Paired
    - Altruistic
Technical.

- Heterotopic
- Kidney goes onto iliac vessels
- Ureter onto bladder
- Drawbacks:
  - Immunosuppression, NODAT, marginal/ECD, Delayed function, infection/tumours…
- BUT
  - Improved QoL and quantity of life
What about the liver?

- Polycystic disease affects liver and kidneys variably:
  - Liver – size
  - Kidneys poor function plus liver size
    - Combined liver + kidney transplant
What problems do polycystic livers cause?

- Pain
- Infection
- Size effects – 10 to 20kg
  - Physical handicap
  - Liver effects (cholestasis/portal hypertension)
  - Malnutrition
What can we do about it?

- Large cysts
  - Fenestration (“make a window”)  
  - Resection (“remove”)

- Small cysts
  - Little can be done

- Hepatectomy and transplantation
Transplantation of the liver

- Pirenne 2001
  - 16 patients (14 combined liver kidney)
  - Survival 87.5%

- Ueno 2006
  - 14 patients (12 combined liver kidney)
  - 93% survival

- Coquillard C 2015
  - 1, 3, and 5 year survival of 91%, 90%, and 90%, respectively (better than LTx alone other causes)
Combined liver kidney transplant

- Immediately after liver transplant the body isn’t in the best place to receive a kidney transplant.

- BUT allocation policy favours combined Liver Kidney Tx.

- It maybe that liver transplant followed by increased priority on kidney transplant list is better (Germany).
Summary

- Polycystic Disease is rare in society
- It is quite common in kidney clinic

- Surgery is not the best option
  - Desire for alternative treatments

- Transplantation is a good option

- Nephrectomy may be required
  - Various options – but evidence is lacking!
  - Timing is difficult
Malnutrition

- Organomegaly (massive organs) may result in malnutrition

- Small study
  - Muscle mass reduced in PCKD vs controls (Inston 2015)

- More research needed

- Is it infection inflammation?
- Is it space effect?
Thank you

☐ Questions