

Preserving Kidney Function in Spina Bifida

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Spina Bifida
Hydrocephalus
Ireland

Overview

- Function of the kidneys
- Symptoms / stages of kidney disease
- Kidney disorders in spina bifida
- Management kidney disease
- Prevention kidney disease

Function of the Kidneys

- Blood pressure control
- Water and salt balance – urine volume
- Clearance of waste products including some drugs (uraemic toxins)
- Hormone production – EPO / Vitamin D
- Regulation of blood acid levels (pH)

KDOQI Stages of Chronic Kidney Disease

Stage	GFR	Description of kidney function
1	>90	Normal unless urine / structural / genetic abnormality
2	60-89	Normal or mildly reduced if above criteria met
3	30-59	Moderately reduced
4	15-29	Severely reduced
5	<15	Very severe / ESRD

Classification of Chronic Kidney Disease

- MDRD formula
 - calculates eGFR corrected for BSA
 - 4 variables including age, race, sex, creatinine
 - eGFR is roughly equivalent to percent of renal function
 - eGFR 30mls/min approximates to 30% renal function
 - eFGR 50mls/min approximates to 50% renal function
 - Less accurate at higher GFRs

Symptoms of Kidney Disease

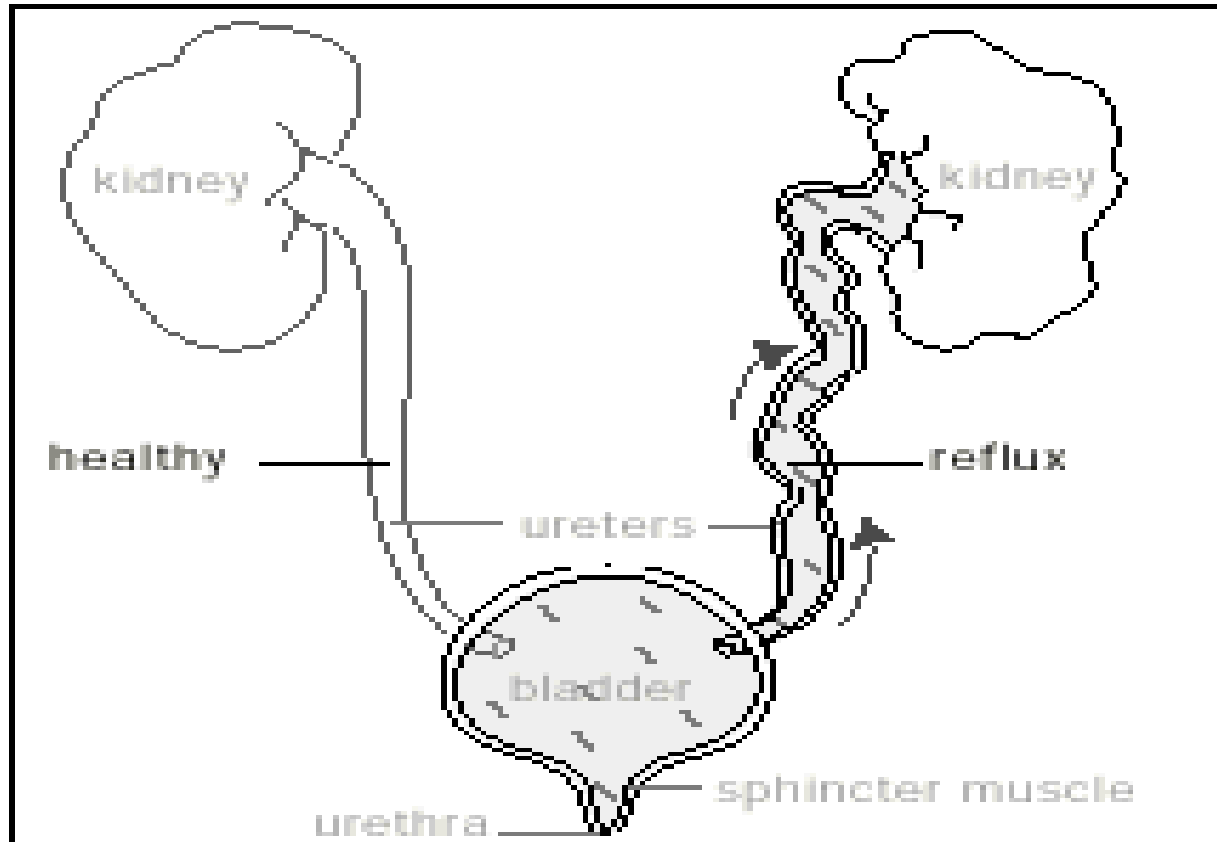
Frequently few symptoms until late in illness

- Tiredness / poor stamina (low Hb)
- Leg swelling / shortness of breath
- Reduced appetite / weight loss
- Itch / bone pain
- Restless legs / muscle twitching
- Nausea / taste in mouth
- Decreased concentration
- Reduced libido

Kidney Disorders in Spina Bifida

- Bladder functions poorly
 - Incontinence
 - Reflux with scarring
 - Obstruction to urine flow
 - Infections
- Kidney stones
- Prostate problems (older males)

Reflux Nephropathy



Kidney Disorders in Spina Bifida

- Usually born with normal kidney function
 - Damage starts within 2-3 months if not managed by urologist
- Kidney disease preventable
 - Anti cholinergics (reduce bladder pressure)
 - Clean self intermittent catheterisation
 - Aggressive mgt of urine infections
 - Take the antibiotics and finish the course!!
 - Treat even if no symptoms!

Kidney Disorders in Spina Bifida

- Further ways to minimise kidney damage
 - Urinary diversion if CIC not possible
 - Restoration of continence
 - Reduce salt intake
 - Avoid toxic drugs ie anti inflammatories
 - Take prescribed meds (espec BP)

Kidney Disorders in Spina Bifida

- **KEEP YOUR CLINIC VISITS**
 - Bloods and BP need to be checked
 - Renal scans to look for damage
 - Ask questions?
 - Know your level of kidney function
- **Spina Bifida patients get lost to follow up**
 - Transfer from child to adult services
 - Independence / reduced parental input

Kidney Disorders in Spina Bifida

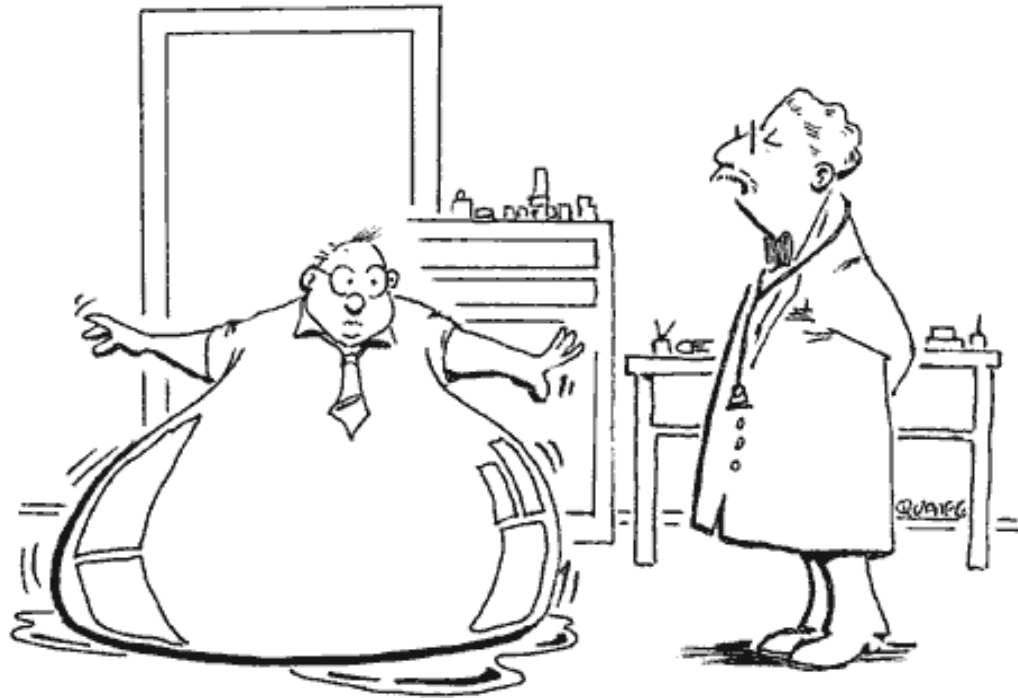
- Kidney injury is preventable in most
- Where kidney injury has already occurred
 - We can slow / stop it from worsening
 - BP control / diet / treating infections
 - Dealing with urology issues
- Where injury is severe
 - Prepare for dialysis and transplant

Dialysis Initiation

- eGFR < 10mls
- Symptoms uraemia
 - Nausea / weight loss / itch / myoclonus etc
- Volume overload
- Metabolic derangements
 - Hyperkalaemia / acidosis
- Ideally start before patient experiences functional decline

Starting Dialysis

www.lightersideofdialysis.com



Your tests reveal that
you are retaining fluids!

Dialysis – General Issues

- Dialysis access required
- Dietary restrictions
- Fluid restrictions
- Large pill burden
- Time consuming (travel / OPD / therapy)
- Huge social cost (family / work)

- SYMPTOM CONTROL – NOT CURE

Haemodialysis – Vascular Access

- **Permcath**

- Tunnelled plastic tube running into large vein
 - Neck or occasionally top of leg
- Infection risk / thrombosis
- Can be difficult to insert as veins small
- Anatomy varied
- Cannot swim / take a bath

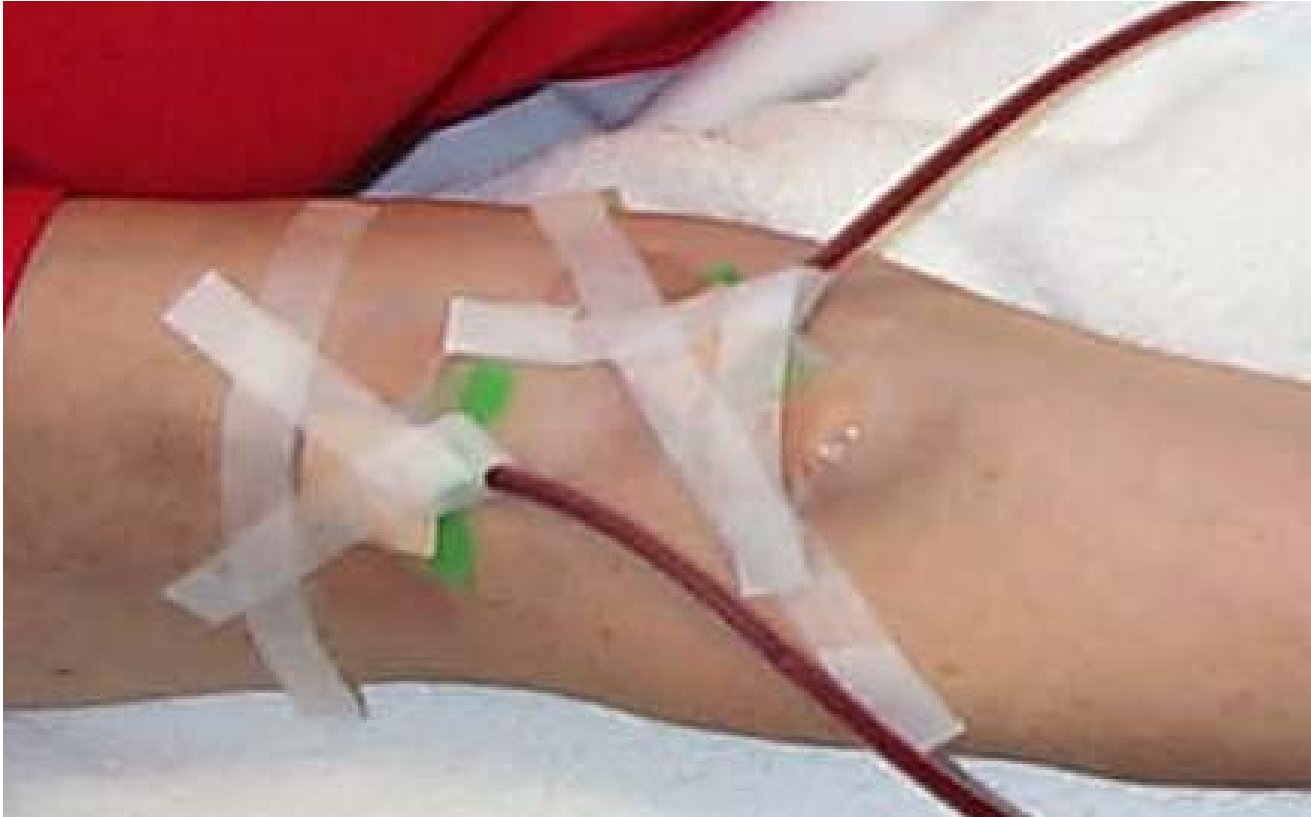
Tunnelled Haemodialysis Catheter



Haemodialysis – Vascular Access

- Arteriovenous fistula (AVF)
 - Surgical connection between an artery and vein allowing needles to be inserted for HD
 - Painful / unattractive to look at / failure
 - Can be difficult to make as veins small
 - Difficulty if wheelchair user

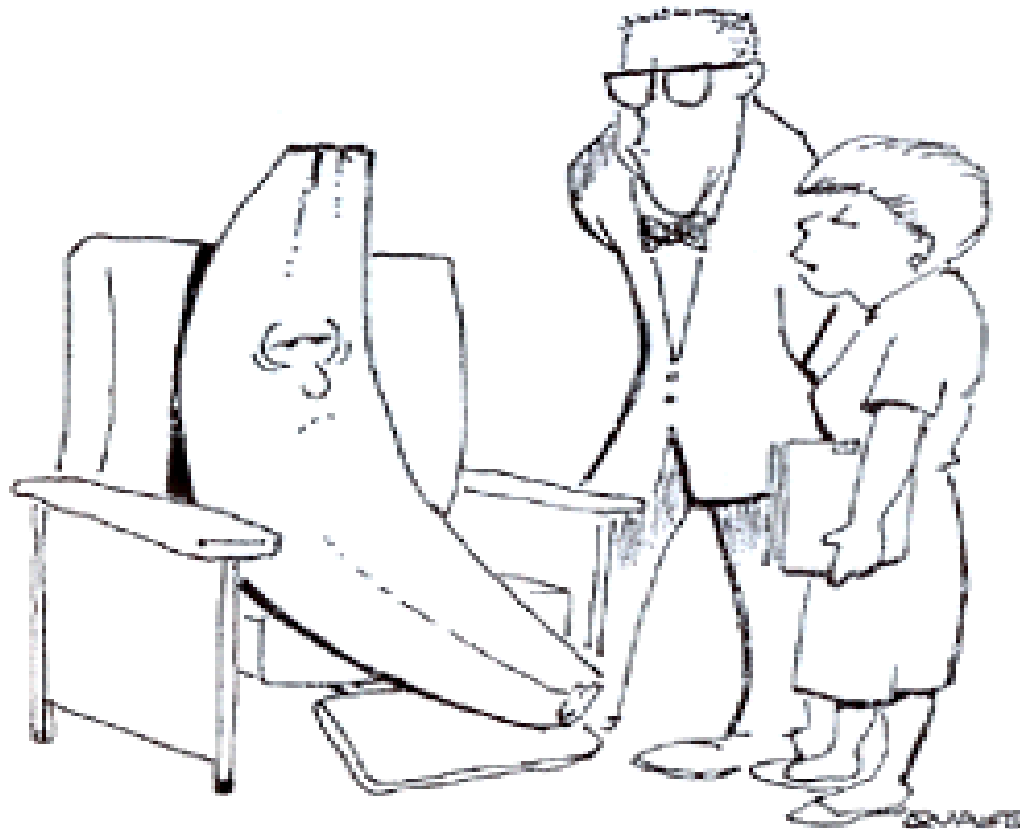
Arteriovenous Fistula in Arm (AVF)



Haemodialysis

- Haemodialysis patients report
 - 3 good days out of 7, generally day post HD
 - 60% would not start dialysis again if they had the choice
 - Travel severely restricted
- Future
 - Moving to community based therapy
 - Home haemodialysis provision
 - Avoiding dialysis in the first place!!

Dietary Restrictions!



We're a little concerned
about your potassium levels.

Peritoneal Dialysis

- **Peritoneal Cavity Access**
 - PD Catheter (Tenckhoff) inserted 10 – 14 days before use
 - Malposition / constipation common
- **Peritonitis main complication**
 - 1 in 29 patient months, admission and catheter loss infrequent
- **Dietary / fluid restrictions**
 - Less extreme

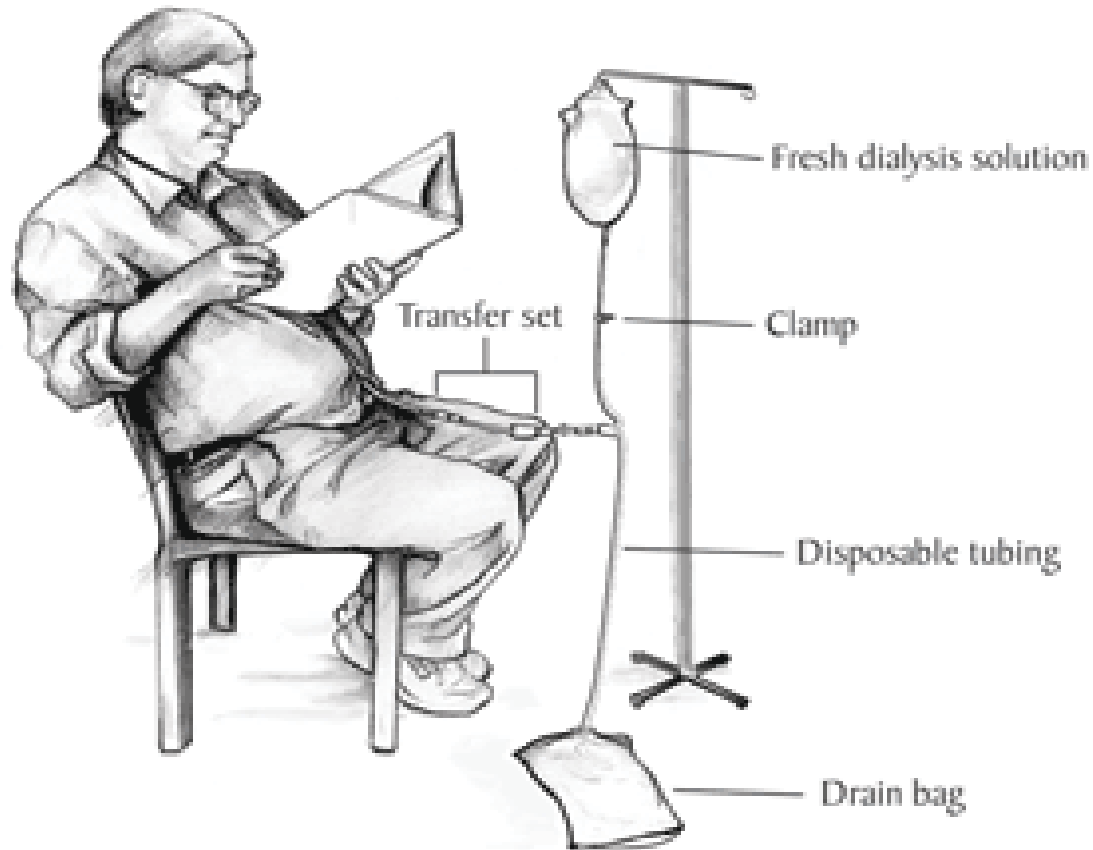
Peritoneal Dialysis

- Peritoneal dialysis procedure
 - CAPD - 4 exchanges daily, 30 mins each
 - APD – 9 hours therapy while you sleep!!
- Pill burden
 - 8 -10 medications on average
- Highly portable therapy
 - Work / travel much less restricted
 - Easier to socialise

Peritoneal Dialysis Catheter



Peritoneal Dialysis Exchange



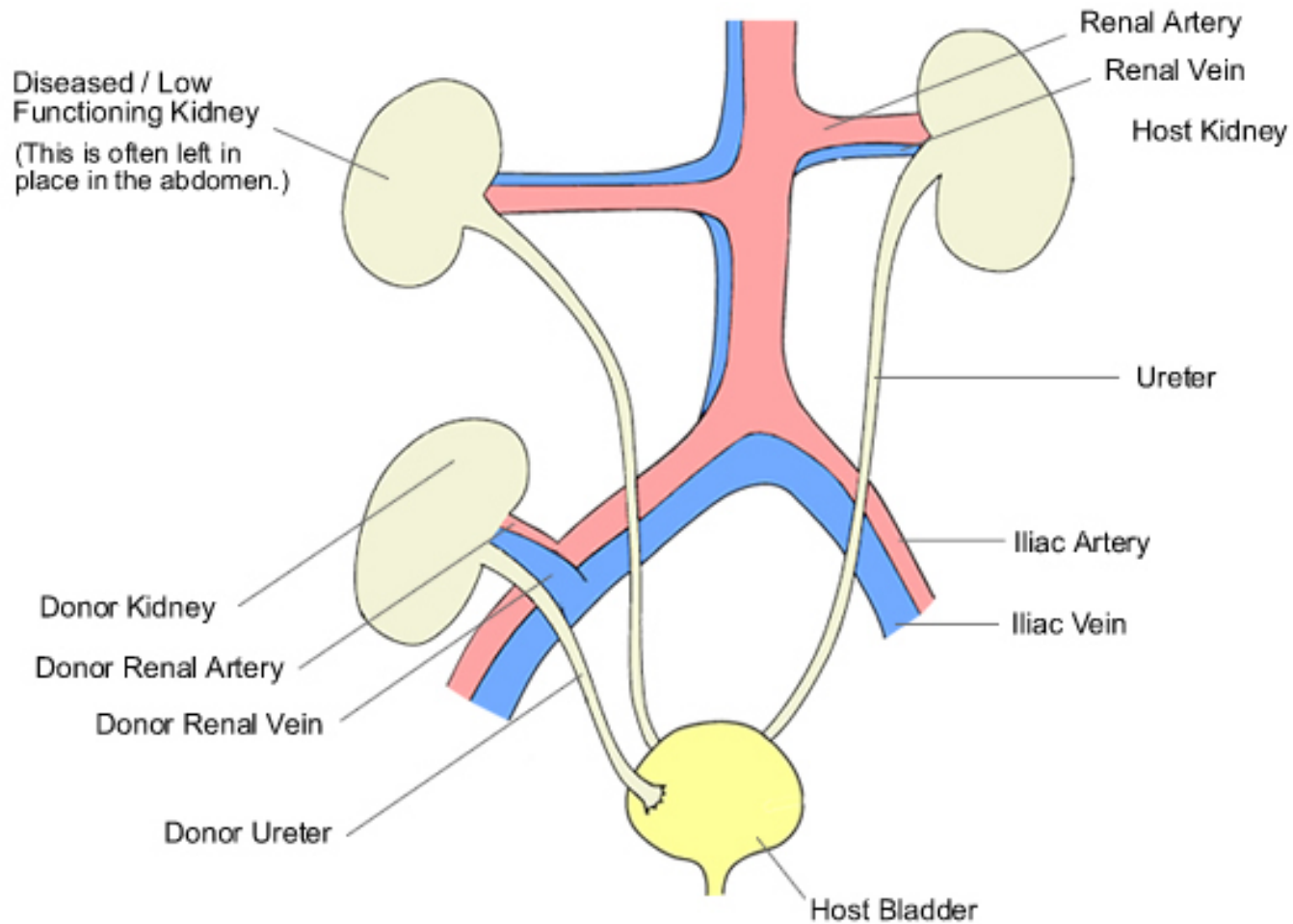
Situations Where PD is Avoided

- Severe scoliosis
- Urinary / bowel openings to abdominal wall
- Presence of VP (ventriculoperitoneal) shunt
- Chronic constipation makes PD difficult
- Carer often performs treatment

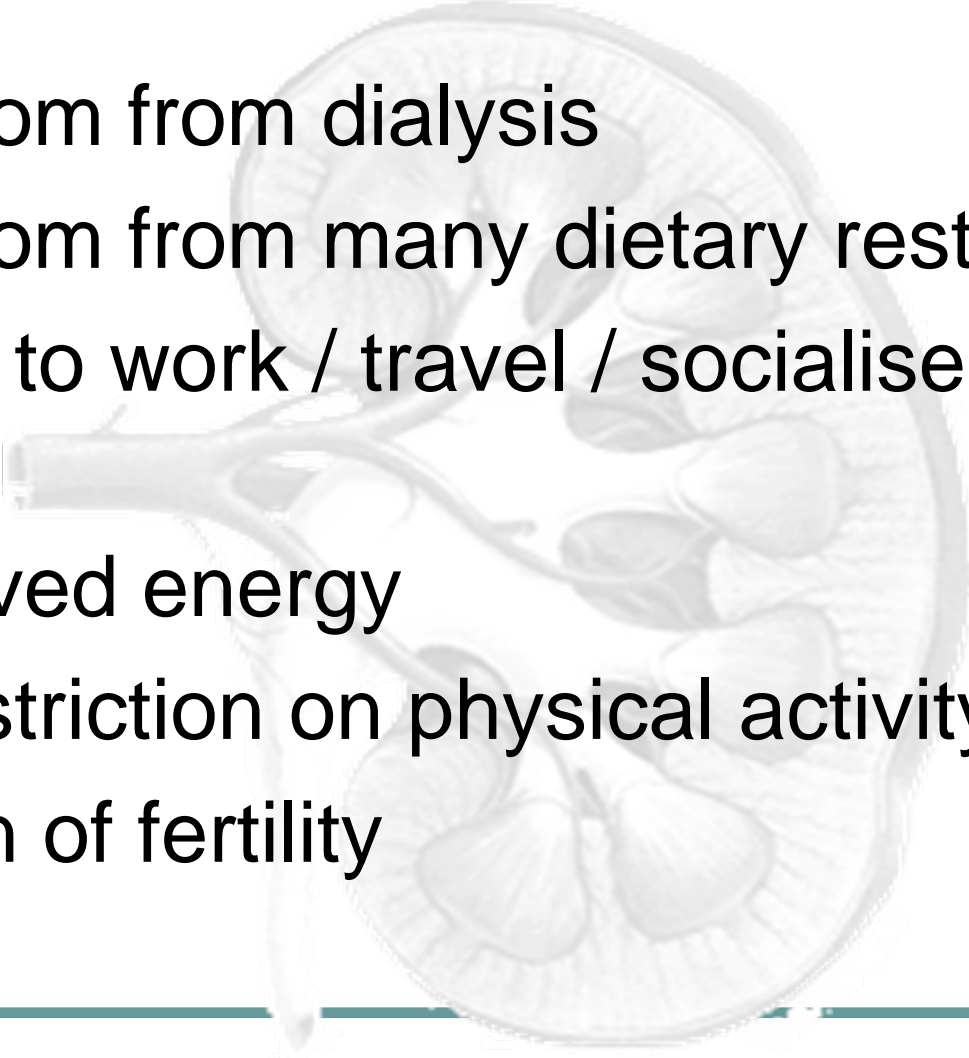
What is Kidney Transplantation?

- Another way of treating kidney failure!
- Most patients already on dialysis
- Some get transplanted before starting
 - Preemptive transplant (child / live donor)
- An operation is needed to place the kidney
- Anti-rejection drugs must be taken for life

The Operation!



The Upside!!

- Freedom from dialysis
 - Freedom from many dietary restrictions
 - Ability to work / travel / socialise to the full
 - Improved energy
 - No restriction on physical activity
 - Return of fertility
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The Downside!!

- Lifelong anti-rejection medication
 - Drug side effects inc diabetes
 - Increased infections
 - Increased risk of some cancers
- Some kidneys fail rapidly
 - Early return to dialysis
- All kidneys fail eventually
 - Many people now awaiting a second kidney

Transplant in Spina Bifida

- It is possible and it does work but there are special challenges
- May need own kidneys removed if recurrent urine infections / stones
- May require surgery to correct bladder
- Transplant operation more difficult
- Urine infections more common post

Why Does Renal Failure Happen?

- Lack of attention
- Lack of compliance
- Lack of follow-up

How Can You Protect Your Kidneys

- Listen to your urologist
 - Catheterise / antibiotics / anti spasmotics
 - Ask questions about your progress
 - Take control
- Don't skip clinic visits - paed to adult
- Adult patients
 - Bloods / BP every 6 to 12 months
 - Continue with good bladder habits
 - Take BP / other meds if prescribed

