Autosomal Recessive Polycystic Kidney Disease (ARPKD) Patient Journey

1. First Symptom

Enlarged, echogenic cystic kidneys

**Note:**
Regular antenatal scans

**Ideally:**
Regular antenatal scans – plan for birth made

2. Diagnosis

High blood pressure, impaired renal function, hypoglycaemia

**Note:**
Bp monitoring, TPN, medication for blood pressure started, dextrose to control blood sugar levels, transferred from local hospital to GOSH.

**Ideally:**
Management of bp – stabilize renal function

3. Treatment...

Decreasing renal function and worsening symptoms of renal failure. Emotional support from psychologist – Rejection twice

4. Surgery...

GFR of 9, severe renal impairment, distended abdomen, regular vomiting, anaemia – kidneys 20 cm each, combined weight 1.5 kg

**Note:**
Requires second renal transplant within the next 12-18 months.

**Ideally:**
Improved quality of life, avoiding dialysis, reduction of medication, regular check ups, psychological input and support

5. Follow-Up

At first, good renal function, as time has progressed, more medication has been reintroduced due to failing transplant – chronic kidney disease, (Glomerulonephropathy). Now anaemic, high PTH levels, high BP, heart disease, acidotic,

**Note:**
Management of bp – stabilize renal function

**Ideally:**
To maintain healthy blood sugar levels, to slow down progression of renal disease, to manage symptoms of renal failure, psychology support.

Improved quality of life, reduction of medication, avoiding dialysis.