

# Autosomal Recessive Polycystic Kidney Disease in the UK National Registry of Rare Kidney Diseases (RaDaR)

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By your side

# ARPKD

- Autosomal Recessive Polycystic Kidney Disease
- Affects 1 in 10,000 to 1 in 40,000
  - UK patient population unknown
- Causes major problems in kidney and liver.
- Wide clinical variability.
- Historically referred to as 'infantile polycystic kidney disease'
- No NHS specialised service/MDT clinics.



- National Registry of Rare Kidney Diseases (RaDaR), launched in 2009.
- UK-wide ethics approval for both **adult** and **paediatric** recruits for over **30** conditions.
- Sep 2017: **14,272** patients in RaDaR from 87 centres.
- Aims:
  - Improve information & support – HCPs & patients & families via **rarerrenal.org**
  - Develop best practice guidelines
  - Develop research cohorts



# ARPKD RaDaR

## ARPKD inclusion criteria:

- ARPKD
- Caroli's disease & Congenital Hepatic Fibrosis

National renal dataset & condition-specific codes  
(SNOMED-CT)

Linked to '**Patient View**' – where patients can view blood  
and other test results directly from NHS labs

1st ARPKD patient recruited October 2012



# Clinic data entry



Birmingham Women's  
and Children's  
NHS Foundation Trust



Cohorts Hospitals Patients Users News Stats Admin

Logged in as admin

GEORGE WRIGHT

ID 70

DOB 13/01/1932

Gender Male

CHI 0158460626

Recruited On 26/06/2010

Recruited By bot

Recruited At RaDaR

Cohort ARPKD

Hospital Glasgow - Queen Elizabeth University Hospital

Hospital Edinburgh Royal Infirmary

Hospital Truro Royal Cornwall Hospital

Hide Demographics

RaDaR Demographics Consultants Cohorts Hospitals More...

ARPKD Primary Diagnosis Comorbidities Genetics Family History Fetal Ultrasounds Fetal Anomaly Scans Renal Imaging Liver Imaging Liver Diseases Lab Results Nutrition Liver Transplants Nephrectomies

## Demographics

Search

	<u>First Name</u>	<u>Last Name</u>	<u>DOB</u>	<u>DOD</u>	<u>Gender</u>	<u>Ethnicity</u>	<u>Data Source</u>
<input type="button" value="View"/> <input type="button" value="Edit"/>	GEORGE	WRIGHT	13/01/1932	-	Male	Black Carribean	RaDaR (RADAR)

## Patient Numbers

New

Search

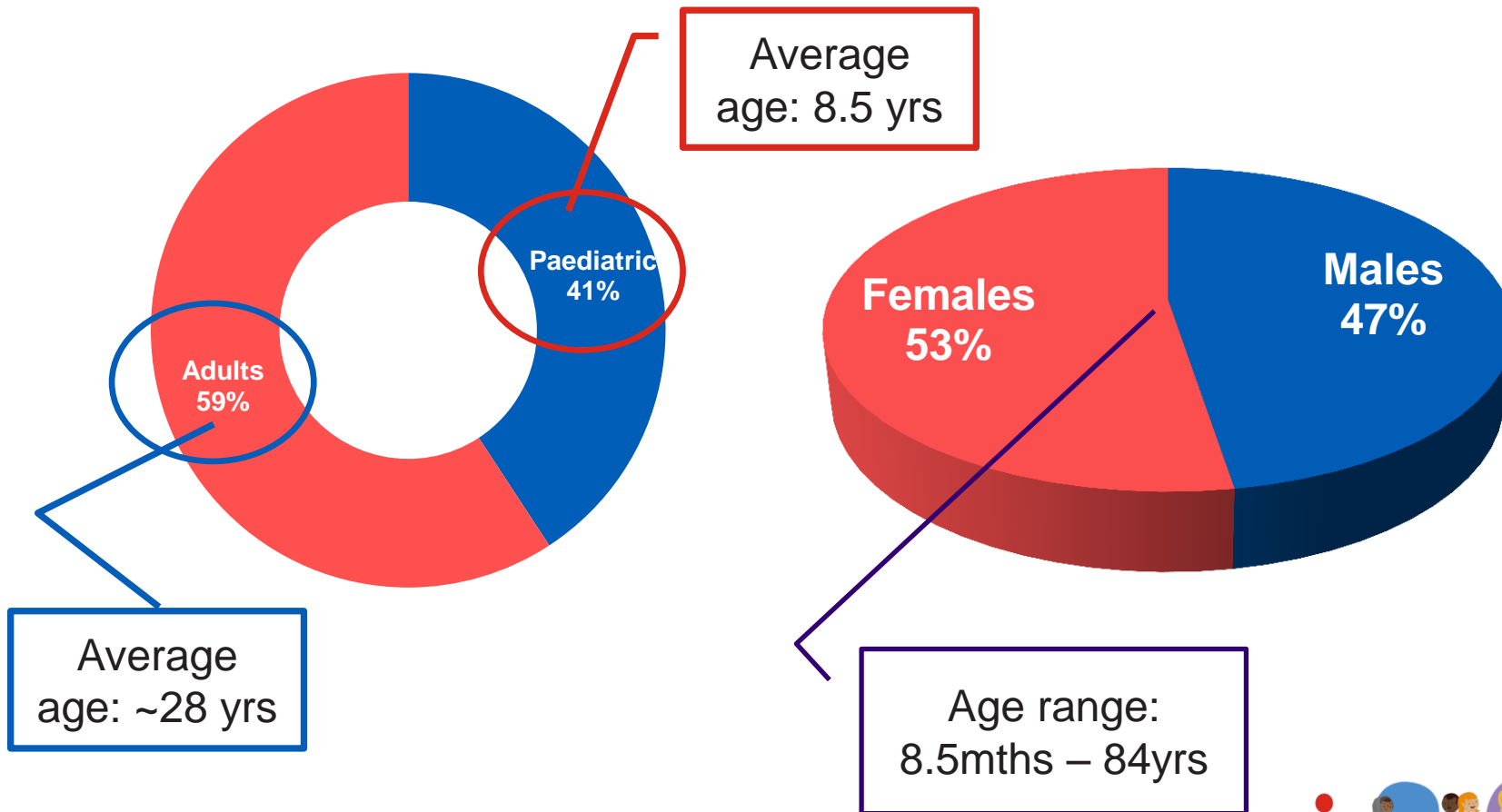
	<u>Number Group</u>	<u>Number</u>	<u>Data Source</u>	
<input type="button" value="View"/> <input type="button" value="Edit"/>	NHS	4503747681	RaDaR (RADAR)	<input type="button" value="Delete"/>
<input type="button" value="View"/> <input type="button" value="Edit"/>	NHS Blood and Transplant	31860	RaDaR (RADAR)	<input type="button" value="Delete"/>

**Demo**  
<http://demo.radar.nhs.uk/>  
Username: admin  
Password: htcsyrnk



# ARPKD RaDaR Cohort\*

- 156 ARPKD patients from 37 UK renal units



\* @ 1/9/2017



# Cohort Governance

- ARPKD RaDaR **Rare Disease Group (RDG)**.
- RDG membership represents national expertise in ARPKD – clinical, scientific, allied health professionals.
- Patient and/or carer representation.
- Representative from a patient support group – PKD Charity.



# International Links

ARPKD RaDaR is UK based & aligned with

- US-based Hepato/Renal Fibrocystic Diseases Core Center (UAB HRFDCC): Prof Lisa Guay-Woodford
- European ARPKD Registry (Cologne)  
ARegPKD: Dr Max Liebau
- ERN (ERKNet)





# Next Steps

- Ongoing data collection and enrichment on each patient on RaDaR for detailed phenotype.
- Potential to link RaDaR to other UK registries, databases and bio-banks that the patient has consented to, eg: HES, UK Bio-bank
- Ability to contact patients directly about future studies.
- Research prioritisation in collaboration with patients, families and HCPs.



# Thank you

- All UK clinicians encouraging patients & families to take part in RaDaR
- Renal Registry/RaDaR, especially Melanie Dillon
- PKD Charity (coding support)



# Thank you



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Walking Together for  
PKD

