The National Registry of Rare Kidney Diseases (RaDaR): cross-sectional analyses of 25,817 adults and children with rare kidney diseases in the UK

Katie Wong[1,2], David Pitcher[1,2], Lewis Downward[1], Retha Steenkamp[1], Fiona Braddon[1], Garry King[1], Kate Osmaston[1], Andrew Atterton[1], Dorothea Nitsch[1,4], Kate Bramham[1,3], Daniel Gale[1,2]

Affiliations: [1] Rare Renal Disease Registry, Bristol, UK; [2]Department of Renal Medicine, University College London; , UK [3] King's College London, UK; [4] London School of Hygiene and Tropical Medicine, London, UK

Background

Rare kidney diseases make a significant contribution to the burden of Chronic Kidney Disease in the UK and globally

- 10% adults
- >50% children

on Renal Replacement Therapy have a rare disease.

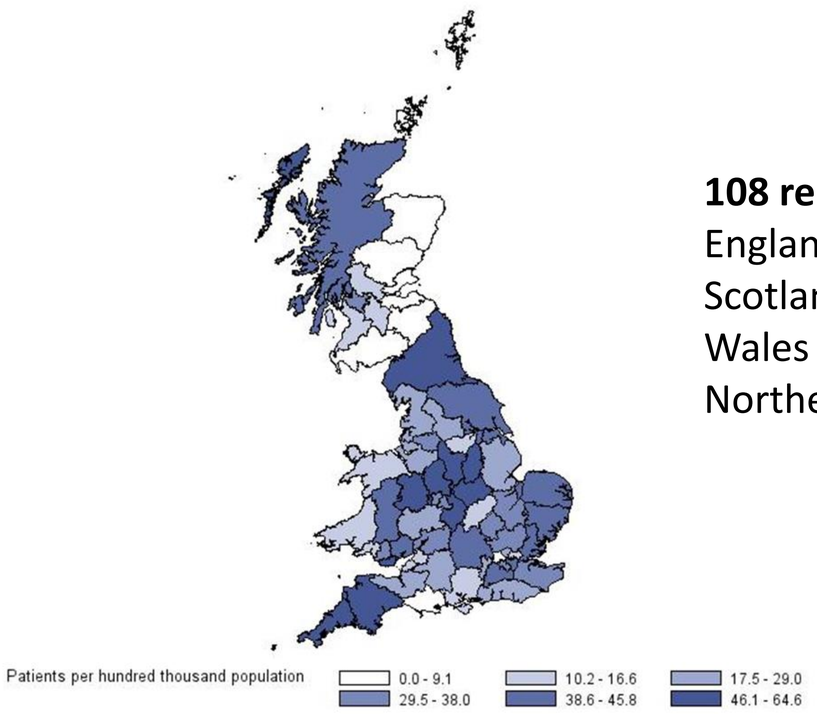
Due to small patient numbers, rare kidney diseases are often poorly characterised and lack data describing the determinants and distribution of these diseases.

RaDaR was formed in 2010 to address this knowledge gap

- It collects longitudinal and retrospective data for individuals with rare kidney diseases, back to the 1980's for some patients

RaDaR is the largest rare renal disease registry in the world

Figure 1: Distribution of recruitment to RaDaR across the UK



108 renal units
England n=91
Scotland n=9
Wales n=3
Northern Ireland n=5

Results

25,817 prevalent patients

1908 children <18yrs (7.4%)	23909 adults (92.6%)
Most common diagnoses	**Most common diagnoses**
• Idiopathic nephrotic syndrome (44%) • Vasculitis (11%) • Alport Syndrome (6%)	• ADPKD (29%) • Vasculitis (16%) • IgA nephropathy (17%)

Children

- More likely to be from **Asian backgrounds** (17.2% vs 7.5%, Chi^2 p<0.0001)
- More likely to live in the **most socially deprived areas** (30.1% vs 17.4%, p<0.0001)

compared to adults, and compared to the general UK population (18% vs 10%, p<0.0001 and 31% vs 24%, p<0.0001, respectively)

Ethnicity and Socioeconomic Status

Ethnicity (all RaDaR participants)

White 86%
Mixed 1%
Asian 9%
Black 3%
Other 1%

Patients with INS, IgA nephropathy, Tubulopathies, Cystinosis and Primary Hyperoxaluria **more likely to be from Asian ethnic backgrounds**

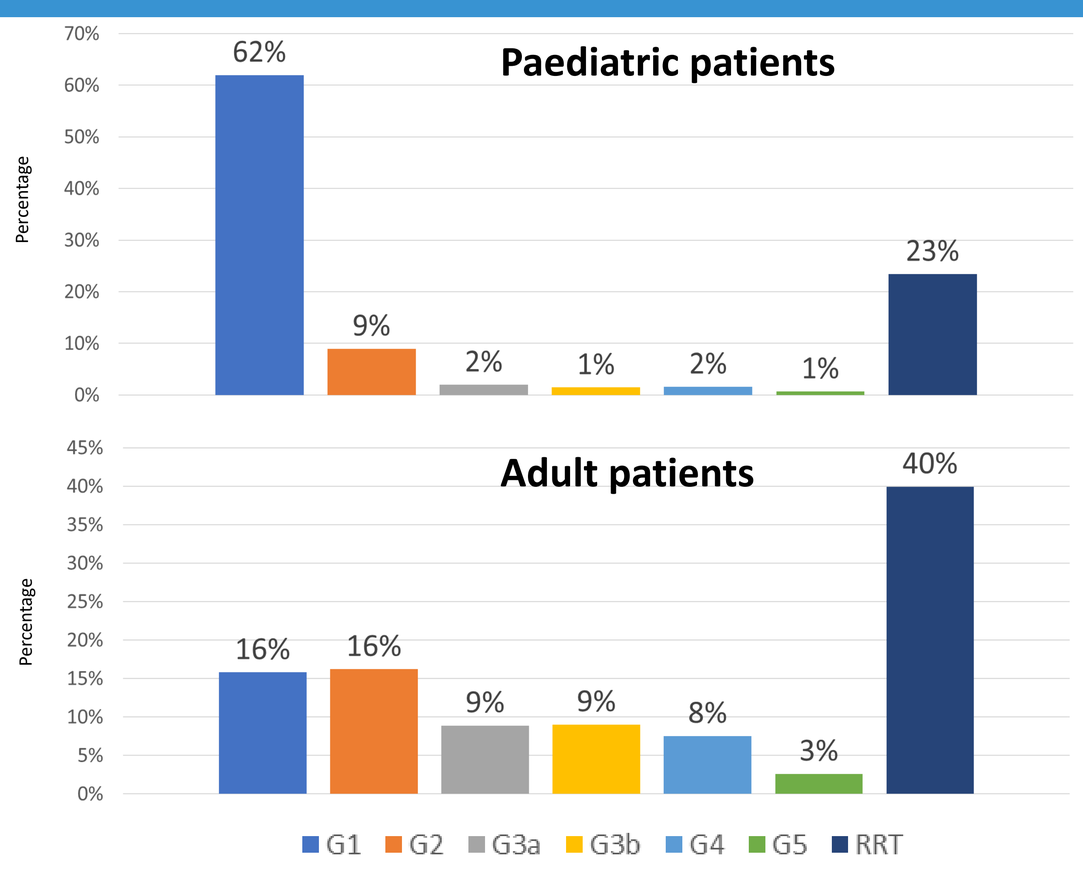
Socioeconomic deprivation (all RaDaR)

Index of Multiple Deprivation Quintile
1- Least deprived- 21%
2- 21%
3- 20%
4- 19%
5- Most deprived- 18%

Patients with Cystinosis, Primary Hyperoxaluria, INS, IgA nephropathy, Membranous Nephropathy and Pregnancy **more likely to be in the most deprived IMD Quintile**

compared to the overall RaDaR population

Renal function and RRT



Methods

- RaDaR is linked with the UKRR for data on RRT initiation and renal IT systems for laboratory data.
- Chi-square or Fishers exact tests were used to compare categorical variables.
- Ethnicity is self-reported. Patient postcodes were used to derive Index of Multiple Deprivation (IMD) scores as an area level measure of socioeconomic status.

Conclusions

- **To our knowledge, this is the largest epidemiological description of a rare disease cohort worldwide**
- We have identified disparities in ethnicity and socioeconomic status between rare diseases, and between children and adults enrolled in RaDaR, which require further study
- Patient numbers, demographics and renal function may be useful in assessing the feasibility of studies or clinical trials in certain rare kidney diseases.

Contact: Katie.Wong@ukkidney.org

UKKIDNEY.ORG

UKKW.ORG