

ADPKD and children

Newcastle

7 March 2015

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ADPKD in CYP

Historical approach

- * It's an adult disease
- * There is no treatment
- * Early scans not everything

Why bother?

Numbers

- 12 million children in the UK
- ADPKD 1 in 1,000
= 12,000 children affected

Is it acceptable to ignore their risk of hypertension and kidney disease?

Referrals to antenatal clinic

1. Family history
2. Babies with cystic kidneys
3. Babies with 'bright' kidneys

1. 50% risk
2. Cystic kidneys are almost never ADPKD
3. Big and Bright increases likelihood
4. Babies are developing so appearances can change (before and after birth)

Anxiety versus uncertainty?

- Great Ormond Street data that there is no difference between children diagnosed before or after birth. About a quarter of each group have high blood pressure
- USA - up to a third have high BP
- USA – small proportion with heart problems

Management of ADPKD in Childhood

A UK survey

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Management of Children with Autosomal Dominant Polycystic Kidney Disease (ADPKD)



We are Paediatric Nephrologists interested in assessing how children affected by ADPKD are currently managed around the UK.

This survey contains questions for two scenarios, followed by a small number of general queries:

- **Scenario 1 concerns a child referred with likely ADPKD**
- **Scenario 2 outlines a similar case where the diagnosis is less certain**

Please feel free to give frank answers – there is probably no correct response for most of these questions at the moment but we hope to develop better guidelines for future management, particularly as new treatments are developed. Free text comments also appreciated.

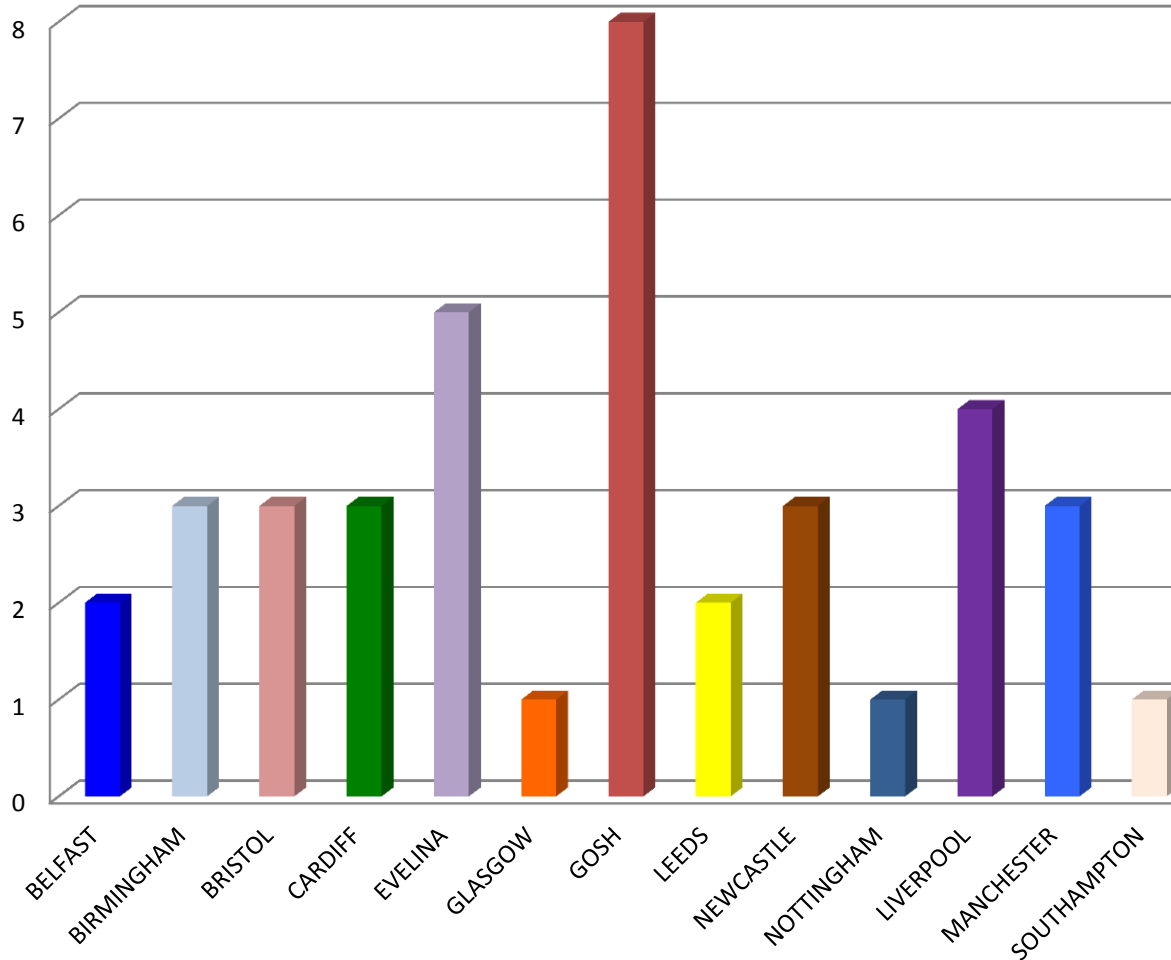
Yours sincerely

Amanda Richardson, Maanasa Polubothu, Manish Sinha, Larissa Kerecuk and Paul Winyard, with the assistance of Tess Harris of the PKD Charity

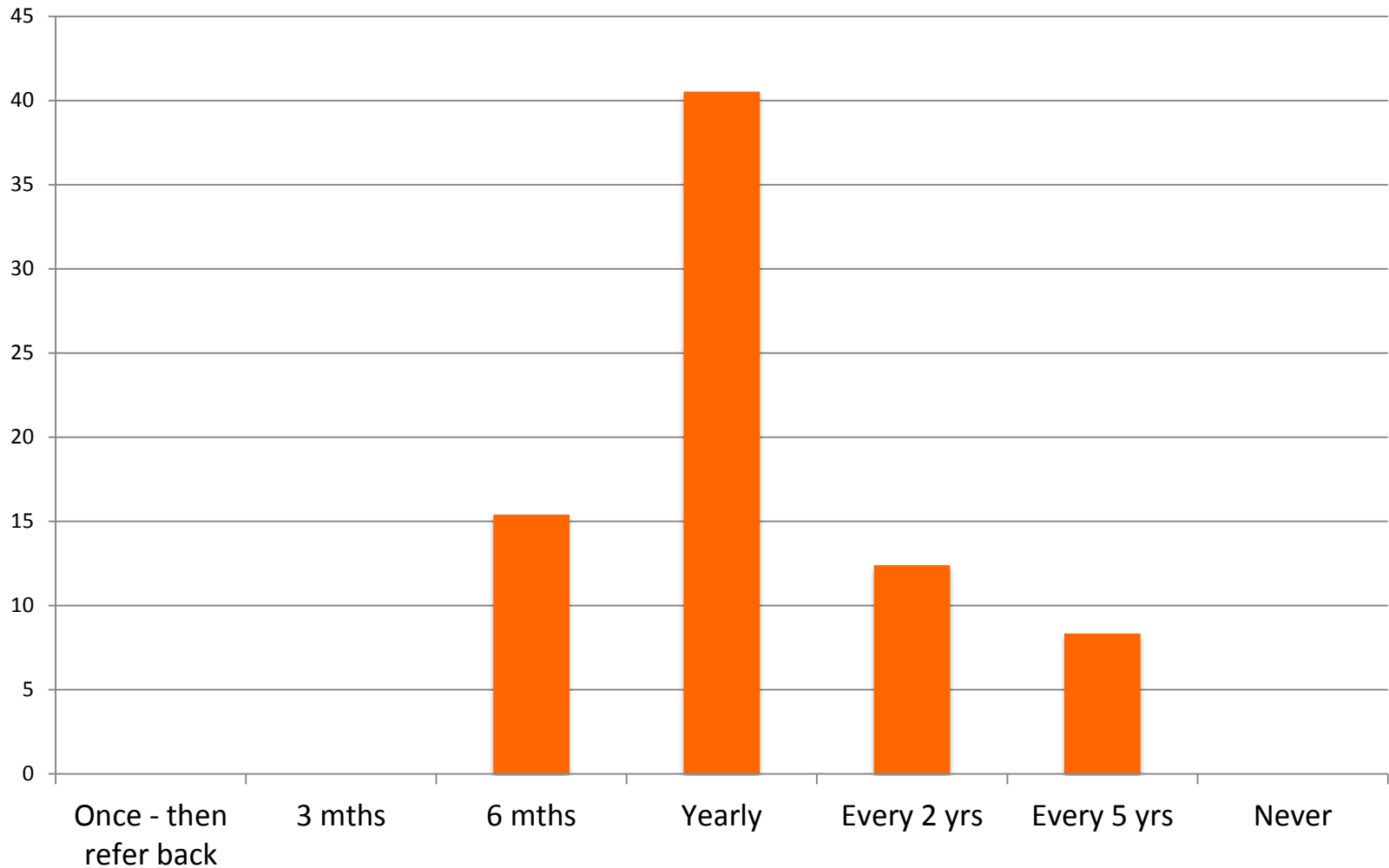
(NB: this is a confidential survey, not open to the public and the results will be kept secure)

Please insert your name or unique study ID to avoid reminders. (optional)

Respondents



How often would you check BP?



Estimated Number of children with at each centre

Centre	Lowest Estimate	Highest Estimate
	15	100
	10	20
	10	15
	20	20
	?	?
	30	50
	5	75
	?	?
	20	50+
	30	40
	25	30
	20	20
	33	33
	20	20
	20 (258)	20 (493)

Summary:

current UK management

- Most centres see children between 1-5 yearly and do routine clinic investigations
- Very few have a set-up to investigate BP or heart status routinely

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Are we doing enough?