

# FH PAEDIATRIC REGISTER NEWSLETTER

THE NEWSLETTER FOR CLINICIANS WHO ARE REGISTERED WITH THE FAMILIAL HYPERCHOLESTEROLAEMIA  
PAEDIATRIC REGISTER WEBSITE

## LATEST FIGURES

We now have **403 children enrolled in the register.**

There are **95 clinicians and 65 hospitals registered** on the system.

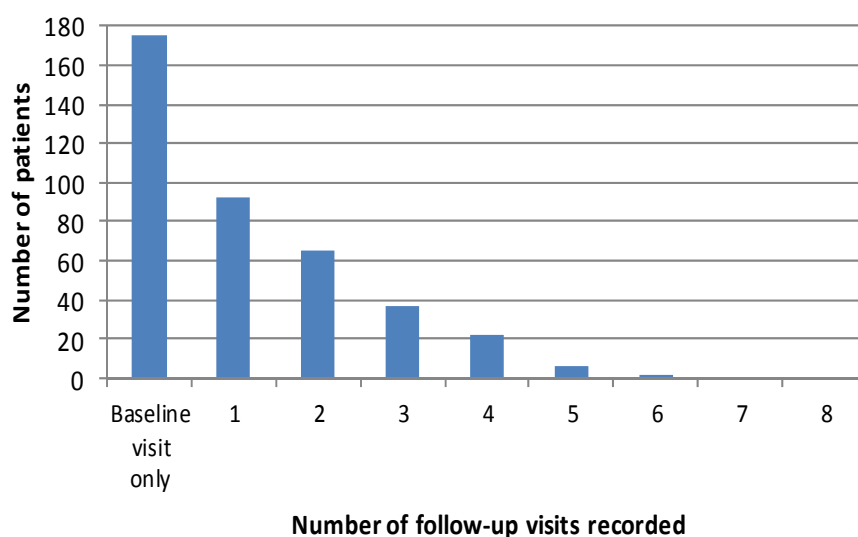
A total of **31 hospitals have entered patient data** into the system.

Since the new introduction of the automated annual update reminder, we continue to see an increase in follow-up data entered. We now have **480 annual updates in the system.**

Do remember that clinicians can continue to enter patient data even once the patient turns 18 years of age, so long as they remain under that clinician's care.

# 403

At Feb 2017, 401 children had been enrolled into the register, of which 226 (56%) had one or more annual follow-up visit recorded. The graph shows the distribution of number of clinic visits recorded in the registry.



## RCPCH Workshop – Birmingham 24<sup>th</sup> May

**Title of Workshop: Paediatric Familial Hypercholesterolaemia - statins - to use or not to use**

**Dr Ramaswami and Lorraine Priestly-Barnham** will be presenting a 50 minute lunchtime workshop at the RCPCH Spring Meeting. This short workshop aims to provide an overview of FH genetics, statins - indication and safety: data from the UK paediatric FH, cascade screening, diagnosis and management of children with FH.

**Session Chair:** Dr Saikat Santra, Consultant in Inherited Metabolic Disorders, Birmingham Children's Hospital

### 13:10 Overview of FH genetics

**Speaker:** **Dr. Robert Cramb**, Director of Chemical Pathology, University Hospitals Birmingham NHS Foundation Trust

### 13.20 to 13.30 Indications and safety of Statins - data from the UK paediatric FH register.

**Speaker:** Dr Uma Ramaswami, Consultant in Inherited Metabolic Disorders; Royal Free Hospital and Clinical Lead for the UK Paediatric FH Register

### 13:30 - 13.45 Managing children with FH and cascade screening: The Harefield experience

**Speaker:** Ms Lorraine Priestley-Barnham, Clinical Nurse Specialist in FH and Cascade Screening Harefield Hospital, London, UK

### 13.45 to 13.55 Q&A

Chaired by Drs Saikat Santra and Uma Ramaswami

### 13.55: Session close

## What information and apps would children with FH find useful?

As you know, the Register is funded for the next 2 years by a grant from the International Atherosclerosis Society, and part of the work will support the production of information materials for children and young people. However we realised that we don't really know what sort of things young people would like! To try to get this information, a completely secure and anonymous on-line survey has been developed to gather views from children on what they would like to have. Children and young people are needed to share their views on what information they want about FH, such as games, apps, booklets. Please encourage any children with FH to complete this online survey to help gather views. For 7-11 year olds link here: <http://dpll.co/adb796a> and 12-16 year olds link here: <http://dpll.co/5285167>. You can also try these surveys yourselves to see what sorts of questions are being asked, or you can email [Steve.humphries@ucl.ac.uk](mailto:Steve.humphries@ucl.ac.uk) to send you the questions in a printable one-sheet format that could be filled in at the next clinic visit.

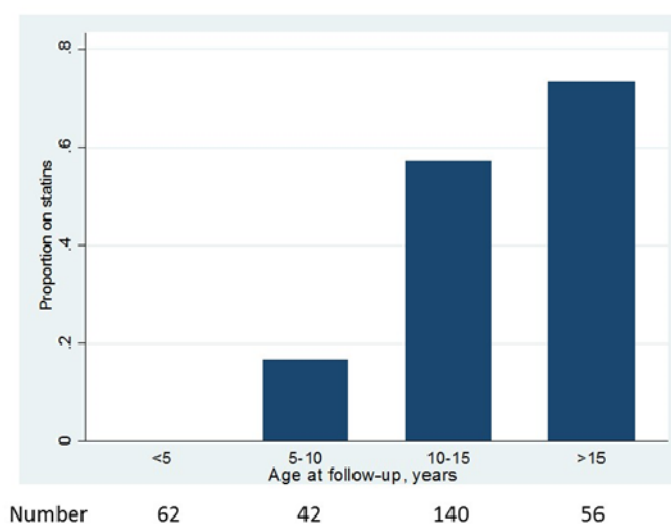
## Statin Information Sheet

We are aware that one of the reasons why an eligible child may not be on a statin is because of parental concern about the safety record of the drugs, especially if they have been reading reports in the lay press. To try to counter this we have developed a statin information sheet specifically to answer parent's questions and address their concerns. We have had very useful comments from colleagues on the Steering Committee including parent representative and our Paediatric Nurse. The final version is currently undergoing scrutiny by the Information Committee at the British Inherited Metabolic Disease Group, and once approved we will let you know and put it on the Register website so you can download it and use it in your clinics.

## Statin treatment in the Register children

Analysis of the current data shows that overall 52.5% of the children were on statins, but this varied significantly by age group, being 0% in those under 5 years, 16.7% in those between 5 and 10 years, 57.1% in those between 10 and 15 years and 73.2% in those over 15 years. Overall, statin treatment reduced LDL-C level by 31% (1.84mmol/L), but with 55.6% still having levels over the suggested target of 3.5mmol/l. In children over 10 years, of those not on statin treatment, 36% had evidence of a family history of early coronary heart disease (CHD), and 89.9% had LDL-C over 3.5mmol/l.

Proportion of children on statin therapy by age group.



Although these results are encouraging they suggest that there are still a significant proportion of children over the age of 10 years who are at high future risk of early CHD and where initiation of statin therapy should be actively considered.