

# FH PAEDIATRIC REGISTER NEWSLETTER

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

## The interactive map is here...

An interactive map which displays the **names of our UK registered clinicians**, along with their hospital contact details is now available on the register home page (before log-in). We hope that this will be a useful resource for other clinicians with an interest in FH. If you wish to **opt-out of having your name/hospital contact details** displayed on the map, please contact us by email: [fh@rcplondon.ac.uk](mailto:fh@rcplondon.ac.uk)



## Pfizer Grant awarded to the Register team!

We are delighted to announce that we have been successful in our application for an International Atherosclerosis Society (IAS) and Pfizer Independent Grant For Learning & Change (IGLC)! The grant is for \$200,000 US dollars and will be used to extend the UK Register into a fully integrated and collaborative International Register. We have already been approached by an interested site in New Zealand who heard our news, and we are now working hard to commence roll out this hugely important project to interested countries **this year**. We're going international!

## STOP PRESS

The first publication of register data has been achieved! The article entitled 'The UK Paediatric Familial Hypercholesterolaemia Register: preliminary data' was published in Archives of Disease in Childhood earlier this year. The citation and a copy of the abstract can be found on page two of the newsletter.

## LATEST FIGURES

We now have **361 children enrolled in the register**, an 11% increase from the 326 patients enrolled at the end of 2015.

There are **95 clinicians and 62 NHS Trusts registered** on the system, and we welcome 2 new sites in 2016: Royal Victoria Hospital, Belfast and Ysbyty Gwynedd, Bangor.

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 **343 annual updates in the system**, an increase from 248 (38%) reported in the Dec 2015 newsletter.

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.



**Heart UK's 30th Annual conference** will be held in Edinburgh on **6-8th July**. Prof. Humphries & Dr Ramaswami will be leading a Paediatrics and Hyperlipidaemia study day including sessions on:

- ◆ Paediatric FH—Statins: to use or not to use?
- ◆ Statin Treatment in children: when to start and how low is low enough?
- ◆ What proportion of the UK FH register children are treated and how low are their LDL levels?
- ◆ Managing children with FH—in England and in Wales

## Reminder

Prof Humphries laboratory is no longer able to offer a free DNA testing service due to staff retirement. He apologises for the slow turnaround of results for samples received already and hopes to complete analyses in the next few weeks.

See page 2 of the newsletter for the first publication of UK Register data...

**The first publication of FH Paediatric Register data:**

**The UK Paediatric Familial Hypercholesterolaemia Register: preliminary data.** *Arch Dis Child.* 2016 Mar 6. pii: archdischild-2015-308570. doi: 10.1136/archdischild-2015-308570. [Epub ahead of print].

**Abstract:**

**BACKGROUND:**

The National Institute for Health and Care Excellence 2008 guidelines on the treatment and management of familial hypercholesterolaemia (FH) recommend that children with FH should be considered for statin treatment by the age of 10 years. The Paediatric FH Register was established in 2012 to collect baseline and long-term follow-up data on all children with FH in the UK.

**METHODS:**

Paediatricians and adult lipidologists have been invited to enter baseline data on any child with a clinical diagnosis of FH using an electronic capture record.

**RESULTS:**

Baseline data is on 232 children (50% boys, 80% Caucasian), with an untreated mean (SD) total cholesterol of 7.61 (1.48) mmol/L and low-density lipoprotein cholesterol (LDL-C) of 5.67 (1.46) mmol/L. Overall 111/232 (47.8%) of the children were on statins. Children over the age of 10 years at the most recent follow-up were twice as likely to be on statin treatment than those under 10 years (57.6% (102/177) vs 23.1% (9/39),  $p=0.00009$ ). In both age groups, those subsequently on statin treatment had significantly higher diagnostic total and LDL-C (overall 6.01 (1.46) mmol/L vs 5.31 (1.37) mmol/L,  $p=0.00007$ ), and had stronger evidence of a family history of early coronary heart disease (CHD) in parent or first-degree relative (overall 28.4% vs 19.0%,  $p=0.09$ ). In statin-treated children LDL-C level was reduced by 35% (2.07 (1.38) mmol/L) compared with a reduction of 5.5% (0.29 (0.87) mmol/L),  $p=0.0001$  in those not treated. None of those on statin had measured plasma levels of creatine kinase, alanine aminotransferase and AST indicative of statin toxicity (ie,  $>2.5$  times the upper limit of the normal range).

**CONCLUSIONS:**

The data indicates that treatment decisions in children with FH are appropriately based on a stronger family history of CHD and higher LDL-C.