

## Paediatric Familial Hypercholesterolaemia Pathway

For Children and Young People <16 years

FH identified in 1st degree relative  
(parent or sibling)

**Offer Specific Mutation DNA  
test from age 5 (4 cheek  
swabs or blood test)**

**FH Genotype  
NEGATIVE**

No further investigations required  
**INFORM GP**

The aim of management of FH is to reduce the risk of cardiovascular disease whilst ensuring adequate growth and development.

Lifestyle modifications alone are unlikely to lower cholesterol concentration adequately in heterozygous FH and drug treatment is often required. **Lipid regulating drugs should be considered by age of 10.**

When deciding to defer or offer lipid-modifying drug therapy, take into account:

- Their age
- Age of onset of CHD within the family
- The presence of other CVD risk factors

In exceptional circumstances (where there is a family history of CHD in early adulthood) consider offering:

- A higher dose of statin than is licensed
- for use in appropriate age group and/or More than 1 lipid-modifying drug therapy

**REFER TO A SPECIALIST IF STATINS NOT TOLERATED**

**FH Genotype  
POSITIVE**

Take **fasting lipids** and **LFTs** at **diagnosis** (if not taken with DNA test) and again **pre-treatment**

Inform Metabolic Paediatric Specialist Nurse at NHSGGC and agree treatment plan before meeting with family (see treatment advice)

FH Nurse to arrange Family appointment to discuss diet and lifestyle and provide treatment advice

### **PRE-TREATMENT YEARS**

GP to monitor growth, development and lifestyle annually

Consider statin by the age of 10 years with advice of Metabolic Paediatric Nurse. (Measure fasting lipids & LFTs)

### **POST-TREATMENT**

Fasting lipids & LFTs at 3 months to achieve reduction in LDL of at least 50% (CK if reported muscle aches)

**REVIEW ANNUALLY** measuring growth, development & lifestyle

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