

## F) Children

### If I have FH will my child have it as well?

FH is a genetic disorder that affects 1 in 500 of the population. It is an autosomal dominant disorder, which means that if you or your partner has the disorder, and then your child has a fifty/fifty chance of inheriting it. This means that on average one in two children would be expected to be affected, although sometimes none of the children will inherit the disorder.

If you and your partner do not have the disorder then your child will not have it either (unless of course the child is adopted).

### At what age can my child be tested?

The British Hyperlipidaemia Association (BHA) recommends that children from affected families should be tested before the age of ten, but should not normally be tested before the age of two years.

### Will my child have to be tested again if the result is normal for his or her age?

During his or her early years your child should not require testing again if the test result is normal. Your child may need testing again in early adulthood if familial combined hyperlipidaemia is suspected because the lipid abnormalities in this case only present later in life. Also, it may be a good idea to have occasional checks on cholesterol levels in later life, certainly if changes in risk for developing heart disease occur. This could include smoking or developing high blood pressure for instance.

### Do we need to have a genetic diagnosis to be absolutely sure?

Genetic diagnosis may become more important as different methods of treating the disorder are explored. Genetic testing can be performed at specialist centres, but it is expensive and not routinely available. It involves one affected family member having a blood sample taken to identify the specific genetic mutation. Once the defect has been identified, other family members need only supply a sample taken from a swab of the inside of the mouth (the buccal mucosa). This sample provides enough information to determine whether the genetic defect is there or not. However, if the result is negative this does not mean that your child will have no chance of developing coronary heart disease (CHD) in the future. This is because CHD is caused by a multitude of risk factors. If, for example, your child smokes and has diabetes but a negative test for FH, he or she is still at risk for CHD because of the other risk factors.

### Where should my child be cared for?

Affected children and possibly affected children should be referred for specialist hospital follow-up.

### What is the treatment for FH?

Diet is the cornerstone of treatment for this age group. A healthy diet is encouraged from as early as two years of age. This should include a variety of foods from all food groups, including lean meats, low fat dairy products, fish, nuts, legumes, cereals, fruit and vegetables. (See H·E·A·R·T UK diet sheet for further information.) However, care must be taken to ensure that enough calories are taken to ensure healthy growth and development. Parents should seek advice from a state registered dietitian experienced in lipid management.

Plant sterol and stanol spreads have recently been introduced and early clinical trial results have indicated that these products are safe, effective and palatable for children to use and help to reduce LDL cholesterol by up to 15%. Your doctor may therefore recommend their use as part of a cholesterol-lowering diet, but you should seek advice before using them.

General healthy lifestyle options should be encouraged such as discouraging smoking and taking physical exercise above and beyond what is offered at school.



## What are resins?

Usually the main drug used to help treat children to lower their cholesterol is called a resin. This works by binding with the excess cholesterol as it passes through the gut. The resin plus excess cholesterol is then excreted in the stools. Unfortunately the resins are not particularly palatable and sometimes cause gastro-intestinal side effects, which can include diarrhoea and flatulence.

## What are statins?

Later on, after puberty, statin drug therapy may be required for older children, when there is particular risk of coronary heart disease in the family.

Statin therapy is less often indicated in girls than boys since coronary risk in early adulthood is much lower in women. Later in adulthood drug treatment will almost invariably be required in all patients.

## How do they work?

90% of cholesterol is made in the liver. Statin drugs work by inhibiting the enzyme reaction that occurs in the liver in this production process. Therefore, whilst taking the tablets production is reduced and if you stop taking them cholesterol manufacture goes back up to its' previous levels.

## What are the side effects of these drugs?

Clinical trials in adults indicate that serious side effects are very rare and the drugs are well tolerated. The minor side effects of these drugs can include headaches and tummy upsets (diarrhoea and constipation). However, should your child develop general muscle aches and pains then he/she should report this to the doctor. It may be necessary to stop the medication and possibly select an alternative tablet at a later date.

Parents should be aware that there is not a great deal of information available on the safety and use of these tablets in children. Your hospital specialist should discuss this with you.

Pregnancy should be avoided whilst taking statins and appropriate contraceptive therapy recommended.

## Will he or she always have to take them?

Yes, until gene therapy or something similar in the future is developed.

## Will my children's children inherit the disorder?

If your child has not inherited the problem then your grandchildren will not either. However, if your child does have FH then their children will have a 50/50 chance of inheriting the disorder.

## What if he/she doesn't want the test?

It is important to know whether your child is affected or not. It is often possible in specialist clinics to make the diagnosis with a finger prick test if your child is particularly frightened by the prospect of a needle and syringe

## What if my GP won't test my child?

Most GPs are sympathetic of parents' concerns. Usually taking along information about the condition and information (such as this leaflet) can help GPs decide to test your child or refer you to a specialist centre for testing.

