Familial hypercholesterolaemia: identification and management

NICE guideline: short version

Draft for consultation, May 2017

This guideline covers the identification and management of familial hypercholesterolaemia in children, young people and adults. It does not cover other forms of hypercholesterolaemia that are not genetic (inherited) or that are due to other genetic conditions.

Who is it for?

- Healthcare professionals who care for people with familial hypercholesterolaemia
- People with familial hypercholesterolaemia, and their families and carers

This guideline will update NICE guideline CG71 (published August 2008).

We have updated or added new recommendations on the identification, diagnosis and treatment of familial hypercholesterolaemia.

You are invited to comment on the new and updated recommendations in this guideline. These are marked as [2017] if the evidence has been reviewed, and the recommendation has been added or updated, or the evidence has been reviewed but no change has been made to the recommended action. Where recommendations end [2008], [2008, amended 2017] or [2016], the evidence has not been reviewed.
You are also invited to comment on recommendations that NICE proposes to delete from the 2008 guideline.

We have not updated recommendations shaded in grey, and cannot accept comments on them. In some cases, we have made minor wording changes for clarification.

See Update information for a full explanation of what is being updated.

This version of the guideline contains the draft recommendations, context and recommendations for research. Information about how the guideline was developed is on the guideline’s page on the NICE website. The supporting information and evidence for the 2017 recommendations is contained in the addendum.
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### Recommendations

People have the right to be involved in discussions and make informed decisions about their care, as described in your care.

[Making decisions using NICE guidelines](#) explains how we use words to show the strength (or certainty) of our recommendations, and has information about prescribing medicines (including off-label use), professional guidelines, standards and laws (including on consent and mental capacity), and safeguarding.

#### 1.1 Case finding and diagnosis

See also section 1.4 on ‘Information needs and support’.

1.1.1 Think about familial hypercholesterolaemia (FH) as a possible diagnosis in adults with:

- a total cholesterol level greater than 7.5 mmol/l, and/or
- a personal or family history of premature coronary heart disease (a coronary event before 60 years in an index individual or first-degree relative). [2008, amended 2017]

1.1.2 Systematically search primary care records for people with a total cholesterol concentration greater than 9.3 mmol/l, as these are the people who are at highest risk of FH. [2017]

1.1.3 For people with a personal or family history of premature coronary heart disease (a coronary event before 60 years in an index individual or first-
degree relative), but whose total cholesterol is unknown, offer to measure their total cholesterol. [2017]

1.1.4 Healthcare professionals should exclude secondary causes of hypercholesterolaemia before a diagnosis of FH is considered. [2008]

1.1.5 Use the Simon Broome or Dutch Lipid Clinic Network (DLCN) criteria to make a clinical diagnosis of FH in primary care settings. This should be done by a healthcare professional competent in using the criteria. [2017]

1.1.6 Refer the person to an FH specialist service for DNA testing if they meet the Simon Broome criteria for possible or definite FH, or they have a DLCN score greater than 5. [2017]

1.1.7 Healthcare professionals should consider a clinical diagnosis of homozygous FH in adults with a low-density lipoprotein cholesterol (LDL-C) concentration greater than 13 mmol/l and in children/young people with an LDL-C concentration greater than 11 mmol/l. All people with a clinical diagnosis of homozygous FH should be offered referral to a specialist centre. [2008]

1.1.8 To confirm a diagnosis of FH, healthcare professionals should undertake two measurements of LDL-C concentration because biological and analytical variability occurs. [2008]

1.1.9 Healthcare professionals should be aware that the absence of clinical signs (for example, tendon xanthomata) in adults and children/young people does not exclude a diagnosis of FH. [2008]

1.1.10 When considering a diagnosis of FH, healthcare professionals with expertise in FH should use standardised pedigree terminology to document, when possible, at least a three-generation pedigree. This should include relatives' age of onset of coronary heart disease, lipid concentrations and smoking history. For deceased relatives, the age and cause of death, and smoking history should be documented. If possible,
1. the index individual should verify this information with other family members. [2008]

1.1.11 Ultrasonography of the Achilles tendon is not recommended in the diagnosis of FH. [2008]

1.1.12 Coronary heart disease risk estimation tools such as those based on the Framingham algorithm should not be used because people with FH are already at a high risk of premature coronary heart disease. [2008]

1.1.13 Inform all people who have an identified mutation diagnostic of FH that they have an unequivocal diagnosis of FH even if their LDL-C concentration does not meet the diagnostic criteria (see recommendation 1.1.5). [2008, amended 2017]

1.1.14 In a family where a DNA mutation is identified, not all family members may have inherited the mutation. When DNA testing has excluded FH in a member of a family, healthcare professionals should manage the person’s coronary heart disease risk as in the general population1. [2008]

1.1.15 In children at risk of FH because of one affected parent, offer a DNA test by the age of 10 years or at the earliest opportunity thereafter. [2017]

1.1.16 In children at risk of homozygous FH because of two affected parents or because of the presence of clinical signs, for example, cutaneous lipid deposits (xanthomata), LDL-C concentration should be measured before the age of 5 years or at the earliest opportunity thereafter. If the LDL-C

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1 ‘Cardiovascular disease: risk assessment and reduction, including lipid modification’ (NICE clinical guideline 181).
concentration is greater than 11 mmol/l then a clinical diagnosis of homozygous FH should be considered. [2008]

1.2 Identifying people with FH using cascade testing

1.2.1 Carry out cascade testing using DNA testing to identify affected first- and second- and, when possible, third-degree biological relatives of people with a diagnosis of FH. [2017]

1.2.2 Healthcare professionals should offer all people with FH a referral to a specialist with expertise in FH for confirmation of diagnosis and initiation of cascade testing. [2008]

1.2.3 Healthcare professionals with expertise in FH should explain what is meant by cascade testing, and discuss its implications with all people with FH. [2008]

1.2.4 Healthcare professionals should be aware of the latest guidance on data protection when undertaking cascade testing. [2008]

1.3 Management

1.3.1 Drug treatment

Adults

1.3.1.1 When offering lipid-modifying drug therapy to adults with FH, healthcare professionals should inform the person that this treatment should be lifelong. [2008]

1.3.1.2 Offer a high-intensity statin with the lowest acquisition cost as the initial treatment for all adults with FH and aim for at least a 50% reduction in LDL-C concentration from the baseline measurement. [2017]

1.3.1.3 The dose of statin should be increased to the maximum licensed or tolerated dose to achieve a recommended reduction in LDL-C concentration of greater than 50% from baseline (that is, LDL-C concentration before treatment). [2008]
1.3.1.4 Ezetimibe monotherapy is recommended as an option for treating primary heterozygous-familial hypercholesterolaemia in adults in whom initial statin therapy is contraindicated. [2016]

1.3.1.5 Ezetimibe monotherapy is recommended as an option for treating primary heterozygous-familial hypercholesterolaemia in adults who cannot tolerate statin therapy (as defined in recommendation 1.3.1.9). [2016]

1.3.1.6 Ezetimibe, co-administered with initial statin therapy, is recommended as an option for treating primary (heterozygous-familial) hypercholesterolaemia in adults who have started statin therapy when:

- serum total or low-density lipoprotein (LDL) cholesterol concentration is not appropriately controlled (as defined in recommendation 1.3.1.8)

  either after appropriate dose titration of initial statin therapy or because dose titration is limited by intolerance to the initial statin therapy (as defined in recommendation 1.3.1.9)

  and

- a change from initial statin therapy to an alternative statin is being considered. [2016]

1.3.1.7 When prescribing ezetimibe co-administered with a statin, ezetimibe should be prescribed on the basis of lowest acquisition cost. [2016]

1.3.1.8 For the purposes of this guidance, appropriate control of cholesterol concentrations should be based on individualised risk assessment according to national guidance on managing cardiovascular disease in the relevant populations. [2016]

1.3.1.9 For the purposes of this guidance, intolerance to initial statin therapy is defined as the presence of clinically significant adverse effects that represent an unacceptable risk to the patient or that may reduce compliance with therapy. [2016]

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2 This recommendation has been adapted from ‘Ezetimibe for treating primary (heterozygous-familial and non-familial) hypercholesterolaemia’ (NICE technology appraisal guidance 385).
1.3.1.10 Prescribing of drug therapy for adults with homozygous FH should be undertaken within a specialist centre. [2008]

1.3.1.11 Healthcare professionals should offer adults with FH a referral to a specialist with expertise in FH if treatment with the maximum tolerated dose of a high-intensity statin and ezetimibe does not achieve a recommended reduction in LDL-C concentration of greater than 50% from baseline (that is, LDL-C concentration before treatment). [2008]

1.3.1.12 Healthcare professionals should offer adults with FH a referral to a specialist with expertise in FH for consideration for further treatment if they are assessed to be at very high risk of a coronary event, that is, if they have any of the following.

- Established coronary heart disease.
- A family history of premature coronary heart disease.
- Two or more other cardiovascular risk factors (for example, they are male, they smoke, or they have hypertension or diabetes). [2008]

1.3.1.13 Adults with FH with intolerance or contraindications to statins or ezetimibe should be offered a referral to a specialist with expertise in FH for consideration for treatment with either a bile acid sequestrant (resin), nicotinic acid, or a fibrate to reduce their LDL-C concentration. [2008]

1.3.1.14 The decision to offer treatment with a bile acid sequestrant (resin), nicotinic acid or a fibrate in addition to initial statin therapy should be taken by a specialist with expertise in FH. [2008]

1.3.1.15 Healthcare professionals should exercise caution when adding a fibrate or nicotinic acid to a statin because of the risk of muscle-related side effects (including rhabdomyolysis). Gemfibrozil and statins should not be used together. [2008]

1.3.1.16 Adults with FH who are prescribed nicotinic acid should be offered advice on strategies that reduce flushing. Such advice should include taking low
initial doses with meals and/or aspirin 30 minutes before the first daily
dose. [2008]

Children and young people

1.3.1.17 Healthcare professionals should offer all children and young people
diagnosed with, or being investigated for, a diagnosis of FH a referral to a
specialist with expertise in FH in children and young people. This should
be in an appropriate child/young person-focused setting that meets the
standards within the ‘National service framework for children, young
people and maternity services’. [2008]

1.3.1.18 Lipid-modifying drug therapy for a child or young person with FH should
usually be considered by the age of 10 years. The decision to defer or
offer lipid-modifying drug therapy for a child or young person should take
into account:

- their age
- the age of onset of coronary heart disease within the family, and
- the presence of other cardiovascular risk factors, including their LDL-C
concentration. [2008]

1.3.1.19 When offering lipid-modifying drug therapy for children or young people,
healthcare professionals should inform the child/young person and their
parent/carer that this treatment should be lifelong. [2008]

1.3.1.20 Offer statins to children with FH by the age of 10 years or at the earliest
opportunity thereafter. [2017]

1.3.1.21 For children and young people with FH, consider a statin that is licensed
for use in the appropriate age group. [2017]

1.3.1.22 Statin therapy for children and young people should be initiated by a
healthcare professional with expertise in treating children and young
people with FH, and in a child-focused setting. [2008, amended 2017]
1.3.1.23 Statin therapy for children and young people with FH should usually be prescribed at the doses specified in the ‘British national formulary (BNF) for children’. [2008]

1.3.1.24 In exceptional instances, for example, when there is a family history of coronary heart disease in early adulthood, healthcare professionals with expertise in FH in children and young people should consider offering:

- a higher dose of statin than is licensed for use in the appropriate age group, and/or
- more than one lipid-modifying drug therapy, and/or
- lipid-modifying drug therapy before the age of 10 years. [2008]

1.3.1.25 In children and young people with homozygous FH, LDL-C concentration may be lowered by lipid-modifying drug therapy and this should be considered before LDL apheresis (see section 1.3.3). [2008]

1.3.1.26 In children and young people with FH who are intolerant of statins, healthcare professionals should consider offering other lipid-modifying drug therapies capable of reducing LDL-C concentration (such as bile acid sequestrants [resins], fibrates or ezetimibe). [2008]

1.3.1.27 Routine monitoring of growth and pubertal development in children and young people with FH is recommended. [2008]

Adults and children/young people

1.3.1.28 Decisions about the choice of treatment should be made following discussion with the adult or child/young person and their parent/carer, and be informed by consideration of concomitant medication, comorbidities, safety and tolerability. [2008]

1.3.1.29 Healthcare professionals should consider offering fat-soluble vitamin (vitamins A, D and K) and folic acid supplementation for adults or children/young people with FH who are receiving long-term treatment with bile acid sequestrants (resins). [2008]
1.3.1.30 Healthcare professionals should offer people with FH a referral to a specialist with expertise in FH if they are experiencing side effects that compromise concordance with lipid-modifying drug therapy. [2008]

1.3.1.31 When the decision has been made to offer adults or children/young people with FH treatment with a statin, baseline liver and muscle enzymes (including transaminases and creatine kinase, respectively) should be measured before initiation of therapy. However, people with raised liver or muscle enzymes should not routinely be excluded from statin therapy. [2008]

1.3.1.32 Routine monitoring of creatine kinase is not recommended in asymptomatic adults or children/young people with FH who are receiving treatment with a statin. [2008]

1.3.2 Lifestyle interventions

1.3.2.1 Healthcare professionals should regard lifestyle advice as a component of medical management, and not as a substitute for lipid-modifying drug therapy. [2008]

Diet

1.3.2.2 All people with FH should be offered individualised nutritional advice from a healthcare professional with specific expertise in nutrition. [2008]

1.3.2.3 People with FH should be advised to consume a diet in which:

- total fat intake is 30% or less of total energy intake
- saturated fats are 10% or less of total energy intake
- intake of dietary cholesterol is less than 300 mg/day
- saturated fats are replaced by increasing the intake of monounsaturated and polyunsaturated fats.

It may be helpful to suggest they look at Live Well for further practical advice. [2008]
1.3.2.4 Healthcare professionals should advise people with FH to eat at least five portions of fruit and vegetables a day, in line with national guidance for the general population. Examples of what constitutes a portion can be found at Live Well. [2008]

1.3.2.5 Healthcare professionals should advise people with FH to consume at least two portions of fish a week (one of which should be oily fish). Pregnant women with FH should be advised to limit their oily fish to two portions a week. Further information and advice on healthy cooking methods can be found at Live Well. [2008]

1.3.2.6 Healthcare professionals should advise people with FH that if they wish to consume food products containing stanols and sterols these need to be taken consistently to be effective. [2008]

1.3.2.7 People with FH should not routinely be recommended to take omega-3 fatty acid supplements. For people with FH who have already had a myocardial infarction (MI), refer to the NICE guideline on myocardial infarction. [2008]

Physical activity

1.3.2.8 Healthcare professionals should advise people with FH to take at least 30 minutes of physical activity a day, of at least moderate intensity, at least 5 days a week, in line with national guidance for the general population. [2008]

1.3.2.9 Healthcare professionals should encourage people with FH who are unable to perform moderate-intensity physical activity at least 5 days a week because of comorbidity, disability, medical conditions or personal circumstances to exercise at their maximum safe capacity. [2008]

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3 See ‘At least five a week: evidence on the impact of physical activity and its relationship to health. A report from the Chief Medical Officer’ (2004).
1.3.2.10 Recommended types of physical activity include those that can be incorporated into everyday life, such as brisk walking, using stairs and cycling. [2008]

1.3.2.11 Healthcare professionals should advise people with FH that bouts of physical activity of 10 minutes or more accumulated throughout the day are as effective as longer sessions. [2008]

**Weight management**

1.3.2.12 Healthcare professionals should offer people with FH who are overweight or obese appropriate advice and support to achieve and maintain a healthy weight in line with NICE guidance on obesity. [2008]

**Alcohol consumption**

1.3.2.13 As for the general population, alcohol consumption for adult men with FH should be limited to up to 3–4 units a day, and for adult women with FH up to 2–3 units of alcohol a day. Binge drinking should be avoided. Further information can be found at Live Well. [2008]

**Smoking advice**

1.3.2.14 People with FH, especially children, who do not smoke should be strongly discouraged from starting because of their already greatly increased risk of coronary heart disease. [2008]

1.3.2.15 People with FH who smoke should be advised that, because of their already greatly increased risk of coronary heart disease, they should stop. [2008]

1.3.2.16 Healthcare professionals should offer people who want to stop smoking support and advice, and referral to an intensive support service, in line with the NICE guidance on smoking cessation. [2008]

1.3.2.17 People with FH who are unwilling or unable to accept a referral to an intensive support service should be offered pharmacotherapy in line with

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4 ‘Obesity prevention’ (NICE guideline CG43).
5 ‘Smoking: brief interventions and referrals’ (NICE guideline PH1).
1.3.3 Specialist treatment

LDL-lowering apheresis

1.3.3.1 Healthcare professionals should consider offering LDL apheresis for the treatment of adults and children/young people with homozygous FH (see recommendations 1.1.7 and 1.1.16). The timing of initiation of LDL apheresis should depend on factors such as the person’s response to lipid-modifying drug therapy and presence of coronary heart disease. [2008]

1.3.3.2 In exceptional instances (such as when there is progressive, symptomatic coronary heart disease, despite maximal tolerated lipid-modifying drug therapy and optimal medical and surgical therapy), healthcare professionals should consider offering LDL apheresis for the treatment of people with heterozygous FH. This should take place in a specialist centre on a case-by-case basis and data recorded in an appropriate registry. [2008]

1.3.3.3 Healthcare professionals should recommend arterio-venous fistulae as the preferred method of access for people with FH who are offered treatment with LDL apheresis. People should be counselled about possible benefits and complications of this procedure. [2008]

1.3.3.4 Routine monitoring of the person’s iron status should be carried out and iron supplementation initiated as required for people with FH who are receiving treatment with LDL apheresis. [2008]

1.3.3.5 Angiotensin-converting enzyme (ACE) inhibitors should not be used in people with FH who are being treated with LDL apheresis. Instead, ACE

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6 ‘The clinical effectiveness and cost effectiveness of bupropion (Zyban) and nicotine replacement therapy for smoking cessation’ (NICE technology appraisal guidance 39).
7 ‘Varenicline for smoking cessation’ (NICE technology appraisal guidance 123).
inhibitors should be substituted with angiotensin-receptor blocking agents. [2008]

1.3.3.6 People with FH who are receiving blood pressure-lowering drug therapy should have this reviewed and considered for discontinuation on the morning of the day of LDL apheresis. [2008]

1.3.3.7 People with FH who are taking warfarin should have this discontinued approximately 4 days before LDL apheresis and substituted with low molecular weight heparin. [2008]

1.3.3.8 People with FH who are receiving anti-platelet therapy should have this continued if they are receiving treatment with LDL apheresis. [2008]

Liver transplantation

1.3.3.9 Healthcare professionals should consider offering liver transplantation as an option for the treatment of people with homozygous FH after treatment with lipid-modifying drug therapy and LDL apheresis. [2008]

1.3.3.10 The decision to refer for liver transplantation should take place in partnership with the patient and/or their relatives in an appropriate specialist setting, following a discussion of the benefits and potential harms of undertaking or declining transplantation. [2008]

1.4 Information needs and support

1.4.1 General information and support

1.4.1.1 During the assessment and communication of familial risk, people should receive clear and appropriate educational information about FH, the process of family testing, DNA testing and the measurement of LDL-C concentration. [2008]

1.4.1.2 A healthcare professional with expertise in FH should provide information to people with FH on their specific level of risk of coronary heart disease, its implications for them and their families, lifestyle advice and treatment options. [2008]
1.4.1.3 Healthcare professionals with expertise in FH should encourage people with FH to contact their relatives to inform them of their potential risk and so that cascade testing can take place. [2008]

1.4.1.4 When considering cascade testing, a healthcare professional with expertise in FH should offer to facilitate the sharing of information about FH with family members. [2008]

1.4.1.5 Healthcare professionals should offer people with FH and their families written advice and information about patient support groups. [2008]

1.4.2 Information and counselling on contraception for women and girls with FH

1.4.2.1 When lipid-modifying drug therapy is first considered for women and girls, the risks for future pregnancy and the fetus while taking lipid-modifying drug therapy should be discussed. This discussion should be revisited at least annually. [2008]

1.4.2.2 Healthcare professionals should give women and girls with FH specific information tailored to their needs and should offer a choice of effective contraceptive methods. [2008]

1.4.2.3 Combined oral contraceptives (COCs) are not generally contraindicated for women and girls being treated with lipid-modifying drug therapy. However, because there is a potential small increased risk of cardiovascular events with the use of COCs, healthcare professionals should consider other forms of contraception. Prescribers should refer to the summary of product characteristics of COCs and the relevant lipid-modifying drugs for their specific contraindications. [2008]

1.4.3 Information for pregnant women with FH

1.4.3.1 Healthcare professionals should be aware that, in general, there is no reason to advise against pregnancy or breastfeeding in women with FH. [2008]
1.4.3.2 Healthcare professionals should advise women with FH that lipid-modifying drug therapy should not be taken if they are planning to conceive or during pregnancy, because of the potential risk of fetal abnormality. Women should be advised that lipid-modifying drug therapy should be stopped 3 months before they attempt to conceive. [2008]

1.4.3.3 Women with FH who conceive while taking statins or other systemically absorbed lipid-modifying drug therapy should be advised to stop treatment immediately and they should be offered an urgent referral (see appendix D) to an obstetrician for a fetal assessment. Women should be fully informed about the nature and purpose of the assessment. [2008]

1.4.3.4 Women with FH who have conceived while taking statins or other systemically absorbed lipid-modifying drug therapy and have had a fetal assessment should be given time, opportunity and full information to consider their options (including the advantages and disadvantages) of continuing with their pregnancy. [2008]

1.4.3.5 Shared-care arrangements, to include expertise in cardiology and obstetrics, should be made for women with FH who are considering pregnancy or are pregnant. Such care should include an assessment of coronary heart disease risk, particularly to exclude aortic stenosis. This is essential for women with homozygous FH. [2008]

1.4.3.6 Serum cholesterol concentrations should not be measured routinely during pregnancy. [2008]

1.4.3.7 Women with FH who are pregnant should be advised on the potential risks and benefits of re-starting lipid-modifying drug therapy for the mother and breastfed infant. Resins are the only lipid-modifying drug therapy that should be considered during lactation. [2008]
1.5 Ongoing assessment and monitoring

1.5.1 Review

1.5.1.1 All people with FH should be offered a regular structured review that is carried out at least annually. [2008]

1.5.1.2 A baseline electrocardiogram (ECG) should be considered for adults with FH. [2008]

1.5.1.3 Healthcare professionals should record the progress of cascade testing among the relatives of a person with FH as part of the structured review. This should include at least the first- and second- and, when possible, third-degree biological relatives. If there are still relatives who have not been tested, further action should be discussed. [2008]

1.5.1.4 Healthcare professionals should update the family pedigree of a person with FH and note any changes in the coronary heart disease status of their relatives as part of the structured review. This should include at least the first- and second- and, when possible, third-degree biological relatives. [2008]

1.5.1.5 Structured review should include assessment of any symptoms of coronary heart disease and smoking status, a fasting lipid profile, and discussion about concordance with medication, possible side effects of treatment the patient may be experiencing, and any changes in lifestyle or lipid-modifying drug therapy that may be required to achieve the recommended LDL-C concentration (see section 1.3). [2008]

1.5.2 Referral for evaluation of coronary heart disease

1.5.2.1 Healthcare professionals should offer people with FH an urgent referral (see appendix D) to a specialist with expertise in cardiology for evaluation if they have symptoms or signs of possible coronary heart disease which are not immediately life-threatening. A low threshold for referral is recommended. [2008]
1.5.2.2 A person with FH with symptoms or signs of possible coronary heart disease which are immediately life-threatening (for example, acute coronary syndrome) should be referred to hospital as an emergency in line with advice for the general population. [2008]

1.5.2.3 Healthcare professionals should consider offering people with FH a referral for evaluation of coronary heart disease if they have a family history of coronary heart disease in early adulthood, or two or more other cardiovascular risk factors (for example, they are male, they smoke, or they have hypertension or diabetes). [2008]

1.5.2.4 Upon diagnosis, healthcare professionals should offer all adults and children/young people with homozygous FH a referral for an evaluation of coronary heart disease. [2008]

1.5.2.5 In asymptomatic children and young people with heterozygous FH, evaluation of coronary heart disease is unlikely to detect clinically significant disease and referral should not be routinely offered. [2008]

Terms used in this guideline

Adults with FH

For the purposes of this guideline, ‘adults’ includes all persons with familial hypercholesterolaemia (FH; heterozygous or homozygous) who are 16 years and older.

Cascade testing

Cascade testing is a mechanism for identifying people at risk of a genetic condition by a process of family tracing. For FH the test employed is a DNA test where a disease-causing mutation has been identified in the index individual/proband.

Children/young people

For the purposes of this guideline, ‘children’ refers to persons younger than 10 years; ‘young people’ refers to persons from 10 years of age up to the age of 15 years. The definitions used here are not prescriptive and healthcare professionals are expected
to exercise their judgement and consider the wishes of the patients, and their families or carers when interpreting these terms in individual instances.

**Child-focused setting**

Child-focused refers to valuing the child’s view and validating their voice in making decisions impacting their lives. A child-focused facility or space is one designed from the viewpoint of the service recipients.

**Dutch lipid Clinic Network (DLCN) criteria/score**

A method of assessing whether a person has FH. It is based on personal and family medical history, clinical signs, LDL-C concentration and DNA testing. A score is attributed to each component; the higher the score, the higher the likelihood of the person having FH.

**Family history**

The structure and relationships within the family that relates information about diseases in family members.

**First-degree relative**

A person’s biological parents, brothers and sisters, and children.

**Heterozygous FH**

High LDL-C concentration in the blood caused by an inherited mutation from one parent only. People with FH are at increased risk of cardiovascular disease.

**High-intensity statin**

Statins are classified as high intensity if they produce greater LDL-C reductions than simvastatin 40 mg (for example, simvastatin 80 mg and appropriate doses of atorvastatin and rosuvastatin).

**Homozygous FH**

Very high LDL-C concentration in the blood caused by an inherited mutation from both parents. When a person inherits exactly the same affected gene from both parents this is called truly ‘homozygous’ FH. When the mutations in the LDL receptor gene (or equivalent) are different, this state is called ‘compound heterozygous’. In
general, the overall effect in both states is similar, in that LDL-C concentrations are very high. Both groups of patients have the same clinical pattern and high risk of cardiovascular disease.

For clinical purposes, both homozygous FH and compound heterozygous FH can be regarded as behaving in a similar manner. Therefore, for the purposes of this guideline the term ‘homozygous FH’ is used to also encompass compound heterozygous FH.

**Index individual (synonymous with ‘proband’)**

The original patient who is the starting point for follow-up of other members of a family when investigating for possible causative genetic factors of the presenting condition.

**Lipid measurements/concentrations/levels**

These terms refer to the measurement of total cholesterol (TC), triglycerides (TGs), high-density lipoprotein cholesterol (HDL-C), and LDL-C. LDL-C is not usually measured directly but calculated from the TC, TGs and HDL-C, ideally using a fasting sample.

Such tests are usually done in a clinical biochemistry laboratory.

**Mutation**

An identified change in the DNA sequence of a gene that is predicted to damage the normal function of the gene and so cause disease.

**Pedigree**

A method of characterising the relatives of an index individual/case and their family relationship as well as problems or illnesses within the family. This information, often represented graphically as a family tree, facilitates analysis of inheritance patterns. Study of a trait or disease begins with the affected person (the index individual). The pedigree is drawn as the relatives are described. One begins with the siblings of the index individual and proceeds to the parents; relatives of the parents, including brothers, sisters, nephews, nieces, grandparents, and so on. At least three generations are usually included. Illnesses, hospitalisations, causes of death,
miscarriages, abortions, congenital anomalies, and any other unusual features are recorded.

Premature coronary heart disease
For the purpose of this guideline, this refers to a coronary event that has occurred before 60 years of age in an index individual or first-degree relative.

Proband
The affected (index) individual through whom a family with a genetic disorder is ascertained.

Second-degree relative
A person’s biological grandparent, grandchild, uncle, aunt, niece, nephew, half sister or half brother.

Secondary causes of hypercholesterolaemia
Causes of hyperlipidaemia other than familial, including uncontrolled diabetes mellitus, obesity, excess alcohol consumption, untreated hypothyroidism and some medications, for example, thiazide diuretics and ciclosporin.

Specialist centre
The definition of a specialist centre is not rigid and is based on a combination of patient treatment services, numbers and ages of people attending there, the presence of a multi-disciplinary team (which may include, for example, physicians, lipidologists, specialist nurses, pharmacists and dietitians), the ability to manage the more unusual manifestations of the condition and the additional functions such as research, education and standard setting. Care is supervised by expert healthcare professionals but shared with local hospitals and primary care teams. Although details of the model may vary between patients and areas, the key is that specialist supervision oversees local provision with the patient seen at diagnosis for initial assessment and then at least annually for review.

Tendon xanthomata
A clinically detectable nodularity and/or thickening of the tendons caused by infiltration with lipid-laden histiocytes (macrophages in connective tissue).
A distinctive feature of FH that most frequently affects the Achilles tendons but can also involve tendons on the back of the hands, elbows and knees.

**Third-degree relative**
A person’s biological great grandparent, great grandchild, great aunt, great uncle, first cousin, grand nephew or grand niece.

**Urgent referral**
For the purposes of this guideline, urgent referral is as soon as possible with a maximum of 14 days.

**Putting this guideline into practice**

NICE has produced tools and resources [link to tools and resources tab] to help you put this guideline into practice.

Putting recommendations into practice can take time. How long may vary from guideline to guideline, and depends on how much change in practice or services is needed. Implementing change is most effective when aligned with local priorities.

Changes recommended for clinical practice that can be done quickly – like changes in prescribing practice – should be shared quickly. This is because healthcare professionals should use guidelines to guide their work – as is required by professional regulating bodies such as the General Medical and Nursing and Midwifery Councils.

Changes should be implemented as soon as possible, unless there is a good reason for not doing so (for example, if it would be better value for money if a package of recommendations were all implemented at once).

Different organisations may need different approaches to implementation, depending on their size and function. Sometimes individual practitioners may be able to respond to recommendations to improve their practice more quickly than large organisations.

Here are some pointers to help organisations put NICE guidelines into practice:
1. **Raise awareness** through routine communication channels, such as email or newsletters, regular meetings, internal staff briefings and other communications with all relevant partner organisations. Identify things staff can include in their own practice straight away.

2. **Identify a lead** with an interest in the topic to champion the guideline and motivate others to support its use and make service changes, and to find out any significant issues locally.

3. **Carry out a baseline assessment** against the recommendations to find out whether there are gaps in current service provision.

4. **Think about what data you need to measure improvement** and plan how you will collect it. You may want to work with other health and social care organisations and specialist groups to compare current practice with the recommendations. This may also help identify local issues that will slow or prevent implementation.

5. **Develop an action plan**, with the steps needed to put the guideline into practice, and make sure it is ready as soon as possible. Big, complex changes may take longer to implement, but some may be quick and easy to do. An action plan will help in both cases.

6. **For very big changes** include milestones and a business case, which will set out additional costs, savings and possible areas for disinvestment. A small project group could develop the action plan. The group might include the guideline champion, a senior organisational sponsor, staff involved in the associated services, finance and information professionals.

7. **Implement the action plan** with oversight from the lead and the project group. Big projects may also need project management support.

8. **Review and monitor** how well the guideline is being implemented through the project group. Share progress with those involved in making improvements, as well as relevant boards and local partners.
NICE provides a comprehensive programme of support and resources to maximise uptake and use of evidence and guidance. See our into practice pages for more information.

Also see Leng G, Moore V, Abraham S, editors (2014) Achieving high quality care – practical experience from NICE. Chichester: Wiley.

**Context**

In some people, a high cholesterol concentration in the blood is caused by an inherited genetic defect known as familial hypercholesterolaemia (FH). A raised cholesterol concentration in the blood is present from birth and may lead to early development of atherosclerotic disease like coronary heart disease. The disease shows an autosomal dominant pattern of inheritance, being transmitted from generation to generation in such a way that siblings and children of a person with FH have a 50% risk of inheriting FH.

Most people with FH have inherited a defective gene for FH from only one parent and are therefore heterozygous. Rarely, a person will inherit a genetic defect from both parents and will have homozygous FH or compound heterozygous FH, which will be collectively termed homozygous FH for the purpose of this guideline.

The prevalence of heterozygous FH in the UK population is estimated to be 1 in 500, which means that approximately 110,000 people are affected. The elevated serum cholesterol concentration that characterises heterozygous FH leads to a greater than 50% risk of coronary heart disease in men by the age of 50 years and at least 30% in women by the age of 60 years.

Homozygous FH is rare, with symptoms appearing in childhood, and is associated with early death from coronary heart disease. Homozygous FH has an incidence of approximately one case per one million.

The guideline will assume that prescribers will use a drug’s summary of product characteristics to inform their decisions for individual patients.

In 2017 the areas on case-finding, diagnosis and pharmacological monotherapy (statin v placebo) were updated. Since the original guideline was published in 2008,
cascade testing may now be more cost-effective, and DNA diagnosis technology has changed greatly. In addition, more evidence has been identified on the use of high-intensity statins, and on the safety profile of statins in children and young people.

More information

To find out what NICE has said on topics related to this guideline, see our web page on lipid disorders.

Recommendations for research

The guideline committee has made the following recommendations for research.

As part of the 2017 update, the standing committee made research recommendations on using different thresholds of low-density lipoprotein cholesterol (LDL-C) concentration in primary care case finding and on long-term monitoring of sub-clinical atherosclerosis in children with familial hypercholesterolaemia (FH) who are treated with statins (see below). The committee also made 3 other research recommendations, on secondary care case finding, cascade testing and the use of clinical scoring criteria. One research recommendation on identification using clinical registers was removed. Details can be found in the addendum.

1 Using different thresholds of low-density lipoprotein cholesterol concentration in primary care case finding

What is the clinical and cost effectiveness of using different thresholds of LDL-C concentration in primary care case finding?

Why this is important

The clinical community recognises that FH is underdiagnosed, with prevalence more likely to be approximately 1 in 250 rather than the widely cited 1 in 500. Searching electronic primary care databases is an effective way of identifying people with FH. One of the ways in which people are identified through electronic primary care database searching is to search using total cholesterol or LDL-C concentration. Currently, the entire evidence base for identifying cohorts of people with FH through primary care case finding uses a total cholesterol concentration cut-off of 9.3 mmol/l.
This is a very high concentration and anecdotal evidence suggests that this identifies older people but may miss younger people with FH. This could lead to missed opportunities to identify and treat people with FH at an earlier age. Research is needed to identify whether using different total cholesterol and LDL-C concentrations to identify people with FH through primary care database searching affects the diagnostic yield of FH. Additionally, there is a lack of data on the ethnicity, age and triglyceride concentration of people with FH identified through primary care database searching. These should be included as outcomes in future research. [2017]

2 Long-term monitoring of sub-clinical atherosclerosis in children with FH who are treated with statin therapy

What are the long-term effects of statin therapy on sub-clinical atherosclerosis in children with FH who are treated with statin therapy?

Why this is important

Although statins are increasing in use, there is still a lack of data on the long-term effects of statins in children. It is particularly important to determine any long-term adverse effects of statin treatment in a population with FH, as people generally take statins for the rest of their lives once treatment starts. [2017]

3 Lipid-modifying drug therapy in children

What is the clinical effectiveness and safety of differing doses of lipid-modifying therapy in children with FH?

Why this is important

There have been no published studies to establish target serum LDL-C concentration in treated children with FH receiving lipid-modifying drug therapy. Treatment is recommended from 10 years onwards, however this lack of data prevents a recommendation regarding the aim of pharmacological treatment on serum LDL-C concentrations.

Research (both cross-sectional and longitudinal) should assess the evidence of end-organ involvement (for example, carotid intima medial thickness [IMT]) to determine at which age abnormalities can first be seen in children. The aim would be to identify...
a threshold effect, with an LDL-C concentration below which carotid IMT is normal and where thickening is absent, and above which it is abnormal and where thickening is observed. Outcomes should include fasting serum total and LDL-C concentration, carotid artery IMT, and growth and pubertal development. [2008]

4 LDL apheresis for people with heterozygous FH

What are the appropriate indications, effectiveness and safety of LDL apheresis in people with heterozygous FH?

Why this is important

There is limited evidence to inform specific indications for LDL apheresis in people with heterozygous FH. In addition, there is limited published evidence on the cardiovascular outcome of such patients treated with LDL apheresis.

Evidence on the value of investigations (various measures of vascular status, considered to reflect the extent or activity of atherosclerotic vascular disease of the coronary arteries) in predicting outcome from LDL apheresis should ideally be based on evidence from randomised controlled trials with clinical outcomes. It is difficult to identify a suitable alternative treatment because LDL apheresis is generally only considered in people for whom no other treatment is available. One comparator may be novel therapies with antisense oligonucleotides (ApoB).

A national register should be established for all people with FH who are referred for and/or are undergoing LDL apheresis. Data should be collected on the natural history of FH and the temporal relationship of clinical and vascular features in relation to treatments and other parameters. [2008]

5 Pregnancy in women with FH

What are the implications of FH for the safety of a mother during pregnancy and what are the risks of fetal malformations attributable to pharmacological therapies?

Why this is important

There is little information on the outcomes of pregnancy in women with FH. A small number of conflicting studies have suggested a small increase in fetal abnormalities...
if the mother has taken statins during the first trimester, but there are not sufficient
data to provide an accurate estimate of the level of risk. There is also limited
information on the risk of pregnancy (including cardiac death) in a woman with FH.

Data on the incidence of cardiac problems in pregnancy and incidence of fetal
malformation would inform future recommendations. This could reduce uncertainty
for women, and help to identify risks during the pregnancy that could be better
managed. The only feasible research method to address these questions is an
observational longitudinal study following women with FH and other women (not
diagnosed with FH) using statins through their pregnancies using a national register.

[2008]

6 Cardiovascular evaluation for people with FH

What is the utility of routine cardiovascular evaluation for asymptomatic people with
FH?

Why this is important

Because of their inherent high risk of developing premature coronary heart disease,
a low threshold of suspicion for coronary disease is recommended for people with
FH. Routine monitoring to detect sub-clinical atherosclerosis should be non-invasive,
sensitive, specific and cost effective. Research to assess the prevalence of both
asymptomatic coronary and non-coronary atherosclerosis in people with definite
heterozygous FH is required.

As well as exercise ECG testing followed by stress echocardiography before
possible angiography in people with an abnormal exercise test and ankle brachial
pressure measures, research should include magnetic resonance imaging (MRI) in
addition to other modalities such as carotid IMT and coronary calcification.
Outcomes should include changes in exercise ECG/ankle brachial pressure
testing/IMT/calcification over time.

Consideration should also be given to the feasibility of conducting a long-term
randomised trial to compare the differences in morbidity or mortality attributable to
early diagnosis using routine monitoring or symptom-based investigation. [2008]
Update information

New recommendations have been added for the identification, diagnosis and treatment of familial hypercholesterolaemia.

These are marked as [2017] if the evidence has been reviewed and the recommendation has been added or updated, or the evidence has been reviewed but no change has been made to the recommended action.

NICE proposes to delete some recommendations from the 2008 guideline, because either the evidence has been reviewed and the recommendations have been updated, or NICE has updated other relevant guidance and has replaced the original recommendations. Recommendations that have been deleted or changed sets out these recommendations and includes details of replacement recommendations.

Where there is no replacement recommendation, an explanation for the proposed deletion is given.

Where recommendations are shaded in grey and end [2008] or [2016], the evidence has not been reviewed.

Where recommendations are shaded in grey and end [2008, amended 2017], the evidence has not been reviewed but changes have been made to the recommendation wording that change the meaning (for example, because of equalities duties or a change in the availability of medicines, or incorporated guidance has been updated). These changes are marked with yellow shading, and explanations of the reasons for the changes are given in ‘Recommendations that have been deleted or changed’ for information.

See also the original NICE guideline and supporting documents.

Recommendations that have been deleted or changed

Recommendations to be deleted

<table>
<thead>
<tr>
<th>Recommendation in 2008 guideline</th>
<th>Comment</th>
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<tbody>
<tr>
<td>1.1.3 A diagnosis of FH should be made using the Simon Broome criteria, which include a combination of family history, clinical signs (specifically tendon</td>
<td>This recommendation has been deleted and replaced by: 1.1.5 Use the Simon Broome or Dutch Lipid Clinic Network (DLCN) criteria to</td>
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Familial Hypercholesterolaemia: NICE guideline short version DRAFT (May 2017)
### 1.1.4 Healthcare professionals should inform people with a diagnosis of FH based on the Simon Broome criteria (see appendix E of the NICE guideline) that they have a clinical diagnosis of FH.  

This recommendation has been deleted because diagnosis of FH is now based on genetic testing for the presence of mutation in APOB, LDLR or PCS9 genes.

### 1.1.8 A family history of premature coronary heart disease should always be assessed in a person being considered for a diagnosis of FH (see Simon Broome criteria in Appendix E of the NICE guideline).  

This recommendation has been deleted because a family history of premature heart disease is a component of both the Simon Broome and DLCN criteria, which are now recommended. Information is captured by recommendation 1.1.1 (amended).

### 1.1.12 Healthcare professionals should offer people with a clinical diagnosis of FH a DNA test to increase the certainty of their diagnosis and to aid diagnosis among their relatives.

This recommendation has been deleted because DNA testing is required to confirm a diagnosis of FH. DNA testing is covered by new recommendation 1.1.6.

### 1.1.15 In children at risk of FH because of one affected parent, the following diagnostic tests should be carried out by the age of 10 years or at the earliest opportunity thereafter.

- A DNA test if the family mutation is known.
- LDL-C concentration measurement if the family mutation is not known. When excluding a diagnosis of FH a further LDL-C measurement should be repeated after puberty because LDL-C concentrations change during puberty.

Relatives of people with FH are now diagnosed using mutation only, not LDL-C concentrations. Replaced by recommendation 1.1.15:

*1.1.15 In children at risk of FH because of one affected parent, offer a DNA test by the age of 10 years or at the earliest opportunity thereafter. [2017]*

### 1.2.1 Healthcare professionals should use systematic methods (that is, cascade testing) for the identification of people with FH

This recommendation has been removed as the committee agreed that the relevant information was now contained within new recommendation 1.2.1:

*1.2.1 Carry out cascade testing using DNA testing to identify affected first- and second- and, when possible, third-degree relatives of people with a diagnosis of FH. [2017]*
1.2.4 Cascade testing using a combination of DNA testing and LDL-C concentration measurement is recommended to identify affected relatives of those index individuals with a clinical diagnosis of FH. This should include at least the first- and second- and, when possible, third-degree biological relatives.

This recommendation has been deleted and replaced by recommendation 1.2.1 as it is no longer recommended to use LDL-C concentration in cascade testing:

1.2.1 Carry out cascade testing using DNA testing to identify affected first- and second- and, when possible, third-degree biological relatives of people with a diagnosis of FH. [2017]

1.2.5 In families in which a mutation has been identified, the mutation and not LDL-C concentration should be used to identify affected relatives. This should include at least the first- and second- and, when possible, third-degree biological relatives.

This recommendation has been deleted because the recommendation is covered by new recommendation 1.2.1:

1.2.1 Carry out cascade testing using DNA testing to identify affected first- and second- and, when possible, third-degree relatives of people with a diagnosis of FH. [2017]

1.2.6 In the absence of a DNA diagnosis, cascade testing using LDL-C concentration measurements should be undertaken to identify people with FH.

This has been deleted because only DNA testing is recommended to identify people with FH through cascade testing. It is no longer practice to use LDL-C concentration the new recommendation. This is covered by new recommendation 1.2.1:

1.2.1 Carry out cascade testing using DNA testing to identify affected first- and second- and, when possible, third-degree relatives of people with a diagnosis of FH. [2017]

1.2.7 To diagnose FH in relatives of an index individual, the gender- and age-specific criteria for LDL-C concentration in appendix E of the NICE guideline should be used. The Simon Broome LDL-C criteria for index individuals should not be used because this will result in under diagnosis.

This has been deleted because new recommendation 1.2.1 advises only DNA should be used in cascade testing. It is no longer recommended that LDL-C concentration should be used to identify people with FH through cascade testing, as this does not provide a definitive diagnosis of FH.

1.2.8 The use of a nationwide, family-based, follow up system is recommended to enable comprehensive identification of people affected by FH.

This recommendation has been deleted because it is ambiguous and lacks clarity. The committee discussed that it refers to cascade testing, which is covered in the new recommendation 1.2.1; therefore this recommendation is unnecessary and was deleted.

1.3.1.2 Statins should be the initial treatment for all adults with FH.

This recommendation was deleted because it is now recommended to prescribe a high-intensity statin, therefore
the recommendation has been replaced by:

1.3.1.2 Offer a high-intensity statin with the lowest acquisition cost as the initial treatment for all adults with FH and aim for at least a 50% reduction in LDL-C concentration from the baseline measurement. [2017]

1.3.1.3 Healthcare professionals should consider prescribing a high-intensity statin to achieve a recommended reduction in LDL-C concentration of greater than 50% from baseline (that is, LDL-C concentration before treatment).

This recommendation has been deleted because the wording about 50% LDL-C reduction has been combined with new recommendation 1.3.1.2.

1.3.1.2 Offer a high-intensity statin with the lowest acquisition cost as the initial treatment for all adults with FH and aim for at least a 50% reduction in LDL-C concentration from the baseline measurement. [2017]

1.3.1.5 Healthcare professionals should offer treatment with a statin with a low acquisition cost for adults with FH in whom the diagnosis is made after the age of 60 and who do not have coronary heart disease

This has been deleted because it is now recommended that all adults with FH should be prescribed a high-intensity statin (new recommendation 1.3.1.2). There is no requirement for a separate recommendation for those over 60 years of age.

1.3.1.22 When the decision to initiate lipid-modifying drug therapy has been made in children and young people, statins should be the initial treatment. Healthcare professionals with expertise in FH in children and young people should choose a statin that is licensed for use in the appropriate age group.

This recommendation has been deleted because the committee made a new recommendation to 'offer statins', which replaces the old text in 1.3.1.22.

1.3.1.20 Offer statins to children with FH by the age of 10 years or at the earliest opportunity thereafter. [2017]

Furthermore, the committee considered it essential that the other elements of the old deleted recommendation 1.3.1.22 should be retained; therefore additional recommendations were made (1.3.1.21 and 1.3.1.22) which highlight that an appropriate statin should be used for each age group and that the healthcare professional should have expertise in treating children and young people with FH.

1.3.1.21 For children and young people with FH, consider a statin that is licensed for use in the appropriate age group. [2017]
1.3.1.23 Statin therapy for children and young people should be initiated by a healthcare professional with expertise in treating children and young people with FH, and in a child-focused setting. [2008, amended 2017]

### Amended recommendation wording (change to meaning)

<table>
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<tr>
<th>Recommendation in 2008</th>
<th>Recommendation in current guideline</th>
<th>Reason for change</th>
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| 1.1.1 Healthcare professionals should consider the possibility of FH in adults with raised cholesterol (total cholesterol typically greater than 7.5 mmol/l), especially when there is a personal or a family history of premature coronary heart disease.  | 1.1.1 Think about familial hypercholesterolaemia (FH) as a possible diagnosis in adults with:  
• a total cholesterol level greater than 7.5 mmol/l, and/or  
• a personal or family history of premature coronary heart disease (a coronary event before 60 years or, in an index individual or first-degree relative)  | The wording has been updated to reflect the current wording style of recommendations, and to emphasise that FH should be thought about as a potential diagnosis when people present with the characteristics outlined |
| 1.1.13 Healthcare professionals should inform all people who have an identified mutation diagnostic of FH that they have an unequivocal diagnosis of FH even if their LDL-C concentration does not meet the diagnostic criteria (see appendix E of the NICE guideline) | 1.1.13 Inform all people who have an identified mutation diagnostic of FH that they have an unequivocal diagnosis of FH even if their LDL-C concentration does not meet the diagnostic criteria (see recommendation 1.1.5) | Appendix E has been removed and the wording has been updated to current style. |
| 1.3.1.22 When the decision to initiate lipid-modifying drug therapy has been made in children and young people, statins should be the initial treatment. Healthcare professionals with expertise in FH in children and young people should choose a statin that is licensed for use in the appropriate age group. | 1.3.1.20 Offer statins to children with FH by the age of 10 years or at the earliest opportunity thereafter. [2017]  
1.3.1.21 For children and young people with FH, consider a statin that is licensed for use in the appropriate age group. [2017]  
1.3.1.22 Statin therapy for children and young people | This recommendation has been deleted/amended because the committee made a new recommendation to ‘offer statins’, which replaces the old text in 1.3.1.21. Furthermore, the committee considered it essential that the other elements of the old deleted recommendation 1.3.1.22 should be retained; therefore additional recommendations were made (1.3.1.21) that highlight that an
should be initiated by a healthcare professional with expertise in treating children and young people with FH, and in a child-focused setting. [2008, amended 2017]

appropriate statin should be used for each age group and that the healthcare professional should have expertise in treating children and young people with FH. This recommendation was based on an evidence review.

An evidence review was not undertaken for 1.3.1.22, but the wording was amended to take account of current NICE style and clinical practice.

Changes after publication

July 2016: Recommendations 1.3.1.4–1.3.1.9 have been replaced and are adapted from Ezetimibe for treating primary (heterozygous-familial and non-familial) hypercholesterolaemia (NICE technology appraisal 385). TA385 has replaced TA132, the original source for these recommendations. They have been changed to remove reference to non-familial hypercholesterolaemia, which TA385 also covers.

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