NICE National Institute for Health and Care Excellence

NICE quality standard

# Familial hypercholesterolaemia

Quality standard
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This standard is based on CG71.

This standard should be read in conjunction with QS15, QS43, QS100, QS99, QS140, QS120, QS111, QS94 and QS84.

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# Quality statement 1: Diagnosis

## Quality statement

Adults with a baseline total cholesterol above 7.5 mmol/l are assessed for a clinical diagnosis of familial hypercholesterolaemia (FH).

## Rationale

Most of the 120,000 people estimated to have FH are undiagnosed and untreated. Because untreated FH carries a very high risk of cardiovascular disease, it is important that every opportunity is taken to identify people with FH and offer them treatment. Considering a clinical diagnosis of FH in people with high cholesterol will result in greater identification of FH and support cascade testing of their relatives. This will lead to more treatment to reduce cholesterol levels and prevention of coronary events among people with FH.

## Quality measures

The following measures can be used to assess the quality of care or service provision specified in the statement. They are examples of how the statement can be measured, and can be adapted and used flexibly.

### Structure

Evidence of local arrangements to ensure that adults with a baseline total cholesterol above 7.5 mmol/l are assessed for a clinical diagnosis of FH.

**Data source:** Local data collection.

### Process

Proportion of adults with a baseline total cholesterol above 7.5 mmol/l who are assessed for a clinical diagnosis of FH.

Numerator – The number of people in the denominator assessed for a clinical diagnosis of FH.

Denominator – The number of adults with a baseline total cholesterol above 7.5 mmol/l.

**Data source:** Local data collection.

# What the quality statement means for different audiences

**Service providers** ensure that systems are in place for adults with a baseline total cholesterol above 7.5 mmol/l to be assessed for a clinical diagnosis of FH.

**General practitioners** assess adults with a baseline total cholesterol above 7.5 mmol/l for a clinical diagnosis of FH.

**Commissioners** ensure that they commission services that identify and assess adults with a baseline total cholesterol above 7.5 mmol/l for a clinical diagnosis of FH.

**Adults with a total cholesterol above 7.5 mmol/l before treatment** have an assessment for FH.

# Source guidance

Familial hypercholesterolaemia: identification and management. NICE guideline CG71 (2008, updated 2019), recommendations 1.1.1, 1.1.2, 1.1.4, 1.1.5 and 1.1.9

# Definitions of terms used in this quality statement

## Adults

People aged 16 and older.

## Baseline total cholesterol

The total cholesterol concentration before treatment.

## Familial hypercholesterolaemia (FH)

FH relates to heterozygous FH only.

## Clinical diagnosis of FH

Assessment for a clinical diagnosis of FH should use all 3 criteria below:

- exclusion of secondary causes of hypercholesterolaemia
- 2 measurements of LDL-C concentration
- assessment against Simon Broome or Dutch Lipid Clinic Network criteria to make a clinical diagnosis of FH in primary care settings.

[Adapted from NICE's guideline on familial hypercholesterolaemia, recommendations 1.1.4, 1.1.5 and 1.1.9]

# Equality and diversity considerations

The statement has been restricted to adults because the criteria for assessment for a clinical diagnosis of FH are not appropriate for children and young people under 16 years.

# Quality statement 2: Specialist referral

## Quality statement

People with a clinical diagnosis of familial hypercholesterolaemia (FH) are referred for specialist assessment.

## Rationale

Diagnosing and managing FH in an individual and their relatives can be complex, and is best achieved when there is access to specialist services. Specialist assessments, which include DNA testing, can confirm a diagnosis. Once an accurate diagnosis has been made, people with FH can receive appropriate treatment, and cascade testing can be started to identify affected family members.

## Quality measures

The following measures can be used to assess the quality of care or service provision specified in the statement. They are examples of how the statement can be measured, and can be adapted and used flexibly.

### Structure

a) Evidence of local arrangements to ensure that people with a clinical diagnosis of FH are referred for specialist assessment.

**Data source:** Local data collection.

b) Evidence of local arrangements to ensure that a protocol for referral for a specialist assessment is agreed between primary and secondary care.

**Data source:** Local data collection.

### Process

Proportion of people with a clinical diagnosis of FH referred for specialist assessment.

Numerator – The number of people in the denominator referred for specialist assessment.

Denominator – The number of people with a clinical diagnosis of FH.

**Data source:** Local data collection.

### Outcome

Ratio of observed to estimated numbers of people with FH, using an estimate based on the area's estimated prevalence of FH (based on 1 in 500) and population size.

**Data source:** Local data collection using a dedicated database.

## What the quality statement means for different audiences

**Service providers** ensure that systems are in place for people with a clinical diagnosis of FH to be referred for specialist assessment.

**Healthcare practitioners** refer people with a clinical diagnosis of FH for specialist assessment.

**Commissioners** ensure that they commission services that can offer specialist assessment for people with a clinical diagnosis of FH.

**People who are given a clinical diagnosis of FH** because they have high cholesterol, family history or other signs are referred for specialist assessment.

## Source guidance

Familial hypercholesterolaemia: identification and management. NICE guideline CG71 (2008, updated 2019), recommendations 1.2.2 and 1.3.1.17

# Definitions of terms used in this quality statement

## Familial hypercholesterolaemia (FH)

FH relates to heterozygous FH only.

## Clinical diagnosis of FH

Assessment for a clinical diagnosis of FH should use all 3 criteria below:

- exclusion of secondary causes of hypercholesterolaemia
- 2 measurements of LDL-C concentration
- assessment against Simon Broome or Dutch Lipid Clinic Network criteria to make a clinical diagnosis of FH in primary care settings.

[Adapted from NICE's guideline on familial hypercholesterolaemia, recommendations 1.1.4, 1.1.5 and 1.1.9]

## Specialist assessment

This should include:

- confirmation of the clinical diagnosis of FH made by a GP or other healthcare professional
- an offer of DNA testing to increase the certainty of the diagnosis
- initiation of cascade testing if a diagnosis is confirmed.

A specialist assessment should be performed by a healthcare professional with expertise in FH who has access to the wider skills of a multidisciplinary team. This team should include a dietitian, cardiologist and paediatrician, and a clinical genetic specialist to take a family history and obtain informed consent for a DNA test. For children and young people, this should be a specialist with expertise in FH in children and young people.

Children refers to people younger than 10, young people refers to those aged 10 up to and including age 15, and adults refers to people aged 16 and older.

# Quality statement 3: DNA testing

## Quality statement

People with a clinical diagnosis of familial hypercholesterolaemia (FH) are offered DNA testing as part of a specialist assessment.

## Rationale

DNA testing is important because it increases the certainty of a diagnosis of FH and allows the identification of affected and unaffected relatives through cascade testing.

## Quality measures

The following measures can be used to assess the quality of care or service provision specified in the statement. They are examples of how the statement can be measured, and can be adapted and used flexibly.

### Structure

Evidence of local arrangements to ensure that people with a clinical diagnosis of FH are offered DNA testing as part of a specialist assessment.

**Data source:** Local data collection.

### Process

a) Proportion of people with a clinical diagnosis of FH who receive DNA testing as part of a specialist assessment.

Numerator – The number of people in the denominator receiving DNA testing as part of a specialist assessment.

Denominator – The number of people with a clinical diagnosis of FH.

**Data source:** Local data collection using a dedicated database.

b) Proportion of people with a clinical diagnosis of FH receiving DNA testing as part of a specialist assessment who give informed consent for the test.

Numerator – The number of people in the denominator who give informed consent for the test.

Denominator – The number of people with a clinical diagnosis of FH receiving DNA testing as part of a specialist assessment.

**Data source:** Local data collection using a dedicated database.

### Outcome

Patient satisfaction with process of informed consent.

**Data source:** Local data collection.

## What the quality statement means for different audiences

**Service providers** ensure that systems are in place for people with a clinical diagnosis of FH to be offered DNA testing as part of a specialist assessment.

**Specialists with expertise in FH** offer DNA testing to people with a clinical diagnosis of FH as part of a specialist assessment.

**Commissioners** ensure that they commission services that offer DNA testing to people with a clinical diagnosis of FH as part of a specialist assessment.

**People who are given a clinical diagnosis of FH** because they have high cholesterol, family history or other signs are offered DNA testing as part of a specialist assessment.

## Source guidance

Familial hypercholesterolaemia: identification and management. NICE guideline CG71

(2008, updated 2019), recommendation 1.1.6

# Definitions of terms used in this quality statement

## Familial hypercholesterolaemia (FH)

FH relates to heterozygous FH only.

## Clinical diagnosis of FH

Assessment for a clinical diagnosis of FH should use all 3 criteria below:

- exclusion of secondary causes of hypercholesterolaemia
- 2 measurements of LDL-C concentration
- assessment against Simon Broome or Dutch Lipid Clinic Network criteria to make a clinical diagnosis of FH in primary care settings.

[Adapted from NICE's guideline on familial hypercholesterolaemia, recommendations 1.1.4, 1.1.5 and 1.1.9]

## DNA testing

DNA testing should test for all gene mutations known to cause FH. Methods should meet the standards set out by the UK Genetic Testing Network. Informed consent should be given for DNA testing.

# Quality statement 4: Diagnosis in children under 10 years

## Quality statement

Children at risk of familial hypercholesterolaemia (FH) are offered diagnostic tests by the age of 10 years.

## Rationale

Children with FH begin to develop cardiovascular disease before clinical signs appear, with thickening of the carotid artery wall identifiable by the age of 10 years. Diagnosis by the age of 10 years allows lifestyle changes and tailored therapy if indicated, which will reduce long-term problems associated with high cholesterol and improve long-term health.

## Quality measures

The following measures can be used to assess the quality of care or service provision specified in the statement. They are examples of how the statement can be measured, and can be adapted and used flexibly.

### Structure

Evidence of local arrangements to ensure that children at risk of FH are offered diagnostic tests by the age of 10 years.

**Data source:** Local data collection.

### Process

Proportion of children at risk of FH who receive a specified diagnostic test by the age of 10 years.

Numerator – The number of people in the denominator who had received a specified

diagnostic test.

Denominator – The number of children aged 10 years at risk of FH.

**Data source:** Local data collection.

### Outcome

Ratio of observed to estimated numbers of children at risk of FH, using an estimate based on the area's estimated prevalence of FH (based on 1 in 500) and population size.

**Data source:** Local data collection.

## What the quality statement means for different audiences

**Serviceproviders** ensure that systems are in place for children at risk of FH to be offered diagnostic tests by the age of 10 years.

**Specialist with expertise in FH in children and young people** offer children at risk of FH diagnostic tests by the age of 10 years.

**Commissioners** ensure that they commission services that offer children at risk of FH diagnostic tests by the age of 10 years.

**Children at risk of FH** because they have 1 parent with the condition are offered diagnostic tests by the age of 10 years.

## Source guidance

Familial hypercholesterolaemia: identification and management. NICE guideline CG71 (2008, updated 2019), recommendation 1.1.15

# Definitions of terms used in this quality statement

## Familial hypercholesterolaemia (FH)

FH relates to heterozygous FH only.

## Children at risk of FH

Children (under 10 years) with 1 affected parent.

## Diagnostic tests

Children at risk of FH because of 1 affected parent receive either of the following diagnostic tests:

- 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.

[Adapted from NICE's guideline on familial hypercholesterolaemia]

## Specialist with expertise in FH in children and young people

A healthcare professional with expertise in FH in children and young people who has access to the wider skills of a multidisciplinary team. This team should include a dietitian, cardiologist and paediatrician, and a clinical genetic specialist to take a family history and obtain informed consent for a DNA test. All children and young people being investigated for a diagnosis of FH should be referred to a specialist with expertise in FH in children and young people in a child-focused setting.

## Child-focused setting

A child-focused setting is defined as valuing the child's view and validating their voice in making decisions impacting their lives. A child-focused facility or space is one designed with the children's needs in mind.

[Adapted from NICE's guideline on familial hypercholesterolaemia, terms used in this guideline]

# Quality statement 5: Cascade testing

## Quality statement

Relatives of people with a confirmed diagnosis of monogenic familial hypercholesterolaemia (FH) are offered DNA testing through a nationwide, systematic cascade process.

## Rationale

Most people in the UK with FH are undiagnosed. Cascade testing has been shown to be effective for identifying people with FH, especially when provided nationwide. Nationwide cascade testing ensures that all family members can access DNA testing wherever they live.

## Quality measures

The following measures can be used to assess the quality of care or service provision specified in the statement. They are examples of how the statement can be measured, and can be adapted and used flexibly.

### Structure

Evidence of local arrangements to ensure that relatives of people with a confirmed diagnosis of monogenic FH are offered DNA testing through a nationwide systematic cascade process.

**Data source:** Local data collection.

### Process

a) Proportion of untested first-degree relatives of people with a confirmed diagnosis of monogenic FH who are offered cascade testing.

Numerator – The number of people in the denominator offered cascade testing.

Denominator – The number of untested first-degree relatives of people with a confirmed diagnosis of monogenic FH.

**Data source:** Local data collection using a dedicated database.

b) Proportion of at-risk, untested, second- and third-degree relatives of people with a confirmed diagnosis of monogenic FH who are offered cascade testing.

Numerator – The number of people in the denominator offered cascade testing.

Denominator – The number of at-risk, untested, second- and third-degree relatives of people with a confirmed diagnosis of monogenic FH.

**Data source:** Local data collection using a dedicated database.

c) Proportion of untested first-degree relatives of people with a confirmed diagnosis of monogenic FH who receive cascade testing.

Numerator – The number of people in the denominator receiving cascade testing.

Denominator – The number of untested first-degree relatives of people with a confirmed diagnosis of monogenic FH.

**Data source:** Local data collection using a dedicated database.

d) Proportion of at-risk, untested, second- and third-degree relatives of people with a confirmed diagnosis of monogenic FH who receive cascade testing.

Numerator – The number of people in the denominator receiving cascade testing.

Denominator – The number of at-risk, untested second- and third-degree relatives of people with a confirmed diagnosis of monogenic FH.

**Data source:** Local data collection using a dedicated database.

### Outcome

Prevalence of FH.

**Data source:** Local data collection using a dedicated database.

## What the quality statement means for different audiences

**Serviceproviders** ensure that systems are in place for relatives of people with a confirmed diagnosis of monogenic FH to be offered DNA testing through a nationwide, systematic cascade process.

**Specialists with expertise in FH** offer DNA testing to relatives of people with a confirmed diagnosis of monogenic FH through a nationwide, systematic cascade process.

**Commissioners** ensure that they commission services that offer DNA testing to relatives of people with a confirmed diagnosis of monogenic FH, through a nationwide, systematic cascade process.

**Relatives of people with a confirmed diagnosis of FH and a known DNA mutation** are offered DNA testing themselves as part of a national scheme.

## Source guidance

Familial hypercholesterolaemia: identification and management. NICE guideline CG71 (2008, updated 2019), recommendation 1.2.1

## Definitions of terms used in this quality statement

### Relatives

At least first- and second-degree biological relatives and third-degree biological relatives if possible.

[Adapted from NICE's guideline on familial hypercholesterolaemia, terms used in this

guideline]

## Familial hypercholesterolaemia (FH)

FH relates to heterozygous FH only.

## Monogenic FH

Present when an autosomal dominant pattern of inheritance of elevated LDL-C levels is seen in the extended family of the proband (for example, on average 50% of first-degree relatives have elevated levels). In relatives, the age- and gender-specific diagnostic cut-offs in NICE's guideline on familial hypercholesterolaemia should be used because the Simon Broome diagnostic cut-offs are not appropriate for relatives. The diagnosis of monogenic FH can also be given when the index case carries a documented FH-causing mutation in the *LDLR*, *APOB* or *PCSK9* genes.

## Confirmed diagnosis of monogenic FH

This requires evidence from DNA testing of an FH-causing mutation in the *LDLR*, *APOB* or *PCSK9* genes. Before cascade testing is initiated in relatives, a diagnosis of monogenic FH in the index person should be confirmed by a specialist with expertise in FH.

## Cascade testing

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.

# Quality statement 6: Drug treatment in adults

## Quality statement

Adults with familial hypercholesterolaemia (FH) receive lipid-modifying drug treatment to reduce LDL-C concentration by more than 50% from baseline.

## Rationale

Lipid-modifying drug treatment reduces LDL-C levels and prevents the development of cardiovascular disease. Studies indicate that treatment that lowers LDL-C levels by more than 50% from baseline offers greater benefit for plaque stabilisation than treatment that is less effective at reducing LDL-C.

## Quality measures

The following measures can be used to assess the quality of care or service provision specified in the statement. They are examples of how the statement can be measured, and can be adapted and used flexibly.

### Structure

Evidence of local arrangements to ensure that adults with FH receive lipid-modifying drug treatment to reduce LDL-C concentration by more than 50% from baseline.

**Data source:** Local data collection.

### Process

Proportion of adults with FH who receive appropriate lipid-modifying drug treatment.

Numerator – The number of people in the denominator receiving appropriate lipid-modifying drug treatment.

Denominator – The number of adults with FH.

**Data source:** Local data collection.

### Outcome

Number of adults with FH whose LDL-C concentration is reduced by more than 50% from baseline within 1 year.

**Data source:** Local data collection using a dedicated database.

## What the quality statement means for different audiences

**Service providers** ensure that systems are in place for adults with FH to receive lipid-modifying drug treatment to reduce LDL-C concentration by more than 50% from baseline.

**Healthcare practitioners** offer adults with FH lipid-modifying drug treatment to reduce LDL-C concentration by more than 50% from baseline.

**Commissioners** ensure that they commission services that offer adults with FH lipid-modifying drug treatment to reduce LDL-C concentration by more than 50% from baseline.

**Adults with FH** are offered drugs to reduce the low-density cholesterol (bad cholesterol) in their blood to less than a half of the level before treatment.

## Source guidance

Familial hypercholesterolaemia: identification and management. NICE guideline CG71 (2008, updated 2019), recommendations 1.3.1.2 to 1.3.1.4, 1.3.1.6 to 1.3.1.8 and 1.3.1.14

# Definitions of terms used in this quality statement

## Familial hypercholesterolaemia (FH)

FH relates to heterozygous FH only.

## Adults with FH

Adults (aged 16 and older) should have a diagnosis of FH made by a specialist with expertise in FH.

## Baseline LDL-C

The concentration before treatment.

## Lipid-modifying drug treatment

Lipid-modifying drug treatment should be given in accordance with the following:

- to achieve the recommended reduction:
  - offer a high-intensity statin with the lowest acquisition cost as the initial treatment
  - increase dose of statin to maximum licensed or tolerated dose
- ezetimibe monotherapy is recommended if the person is intolerant to statin therapy or there are contraindications to initial statin therapy
- co-administer ezetimibe with initial statin therapy when serum total or LDL-C concentration is not appropriately controlled and a change from initial statin therapy to an alternative statin is being considered.

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).

Treatment for FH is usually provided by either a specialist with expertise in FH or a GP through a shared care arrangement.

[Adapted from NICE's guideline on familial hypercholesterolaemia, recommendations 1.3.1.2 to 1.3.1.6, 1.3.1.11, and terms used in this guideline]

# Equality and diversity considerations

The statement has been restricted to adults only because there is currently no evidence on which to base any specific target for lowering LDL-C in children and young people under 16 years. However, lipid-modifying drug treatment should be considered by the age of 10 years in line with NICE's guideline on familial hypercholesterolaemia.

Women with FH should be advised that lipid-modifying drug treatment should not be taken if they are planning to conceive or during pregnancy because of the risk of fetal abnormality. Women should be advised that lipid-modifying drug treatment should be stopped 3 months before they attempt to conceive. Women with FH should be advised about the potential risks and benefits of re-starting lipid-modifying drug treatment for the mother and breastfed infant. Resins are the only lipid-modifying drug treatment that should be considered during breastfeeding.

# Quality statement 7: Drug treatment in children

## Quality statement

Children with familial hypercholesterolaemia (FH) are assessed for lipid-modifying drug treatment by a specialist with expertise in FH in a child-focused setting by the age of 10 years.

## Rationale

Children with FH begin to develop cardiovascular disease before clinical signs appear, with thickening of the carotid artery wall identifiable by the age of 10 years. Once a child is diagnosed as having FH, it is important they should be assessed for lipid-modifying drug treatment as soon as possible. The assessment should include a discussion of the harms and benefits of different treatments. This allows children to start treatment as soon as it is appropriate and before significant atherosclerosis has developed.

## Quality measures

The following measures can be used to assess the quality of care or service provision specified in the statement. They are examples of how the statement can be measured, and can be adapted and used flexibly.

### Structure

Evidence of local arrangements to ensure that children with FH are assessed for lipid-modifying drug treatment by a specialist with expertise in FH in a child-focused setting by the age of 10 years.

**Data source:** Local data collection.

### Process

Proportion of children with FH who are assessed for lipid-modifying drug treatment by a specialist with expertise in FH in a child-focused setting by the age of 10 years.

Numerator – The number of people in the denominator assessed for lipid-modifying drug treatment by a specialist with expertise in FH in a child-focused setting.

Denominator – The number of children with FH aged 10 years.

**Data source:** Local data collection.

## What the quality statement means for different audiences

**Service providers** ensure that systems are in place for children with FH to be assessed for lipid-modifying drug treatment by a specialist with expertise in FH in a child-focused setting by the age of 10 years.

**Specialists with expertise in FH in children and young people** assess children with FH for lipid-modifying drug treatment in a child-focused setting by the age of 10 years.

**Commissioners** ensure that they commission services in which specialists with expertise in FH assess children with FH for lipid-modifying drug treatment, in a child-focused setting, by the age of 10 years.

**Children with FH** have an assessment for possible drug treatment to reduce the low-density cholesterol (bad cholesterol) in their blood by a specialist in a children's department, by the age of 10 years.

## Source guidance

Familial hypercholesterolaemia: identification and management. NICE guideline CG71 (2008, updated 2019), recommendation 1.3.1.20

# Definitions of terms used in this quality statement

## Familial hypercholesterolaemia (FH)

FH relates to heterozygous FH only.

## Assessment for lipid-modifying drug treatment

The decision to offer or defer lipid-modifying drug treatment for a child or young person should take into account their age, age at onset of coronary heart disease within the family and the presence of other cardiovascular risk factors, including their LDL-C concentration.

[Adapted from NICE's guideline on familial hypercholesterolaemia, recommendation 1.3.1.18]

## Specialist with expertise in FH in children and young people

A healthcare professional with expertise in FH in children and young people who has access to the wider skills of a multidisciplinary team. This team should include a dietitian, cardiologist and paediatrician, and a clinical genetic specialist to take a family history and obtain informed consent for a DNA test.

## Child-focused setting

A child-focused setting is defined as valuing the child's view and validating their voice in making decisions impacting their lives. A child-focused facility or space is one designed with the children's needs in mind.

[Adapted from NICE's guideline on familial hypercholesterolaemia, terms used in this guideline]

# Equality and diversity considerations

Boys and girls should have equal access to lipid-modifying drug treatment for FH. There is anecdotal evidence that some clinicians are less likely to recommend statins in a girl than a boy at the same level of individual risk, arguing that because the age at onset of

coronary heart disease in women is later than in men, the start of drug treatment can be 'safely' delayed until adulthood. However, many young women will need to stop statins for several years when they are trying to conceive, and during pregnancy and breastfeeding. Because the accumulation of LDL-C burden is similar in boys and girls, this will result in the exposure to high LDL-C being greater in early adulthood in young women than in their male siblings. Gender should not influence a clinician's decision to offer treatment; the decision should be made in accordance with the recommendations in NICE's guideline on familial hypercholesterolaemia, which indicate that lipid lowering with statins should be considered by the age of 10 years.

# Quality statement 8: Annual review

## Quality statement

People with familial hypercholesterolaemia (FH) are offered a structured review at least annually.

## Rationale

Regular structured review enables treatment to be monitored and adjusted to achieve the recommended LDL-C concentration. It also enables monitoring for the possible development of symptoms and signs of coronary heart disease and optimising management. Records can be maintained of affected family members and information can be tailored to individual circumstances. Progress with cascade testing of at-risk relatives can be monitored.

## Quality measures

The following measures can be used to assess the quality of care or service provision specified in the statement. They are examples of how the statement can be measured, and can be adapted and used flexibly.

### Structure

Evidence of local arrangements to ensure that people with FH are offered a structured review at least annually.

**Data source:** Local data collection.

### Process

Proportion of people with FH who receive a structured review at least annually.

Numerator – The number of people in the denominator who had a structured review within

12 months of the last review or diagnosis.

Denominator – The number of people with FH.

**Data source:** Local data collection using a dedicated database.

# What the quality statement means for different audiences

**Serviceproviders** ensure that systems are in place for people with FH to be offered a structured review at least annually.

**Healthcarepractitioners** offer people with FH a structured review at least annually.

**Commissioners** ensure that they commission services that offer a structured review at least annually to people with FH.

**People with FH** are offered a detailed review of their condition at least once a year.

# Source guidance

Familial hypercholesterolaemia: identification and management. NICE guideline CG71 (2008, updated 2019), recommendations 1.3.1.27, 1.4.2.1, 1.4.3.2, and 1.5.1.1 to 1.5.1.5

# Definitions of terms used in this quality statement

## Familial hypercholesterolaemia (FH)

FH relates to heterozygous FH only.

## People with FH

People should have a diagnosis of FH made by a specialist with expertise in FH.

## Structured review

Structured review should include all the following components if appropriate:

- recording progress of cascade testing among relatives
- updating the family pedigree and noting changes in the coronary heart disease status of relatives
- assessing any symptoms of coronary heart disease
- assessing smoking status, and offering advice and information on smoking cessation services
- measuring fasting lipid profile
- discussing adherence to treatment, and possible side effects of treatment the person may be experiencing
- discussing any changes in lifestyle or lipid-modifying drug treatment that may be needed to achieve the recommended LDL-C concentration
- giving advice on contraception and pregnancy (to women and girls only)
- monitoring growth and pubertal development (in children and young people only).

# Update information

**November 2017:** Changes were made to definitions and source guidance sections throughout to ensure alignment with the updated NICE guideline on familial hypercholesterolaemia: identification and management.

# About this quality standard

NICE quality standards describe high-priority areas for quality improvement in a defined care or service area. Each standard consists of a prioritised set of specific, concise and measurable statements. NICE quality standards draw on existing NICE or NICE-accredited guidance that provides an underpinning, comprehensive set of recommendations, and are designed to support the measurement of improvement.

Expected levels of achievement for quality measures are not specified. Quality standards are intended to drive up the quality of care, and so achievement levels of 100% should be aspired to (or 0% if the quality statement states that something should not be done). However, this may not always be appropriate in practice. Taking account of safety, shared decision-making, choice and professional judgement, desired levels of achievement should be defined locally.

Information about how NICE quality standards are developed is available from the NICE website.

See our webpage on quality standards advisory committees for details about our standing committees. Information about the topic experts invited to join the standing members is available from the webpage for this quality standard.

NICE has produced a quality standard service improvement template to help providers make an initial assessment of their service compared with a selection of quality statements. This tool is updated monthly to include new quality standards.

NICE guidance and quality standards apply in England and Wales. Decisions on how they apply in Scotland and Northern Ireland are made by the Scottish government and Northern Ireland Executive. NICE quality standards may include references to organisations or people responsible for commissioning or providing care that may be relevant only to England.

# Diversity, equality and language

Equality issues were considered during development and equality assessments for this quality standard are available. Any specific issues identified during development of the quality statements are highlighted in each statement.

Good communication between healthcare practitioners and people with familial hypercholesterolaemia (FH) is essential. Treatment, care and support, and the information given about it, should be culturally appropriate. It should also be accessible to people with additional needs such as physical, sensory or learning disabilities, and to people who do not speak or read English. People with FH should have access to an interpreter or advocate if needed.

Commissioners and providers should aim to achieve the quality standard in their local context, in light of their duties to have due regard to the need to eliminate unlawful discrimination, advance equality of opportunity and foster good relations. Nothing in this quality standard should be interpreted in a way that would be inconsistent with compliance with those duties.

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# Endorsing organisation

This quality standard has been endorsed by NHS England, as required by the Health and Social Care Act (2012)

# Supporting organisations

Many organisations share NICE's commitment to quality improvement using evidence-based guidance. The following supporting organisations have recognised the benefit of the quality standard in improving care for patients, carers, service users and members of the public. They have agreed to work with NICE to ensure that those commissioning or providing services are made aware of and encouraged to use the quality standard.

- HEART UK

- Royal College of General Practitioners (RCGP)
- Royal College of Nursing (RCN)