Lipid Management for Familial Hyperlipidaemia in Adults

The guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer.

Familial Hyperlipidaemia (FH) is a genetic condition resulting in exceptionally high total cholesterol and LDL levels. People with FH are at high risk of premature CV disease and therefore require aggressive lipid-lowering therapy.
The diagnosis of FH should be confirmed by a specialist in line with the SLCSN FH Pathway

Lifestyle Advice and Blood Pressure Control

The following lifestyle issues should be addressed alongside statin therapy:

- Smoking cessation
- Diet (reduce saturated fats, include Mediterranean diet and oily fish twice a week, aim for body mass index (BMI) of 19 – 25kg/m^2, or a minimum of a 10% reduction in body weight)
- Alcohol moderation to within safe limits (up to 21 units per week for men and 14 units per week for women)
- Exercise (aim for a total of 30 minutes of moderate intensity physical activity (eg, brisk walking) at least 5x a week)

Blood pressure control - Treat if BP consistently over 140/90mmHg to achieve a BP of less than 140/90mmHg; more aggressive targets apply in patients with chronic kidney disease and diabetes

Initiating Therapy

Initiate a statin first-line in all patients with a diagnosis of familial hyperlipidaemia

First line choice: Simvastatin at a dose of 40mg with the evening meal.
Where simvastatin 40mg is contraindicated or not tolerated or there are drug interactions which limit the dose*, initiate a lower dose of simvastatin or consider an alternative agent, such as atorvastatin 20mg daily initially.

**Max dose 10mg daily with concomitant lipid-lowering dose of niacin*
**Max dose 20mg daily with concomitant amlodipine, diltiazem, amiodarone or verapamil*

Next Steps

- Repeat fasting lipid levels within three months of initiation
- Reinforce lifestyle issues and check adherence to medication
- **The aim of lipid lowering in FH is to achieve <u>at least</u> a 50% reduction in LDL from baseline. In patients not achieving this on simvastatin 40mg daily, consider switching to a higher intensity statin – i.e. atorvastatin 40mg daily, increasing to atorvastatin 80mg daily**
- In patients requiring additional LDL lowering despite higher intensity statin at maximal dose (or maximum tolerated
- dose), consider the addition of ezetimibe 10mg daily
- If at least a 50% reduction in LDL cholesterol is not achieved on higher intensity statin at maximal dose (or maximum tolerated dose) in combination with ezetimibe - refer for specialist advice
- If statin therapy is contraindicated or not tolerated, consider offering a fibrate, anion exchange resin or ezetimibe as an alternative
- FH patients should be reviewed annually, with lipid monitoring, to check efficacy and on-going adherence to therapy, and every five years should have a formal review with consideration given to seeking specialist advice if appropriate

For more information on contraindications and cautions to statin therapies, common drug interactions with statins and for guidance on safety monitoring – see SLCSN Guidance on Prescribing Statin Therapies, at www.slcsn.nhs.uk/prescribing.html. A lipid modification FAQ document can also be found on the website.

References

1. NICE Clinical Guideline 71: Familial Hyperlipidaemia. Aug 2008
2. MHRA Drug Safety Update Vol 6 Issue 1 - Simvastatin: updated advice on drug interactions – updated contraindications Aug 2012

South London Cardiac and Stroke Networks

Management of Lipids in Familial Hyperlipidaemia

This algorithm applies only to adults with Familial Hyperlipidaemia

Identify and address all modifiable risk factors:
Smoking, diet, alcohol intake,
BP control and physical activity

- **Offer SIMVASTATIN 40mg daily**
- If simvastatin 40mg daily is contraindicated or not tolerated or there are potential drug interactions which limit the dose, initiate a lower dose of simvastatin or offer an alternative agent, such as atorvastatin 20mg daily*

***For further information, please see SLCSN guidance Simvastatin: Recommendations following revised MHRA safety advice which is available at www.slcsn.nhs.uk**

- Repeat fasting lipid profile within 3 months and check adherence to therapy
- **In patients not achieving at least a 50% reduction in LDL cholesterol from baseline, consider switching to a higher intensity statin, such as atorvastatin 40mg daily, increasing to 80mg daily**
- Consider the addition of ezetimibe if higher intensity statin does not deliver the required reduction in LDL cholesterol.
- If at least a 50% reduction in LDL cholesterol is not achieved on higher intensity statin at maximal dose (or maximum tolerated dose) in combination with ezetimibe - refer for specialist advice

Routine safety and efficacy monitoring should be undertaken – see SLCSN Guidance on Prescribing Statin Therapies

FH patients should be reviewed annually, with lipid monitoring; to check efficacy and on-going adherence to therapy and have a formal five year review with specialist advice, in line with SLCSN FH pathway

Lifestyle issues should be revisited regularly

If statin therapy is contraindicated or not tolerated, consider offering a fibrate, anion exchange resin or ezetimibe as an alternative