DUTCH LIPID CLINIC NETWORK (DLCN) CRITERIA FOR DIAGNOSING HETEROZYGOUS FAMILIAL HYPERCHOLESTEROLEMIA IN ADULTS[2]

Patient name .....

LDL-C level ..... **Age** .....

Familial hypercholesterolemia (FH) is a common genetic condition characterised by **very high plasma levels of low-density lipoprotein cholesterol (LDL-C)** from birth and **~20-fold higher risk** of premature coronary heart disease (CHD); however, **FH is currently under-diagnosed**.[1-3]

- Formal clinical diagnosis can be made using the DLCN criteria which take into consideration levels of LDL-C, a molecular FH diagnosis, clinical history, family history and clinical signs that may be present in some patients with FH[1,2]
- Add up the score for each category (e.g. family history, clinical history, LDL-C level etc.) to determine the diagnosis
- If a patient has two criteria in one category, only the highest score is counted. For example, when CHD and tendon xanthoma as well as dyslipidaemia are present in a family, the highest score for family history is 2
- 'Premature' is defined as <55 years for men; <60 years for women

Category	Points	Your patient's score
Family history		
First-degree relative with known premature (<55 years, men; <60 years, women) coronary heart disease (CHD) OR	1	
First-degree relative with known LDL-C >95th percentile by age and gender for country	1	
First-degree relative with tendon xanthoma and/or corneal arcus OR	2	
Child(ren) <18 years with LDL-C >95th percentile by age and gender for country	2	
Clinical history		
Subject has premature CHD	2	
Subject has premature cerebral or peripheral vascular disease	1	
Physical examination		
Tendon xanthoma	6	
Corneal arcus in a person <45 y	4	
LDL-C level		
>8.5 mmol/L (>325 mg/dL)	8	
6.5–8.4 mmol/L (251–325 mg/dL)	5	
5.0–6.4 mmol/L (191–250 mg/dL)	3	
4.0–4.9 mmol/L (155–190 mg/dL)	1	
Molecular genetic testing (DNA analysis)		
Causative mutation shown in the *LDLR, APOB* or *PCSK9* genes	8	
Your patient's total score		

LDLR = LDL receptor; *APOB* = apolipoprotein B; *PCSK9* = proprotein convertase subtilisin/kexin type 9

Total score	Diagnosis
>8 points	Definite FH
6–8 points	Probable FH
3–5 points	Possible FH
0–2 points	Unlikely FH

Recommendations for further screening and testing[2]

- **Genetic testing is strongly recommended** among individuals with a definite or probable diagnosis of FH (DLCN score >5) and particularly in those with an obvious clinical diagnosis with xanthoma and/or high cholesterol plus a family history of premature CHD
- In patients with a confirmed diagnosis of FH, **cascade screening of family members is recommended** using LDL-C levels or genetic testing if the causative mutation is found, in conjunction with DLCN system
- All subjects with FH and their families should undergo **intensive education targeting lifestyle** management
- **Most FH patients will need active treatment** as their risk is significantly higher than the rest of the population due to their long exposure to high LDL-C levels

Your patient's total score	
Your patient's diagnosis	
Family screening initiated?	YES / NO

Further information on FH for the patient

- Please refer the patient to the booklet entitled 'A guide to familial hypercholesterolemia (FH) for patients'
- Provide details of local patient associations

NOTES

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References

1. World Health Organization. Familial hypercholesterolemia Report of a second WHO Consultation Available at: http://whqlibdoc.who.int/hq/1999/WHO_HGN_FH_CONS_99.2.pdf
2. Nordestgaard B et al. Eur Heart J. 2013;34:3478-90a
3. Goldberg et al. J Clin Lipidol. 2011;5(3 Suppl):S1-8