



SEVERE FH

A proportion of patients with familial hypercholesterolemia present with severe metabolic disorders during the clinical course. In some cases, this may obscure the clinical picture of the heterozygous and homozygous forms of familial hypercholesterolemia. This disease, known as Severe FH, requires exceptional attention. Patients with severe FH should be targeted for maximally intensive treatment which, after diagnosis, should commence as soon as possible (regardless of age). The diagnostic criteria for Severe FH are presented below¹.

Criteria for Severe FH

Lipidogram without treatment:

1. **LDL-C > 10 mmol / L**
2. **Or LDL-C > 8 mmol / L + 1 of the risk factors (see below)**
3. **Or LDL-C > 5 mmol / L + 2 of the risk factors (see below)**
(LDL-C = LDL cholesterol)

The risk factors that contribute to the diagnosis of Severe FH are presented below. They are all common risk factors for development of the atherosclerotic process, although some (e.g. reduced glomerular filtration), are not widely known as such. An additional risk includes patients who have not been treated prior to the age of 40, by which time atherosclerotic processes may have already developed.



Risk factors:

40 years of age without any prior treatment

Smoking

Male

Lp(a) > 50 mg / dL

HDL < 1 mmol / L

Arterial hypertension

Diabetes mellitus

Men < 55 years and women < 60 years, with first-degree relatives having a family history of atherosclerosis

A family history of early atherosclerosis in first-degree relatives (men < 55 years and women < 60 years)

BMI > 30 kg / m²

GFR < 60 mL / min / 1.73 m²

¹ Defining severe familial hypercholesterolaemia and the implications for clinical management: a consensus statement from the International Atherosclerosis Society Severe Familial Hypercholesterolemia Panel. Santos RD, Gidding SS, Hegele RA et al. *Lancet Diabetes Endocrinol.* 2016 May 27. pii: S2213-8587(16)30041-9. doi: 10.1016/S2213-8587(16)30041-9.