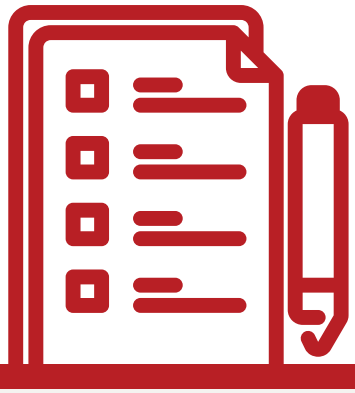
Calculate a familial chylomicronemia syndrome (FCS) score for your patient[1]



Published in 2024 by the *Journal of Clinical Lipidology*, the North American FCS (NAFCS) scoring criteria were developed and validated by a panel of experts with the intent of minimizing diagnostic delays, expediting access to new management options, and improving patient care for FCS in the United States and Canada. The calculator uses signs, symptoms, and biochemical traits of FCS to help determine a score that predicts the likelihood of FCS in a patient and may potentially distinguish FCS from multifactorial chylomicronemia syndrome, another genetic form of severe hypertriglyceridemia. Look inside for more details about calculating an FCS score.

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Calculating a score for FCS[1]

Fill in values for each category below and add category scores to determine the NAFCS score and help facilitate a diagnosis of FCS. This calculator is designed for use in patients ≥1 year old with hypertriglyceridemia (>440 mg/dL). In patients ≥10 years old, the calculator is intended for patients who are not responsive to fibrates and high-dose omega-3 fatty acids even when the patient is adherent to therapy (ie, triglycerides do not decrease by 20% or more from these treatments and do not remain reduced). The NAFCS Score does not provide a value for pregnant patients.

Characteristic	Patient data	Category score
Current age <1* ; ≥1-9 **[+12]** ; ≥10 **Hypertriglyceridemia onset (years)** Only asked of patients ≥10 <10 **[+12]** ; ≥10 **[+0]**		+
Body mass index (percentile for children/adolescents) <25 kg/m^2 or <85th percentile **[+9]** ; ≥25 kg/m^2 or ≥85th percentile **[+0]**		+
History of pancreatitis Pancreatitis **[+16]** ; Abdominal pain but no pancreatitis **[+9]** ; Neither abdominal pain nor pancreatitis **[+0]**		+
Presence of secondary factors[2†] that may contribute to hypertriglyceridemia None **[+11]** ; ≥1 **[+0]**		+
Laboratory values (select if true) All triglycerides >880 mg/dL **[+13]** ; Ratio triglycerides/total cholesterol >8‡ **[+8]** ; Apolipoprotein B (apoB) <1.0 g/L **[+12]**		+
Additional points added: If patient is ≥1-9 years old AND has no secondary factors **[+7]** If triglycerides/total cholesterol >8‡ AND apoB <1.0 g/L **[+7]** If triglycerides/total cholesterol >8‡ AND has no secondary factors **[+5]**		=
Interpreting results **≥60** strongly indicates definite FCS **≥45** suggests likely FCS rather than MCS **≥30-44** suggests rationale to confirm FCS with genetic testing **<30** deems unlikely FCS, but genetic testing may still be needed	**Result**	

Adapted from Hegele et al. *J Clin Lipidol*. Published online November 12, 2024. doi:10.1016/j.jacl.2024.09.008[1]

*Calculator cannot be used for patients <1 year old. If infant presents with no secondary factors that may contribute to hypertriglyceridemia, consider a diagnosis of FCS. If infant presents with ≥1 secondary factor that may contribute to hypertriglyceridemia, but with 2 triglyceride readings >880 mg/dL and unexplained failure to thrive, consider a diagnosis of FCS.[1]

†See next page for complete list.[2]

‡When measured in mg/dL.[1]

FCS=familial chylomicronemia syndrome; MCS=multifactorial chylomicronemia syndrome.

Secondary factors that may contribute to hypertriglyceridemia[2]

	Adolescents/adults	Children	Infants
Lifestyle	• High alcohol intake • Reduced physical activity • High fat or sugar intake • Ultraprocessed (NOVA4) food diet	• High fat or sugar intake • Ultraprocessed (NOVA4) food diet	
Clinical conditions	• Autoimmune chylomicronemia (eg, systemic lupus erythematosus, anti-lipoprotein lipase antibodies) • Uncontrolled diabetes type 1 or 2 (non-pancreatitis induced) • Endocrine causes (eg, Cushing's syndrome, polycystic ovarian syndrome, growth hormone deficiency, acromegaly) • Human immunodeficiency virus (HIV) • Hypothyroidism • Metabolic syndrome • Rare genetic disorders (eg, glycogen storage disorders) • Lipodystrophies • Renal causes (eg, chronic renal failure, nephrotic syndrome) • Glycerol kinase deficiency • Autoantibodies against GPI-HDL binding protein 1	• Cancer • Congenital nephrotic syndrome • Glycogen storage disease type 1 • Hypothyroidism • Protein-losing enteropathy • Lipodystrophies • Glycerol kinase deficiency	• Cancer • Congenital nephrotic syndrome • Hypothyroidism • Protein-losing enteropathy • Glycerol kinase deficiency
Medications	• Androgen deprivation therapy • Antidepressants (eg, sertraline) • Antiretrovirals (eg, protease inhibitors) • Atypical antipsychotics • Beta-adrenergic blocking agents • Bile acid binding resins • Diuretics • Estrogen, estrogen receptor agonists, estrogen receptor modulators, oral contraceptives • Glucocorticoids (eg, corticosteroids) • Immunosuppressants (eg, cyclosporine) • L-asparaginase • Propofol • Retinoids, retinoid X receptor agonists • Total parenteral nutrition	• Total parenteral nutrition • Glucocorticoids (eg, corticosteroids) • Chemotherapy • Antiretrovirals (eg, protease inhibitors) • Immunosuppressants (eg, cyclosporine) • Propofol • For older children: — Antidepressants (eg, sertraline) — Atypical antipsychotics	• Total parenteral nutrition • Glucocorticoids (eg, corticosteroids) • Chemotherapy

Adapted from Hegele et al. *J Clin Lipidol*. Published online November 12, 2024 (online-only supplementary material). doi:10.1016/j.jacl.2024.09.008[2]

The information in this brochure is intended as educational information for healthcare professionals. It does not replace a healthcare professional's judgment or clinical diagnosis.

Contact your **Ionis Rare Disease Account Specialist** or visit **TGAware.com** to learn more about diagnosing FCS and ways to manage FCS.

Visit **TGAware.com** to learn more about the dangerous consequences of FCS.

FCS=familial chylomicronemia syndrome.

References: 1. Hegele RA, Ahmad Z, Ashraf A, et al. Development and validation of clinical criteria to identify familial chylomicronemia syndrome (FCS) in North America. *J Clin Lipidol*. Published online November 12, 2024. doi:10.1016/j.jacl.2024.09.008 **2.** Hegele RA, Ahmad Z, Ashraf A, et al. Development and validation of clinical criteria to identify familial chylomicronemia syndrome (FCS) in North America. *J Clin Lipidol*. Published online November 12, 2024 (online-only supplementary material). doi:10.1016/j.jacl.2024.09.008

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