A PATIENT'S GUIDE

Familial Chylomicronemia Syndrome (FCS)

The National Pancreas Foundation

What is FCS?

- **FCS is a rare genetic disorder** that affects how your body handles triglycerides, or fat metabolism.
- Someone in your family must be a carrier for you to have FCS. This can be confirmed through **genetic testing or a clinical assessment**.
- Because FCS is so rare, diagnosis is often **delayed or mistaken** for other conditions.

What the name means

Familial — inherited; it runs in families.

Chylomicronemia — large fat droplets, called chylomicrons, build up in the bloodstream.

Syndrome — a group of symptoms that consistently occur together.

Possible signs & symptoms

- Abdominal pain
- Acute pancreatitis
- Brain fog
- Swelling of the liver or spleen
- Lipemic or "milky" blood
- Eruptive xanthomas*

**Eruptive xanthomas are pea-sized, pink bumps (almost like acne) that develop on the hands, feet, arms and legs.*

880+

mg/dL — fasting triglyceride level that may point to FCS

How does FCS put me at risk for pancreatitis?

- With FCS, the lipoprotein lipase (LPL) enzyme is missing, which leads to fat building up in the blood.
- These extremely high triglycerides accumulate around the pancreas, causing inflammation — pancreatitis.

Diagnosing FCS

Because FCS is so rare, diagnosis is often delayed or mistaken for other conditions. Common diagnostic steps include:

1. **Fasting lipid panel** to check for extremely high triglycerides of 880 mg/dL or higher
2. **See a lipidologist**, a specialist in cholesterol and triglyceride disorders
3. **Rule out secondary causes** of high triglycerides, such as diabetes, alcohol use or medications
4. **Genetic testing** using a panel of 5 genes involved in LPL activity to confirm variants causing FCS
5. **The North American FCS Scoring Tool** can help evaluate if someone may have FCS

Scan to view the tool

Living with FCS

The National Pancreas Foundation

Managing FCS

Every patient living with FCS is different — what works for one does not always work for another. One reason living with FCS can be so difficult is that it does not follow any of the typical rules.

1. **Traditional triglyceride-lowering medicines, like fibrates**, don't work for FCS
2. **The FDA has approved the two medications specifically for FCS**

Other therapies may be available in the near future. Consult with your physician, ideally a **lipid specialist**, about the best treatment plan for you.

What nutrition may look like

<10–20 g
of fat per day — about one tablespoon of olive oil

Follow a very low-fat diet
Build meals around fruits, lean meat and vegetables.

All fats count
Even "healthy" or "good" fats can be dangerous and need to be included when tracking daily fat intake.

Limit sugar and carbohydrates
Avoid foods that are high in sugar and carbohydrates.

Ask about supplements
Talk to your care team about vitamin A, magnesium, MCT oil, calcium and other fat-soluble vitamins that can become deficient on a specialized diet.

Review your medications
Avoid medications that will increase your triglyceride levels.

It is VERY important to avoid alcohol and tobacco, as they can increase the likelihood of an acute pancreatitis episode.

Your care team

FCS requires a multidisciplinary care team, which may include an **endocrinologist, geneticist, dietitian, lipidologist, cardiologist, primary care physician, mental health provider and/or pancreatologist.**

Need help finding a lipid specialist? Try the **Find a Lipid Specialist** tool from the Foundation of the National Lipid Association.

Find a lipid specialist

Planning for pregnancy

If a family member has been diagnosed with FCS and you become pregnant, it is **critical to talk to your physician** about a possible diagnosis and management.

You're not alone

Diet limits, stigma, misdiagnosis and fear of pancreatitis can make FCS mentally and socially challenging. **Support groups** can offer community to you and your family.

THANK YOU TO OUR SPONSORS arrowhead pharmaceuticals IONIS

More FCS education & resources: PancreasFoundation.org/disease/familial-chylomicronemia-syndrome-fcs

PancreasFoundation.org info@pancreasfoundation.org 866.726.2737