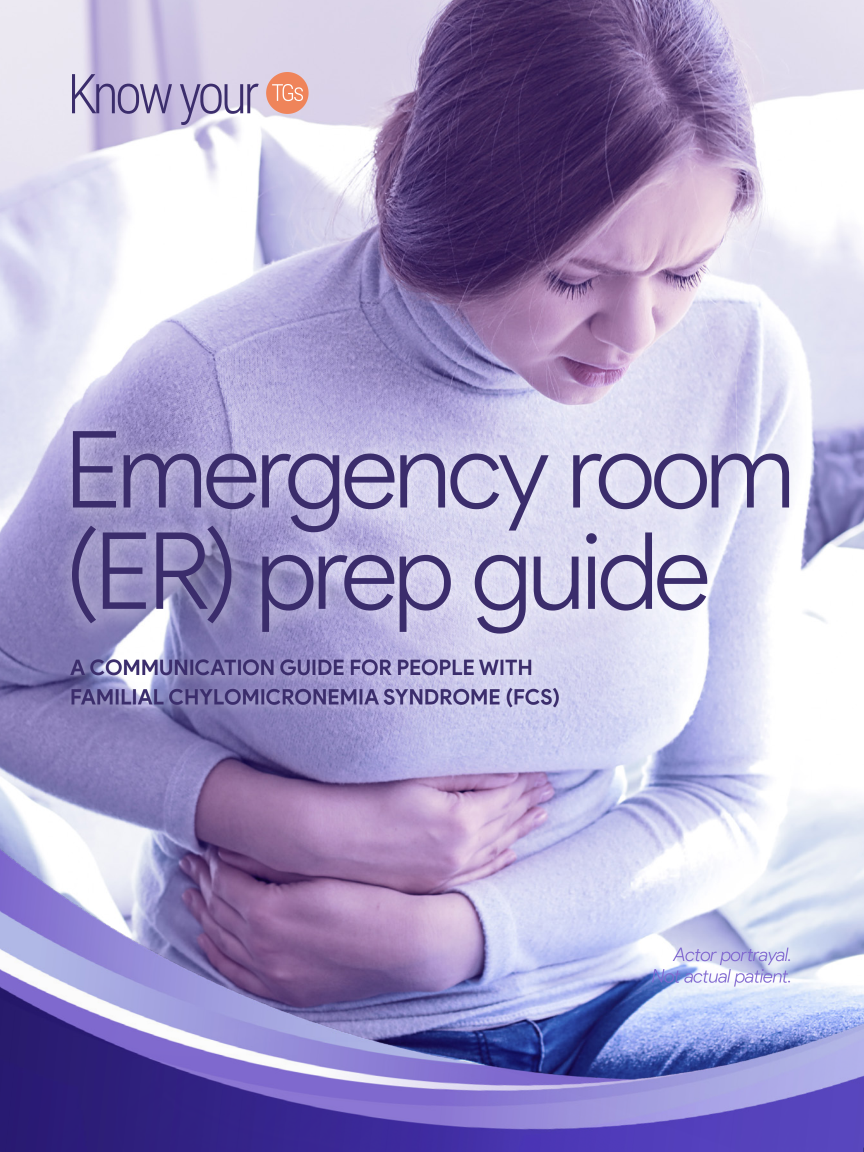
Know your TGs



Emergency room (ER) prep guide

A COMMUNICATION GUIDE FOR PEOPLE WITH FAMILIAL CHYLOMICRONEMIA SYNDROME (FCS)

Actor portrayal.
Not actual patient.

Actor portrayal.
Not actual physician
or patients.

If acute pancreatitis (AP) sends you to the ER, you'll be ready.

Acute pancreatitis caused by FCS can result in unexpected visits to the hospital. While there, you may meet healthcare professionals (HCPs) who do not know about FCS because it is an underdiagnosed rare genetic condition. That means you will have to tell them about FCS and advocate for your own care, all while dealing with the pain of acute pancreatitis (AP).

An ER visit can leave you feeling overwhelmed, confused, and impatient. We developed this guide with input from people living with FCS. It can help you and your caregivers prepare for a medical crisis before one happens, so you have greater confidence and less stress, right when you need it the most.

Know before you go

How does FCS cause acute pancreatitis?

FCS severely impairs the body's ability to remove triglyceride-carrying chylomicrons from the bloodstream. This often causes extremely high triglyceride (TG) levels.

As a result, TGs can build up in the pancreas until they trigger inflammation and damage, a painful condition called acute pancreatitis (often referred to as "AP"). Potentially life-threatening, AP can damage the pancreas and other organs, as well as cause or worsen diabetes.

Know how to recognize acute pancreatitis

Signs and symptoms include:

- Persistent or worsening pain in the upper abdomen, sometimes radiating to the back
- Nausea, vomiting, or the inability to eat
- Unexplained fatigue or feeling unusually weak
- Bloating or tenderness in the abdominal area
- Increased difficulty breathing or rapid shallow breath

If any of these occur, immediately contact your healthcare professional and/or go to the ER.

Before your visit

Actor portrayal.
Not actual physicians.

Have an ER go-bag ready to go

In the middle of an AP event, it's harder to think about what you should bring to the ER. And, if you're admitted to the hospital, you could stay there for 2 or more days – so pre-pack a bag with a clear head and have some items you might need.

Essentials include:

- Your FCS Emergency CareBook available at **KnowYourTGs.com**
- A letter from your doctor explaining your medical condition
- Insurance card/documents
- Phone charger with cable
- Medication bottles
- Hygiene supplies (toothbrush, toothpaste, deodorant, hairbrush, contact lens case and solution, etc.)

Items that may make your hospital stay more comfortable include:

- Loose-fitting clothing, warm socks, fresh underwear, and slip-on footwear
- Eye mask and earplugs
- Lip balm and hand lotion
- Headphones or earbuds
- Books or magazines

You may be tempted to pack food, but should begin fasting as soon as you think you are having an AP event. Also, the ER staff may not want you to even drink, so always check with them before you do. Only pack clear liquids like water, coconut water, or electrolyte drinks. Avoid red drinks during this time, as they can look like blood in vomit.

Plan where you are going before you have to go

Whether at home or away, always know the nearest in-network hospital covered by your insurance, if applicable. Emergency room visits can cost a lot of money, especially if they admit you to the hospital for multiple days. Out-of-network hospitals may result in denied claims or high out-of-pocket expenses.

Whenever possible, skip going to urgent care facilities (i.e. walk-in facilities not attached to a hospital). For acute pancreatitis, they will most likely tell you to go to the ER as they are not equipped to handle such conditions. And if they have already admitted you, they will have to send you by ambulance.

Have transportation on standby

The severe pain, nausea, and weakness of an AP event can make driving unsafe. Cost and hospital choice may make an ambulance less than ideal if you don't really need it.

If possible, pre-arrange transport to and from the hospital with a trusted friend, family member, or caregiver. Prepare them to talk about your condition in case you cannot do it yourself. And have a safe ride home after discharge, when you may be on pain medication that makes driving unwise.

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Not actual physician
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Don’t wait until it is too late: Keep your FCS Emergency CareBook up to date

Available at **KnowYourTGs.com**, this binder holds copies of info that can help ER staff more quickly understand your medical condition, history, and needs.

- Medical history sheet – which lists previous AP events, current medications, allergies, emergency contact, HCP list, and more
- Signed doctor’s letter
- “What is FCS” fact sheet
- QR code that busy ER staff can use to quickly find information about FCS
- Lipid Panel Tracker showing past lab result
- Advanced directive or living will, if applicable

PRO TIP!

Keep multiple copies of these sheets in the binder so ER staff can keep them for charts, medical records, etc. and you will still have the originals.

Make getting a signed doctor’s letter a top priority

The legitimacy of a professional FCS diagnosis from one of your HCPs may help prevent misdiagnosis and lead to faster, more accurate care – especially when it comes to an underdiagnosed condition like FCS that is not well known.

You can give such a letter to ER Registration and then follow-up with both the triage nurse and attending physician to make sure they have seen it and understand your condition. The sample letter is available on **KnowYourTGs.com** and can help guide your HCP in what to include in the letter, such as:

- Your diagnosis and a brief description of FCS
- Symptoms and complications
- Previous treatment history and emergency treatments that work best
- Medications or treatments to avoid
- HCP contact information and signature

This information can also help when you can’t communicate due to pain, confusion, or a neurological issue.

Prepare your healthcare team for an AP event

Develop an action plan with your HCPs for how you’ll handle a future AP event. For example, before heading to the ER, you could call your HCPs to let them you’re having an acute pancreatitis event. If they are willing to call ahead to the ER to provide key medical details about your condition, it may lead to faster, more accurate care, with less chance of misdiagnosis. And when your healthcare team has a record of your AP episodes they have a more complete understanding of the impact FCS is having on your health and quality of life.

Actor portrayal.
Not actual physicians.

During your visit

Set realistic expectations for the ER

The ER is often a fast-paced environment where medical staff must quickly assess and prioritize a high volume of patients based on the severity of their conditions.

1. **Registration.** Staff will collect basic information about your symptoms, medical history, and insurance, if applicable.
2. **Triage.** Triage nurses assess your condition by checking your vital signs, pain level, and symptoms to determine how urgently you need to be seen by a doctor.

- The ER does not operate on a first-come, first-serve basis. While acute pancreatitis is serious, there may be patients in even more critical condition who require attention before you do.
- Waiting times can vary. It may be a while before you see a triage nurse or receive treatment.

3. **Treatment.** After triage, you will either go to a treatment area or wait until a room becomes available. Once an HCP sees you, they may order blood tests, imaging such as an ultrasound or CT scan, and intravenous fluids or pain management. Lab results and imaging can take hours to process.
4. **Admission or discharge.** The ER team will assess the severity of your condition and determine whether you need to stay at the hospital or can leave with treatment recommendations.

Be prepared for bias and self-advocacy

ER staff might mistakenly think your symptoms are caused by alcohol abuse, gallstones, obesity, or lifestyle factors – or that you are just a drug abuser seeking pain medications. Because FCS is a rare genetic condition they may rarely, if ever, see someone in the ER with this disease.

As a result, they may not believe you when you tell them you haven't had any alcohol. They may treat you in a patronizing manner or seem like they blame you for your symptoms. They may look rushed or distracted. All of this is expected, given the stressful, fast-paced workplace where they make life-and-death decisions many times a day, for many days, weeks, and months in a row.

It is up to you to gain and hold their attention, directing it to what is best for you. Be firm, clear, persistent, and polite in helping them understand your condition, so they do not waste time or misdiagnose you. This pamphlet can help you prepare to advocate for yourself.

Guide your care with effective communication

Both you and anyone with you, like a caregiver, must be hyper-aware that language can help or hinder your progress through the ER process.

Practice using specific language and medical terms that will build credibility and grab their attention, such as:

- *"I believe I'm experiencing an acute pancreatitis event.*
- *"This has happened before due to my rare genetic condition."*
- *"I have familial chylomicronemia syndrome, which causes life-threatening hypertriglyceridemia."*
- *"Due to this rare condition, I must regularly monitor my triglyceride levels.*
- *I have had extremely high triglycerides in the past, up to [#] milligrams per deciliter."*
- *"I am not an alcoholic and I have not been drinking."*
- *"It's important that you understand that normal levels of amylase and lipase do not necessarily exclude acute pancreatitis in an FCS patient."*

ER staff often have little time for a lot of patients, so focus on concise, key details, such as:

- FCS being an underdiagnosed, genetic form of hypertriglyceridemia
- Your past history of high triglycerides and AP events
- The specific symptoms or history relevant to the current visit
- A summary of your typical treatment plan (excluding drug-seeking language)

Avoid drug-seeking language that can slow down your care

You (and anyone with you) will only want to see your pain go away. But going straight to pain relief will usually get you labeled as someone who is only seeking drugs and not really ill. That can lead to much longer wait times, so **avoid phrases like**:

"[Pain medication name] has worked for me in the past."

"I need pain relief right now."

"I need [pain medication name]."

"Can't you do anything for the pain now?"

Instead, focus on your condition using some of the suggestions previously mentioned to build credibility with hospital staff. That may lead to getting the pain relief you need more quickly. Try:

- Talking about your condition using specific language and medical terms
- Providing a signed doctor's letter

Try to keep calm & focus on relief

While waiting, stay as still as possible to help reduce pain. Practice slow, deep breathing to calm yourself. Use visualization techniques such as picturing yourself in a serene place or watching the pain leave your body. If allowed, hold a warm blanket for comfort and focus on a loved one's voice. Above all, remind yourself that help is on the way. Stay strong and hold on!

Actor portrayal. Not actual physician or patient.

After your visit

Actor portrayal. Not actual patient.

Continue your care after leaving the hospital

- Talk to your doctor about the potential side effects of any medications they may prescribe.
- Carefully review discharge instructions to understand dietary restrictions, medication changes, and follow-up care.
- Stick to a strict low-fat diet and avoid alcohol.
- Examine what might have caused the recent AP event. Did your diet change? Did you start taking any new medication or supplements?
- Talk to your primary care physician and/or specialist(s) to ensure continuity of care and help minimize the recurrence of AP episodes.
- Work with your healthcare team to confirm all prescribed and over-the-counter medications are correct, safe, and appropriate; adjust as recommended.
- Schedule any needed follow-up appointments and lab tests to help monitor triglyceride levels and overall health.
- Set realistic pain management goals based on what your body is going through.
- Get help from family or caregivers if under the influence of pain medications.

PRO TIP! Get a medical alert bracelet

ER staff are often trained to take medical alert bracelets seriously. You can have one made that lists your condition as familial chylomicronemia syndrome and links to an informative website made for HCPs, such as TGAware.com.


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NOTES

Visit **knowyourtgs.com** to learn more about about FCS symptoms, acute pancreatitis, genetics and more.

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