Sickle Cell Disease – Pain Crises

Pain is one of the most common and distressing symptoms that affects sickle cell patients. The more you know about the causes, prevention, and treatment of pain, the better your chances for an early recovery.

Preventing a Pain Crisis

In general, any situation that increases the body’s requirements for oxygen can cause a pain crisis. This can include illness, physical stress, or being at high altitudes. Pain crisis can also be brought on by extremes in temperature (being very hot or very cold) and dehydration.

While some causes of a pain crisis are beyond your control, most of them can be prevented with proper self-care. Having sufficient rest, warmth, and fluid intake are the most important ways you can prevent a pain crisis.

Important ways you can help prevent a pain crisis:

- Dress appropriately when heading outdoors to exercise, play or work to avoid getting overheated or too cold.
- Drink plenty of healthy fluids and always carry a water bottle with you.
- Listen to your body and rest when you feel you need to.
- Avoid cold swimming pools and very warm hot tubs.
- Avoid excessive physical activity and severe emotional stress.
- Do not smoke, and avoid exposure to second hand smoke. Both active and passive smoking may trigger acute chest syndrome in sickle cell patients.
- Take extra precautions at high altitudes and be sure that you only travel by pressurized airplanes.

Remember the acronym FARMS to help prevent pain crises:

F: Fluids and Fever – Drink plenty of water, and if you get a fever see your health care provider immediately.
A: Air – Make sure you do not get into situations where there is not enough oxygen, like flying in an unpressurized airplane.
R: Rest – Get plenty of sleep. Don’t overdo it and take breaks when your body feels tired.
M: Medications – Take your preventative medications, like daily penicillin to fight infections, hydroxyurea for pain prevention, and folic acid to help make red blood cells.
S: Situations – Avoid situations where it is too hot or too cold, and avoid smoking, alcohol and illegal drugs.

Treating a Pain Crisis

Despite the best attempts at prevention, pain crises can still happen. Pain from sickle cell disease can be acute or chronic, but acute pain is more common. Pain crises can come on very suddenly, can range from mild to severe, and can last from hours to weeks. Depending on the severity of the pain, treatment options vary from home management to hospitalization.

It is best to try to treat the crisis at the earliest sign of pain. The 3 best ways to treat a pain crisis at home are to (1) rest, (2) drink lots of fluids, and (3) take pain medication. At the first sign of pain one should drink plenty of water then lie down to rest. Sometimes taking a warm bath, using a warming pad and using distractions such as music, TV or other relaxation techniques can help.

If your health care provider has given you pain medication, start taking it as prescribed. The different kinds of pain medications include acetaminophen (Tylenol), anti-inflammatory agents (Advil, Naproxen) and narcotics (codeine, morphine, hydromorphone). These pain medications all work in different ways, and sometimes you can combine them to get better pain relief. However, be sure to talk with your hematology team prior to combining medications.

Recording pain in a diary can be helpful. It gives you and your health care providers important information about your pain. The acronym LOCATES will help you remember the information a pain diary should include:

L: Location – where is the pain located? (joints, back, abdomen, etc.)
O: Other Symptoms – any other symptoms besides the pain? (fever, nausea, cough, etc.)
C: Character – is the pain burning, deep, sharp, throbbing?
A: Aggravating and Alleviating measures – what makes the pain better or worse?
T: Timing – when did it start and does it come and go?
E: Environment and Effect – where were you and what were you doing when the pain started? How does the pain affect your daily routine?
S: Severity – rate how much pain you are having on a pain scale from 1-5 or 0-10

If home treatment fails or a warning sign is present, such as fever, weakness, new or different pain, headache, chest pain, or abdominal pain, contact your hematology team and immediately go to the emergency room.

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