Sickle cell disease: avascular necrosis (AVN)

What is avascular necrosis (AVN)?

Avascular necrosis (AVN) is a well-known complication of sickle cell disease. AVN:

- Affects up to 10 percent of persons with sickle cell disease.
- Can happen with all sickle cell types.
- Can also be called osteonecrosis.

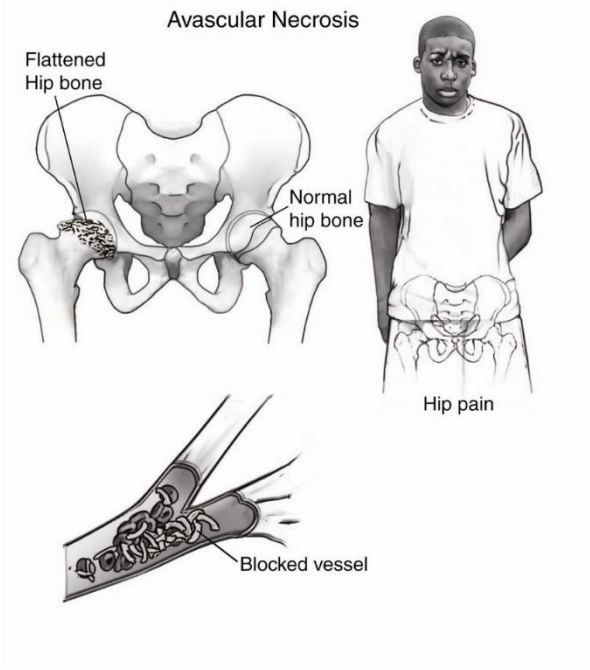
AVN is a type of bone damage that most often happens during the teen years. It is caused by a lack of blood supply to the bone.

- Avascular means no blood flow.
- Necrosis means death and decay of certain tissues in the body.

What causes it?

The exact cause for this type of bone damage is not clear. It is thought to be linked to abnormal blood flow from sickled red blood cells.

- This can cause damage and swelling of the blood vessels.
- Without a good blood supply, the bone tissue starts to die. This causes weakness that worsens over time and changes the normal shape of the bone.
- AVN can cause pain and loss of bone function.



In sickle cell disease, AVN mostly affects the joints of long bones, like the hip and shoulder. AVN may also affect other small bones in the body, like the sternum (breastbone) or vertebrae (spine).

- When AVN affects the hip joints, the round, smooth surface of the bones becomes flat and rough.
- These changes make it hard for the joints to move freely.
- This causes the area to become very sore, swollen and tender.
- Your child may complain of pain when they walk, climb stairs or move.

What are the symptoms?

Many children with early AVN may not have any symptoms. As AVN gets worse, your child may have some or all of these:

- Swelling, tenderness and soreness in the involved bone or joint area
- Pain, which can be sharp, dull or aching

In case of an urgent concern or emergency, call 911 or go to the nearest emergency department right away.

  - It may be different from a regular pain episode.
  - It could worsen with putting weight on the involved area (weight bearing) or moving (such as walking, climbing stairs or sitting for long periods of time).
- Stiffness or tightness in the involved joint
  - AVN can feel like arthritis pain.
  - Your child may limp when they walk.
  - There may be less movement of the joint that is in pain.

How do I know if my child has it?

If your child complains of bone or joint pain often, please call their sickle cell provider who can begin to check for AVN. This may include:

- A complete exam and questions about your child's health history.
- X-rays of the involved bones or joints. This may include the hips, shoulders or spine.
- An MRI, or detailed imaging study, to take pictures of the involved bones or joints.

What is the treatment?

Treatment for AVN is more helpful when started early. It may depend on how severe your child's symptoms are and the extent of bone or joint damage. Treatment may include:

- Anti-inflammatory medicines for joint pain and swelling, such as ibuprofen (Motrin or Advil), naproxen (Aleve) or celecoxib (Celebrex).
- Stronger pain medicines called opioids, such as hydrocodone (Norco or Lortab) or oxycodone (Roxicodone), if anti-inflammatory medicines do not work well.
- Bed rest and crutches or braces to help decrease pressure at large joints like the hip and shoulder.
- Physical therapy (PT) and exercises to help improve muscle strength around the affected joints.

If these treatments do not provide enough pain relief, your child may need to see a bone specialist (orthopedic surgeon).

- The bone specialist can complete an exam, view X-rays and MRIs, and decide if surgery is needed to remove some of the damaged bone.
- In severe cases of AVN, surgery may be needed. This could involve joint replacement, bone reshaping (osteotomy) or core decompression (removal of necrotic tissue).

How can I prevent AVN?

The exact cause of AVN in sickle cell disease is still not known.

- Regular physical activity may help your child. Mild, weight-bearing activity like walking can help to improve bone health.
- Helping your child maintain a healthy weight and normal body mass index (BMI) can decrease stress on the joints, especially the hips.

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- It may also decrease your child's chance of having AVN when you help them follow their sickle cell treatment plan to avoid other problems like pain crises and acute chest syndrome.

What should I do if my child is sick?

- Follow the sickle cell provider's advice for what you should do.
- FEVER IS A MEDICAL EMERGENCY. Your child should be seen by a provider **right away** if they have a fever of 101°F (38.3°C) or higher.
- During the time when the clinic is closed (between 5 p.m. and 8 a.m. during the week and on weekends and holidays), call the hematologist on call for concerns. They can help guide you with your child's care.
- ALWAYS tell the emergency department (ED) doctors and staff that your child has sickle cell disease. Also tell them which sickle cell clinic your child visits.

For more details on sickle cell disease and services, visit the Children's Healthcare of Atlanta website at choa.org/sicklecell.

This teaching sheet contains general information only. Talk with your child's doctor or a member of your child's healthcare team about specific care of your child.

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