Kienböck's Disease

Richard A. Bernstein, MD

Kienböck disease is a condition in which one of the small bones of the hand in the wrist called the lunate dies. We don't know what causes it. Bone is a living tissue that grows and changes, and it requires a regular supply of blood for nourishment. If the blood supply to a bone stops, the bone can die, a condition known as osteonecrosis.

Kienböck's disease is osteonecrosis of the lunate. It usually affects the dominant wrist of men aged 20-40 years, but can affect women, but usually later than in men. The true natural history (what happens if we do not treat it) of this condition is not well understood. Not only do we not know what causes it, we do not know why some get better by themselves and some get worse. We don't even know how many have minimal symptoms and get better by themselves, as doctors usually only see a patient after the symptoms are pretty bad. As you can imagine, this has hampered the determination of the ideal treatment. In addition, as with many conditions that affect the wrist, the patient's pain does not necessarily correlate well with the radiographic appearance: some people with almost normal x-rays hurt a lot, some with bad x-rays hardly hurt at all. (Note: the little "ö" is used because the disease was first described by a Viennese radiologist named Robert Kienböck, and that is how his name is spelled in English. Just thought you might want to know.)

Signs and symptoms

Many people with Kienböck's disease think they have a sprained wrist at first. They may have experienced some form of trauma to the wrist, such as a fall. This type of trauma can disrupt the blood flow to the lunate. In most people, two vessels supply blood to the lunate, but in some people there is only one source. Our best guess is that this puts them at greater risk for developing the disease (but as you can understand, we can't do an experiment and prove this!) As the disease progresses, other signs and symptoms may start, including:

- A painful and sometimes swollen wrist
- Limited range of motion in the affected wrist (stiffness)
- Decreased grip strength in the hand
- Tenderness directly over the lunate (on the top of the hand at about the middle of the wrist)
- Pain or difficulty in turning the hand upward

However, as you can see, these things also can occur with many other things, so they are not very specific.

Stages of Disease

Kienböck's disease follows a specific progressive pattern through five stages.

Stage 0: Symptoms are similar to those of a wrist sprain. X-rays are normal, magnetic resonance imaging (MRI) is normal. (We only know this stage retrospectively, that is, later down the road, when more symptoms occur and more x-rays or MRI's are done.)
Stage I: Symptoms are similar to those of a wrist sprain. X-rays may be normal or show a line indicating a possible fracture. Magnetic resonance imaging (MRI) may also be helpful in making the diagnosis in this early stage.

Stage II: The lunate bone begins to harden. On an X-ray, it may appear brighter or whiter than the surrounding bones. These changes indicate that the bone is dying. Either an MRI or a computed tomography (CT) scan may be used to assess the condition of the bone. Recurrent pain, swelling and wrist tenderness are common.

Stage III: The dead bone begins to collapse and break into pieces. The surrounding bones may begin to shift position. Patients experience increasing pain, weakness in gripping, and limited motion. Stage III is broken down into IIIa (no fixed scaphoid rotation) and IIIb (fixed scaphoid rotation).

Stage IV: The surfaces of adjoining bones are affected, resulting in arthritis of the wrist.

Diagnosis

Diagnosis is usually based on a suggestive history and physical exam, then x-rays or MRI. In its early stages, Kienböck’s disease may be difficult to diagnose because the symptoms are so similar to those of a sprained wrist. Even X-rays of the wrist may appear normal.

Treatment

Treatment is not well-defined, because the natural history (see above) is not well known. However, this is what has worked best:

Medical therapy: Treatment is primarily directed by the level of symptoms. The primary methods of non-operative treatment are immobilization and anti-inflammatory medications. Because our best understanding of the disease is that it is related to loss of blood supply to the lunate, and because we know some patients can be cured if we can just avoid trauma and collapse, this is our first choice in the earlier stages. Younger patients tend to have a better ability to re-establish blood flow to various areas, so in a very young patient, we almost always start with this approach, especially if the disease is early, in hopes of allowing revascularization of the lunate and prevention of disease progression.

Surgical therapy: Surgery is reserved for more advanced disease. Although there is no cure, there are several surgical options for treating the more advanced stages of Kienböck’s disease. The number of options is a bit confusing, so hang in there. The right procedure for you will depend on several factors, including disease progression, your personal activity levels and goals and my experience with various procedures. We will need to discuss this several times. The two most important pieces of information are the stage of your disease and the presence or absence of ulnar variance (the length of the ulna to the length of the radius).

Directly comparing the results of different techniques is difficult because most studies have a fairly small number of patients and short follow-up. However, our experience is that many of the techniques result in very similar rates of good outcomes.

Operative treatment can be classified broadly into six categories, including: (1) joint-leveling procedures; (2) intercarpal fusions; (3) salvage procedures; and (4) others.
Joint-leveling procedures

If the bones of the lower arm are uneven in length, a "joint leveling" procedure (shortening the longer bone or lengthening the shorter bone) has been done. Bones can be made longer with bone grafts, or shortened by removing a section of the bone. This reduces the compressive forces on the lunate and seems to halt progression of the disease. Forces on the lunate, caused by using the hand forcefully, are thought to flatten down the lunate, advancing the stage of the disease. This collapsing force can be reduced by 70% with an appropriate radial shortening or ulnar lengthening. Currently, radial shortening with a 3.5 - mm metal plate is preferred over ulnar lengthening, as there is a lower complication rate and similar good outcomes. In patients with neutral or positive ulnar variance, shortening the radius is a poor idea. In this clinical situation, radial wedge osteotomies designed to decrease the radial inclination have been proposed. If the lunate is severely collapsed or fragmented, it can be removed. The two bones on either side of it are also removed. This procedure is called a proximal row carpectomy and will relieve pain while maintaining partial wrist motion.

Intercarpal fusions

Another way to ease pressure on the lunate is to remove some of the wrist bone joints and make them grow together into one or more bigger bones. This is called a fusion. If you make all of the bones grow together, it is called a complete fusion. Usually, we only do some of the bones; this is called a partial fusion. However, this approach may or may not improve range of motion, depending on whether the fusion is complete or partial, and on the stage of the disease, and on the amount of motion you started with. If the disease has progressed to severe arthritis of the wrist, fusing the bones will reduce pain and help maintain function, although motion is limited. A variety of intercarpal fusions for the treatment of Kienböck disease have been used. The goal is to red uce forces on the lunate, crushing it down, and, in procedures that involve the scaphoid , to correct and maintain proper scaphoid position.

Scaphocapitate (SC) fusion has been used by some surgeons. Biomechanically, this fusion has been shown to reduce compressive forces at the radiolunate joint by about 10%. Some surgeons prefer this fusion because it requires only one fusion site and is technically easier to perform.

What to do if all else fails

Salvage procedures are what we call the surgeries that are used when all else fails. One procedure that has worked rather well, despite how destructive it sounds: cut out three of the little bones of the wrist (scaphoid, lunate, and triquetrum.) This is called a proximal row carpectomy (PRC). It has been shown to provide relatively good results in Kienböck disease by a several very good friends of mine. Our experience with a PRC is also supported by the fact that it works well for other wrist problems, so we have more experience with this surgical approach than w ith many of the others, which are only used for the relatively rare condition of Kienbock's. Complete wrist fusion (arthrodesis) is the final option for patients with global wrist degeneration. Arthrodesis can be achieved successfully following a failed PR C. It always works to stop the pain, and although the patient cannot do wrist flexion and extension, they usually have increased function in their wrist because the pain is gone.

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