Kienböck’s disease: current concepts

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ABSTRACT
Osteonecrosis of the lunate or Kienböck’s disease is a progressive, debilitating condition that can lead to chronic pain and function loss. Despite it was identification almost 100 years ago, the etiology remains unknown, although mechanical, vascular, and traumatic factors have been associated. The natural history of Kienböck’s disease is poorly defined, and the radiographic findings do not always correlate with the clinical findings. There has been some progress in identifying and understanding the progression of the avascular process and its deleterious effects on wrist mechanics. Initial treatment is conservative. Improvement in surgical techniques with vascularized bone grafts from the distal radius may lead to an outcome improvement for patients in the early stages of disease. However, more research is still needed to determine whether this surgical treatment represents an improvement over conventional treatment alternatives. Recent reports of long-term outcomes for radial shortening osteotomy in osteonecrosis early stage patients and for proximal row carpectomy (PRC) in advanced Kienböck’s disease patients reveal that these procedures provide reliable options for the long-term management of this challenging condition.

Key words: Kienböck’s disease

INTRODUCTION
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ized bone grafts from the distal radius may lead to an outcome improvement for patients in the early stages of disease. However, more research is still needed to determine whether this surgical treatment represents an improvement over conventional treatment alternatives. Recent reports of long-term outcomes for radial shortening osteotomy in osteonecrosis early stage patients and for proximal row carpectomy (PRC) in advanced Kienböck’s disease patients reveal that these procedures provide reliable options for the long-term management of this challenging condition.

**HISTORICAL OUTLOOK**

Robert Kienböck, an Austrian radiologist, was the first to describe the avascular necrosis of the lunate.¹ His article “Concerning Traumatic Malacia of the Lunate and Its Consequences” was published in 1910. The article described the condition as an isolated disease of the lunate that was associated with secondary changes in the other carpal bones. Clinical findings included pain, range of motion (ROM) loss, and prominence in the wrist area. Radiographic findings included isolated changes in the proximal lunate, with occasional collapse and fragmentation of the bone. Kienböck believed the cause of this condition to be a “disturbance in the nutrition” of the lunate due to repetitive trauma.

In 1928, Hultén noted that 78% of his study group comprised of 23 Swedish patients with Kienböck’s disease showed negative ulnar variance while the same parameter had a 23% prevalence in the general population.² This study led to the association between ulnar-minus variance and avascular necrosis of the lunate, and the constitution of groups that supported a mechanical etiology of the disease. More recent studies on patients of Asian ethnicity have shown that Kienböck’s disease can occur in patients with ulnar neutral and ulnar positive variance.³

**ETIOLOGY**

The lunate is the central bone of the carpal proximal row, and it articulates proximally with the lunate fossa of the radius and the triangular fibrocartilage complex, and distally with the capitate and the hamate bones. The lunate and the lunate fossa of the radius have a hemispherical shape and a high level of congruence between their articular aspects. This anatomical characteristic is believed to play a major role in the joint between the radius and the lunate being one of the last joints to develop degenerative changes.

As the Kienböck’s disease is mainly an avascular process, the vascular patterns of the lunate have been studied in depth in order to identify the development of osteonecrosis.⁴,⁵ Several vascular patterns have been identified in cadaveric specimens; however, most nutrient vessels belong to the dorsal and palmar vessels.⁵ These vessels enter through the bone non-articular surfaces. In one study, between 7% and 23% of the lunate were supplied by a single dorsal artery.⁴,⁵

The vessel or vessels that enter the bone may result in several intra-osseous branching patterns, including “I”, “X” and “Y” patterns. Lunate with a single vessel or with a limited intra-osseous branching may be at increased risk of osteonecrosis associated with acute or chronic repetitive trauma. A palmar vascular supply may explain why patients with perilunate instability or dislocations rarely develop avascular changes in the lunate, since the capsules volar to the lunate remain intact.⁶

Disruption of venous outflow leading to increased intra-osseous venous pressure may also cause avascular changes and osteonecrosis. In vitro intra-osseous pressure measurements of necrotic lunates showed marked increases in pressure when compared to normal lunates. The study describes dense plexus of small venous vessels that were found at the palmar and dorsal periosteal aspect of the bones.⁷ The authors suggest the plexus as a potential site of venous outflow disruption, secondary to systemic and local compression factors.

Mechanical factors have also been studied as contributing factors to osteonecrosis development.⁸,⁹ As already mentioned, the height ratio between the radius and the ulna at the distal radioulnar joint and the radiocarpal joint (ulnar variance) has long been believed to play a major role in the development of Kienböck’s disease. Other mechanical factors, such as the radial inclination, may also lead to Kienböck’s disease, especially in patients with ulnar-neutral and ulnar-positive variance who would not develop an avascular process as a result from an increase of radiolunate pressure due to ulnar-negative variance. Patients with decreased radial inclination are more likely to have a smaller lunate and to develop Kienböck’s disease.⁸
There have been also systemic factors associated with lunate osteonecrosis. Conditions associated with hypercoagulability, decreased arterial inflow or increased venous congestion may affect this process.\textsuperscript{6} Osteonecrosis has been association with systemic corticosteroid use, sickle cell disease, cerebral palsy and septic embolism, although there is no defined correlation between theses systemic conditions and Kienböck’s disease. To this day, over 100 years after Kienböck study, the condition etiology remains to be defined. Although mechanical, anatomic, and systemic mechanisms have all been implicated in the development of the Kienböck’s disease process, no specific etiologic mechanism has been identified. Our understanding is that a complex combination of all the above factors is likely to be responsible for the Kienböck’s disease process.\textsuperscript{6,8,9}

**CLINICAL AND RADIOGRAPHIC FINDINGS**

Kienböck’s disease patients commonly present with wrist pain and weakness, often without history of acute trauma. Symptoms duration before consultation vary, although patients commonly report progressive symptoms over long periods of time. For this reason there is no clear understanding of the natural history of the disease. Pain ranges from mild and occasional to severe and debilitating. Kienböck’s disease is most common in young adults (between 20 and 40 years) and occurs infrequently in children, although there have been reported cases. Bilateral Kienböck’s disease is rare.

Physical examination reveals swelling and tenderness over the dorsal aspect of the wrist (over the lunate), probably due to synovitis. Typical presentation includes a decreased ROM, with associated loss of flexion and extension; pain at the end of the physiological ROM, especially in extension; and forearm rotation is usually normal. In addition, grip strength may be markedly reduced in comparison to the contralateral wrist.

Kienböck’s disease can be diagnosed by radiographic studies. Depending on the disease stage, radiographs may evidence diffuse sclerosis of the lunate, cystic change, fragmentation, articular surface collapse of the lunate, and perilunate arthritic changes. Radiographic findings in the early stages of Kienböck’s disease commonly include a fracture in the lateral view; however, it is not clear whether the lunate fracture line represents a primary event or whether it occurs later as a consequence of the structural weakness of the necrotic bone. Radiographic studies also play a key role in establishing the affected wrist anatomical and mechanical characteristics, including ulnar variance, radial inclination, carpal height, radioscaphoid angle, and lunate size.

MRI is a key imaging study, particularly instrumental in the early stages of Kienböck’s disease when radiographic imaging is less reliable. On T1-weighted images, bone marrow fat loss results in low signal intensity. Accordingly, T2-weighted sequence images also demonstrate low signal intensity. In order to establish a Kienböck’s disease diagnosis, imaging evidence must include diffuse signal changes that involve the entirety of the lunate. Confinement of signal changes within the lunate should lead to considering other differential diagnoses. For example, if signal changes are confined to the proximal ulnar region of lunate, ulnar carpal impaction should be considered. Other studies may also prove instrumental. Before MRI, bone scintigraphy was the next diagnostic study recommended in cases of non-diagnostic initial radiographs. Finally, computed tomography proves useful in assessing articular surface collapse and fractures.

Cases where plain radiographs fail to provide clear evidence of radiocarpal or midcarpal joint involvement may benefit from the use of arthroscopic diagnosis in staging Kienböck’s disease. Bain and Begg have developed an arthroscopic staging system based on the number of articular surfaces involved.\textsuperscript{10} The system classifies lunate articular surfaces (proximal lunate, distal lunate, lunate facet of the radius) into functional and non-functional, and provides a treatment algorithm based on arthroscopic findings. However, performing a diagnostic arthroscopic procedure should be carefully considered in this setting as plain radiography and other imaging studies commonly provide diagnostic findings when assessing articular surfaces.

**STAGES**

Kienböck’s disease staging is made mainly on radiographic findings and is a critical step in the assessment and management of Kienböck’s disease patients as it establishes the different treatment options for each case. There are several staging systems, but the one described by Lichtman et al. is the most widely used and has been shown to have adequate reproducibility and reliability.\textsuperscript{6}

Stage-1 plain radiographs evidence normal density and articular surfaces of the lunate, while stage-1 MRIs reveal diffuse signal intensity changes. Stage-2 plain radiographs show a diffuse sclerotic appearance of the lu-
Kienböck's disease

Table 1. Lichtman Classification for Kienböck's Disease

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
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<tbody>
<tr>
<td>Stage 1</td>
<td>Normal radiographs, signal intensity changes are present on MRI</td>
</tr>
<tr>
<td>Stage 2</td>
<td>Radiographs evidence lunate sclerosis; fractures lines may be present</td>
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<tr>
<td>Stage 3</td>
<td>Collapse of the articular surface of the lunate.</td>
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<tr>
<td>Stage 3A</td>
<td>Normal carpal alignment and height</td>
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<tr>
<td>Stage 3B</td>
<td>Scaphoid rotation, proximal capitate migration, and loss of carpal height</td>
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<tr>
<td>Stage 3C</td>
<td>Lunate coronal fracture; lunate divided into two (palmar and dorsal) triangular fragments</td>
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<tr>
<td>Stage 4</td>
<td>Lunate collapse with associated radiocarpal or midcarpal osteoarthritis</td>
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Goldfarb et al. conducted a study to assess Lichtman classification reliability and found that its interobserver reliability was substantial. However, they also found the differences set for stages 3A and 3B not to be particularly reliable. Therefore, they proposed a new modification to the staging system: patients with a radioscaphoid angle of greater than 60° belonged to stage 3B. This modification resulted in an interobserver reliability increase both for general staging (kappa coefficient from 0.63 to 0.81) and for stage 3A (kappa coefficient from 0.38 to 0.75). The authors concluded that the use of the radioscaphoid angle increases the interobserver reliability of the Lichtman classification and allows for a better distinction between stage 3A and 3B disease.

TREATMENT

Kienböck's disease treatment is based on the patient's symptoms and functional impairment and should observe the previously mentioned staging system. Most longitudinal studies on the conservative treatment for Kienböck's disease have reported a radiographic progression of the disease through the previously mentioned stages. Keith et al. studied 33 patients treated solely by conservative means. They identified ROM reduction (flexion) and function deterioration, which was evidenced by a reduction in the Disabilities of the Arm, Shoulder and Hand (DASH) scores. Although the radiographic longitudinal study was limited to their patient series, they authors concluded that Kienböck's disease is a progressive condition.

There is no consensus on which approach, whether surgical or conservative, yields better outcomes. Delaere et al. compared management with night splinting versus surgery in patients with stage 1, 2, or 3 Kienböck’s disease. Their results with immobilization were equivalent to those obtained with surgical treatment. However, these results should be considered with reservations as the average stage in the splinted group was lower than that in the surgery group. Other studies have reported surgical treatment to be superior to conservative management. Salmon et al. provided evidence showing that radial shortening osteotomy may be superior to conservative management.
Despite Kienböck’s disease progression, many patients, even in advanced stages of the disease, still have adequate wrist function and bearable pain. Hence, conservative therapy using a splint or plaster cast and analgesic or anti-inflammatory drugs may be considered a valid approach for all patients, irrespectively of the disease stage at presentation. Immobilization should be maintained for 3 months. Surgery should only be considered for patients for whom conservative treatment and remain symptomatic.

There are multiple surgical procedures for Kienböck’s disease; however, we failed to find enough literature data to establish superiority among surgical options. Despite the plurality of surgical procedures, none has been proven superior to the others. Following the previously mentioned staging system, surgeons opt for different procedures according to the different stages. Ulnar variance, radial inclination and other relevant anatomical variables play a key role in establishing which procedure is adequate for each individual patient (Table 2).

**Table 2. Kienböck’s disease treatment according to its stage**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Stage 1</strong></td>
<td>Cast immobilization for 3 months</td>
</tr>
<tr>
<td><strong>Stages 2 and 3A, negative ulnar variance</strong></td>
<td>Radius shortening; ulnar lengthening; capitate shortening</td>
</tr>
<tr>
<td><strong>Stages 2 and 3A, positive ulnar variance</strong></td>
<td>Vascularized bone grafting and external fixation; radial-wedge or dome osteotomy; capitate shortening</td>
</tr>
<tr>
<td><strong>Stage 3B</strong></td>
<td>Scaphotrapeziotrapezoid fusion; lunate excision; radius shortening; proximal-row carpectomy</td>
</tr>
<tr>
<td><strong>Stage 4</strong></td>
<td>Proximal-row carpectomy; wrist fusion; wrist denervation</td>
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**Stage 1**

Splint or plaster cast immobilization remains the first treatment option. External fixation has been used both for immobilization as well as lunate unloading procedures, although this technique data is still limited. Despite there are patients who show improvement with the conservative treatment, most of them are likely to progress to stage 2.

**Stages 2 and 3A**

Stages 2 and 3A are often considered together in terms of treatment options, with the main goal being lunate revascularization in order to prevent the disease progression. The only exceptions to this pairing are the direct revascularization procedures, which are mainly used during stage 2, where no articular collapse has taken place. However, many authors describe the use of this procedures in patients with some degree of collapse and loss of lunate height, a scenario referred to early 3A stage.

Several revascularization procedures have been described for the treatment during stage 2, such as vascularized grafts of the pisiform bone, vascularized pedicled grafts from the distal radius (including the pronator quadratus), direct implantation of the metacarpal arteries and free vascularized grafts, among others. In addition to the vascularized grafts, unloading procedures to decrease mechanical stress on the lunate, such as external fixation, STT or scaphocapitate fusion, are commonly performed.

Distal radius vascularized bone grafts have become popular. Several types of grafts have been described based on their location with respect to the wrist extensor compartment and also based on whether the donor artery is located within or outside the extensor compartment. The most used graft for Kienböck’s disease is based on the 4+5 extracompartmental artery (ECA). Retrograde flow of the fifth ECA is directed in an antegrade direction into the fourth ECA after ligation of the posterior branch of the anterior interosseous artery. This grafting allows for a longer and larger-diameter pedicle of ulnar location. Moran et al. have studied 4+5 ECA bone grafts in a retrospective analysis including 26 patients with a 31-month average follow-up. Most patients experienced pain relief and grip strength improvement. From the 26 patients, 71% evidenced revascularization of the lunate.
Despite the overall success of the procedure, 23% of patients had radiographic evidence of disease progression at last follow-up.

Vascularized grafts of the pisiform bone are performed using the ipsilateral pisiform bone as donor site, along with its blood supply from small dorsal and radial branches of the ulnar artery. The pedicled bone graft is then implanted to the bone defect surgically created in the dorsal surface of the lunate. Daecke et al. described this procedure in 23 patients with stage 2 or 3A Kienböck’s disease, with a 12-year mean follow-up. Pain and function improvement were recorded in 20 out of the 23 patients. Out of 20 patients with preoperative radiographic studies, 6 (33%) evidenced disease progression.

Free vascularized grafts have also been used in the treatment of Kienböck’s disease. Gabl et al. studied 18 patients with stage 3 Kienböck’s disease that were treated with implantation of a free vascularized corticocancellous iliac bone. The wrist was stabilized using external fixation during the healing period. In a 5-year mean follow-up, the graft became incorporated in the lunate in 16 patients with no carpal collapse. The graft did not integrate and was resorbed in the remaining two patients. In their cohort study, Arora et al. reported that after a 10-year follow-up those 16 patients still showed no evidence of Kienböck’s disease progression or carpal collapse.

Other treatment options for patients with these Kienböck’s disease stages include “joint-leveling” procedures. These procedures are believe to produce a mechanical unload of the lunate, resulting in revascularization and preventing progression to collapse of the articular surface. Establishing the adequate procedure for each individual patient is primarily based on the height relationship between the radius and the ulna at wrist level.

The goal in negative ulnar variance is to level the distal radioulnar joint to result in neutral to 1-mm positive ulnar variance. This result can be achieved by shortening the radius or lengthening the ulna. Ulnar lengthening requires bone graft and has a higher rate of nonunion, which explains why is not commonly performed.

Radius shortening osteotomies are technically easier procedures and have low nonunion rates. Several authors have published on long-term follow-up studies, describing this procedure outcomes. Raven et al. studied 12 patients who underwent radius shortening osteotomies, with a 22-year mean follow-up period. One patient had stage 3B Kienböck’s disease and the rest were either stage 2 or 3A. Only 3 patients evidenced mild disease progression at last follow-up while most patients experienced excellent outcomes as regards pain relief and function. In a study of similar characteristics, Watanabe et al. studied 12 patients who underwent radius shortening osteotomies, with a 21-year mean follow-up period. Most patients had mild wrist pain and good ROM and function, but radiographs revealed that the Kienböck’s disease had progressed in 50% of the patients.

Radius shortening osteotomy in stage 3B Kienböck’s disease remains controversial. Altay et al. compared the procedure effectiveness in stage 3A (n = 13) and 3B (n = 10) patients during a 85-month follow-up period. They found no significant differences between the two groups with regard to pain and ROM between the different stages and concluded that the procedure produces similar clinical outcomes irrespective of the disease stage, despite no radiological healing.

Joint leveling options for patients with neutral or positive ulnar variance include radial closing-wedge osteotomy, radial-dome osteotomy, and capitate shortening osteotomy with or without capitate-hamate fusion. Radial osteotomies are used to decrease radial inclination in an attempt to decrease the contact pressure between the radius and the lunate. Leveling is achieved by increasing the area of contact between the radius and the lunate and decreasing the force transmitted through the radiolunate joint and the lunocapitate joint. Koh et al. reported improvements in pain, ROM and function in their 10-year minimum follow-up study. However, osteoarthritic changes were observed in almost 75% of patients at the final follow-up evaluation, suggesting that Kienböck’s disease progression did not stop. The goal of these procedures is to unload the lunate articular surfaces by distributing its load across the adjacent joints. These procedures commonly achieve similar outcomes to those of radial shortening osteotomy, providing good pain relief, motion and function restoration, and potentially stopping or slowing disease progression.

Metaphyseal decompression of the radius is a novel concept that has been described for patients with stages 1 to 3A. Illaramendi et al. originally described the procedure in their report on their experience with this procedure in 22 patients with a 10-year mean follow-up. This procedure is performed by decompressing the distal metaphysis of the radius through small cortical windows, using a curette or other blunt instrument to produce cancellous bone impaction against the medullary canal, leaving cortical bone without cancellous bone through the 3cm distal to the radius (Figure 1).
The authors reported good motion restoration and pain relief, with only 4 patients with complaints of mild pain. Most of the patients (17 out of 22) did not show disease progression at last follow-up, 2 showed radiographical improvements and 3 showed radiographical disease progression. In 2017, De Carli et al. published a series of 15 stage 3A Kienböck’s disease patients, with a 13-year mean follow-up. Based on the Mayo Wrist Score, outcomes were excellent in 6 patients, good in 8 patients, and poor in 1 patient (Figure 2).31

Figure 1. Lateral window for metaphyseal decompression of the radius.

Figure 2. A 45-year old woman with stage 3A Kienböck’s disease. A. Initial X-rays. B. Radiographic control at 12-year follow-up.
Stage 3B

In this stage, there is loss of carpal height due to flexion of the scaphoid as well as articular collapse of the lunate articular surface. Correction of the flexed scaphoid coupled with intercarpal fusion (STT or scaphocapitate) can decrease lunate load, which facilitates midcarpal joint stabilization, and can prevent further carpal collapse. In addition to fusion, the lunate can be removed and replaced with a tendon or a lunate implant to reduce joint irritation. Van den Dungen et al. compared the outcomes of patients treated conservatively (n = 19) and with STT fusion (n = 11), with a 13-year mean follow-up period. The authors state that the groups were statistically comparable and found that STT fusion caused an increased loss of mobility and pain and a longer rehabilitation period as compared to conservative treatment. Tambe et al. compared outcomes of patients treated with partial intercarpal fusion and radiocarpal fusion for advanced Kienböck’s disease, and found that patients treated with radiocarpal fusion had better outcomes at last follow-up.

PRC is a successful salvage procedure, but it requires the capitate head to be free of degenerative changes. In presence of mild arthritic changes are present on the capitate head, an interpositional arthroplasty of dorsal wrist capsule in addition to the PRC may prove effective.

Several studies present detailed long-term PRC outcomes for the treatment of Kienböck’s disease. Croog and Stern described a series of 21 patients treated for stages 3 and 4, with a 10-year mean follow-up. Three patients required radiocapitate fusion due to chronic pain, from which 2 had stage 4 disease at the time of the index procedure. The authors concluded PRC to be a reliable procedure in this setting, but advise caution in performing PRC in advanced Kienböck’s disease patients. In a similar study, DiDonna et al. detailed their experience with CFP in their 10-year minimum follow-up study for patients with degenerative conditions of the wrist, including Kienböck’s disease. Again, the authors concluded PRC to be a reliable procedure in this setting, but advise caution in performing PRC in Kienböck’s disease patients younger than 35 years of age. De Smet et al. reported on 21 patients with advanced Kienböck’s disease, with a 67-month mean follow-up period. Lumsden et al. report on 13 patients, with a 15-year mean follow-up period. Like the previous authors, these authors found PRC to be a reliable longterm procedure for the treatment of advanced Kienböck’s disease.

Stage 4

The treatment pillars for this stage include PRC and wrist fusion. As mentioned above, mild arthritic changes on the capitate head can be addressed at the time of PRC with an interpositional arthroplasty of dorsal wrist capsule. However, arthritic changes in this region may cause chronic pain and PRC failure, resulting in further intervention, this time a fusion.

Wrist denervation alone or in combination with the aforementioned procedures can help with symptom relief. Schweizer et al. reported their long-term experience with complete wrist denervation in a 70-patient series, of which 11 patients had Kienböck’s disease. The average follow-up was 9.6 years. Complete wrist denervation led to subjective improvement in approximately 66% of patients, and approximately 50% reported complete or significant pain relief.

Kienböck’s disease remains a clinical challenge for hand surgeons. The etiologic factors behind the disease process remain to be identified. Surgical advances involving local vascularized grafts from the distal radius have the potential to improve its treatment. Traditional procedures for the treatment of advanced Kienböck’s disease, such as radial shortening osteotomy and PRC, have shown in long-term studies to provide adequate symptom control and functional restoration.

Conflict of interests: Authors claim they do not have any conflict of interest.
REFERENCES


