Kienböck disease, current issues

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ABSTRACT

Kienböck’s disease (KD) is a progressive condition leading to lunate collapse and carpal functionality disorder, with still uncertain etiology. The disease seems initially to affect the vascularity, lately the bone density and finally the cartilage of lunate. Advanced imaging and wrist arthroscopy offer useful tools for its approach. Classification, staging, therapeutic treatment and prognosis mostly depend on their findings. The cure algorithms that are usually followed include load redistribution, revascularization and lunate or wrist reconstruction as salvage surgery. Despite the great variation of operative options the majority of them provide satisfactory results.

KEY WORDS: Kienbock disease, lunate, wrist

Introduction

Kienböck’s disease (KD) was initially described in 1910 by the Austrian radiologist Robert Kienböck (1871 – 1953). Although originally characterized as traumatic softening of the lunate, it is now defined as sedimentation of the whole lunate or part of it, with progressive evolution. The exact cause is not clearly clarified despite the fact that various pathophysiological mechanisms have been proposed. The lunate is the middle bone of the proximal raw carpal bones and is articulated with scaphoid (lateral), capitate (distal), triquetrum (medial), hamate (distal and medial), radius and with the triangular fibrocartilage complex attached to the distal ulna. It is crescent-shaped with a distal concave articular facet and a proximal convex articular facet. Load distribution between the forearm and the carpus is shared in the scaphoid and the lunate playing a crucial role as far as the function of the carpal is concerned.

Epidemiology

Kienböck disease is a rare condition with incidence < 5/10000 in general population but is a common type of avascular necrosis of the carpal bones (1). The characteristically affected population is young, males, aged 15 - 40 years old with unilateral symptoms. No difference between dominant and non-dominant hand seems to occur. Several cases of
Kienböck disease have been described in children where the disease is classified infantile (<12 yo) and juvenile (>12 yo) and in elder patients (2). Among carpal bones is the most common type of avascular necrosis.

Predisposing factors
In literature there is not a clear explanation for the exact cause of KD. Many pathophysiological pathways have been proposed in order to explain the trigger point and the progressive evolution of this condition.

The anatomy of the carpal has been initially complicated. A more radially inclined lunate tilting angle and a flatter radial inclination with or without the combination of a smaller lunate diameter and height seems to play a predisposing role (3). Patients with KD tend to present negative ulnar variance (incidence 3.1:1) in comparison to patients with neutral or positive ulnar variance (4). Blood supply is based in a complex of extraosseous and intraosseous vascular anastomosis arising from the radial, ulnar, and anterior interosseous artery. Three major intraosseous vascular patterns (Fig.1) have been described represented with the symbols Y, I, and X in frequency of occurrence (5). High intraosseous pressure, venous congestion and insufficient vascularity have been proposed as risk factors (5, 6). Joint motion can interfere negatively to blood flow of the lunate bone (7).

Great part of these patients (~70%) has a medical record of recent trauma or fracture, while the theory of repeated microtrauma has been proposed to explain the common presence of the disease among labor workers and athletes.

Higher incidence of this condition is observed in patients with cerebral palsy, scleroderma, systemic lupus erythematosus, rheumatoid arthritis, dermatomyositis and Crohn’s disease. A possible etiology of this correlation may come from the higher levels of antiphospholipid antibodies that are detected in Kienböck disease. Cases of concurrent avascular necrosis of other neighbor carpal bones may indicate a regional identity of the disorder (8).

Some authors propose the theory that this condition is rather a phenomenon that osteoclast prevails over osteoblast activity, during the procedure of reconstruction of microfractures. According to this hypothesis load disposition in the area seems to exacerbate and worsen the symptoms of an underlying situation (9).

Clinical presentation
Patients usually present unilateral carpal pain, located dorsally above lunate with restricted range of motion and various lack of strength. Symptomatology may deteriorate with axial loading during dorsal extension of the wrist. Medical history may reveal recent or former injury but it is not always a precondition.

In many cases the disease is possible to exist without symptoms or any other indications and be discovered as a random finding in radiological examination.

Classification
Lichtman classification, that is the most common
Mitsikostas P K, et al. Kienböck disease, current issues

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proposed to describe KD, is based in radiological display of the lunate in plain X-rays and MRI findings (Table 1). In stage I lunate gives no pathological findings in radiographs, keeps normal architecture and only in MRI marrow pathology is present (Fig. 2). In stage II lunate becomes increasingly radiodense, displays a sclerosis mosaic but still the overall structure is preserved. Stage III is characterized by a collapsed lunate with multi-fragmentation lines present in radiographs as bone disorder progresses. This stage is further divided in two subgroups. In stage IIIA lunate appears collapsed with fragmentation lines but no associated carpal alignment changes are present. For this stage radioscaphoid angle is less than 60°. In stage IIIB there is carpal collapse and fixed scaphoid volar flexion with a radioscaphoid angle more than 60°. Stage IV depicts lunate collapse with midcarpal and/or radiocarpal joints affected, described as secondary pancarpal arthritis (10, 11).

Wider use of MRI offered a more powerful tool allowing more persuasive investigation especially with the use of gadolinium. With this method differential diagnosis is more likely to be sensitive and accurate. Moreover, MRI proves helpful in recognizing the disease in initial stages providing the

Figure 2. Lunate bone marrow edema in T2 and T1-weighted MRI.

<p>| Table 1. The widely used Lichtman classification |</p>
<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
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<tbody>
<tr>
<td>Stage I</td>
<td>normal radiographs, normal architecture, marrow pathology in MRI</td>
</tr>
<tr>
<td>Stage II</td>
<td>lunate increasingly radiodense, sclerosis mosaic, overall structure preserved</td>
</tr>
<tr>
<td>Stage III</td>
<td>collapsed lunate</td>
</tr>
<tr>
<td>Stage IIIA</td>
<td>lunate with fragmentation, no associated carpal alignment changes, radioscaphoid angle &lt; 60°</td>
</tr>
<tr>
<td>Stage IIIB</td>
<td>carpal collapse and fixed scaphoid volar flexion, radioscaphoid angle &gt; 60°</td>
</tr>
<tr>
<td>Stage IV</td>
<td>lunate collapse with secondary pancarpal arthritis (midcarpal and/or radiocarpal joints affected)</td>
</tr>
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advantage to estimate prognosis and healing rate. Additionally, it appears to be a sensitive follow-up examination (12, 13).

Bain and Begg in 2006 proposed a different classification formed on the display of the lunate in arthroscopy. To be more specific, the disease is categorized by the number of joint surfaces that present degenerative changes and seem to be non-functional among the joints that lunate is involved in the wrist (14).

In other words, Lichtman classification focuses in bone integrity of the Lunate, Schmitt classification according to MRI images emphasizes in bone vascularity and Bain and Begg classification gives priority to cartilage condition and functionality.

Treatment
Conservative treatment
Most patients in initial stage tend to be treated conservatively for at least 3 months with immobilization, activity modification and anti-inflammatory medication. In this way, a sufficient time frame is assigned for revascularization and prevention of the progress of the disease. In case symptoms remain or there is deterioration in radiological examinations, radical medical therapeutic measures and operative procedures should be considered (15).

Non operative treatment is a preferable choice in children (<15 yo) and in elder patients (>70 yo) especially in initial stage of the disease taking into consideration that provides better prognosis and healing rates.

Operative treatment
A great variety of operations has been suggested as KD therapy. Most popular among them involve: load redistribution, operations for revascularization, core decompression, arthrodesis of carpal bones, graft placement, wrist arthroscopy, arthrodesis of the wrist and wrist denervation. The main pathways that are commonly followed conclude to lunate unloading procedures, lunate revascularization, lunate or wrist reconstruction as salvage surgery.

Load redistribution operations are used to protect lunate from further collapse. In case of negative ulnar variance or flatter radial inclination or radially inclined lunate tilting angle, correction osteotomy of the distal radius is an established method to prevent the progression of the disease. In case of neutral or positive ulnar variance capitate shortening osteotomy is preferred offering satisfactory radiological and functional results (16, 17). For younger patients, radius shortening osteotomy (Fig. 3), before epiphysial closure observed, may lead to relevant recurrence. In children, with open growth plate, epiphysiodesis of the distal radius may be helpful and temporary scapho-trapezio-trapezoidal
pinning that unloads lunate while anticipating for revascularization is an alternative (18, 19).

Revascularization techniques with the use of intercompartmental or extracompartmental artery bone graft, 2nd or 3rd metacarpal base bone graft, vascularized pisiform bone graft, volar bone grafts from the distal radius or free bone grafts from the medial femoral condyle or iliac crest produce promising results and significant clinical recovery (20).

Core decompression is possible to be performed with an open technique or during arthroscopy. The main goal is to reduce intrasosseous pressure and venous congestion to achieve indirect revascularization. Some authors suggest this method combined with bone grafts and arthroscopic debridement of synovitis (21, 22).

For patients in advanced stages, with higher possibility for lunate collapse, therapy is based on the potential for bone reconstruction. It is crucial to maintain the height of the proximal carpal row and ensure functional joint surfaces. The most essential difficulty is the high failure rates that accompany reconstruction techniques with the use of free vascularized osteochondral grafts (23, 24). Lunate reconstruction may be performed with the use of bone cement to prevent collapse (25).

In cases with lunate collapse, independent the functionality of articular surfaces, various methods have been proposed for interposition. Silicon, metal and pyrocarbon implants, tendonous grafts and Pisiform transfer have been utilized for this purpose (26-30). Lately, 3D printed implants have been produced, corresponding to CT depict of the non-affected hand (31). Capitate lengthening and proximal proximal row carpectomy are alternatives when the lunate is not reconstructable but joint surfaces of the lunate facet and the capitate remain functional (15). In late stages, with midcarpal and radiocarpal articulation compromised, radioscapolunate fusion, scaphocapitate fusion and hemiarthroplasty offer a choice for wrist reconstruction with prerequisite some joint functionality. For patients with established pan-arthritis wrist fusion and wrist arthroplasty may be applied depending (15).

Arthroscopy has been primarily used as a diagnostic tool for staging and classification of KD. Furthermore, it has allowed some therapeutic choices: synovectomy and debridement, lunate forage, radio-scapholunate fusion, scapho-capitate fusion, arthroscopic proximal row carpectomy. Graft placing can also be an arthroscopically assisted procedure (32, 33)

Wrist denervation complete or partial has been chosen as a therapeutic option by some authors. Results range and present a considerable variation despite the fact that there is a trend for improvement in functionality and pain relief. Another advantage is that wrist denervation permits additional surgical operations to be performed as adjuvant therapy (34, 35).

Conclusions
As far as the etiology of KD remains unknown, treatment strategies will present high variation among practitioners. Treatment is directly correlated to the disease staging. With technical advances new procedures are added to conventional tactics. Few treating algorithms are available but the final decision seems to come taking into consideration a main factor: patient’s personal expectations.

Conflict of interest
The authors declared no conflicts of interest.
REFERENCES

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