INTRODUCTION

Kienböck’s disease or lunatomalacia is an aseptic osteonecrosis of the lunate bone initiating a progressive collapse of the lunate bone, followed by total carpal collapse. The first description was made one hundred years ago (107). Thus far, its exact cause and natural history remain unknown (44).

Several extrinsic and intrinsic factors have been investigated as to their role in the aetiology of the bone necrosis. Kienböck’s disease is a slowly progressive disorder with well recognised stages leading to various treatment options. However, little is known about their effectiveness, mainly because Kienböck’s disease is not very common and a long follow-up is needed to evaluate the ultimate outcome (1,84).

EVALUATION

Kienböck’s disease is seen most often in young adults (between 20 and 45 years of age), but it is also diagnosed in children as well as in older age groups. It is somewhat more common in males. Kienböck’s disease is usually unilateral and isolated. A history of trauma is often reported by the patients, often minor or repetitive trauma. The disease is slowly progressive with unspecific signs and symptoms. Patients complain of dorsal wrist pain, weakness and restricted motion. On physical examination, dorsal tenderness is obvious and is often well localized over the dorsal side of the lunate.

More advanced cases may show swelling due to synovitis and a loss of wrist motion with decreased grip strength due to underlying degenerative arthritis. Early signs of synovitis include a restricted anteroposterior drawer test of the wrist and an obliteration of the anatomical snuffbox. The anteroposterior drawer test is done by the examiner grasping the forearm of the patient with one hand, just proximal to the wrist joint and with the other hand just distal to the wrist joint. The patient’s hand is then moved in a dorsopalmar direction. Normally, some translation is possible, but not in the presence of synovitis (pseudo stability).

Occasionally, the patient has no symptoms at all and lunate collapse is detected on a routine radiograph of the wrist (96).

The diagnosis and staging of Kienböck’s disease are based on the x-ray findings. In early stages, plain radiographs may be normal and the diagnosis is suspected on an abnormal bone scan or MRI. CT scan may reveal an occult lunate fracture. In a later stage, sclerosis of the lunate and collapse are observed. Lichtman et al (56) classified Kienböck’s disease into four stages:

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The pathogenesis of the wrist pain in Kienböck’s disease is not always apparent. The bone necrosis stricto sensu can be a source of dorsal wrist pain. Radial side wrist pain, in the presence of a positive Watson scaphoid shift test (109) may indicate rotatory subluxation of the scaphoid. Generalized wrist pain and swelling are signs of osteoarthritis.

### Aetiology

The blood supply of the lunate is probably the key factor in the pathogenesis of the disorder (44). The extra-osseous and intra-osseous vascular anatomy has been well studied (34). The carpal bones with type 1 vascularity are “at risk”. These are carpal bones which are mostly supplied by one single vessel without additional anastomoses, hence occlusion of this vessel will probably lead to necrosis. According to Gelberman et al (34), all scaphoids and capitates and 8 to 20% of the lunates have this vascular pattern. Besides arterial insufficiency, venous stasis has also been suggested as a possible cause of (carpal) bone necrosis. Extrinsic factors such as fractures or repetitive minor trauma can damage the intra-osseous blood supply leading to osteonecrosis. However, rather than causing the necrosis, the fracture may also be a consequence of the necrosis.

Hulten (39) noted an association between Kienböck’s disease and an ulnar minus variance. The relevance of this finding in the aetiology of Kienböck’s disease has however been questioned (26,51). Several authors have found a change in the ulnar variance with age, sex and position of the wrist as well as osteoarthritis secondary to Kienböck’s disease (71).

Morphological factors may also play a role in the aetiology (64,103). In our series we found that a negative ulnar variance was not predisposing to Kienböck’s disease (99). However, when we compared the contralateral wrist with an age - and sex matched control group, we found that in patients with Kienböck, the lunates were smaller and had more radial tilt than the controls and the radial slope of the distal radius was less. A recent biomechanical study with finite element modelling demonstrated that ulnar minus variance was important for further progression of the collapse of the lunate (55,85).

### Management

A thorough evaluation of the involved wrist is necessary prior to instituting treatment. The stage of the disease must first be determined, followed by an evaluation of the biomechanical effects of the collapse of the lunate on the the carpus. The biological effects of the diseased bone, which may or may not be associated with synovitis and/or fibrosis resulting in wrist stiffness, and their effects on the patient’s disability have important implications with regards to future management. Finally the presence or absence of osteoarthritis may help to choose between reconstructive versus salvage procedures.

Treatment options include both conservative and operative methods. As a rule (except in children) surgical treatment is preferred since it generally leads to a quicker improvement of the symptoms and a better outcome. Many operative treatment methods have been suggested for the various stages of Kienböck’s disease and they can be grouped into three categories (fig 1a-s).

1) **Revascularisation.** This can be achieved either directly by vessel implantation or indirectly by a vascularised bone graft (VBG) in stage 1 and 2. The graft may be free or pedicled (fig 11,m).
2) **Joint decompression.** This is done not only to relieve symptoms but also to allow spontaneous revascularisation of the lunate while diminishing the compressive forces acting on it to prevent further collapse in stages 1, 2 and 3A. This can be accomplished by levelling the distal radioulnar joint (DRUJ) by shortening of the radius, closing wedge osteotomy of the radius or lengthening of the ulna (fig 1i,j), intercarpal arthrodesis (fig 1b,c,e) or capitate shortening (fig 1d). Restoring the normal carpal height by either scaphotrapezialtrapezoidal (STT) or scaphocapitate arthrodesis is a key element in preventing osteoarthritis. Arthrodeses on the ulnar side of the carpus have also been proposed (hamate-capitate fusion). There exists a large amount of biomechanical evidence to demonstrate that all these procedures significantly reduce the pressure on the lunate (4,14,38,112).

3) **Salvage procedures for pain relief.** These include wrist denervation (fig 1r), arthroplasty or arthrodesis (fig 1n,o,q) in stages 3B and 4. Arthroplasty can be done in different ways: resection of the lunate (fig 1a), replacement of the lunate by a tendon roll, the pisiform bone (fig 1k), a silicone spacer (fig 1g), the head of the capitate (Graner’s procedure) (fig 1f) with or without associated intracarpal arthrodesis, and more aggressive arthroplasties such as partial or complete prosthetic replacement, or proximal row carpectomy (fig 1p).

**Non-operative treatment**

Non-operative treatment has its proponents. The rationale for this approach lies in the observation that spontaneous regression of the signs and symptoms has been observed in early Kienböck. Moreover, cases of asymptomatic patients with radiographic evidence of longstanding Kienböck’s disease have been reported. Previous reports suggest that the pain can subside over several years. Kristensen et al (52) reported good results after short periods of immobilisation, whereas Mikkelsen and Gelineck (61) noted poor outcomes after nonoperative treatment. The debate continues, with some authors advocating a conservative approach (9,25) whereas others have observed progressive clinical and radiographic deterioration (48) or have demonstrated better outcomes with a surgical approach (80). In children, however, Kienböck’s disease is uncommon and a non-operative approach is preferred (23,37). We and others have observed some adaptation of the carpus due to remodelling rather than collapse. A careful neglect policy or, in very painful wrists, temporary immobilization by cast or bracing, often results in an asymptomatic wrist.

**Revascularisation**

The lunate is the keystone of the proximal carpal row. Simple resection leads to immediate and severe collapse of the carpus with subsequent osteoarthritis. The basic principles involve: removing the necrotic bone, replacing it with living bone and protecting the lunate during the revascularisation period (usually with temporary stabilization of the carpus). This is done either by direct revascularisation or indirectly by replacing the necrotic bone by vascularised bone.

Historically, the lunate has been replaced by the pisiform bone pedicled on the ulnar artery (78). This procedure is usually called Saffar’s technique. Daecke et al (16) and Kuhlman et al (53) demonstrated favourable results in 14 and 23 patients respectively (fig 1k).

Some authors have removed the necrotic bone by curettage, followed by cancellous bone grafting and vessel implantation, such as the posterior interosseous artery (fig 1l) (12). There is clear evidence that the bone grafts were revascularised using this technique. We found only one paper by another group (66) reporting results with this technique in 11 wrists. They found a radiological stabilisation of the lunate with good pain relief in 9 patients. Recently, Jones et al (45) reported one case in which vessel implantation was combined with the introduction of Bone Morphogenetic Protein (BMP) into the lunate after scooping out the necrotic bone.

Vascularised bone grafts (VBG) (fig 1m) have the advantage of immediate implantation of viable bone which simplifies matters by substituting a healing fracture to a bone defect. This obviates the need to wait for secondary revascularisation of a
cancellous bone graft and it avoids the period of temporary weakening that occurs with nonvascularized bone grafts. The Mayo group studied the vascularity of the distal radius (88). Based on anatomic studies and animal experiments they developed the technique of the so-called 4,5 extensor compartment vascularized bone graft (67). In their clinical series of 26 patients they reported pain relief in 92%, a significant improvement in grip strength and maintenance of carpal height in 77%. We have applied this technique for the treatment of early Kienböck with a painful but still mobile wrist in eight patients (36). Postoperatively, patients had considerably less pain (45% pain reduction during activity and 74% at rest). Postoperative mean range of movement was 65% of the unaffected side. Three patients had an excellent result, three had good results, one had a fair result and one patient was unsatisfied. The mean postoperative disabilities of the arm, shoulder, and hand (DASH) score was 29.

The metacarpal head of the index ray is another donor site for VBG. It was used by Bengoechea et al (1 case) (11), Makino (1 case) (59), and Zafra (5 cases) (115). In all cases a satisfactory outcome was obtained, but an additional procedure on the radius (shortening or wedge osteotomy) was combined in all these cases.

Free VBGs from the iliac crest have also been used. Arora et al (6) and Gable et al (31) used this technique in 18 wrists each; good bony integration was noted in 16 wrists in both studies. Good clinical results were obtained in 15 cases by Arora et al and in 16 cases by Gabl et al.

Stages I and II are good indications for a VBG, provided the lunate is not fractured. Smoking is considered a general contra-indication for all VBG’s and this procedure is not suitable in older patients.

Decompression of the lunate

1. Levelling procedures of the DRUJ: radial shortening or ulnar lengthening?

Levelling the ulna to the radius by shortening the radius or lengthening the ulna (fig 1i) is based on Hulten’s finding (39) that Kienböck’s disease was more frequent in ulna minus variant wrists. Despite the fact that several authors have questioned this finding, good clinical results were reported with these levelling procedures (2,3,5,7,13,18,22,27,28,32,35, 50,70,74-77,83,88,92,98,100,102,111,114) (table I). The basic mechanism appears to be unloading of the lunate preventing further collapse. These procedures have been used for several decades and they...
probably have the best documented series in the treatment of Kienböck’s disease. Good outcome is reported in 69 to 100% of cases.

Proponents of more sophisticated procedures including lateral closing, lateral opening wedge osteotomies and medial closing wedge osteotomies of the radius (fig 1j), claim that the osteotomy changes the morphology of the distal radius (33,47,54,65,90,97,106). In general the results are good. Radial opening osteotomies produce a better decompression than closing wedge osteotomies (108). However the clinical outcome was not significantly different in one comparative series (43).

Some authors believe that the biological decompression effect of the osteotomy is responsible for the pain relief. Based on this, Illarramendi et al proposed a pure fenestration of the distal radius and ulna rather than changing their length or orientation (40) (fig 1h). In their series of 22 patients, 16 were pain free and 4 had only moderate pain. Schultz et al(86) confirmed these findings in their series of 10 patients.

Most papers and handbooks still recommend an osteotomy for stages I and II, some authors even for stage III, provided there is an ulna minus variance. The morphology of the sigmoid notch and the ulnar head must be evaluated prior to any joint leveling procedure. In morphological studies of the DRUJ (19,30,79) three different types of sigmoid notch of the radius were distinguished; in some cases, there may be a risk of creating DRUJ incongruity or impingement following a change in the length of one of the forearm bones. In these cases, some other type of decompressive procedure should be considered.

Although adequate pain relief has been obtained with levelling procedures (radial shortening and ulnar lengthening), the fate of the lunate following these procedures is not clear. Weiss et al (111) found no progression but Wada et al (106) saw further progression of the disease as we did in a previous study (22). The outcome was also determined by the occurrence of a revascularisation process (64).

Based on our personal experience with this technique (22) and the fact that intracarpal procedures can produce a similar unloading on the lunate (38,112) with similar clinical outcomes (69), we only have limited indications for these procedures, i.e. stage II and sometimes IIIa in wrists of non-smokers with a marked ulna minus and a parallel DRUJ (type 2).

2. Intercarpal procedures

One of the important consequences of lunate necrosis and the subsequent collapse is the disturbance of the carpal architecture, in particular rotational subluxation of the scaphoid. Based on the work of Watson et al (109) on the treatment of scapholunate ligament tears, some authors have reported good results with an STT arthrodesis for treating Kienböck’s disease (fig 1b) (82,109). It has been shown that this procedure unloads the lunate to a similar degree as a joint levelling procedure. The STT arthrodesis can also be combined with a VBG or other revascularisation procedure (Stage 3a). Most authors (mostly German and

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Ulnar lengthening

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Japanese) recommend this procedure as a primary treatment. The outcomes are good with Mayo wrist scores between 62 and 66 and a DASH score of 24.8. There were no significant differences between the STT fusion and radial shortening (18,97) and the outcome was better than following full radiocarpal fusion (95). In contrast however Van Der Dungen et al (104) reported better outcomes with a non-operative treatment than with an STT fusion.

Other intracarpal procedures include scaphocapitate arthrodesis (fig 1c), a capitate shortening (fig 1d) osteotomy and triquetro-capito-hamate fusion combined with lunate resection (fig 1e). Although capitate shortening decompresses the lunate, clinical and radiological results have been poor (8).

Scaphocapitate fusion has a similar biomechanical effect as the STT fusion. Sennwald and Ufenast (87) also found satisfactory results in 10 of their 11 procedures. It is also the primary choice for Moy and Peimer (68).

Capitatohamate fusion resulted in excellent outcome in the series of Inoue (42) (8 patients, all pain-free) and Oishi et al (45 patients, 42 painfree) (72).

A more sophisticated procedure was proposed by Wilhelm et al (113). After resection of the lunate, a transverse osteotomy of the capitare is performed and the proximal fragment of the capitare is brought into the empty space by callus distraction (fig 1f). They performed this in 14 patients, with a relatively short follow-up. Lu et al (57) had a larger series (30 patients) with reasonable results. Personally, we do not see any advantage as compared with a simple PRC. The procedure is contraindicated in pre-existing wrist stiffness. Main complications for this procedure are non-union, hardware failure and last but not least impingement between the radial styloid and the scaphoid with the development of osteoarthritis. This complication is hard to deal with since it appears to progress even following a radial styloidectomy.

Salvage procedures

Once osteoarthritis is present (stage IV) or in stiff wrists (stages IIIB), a reliable salvage procedure is preferred. As in other joints the options are arthrodesis, arthroplasty or denervation. Our experience with a total wrist fusion was less satisfying. We noted a high percentage of complications and unsatisfactory results (21) similar to Dap’s series (17). Our follow-up studies have demonstrated that preservation of some wrist motion is needed in order to achieve a good outcome, but the amount of ROM is not very important (24). Partial radiolunate (fig 1n) and radioscapulunate (fig 1o) fusions have been proposed to overcome the poor results of full wrist arthrodesis. Kilgus et al (49) found acceptable results in 5 cases of radioscapulunate fusion but the Wrightington group had 50% failures of radiolunate fusions (94).

Full radiocarpometacarpal arthrodesis (fig 1q) resulted in 55% pain reduction, a DASH of 51.4 and 70% return to previous occupations in the large series of Sauerbier (81). This was confirmed in a smaller series by Tambe et al (95).

Arthroplasty by simple resection is obviously the first step in a complete carpal collapse (fig 1a). Replacement by a silastic spacer was proposed by Swanson and de Groot Swanson (93) (fig 1g). The appearance of intraosseous cysts is the major drawback for this technique. Kaarela et al (46) had to remove 41% of the spacers in 39 patients. Wachtl and Sennwald (105) found similar problems and both authors do not recommend this technique any more.

Partial denervation of the wrist joint by sectioning the posterior and anterior interosseous nerve is a common adjunctive procedure. An isolated full denervation of the wrist joint is possible and is indicated in specific situations such as in older patients, moderate pain, and in patients desiring to retain full range of motion (ROM) and accepting only partial pain relief and probable further deterioration of the wrist joint (fig 1r).

In most series however the first choice is the PRC (fig 1p) in stiff stage III and in all stage IV wrists. Despite the theoretical possibility of damage to the lunate fossa, a proximal row carpectomy has led to very satisfying outcomes in most series and also in our hands (20).

Severe damage of the cartilage in the lunate fossa, ulnar translocation of the wrist (caution with
Kienböck’s disease is a progressive chronic wrist disorder due to aseptic necrosis of the lunate, leading to osteoarthritis of the wrist. It usually affects young and active adults.

Non-operative treatment is recommended for children or juvenile patients and in older patients with mild symptoms. Otherwise, surgical treatment is the first choice. However, surgical options are numerous. Based on current literature, it is impossible to clearly define specific indications for each procedure. What appears to be an absolute contraindication for a certain technique for one author does not seem to be important for another. Contradiction prevails. No obvious conclusions can be drawn and no evidence based recommendations can be given. Case series are small without randomized control trials, follow-up is generally short, and outcome measures are different and not comparable.

The choice of the procedure depends on the stage of the disease, the ROM of the wrist, the length of the ulna, the shape of the sigmoid notch of the radius and the presence of a lunate fracture. The surgeon’s experience, preference and skill also are important in decision-making.

For stage I and II we prefer a VBG with temporary scapho-capitate fixation. An alternative is a radial shortening osteotomy provided that the ulna is short and that the geometry of the DRUJ is convenient.

For stage IIIa, a VBG with an STT arthrodesis is our first choice, provided that the ROM is acceptable and there is no fracture of the lunate.

For stage IIIb and for stage IIIa with a fractured lunate and acceptable ROM, a solitary STT arthrodesis or a scaphocapitate fusion is recommended.

For stage IV and a stiff stage III wrist, a PRC gives excellent outcomes, even better than reconstructive procedures for earlier stages.

For those in which a PRC seems impossible due to severe damage to the lunate fossa of the distal radius, a full wrist arthrodesis in younger patients and a denervation in the older low-demand patients are proposed.

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