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A Case Report of Kienbock's Disease in a Thirteen-Year-Old Girl

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Abstract

Introduction: Kienbock's disease is characterised by progressive collapse of the lunate caused by avascular necrosis. It is rare and most commonly affects the dominant wrist of males between twenty and forty. It is extremely rare in children and there is little evidence discussing optimal management. Management in adults is based on classification, with less advanced disease (stage 1) treated conservatively with a period of immobilisation and more advanced disease (stages 2-4) treated surgically.

Case Report: A thirteen-year-old girl presented with a three-week history of left wrist pain following a hyperflexion injury. She complained of pain with activity, reduced range of movement and reduced power. Although xray was normal, MRI showed advanced avascular necrosis of the lunate confirming Kienbock's disease. She was managed with a Futura Splint. After two months her pain had improved and on examination she had an improved range of movement. Repeat xray showed no further lunate collapse.

Comments: Our case study has demonstrated that it is important to consider Kienbock's disease in children presenting with persistent wrist pain. An early diagnosis is essential due to its progressive degenerative nature. Our case study has confirmed that children with stage 1 disease can be successfully managed conservatively through immobilisation.

Key Words

Kienbock's; Teenager; Treatment; Case Report

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Introduction

Kienbock's disease is characterised by progressive collapse of the lunate caused by avascular necrosis. It was first described by Austrian radiographer Dr Robert Kienbock in 1910¹. It is a rare disease which most commonly affects the dominant wrist of male manual workers between the ages of twenty and forty². Although its aetiology is unknown, it has a higher incidence in those who have sustained trauma to the wrist. An increased incidence of

Kienbock's disease in people with a negative ulnar variance has also been described in the literature³. It is characterised by wrist pain which commonly worsens with activity, a reduced range of movement and decreased grip strength².

The disease is classified into four stages based on plain radiography according to Lichtman et al (Figure 1)⁴. The first stage of the disease can be diagnosed using magnetic resonance imaging (MRI).

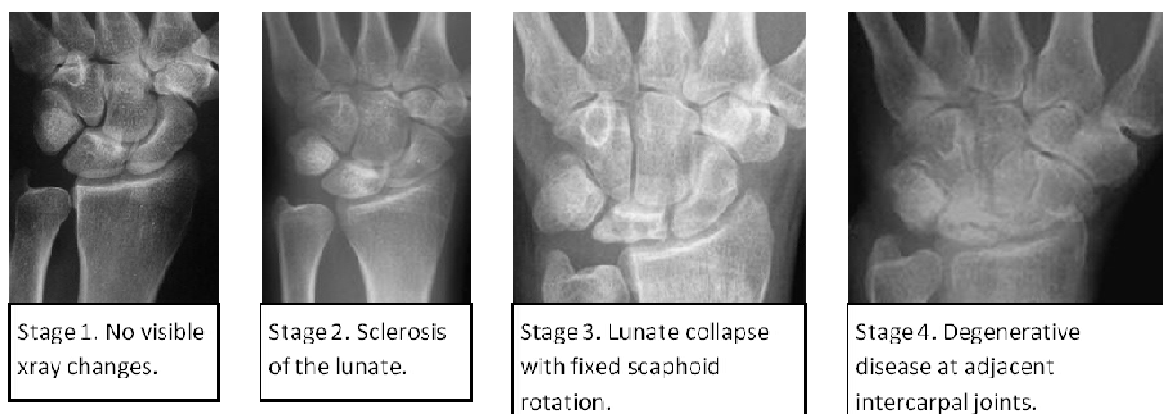


Figure 1: Radiographs demonstrating the Lichtman's four stages of Kienbock's disease

However, there are no changes on plain film. Progressive radiological findings include sclerosis of the lunate, lunate collapse, fixed scaphoid rotation and degeneration of the adjacent intercarpal joints⁴. Management is based on its classification, with less advanced disease (stage 1) usually treated conservatively with a period of immobilisation and more advanced disease (stages 2-4) occasionally treated surgically. Surgical options include temporary scaphotrapeziotrapezoidal pinning, joint levelling procedures and radial wedge procedures for those with stage 2-3 of the disease, and proximal row carpectomy, arthrodesis and arthroplasty for those with stage 4 of the disease⁵.

Kienbock's disease is extremely rare in children, although some cases are reported in the literature^{6,7,8}. There is little evidence discussing the optimal management of Kienbock's disease in children. It has been suggested that younger patients have a greater potential for remodelling and revascularisation, suggesting increased success rates with conservative immobilisation in this age group^{2,8,9}. We present a case of Kienbock's disease in a thirteen-year-old girl managed conservatively.

Case Report

A thirteen-year-old girl presented with a three week history of left wrist pain following a hyperflexion injury when landing awkwardly whilst dancing. She complained of persistent pain in the wrist particularly with activity, reduced range of movement and reduced power. On examination she was tender on palpation around the distal radius, distal ulna, scaphoid and lunate. She had limited flexion to 20 degrees and extension to 45 degrees due to pain.

A radiograph was suspicious of a fracture through the radial styloid with minimal displacement (Figure 2). She was treated for a fracture of her radial



Figure 2: A radiograph of the left wrist three weeks after initial injury (lunate highlighted).

styloid and managed conservatively with a Futura Splint. However, an MRI scan was arranged to rule out any scaphoid injury.

The MRI showed advanced avascular necrosis of the lunate confirming the diagnosis of Kienbock's disease (Figure 3). She was managed conservatively with a Futura Splint and analgesia for a further two months.



Figure 3: An MRI two months after the initial injury demonstrating advanced avascular necrosis of the lunate. (T1 image left and T2 image right).

On follow up after two months her pain had improved and on examination she had an improved range of movement. A repeat xray showed no further collapse of the lunate (Figure 4).

Discussion

The cause of Kienbock's disease is poorly understood. Although trauma tends to be a preceding factor, there are a number of risk factors that are thought to be contributory². Negative ulnar variance of the wrist has shown to be a significant risk factor for the development of the disease³ as have anatomical differences in the blood supply to the lunate leading to reduction in venous outflow¹⁰.

There are a number of imaging modalities that are useful in diagnosing Kienbock's disease. Initial



Figure 4: A radiograph of the left wrist four months after initial injury.

diagnosis is usually made based on plain radiographs. Radiographs are also used for staging of the disease, guiding appropriate treatment and evaluating the success of treatment. However, radiographs are only useful in diagnosing Lichtman's stages two to four of the disease, as there are no visible changes in stage one. Progressive radiograph findings include sclerosis of the lunate, lunate collapse, fixed scaphoid rotation and degeneration of the adjacent intercarpal joints. Computed tomography (CT) scanning may also be useful in demonstrating fracture or fragmentation of the lunate that may be difficult to see on plain radiograph².



Figure 5: A T2-weighted MRI showing bone marrow oedema in the lunate in the acute stage of Kienbock's disease¹⁷.

MRI is widely thought to be the most sensitive and reliable imaging modality in the diagnosis of Kienbock's disease. Unlike plain radiographs, it is useful in diagnosing the acute stages of the disease with bone oedema, particularly on the radial side of the lunate, clearly visible within a T2-weighted image. Decreased signal intensity can also be seen in

a T1-weighted image. However, this can also be seen in ulnar compaction syndrome or following wrist trauma¹¹.

Isotope bone scans also have a place in investigating Kienbock's disease. Bone scans expose the patient to significant lower doses of radiation than a CT and cost roughly half as much as an MRI¹². Negative bone scans will reliably rule out the condition. However, positive scans are not specific to Kienbock's disease and are therefore not diagnostic.

In adults, treatment of the disease is based on Lichtman's radiographic staging. It is recommended that adults with stage 1 Kienbock's disease are managed with a period of immobilisation⁵. Studies have however shown that adults in the later stages of the disease can have better outcomes when managed operatively¹³. These procedures include joint levelling procedures and radial wedge procedures for those with stage 2-3, and wrist fusion and proximal row carpectomy for those with stage 4. Several studies have suggested improved outcomes in adults treated with radial shortening compared to immobilisation^{13,14}. In some long-term studies, conservative management resulted in progressive deterioration of the lunate radiologically, but this did not necessarily correlate with a deterioration in symptoms^{4,15}. One particular study suggested that surgery should only be performed on symptomatic patients after an attempt of immobilisation and analgesia¹⁶.

Kienbock's disease is rare in children and there are few studies that attempt to compare outcomes in children treated conservatively or surgically. One literature review suggests that outcomes are better in children less than 14 following conservative management with a period of immobilisation as they have greater potential for revascularisation and remodelling⁸. Further studies also recommend conservative management in children with a period of prolonged immobilisation⁹.

Our case study has demonstrated that it is important to consider Kienbock's disease as a differential in children presenting with persistent wrist pain. An early diagnosis is essential due to the progressive degenerative nature of the disease. The majority of patients with stage 1 disease will go on to worsen even with prompt diagnosis and immobilisation of the wrist^{4,15}. Our case study has confirmed that children with stage 1 disease can be successfully managed conservatively through immobilisation of the affected wrist. This supports previous studies that suggest that children with Kienbock's disease have a better outcome with conservative management when compared to adults possibly due to their greater potential for

revascularisation and remodelling. To increase the chances of successful conservative management in children, it is important to consider a differential of Kienbock's disease and investigate appropriately to ensure an early diagnosis.

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