CASE REPORT

# Stage III Kienböck's disease treated with hyperbaric oxygen: the role of an unusual approach to a rare condition

Paulo Jorge Figueira,[1,2] Diogo Alpuim Costa,[3,4] Nunzio Barbagallo,[5] Francisco Gamito Guerreiro[4,6]

[1]Orthopaedic Department, Centro Hospitalar de Lisboa Central, Hospital Curry Cabral, Lisbon, Portugal
[2]Centro de Medicina Naval, Marinha Portuguesa, Almada, Portugal
[3]Haematology and Oncology Department, Instituto CUF de Oncologia (I.C.O.), Lisbon, Portugal
[4]Centro de Medicina Subaquática e Hiperbárica, Marinha Portuguesa, Marinha Portuguesa, Lisbon, Portugal
[5]Physics and Astronomy Department, Università di Bologna, Bologna, Italy
[6]Centro de Investigação Naval (CINAV), Marinha Portuguesa, Almada, Portugal

**Correspondence to**
Dr Diogo Alpuim Costa, diogo.costa@jmellosaude.pt

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## SUMMARY

Kienböck's disease is a rare condition characterised by avascular necrosis of the lunate bone. Its natural history and aetiopathogenesis have not yet been clarified, nor are its triggering factors identified. We present a case of a 17-year-old male gymnast, without relevant medical/family history, with stage IIIA Kienböck's disease diagnosed in 2016. Initially, submitted to conservative treatment that proved to be insufficient. Consequently, surgical treatment was proposed, but refused. The patient instead underwent experimental treatment with hyperbaric oxygen (120 sessions, 100% oxygen at 2.5 atm, for 70 min periods, once daily, five times per week). In April 2018, a favourable clinical and radiological evolution was observed, with an improvement in the patterns of pain, motion and strength and an almost complete involution of the process of aseptic necrosis of the semilunar. To the best of our knowledge, this is the first report of Kienböck's disease treated with hyperbaric oxygen.

## BACKGROUND

Kienböck's disease is characterised by avascular necrosis of the lunate bone. The complete pathophysiology of this disease is yet to be understood, but it seems to relate to an apparent combination of vascular, anatomic and traumatic insults, resulting in progressive bone infarction and collapse with biomechanical compromise. Generally, it afflicts mostly adults between 20 and 40 years of age, typically male manual workers. Its natural history is not well known, and the clinical features do not necessarily correlate with the radiological findings that are most times the cornerstone of the diagnosis.

The main objective of the treatment is to relieve pain. However, despite the different therapeutic approaches mainly based on the radiological stage, there is no gold standard for Kienböck's disease. For stages II and IIIA of the disease, the treatment is centred on joint levelling and/or revascularisation procedures. We present a case of a young patient with Kienböck's disease stage IIIA who underwent treatment with hyperbaric oxygen therapy (HBOT). As a complementary treatment, this strategy has been reported to improve the outcomes in patients suffering from femoral head necrosis. To the best of our knowledge, this is the first report of Kienböck's disease treated with this non-surgical approach.

## CASE PRESENTATION

A 17-year-old male gymnast, without relevant medical and family history, reported in February 2016 pain in his left wrist with 2 weeks of evolution and no associated traumatic event. The pain was constant (5 points at Visual Analogue Scale, VAS), located at the level of the scaphoid–lunate space, with no irradiation and worsened by the dorsiflexion of the wrist.

The patient performed a wrist radiograph that revealed no acute osteoarticular lesions or degenerative changes. Analgesics and eviction of manual physical efforts were prescribed. After 3 months, there was a recurrence and worsening of pain with crepitus and repercussions in daily life (7 points at VAS) and sports activities (10 points at VAS). The wrist's mobility was minimal because it triggered maximum pain. Severe night and light touch pain were present. A new radiographic study revealed a subchondral sclerosis of the lunate (figure 1). MRI confirmed the diagnosis of Kienböck's disease, stage IIIA of Lichtman classification (figure 2A–C). An elective hospitalisation was proposed for surgical treatment with radius shortening osteotomy. However, the patient and his family were sceptical of this invasive approach. Therefore, this procedure was postponed in favour of a conservative treatment with a plaster cast forearm immobilisation for 50 days and a potential treatment with HBOT. After this, the use of a forearm orthosis for pain relief was recommended. Due to bureaucratic and logistical issues, there was a 5-month gap until starting HBOT. In January 2017, the patient started HBOT in a multiplace hyperbaric chamber, exposed to 100% oxygen at 2.5 atm, for 70 min periods, once daily, five times per week. In a first stage, 30 sessions were prescribed, after which the patient attended an evaluation. During this consultation, the patient reported significant improvement in mobility, pain intensity and oedema resolution, with a progressive increase in grip strength and the need for further treatment was decided. After an evaluation MRI (session 77th) showing an important radiological response (figure 2D), the decision was to extend the treatment to 120 sessions.

## OUTCOME AND FOLLOW-UP

In the last follow-up consultation (April 2018), the patient presented mild pain, only for activities of

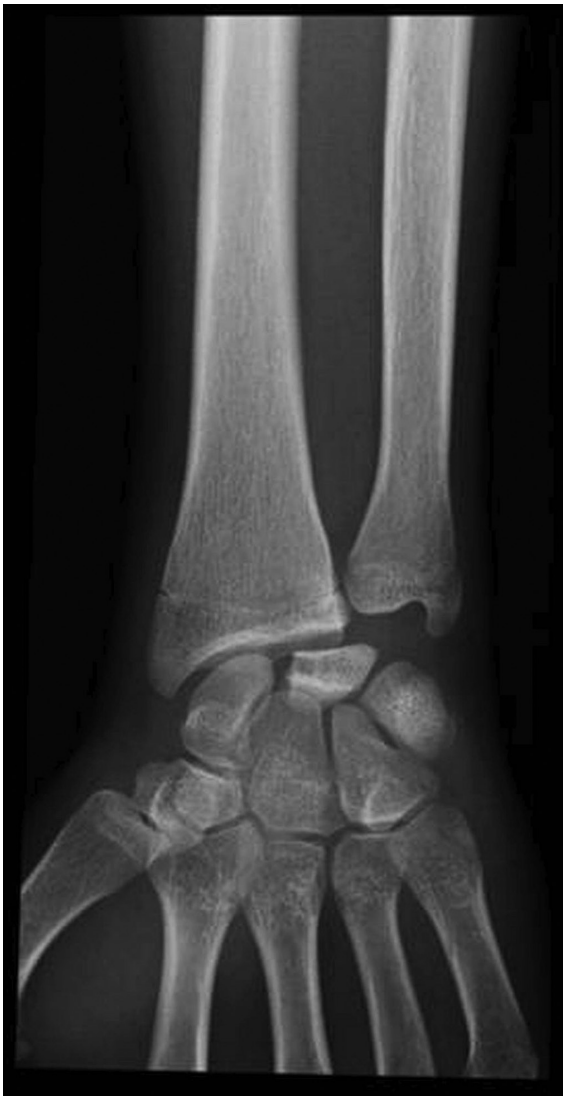

**Figure 1** Frontal radiograph of the left wrist before HBOT. There are diffuse heterogeneity and sclerosis of the lunate bone. Negative ulnar variance is noted. HBOT, hyperbaric oxygen therapy.

axial load sport's related (VAS 4), carpus's crepitation with wrist ulnar deviation and a slight pain and decrease of the extension of the fist in the last degrees of extension (joint painless range of motion for 90°—figure 3). Functional evaluation was performed, using the Patient-Rated Wrist Evaluation score (7 points), the disabilities of the arm, shoulder and hand score—QuickDASH (0 points) and QuickDASH sports performing (75 points). Grip strength and mobility were evaluated by comparison with the unaffected side. These parameters did not reveal significant alterations compared with the contralateral side. Notwithstanding, the absence of pain and good mobility for daily activities, the patient maintains a limitation to the previous sporting practice. The MRI revealed (figure 2E–F) almost complete involution of the process of aseptic necrosis of the semilunar.

We believe that the follow-up period should include a semiannual consultation during the first 2 years, followed by an annual appointment.

## DISCUSSION

Kienböck's disease is a rare condition. Its natural history and aetiopathogenesis have not yet been clarified, nor its triggering

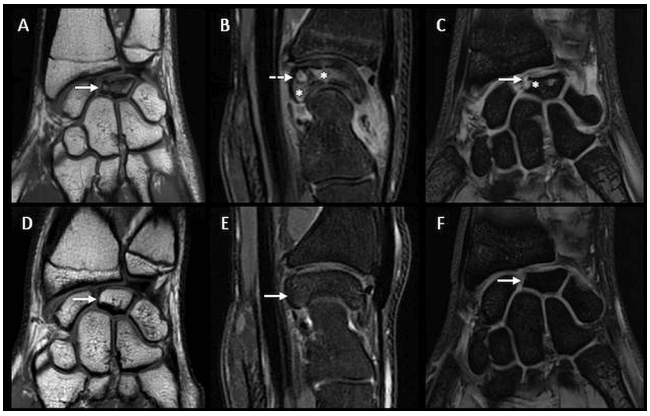


**Figure 2** Left wrist MRI images before (A–C), during (D) and after (E and F) HBOT. (A) T1-weighted coronal image shows diffuse hypointensity, heterogeneity and altered trabeculation of the lunate bone (arrow). (B) Fat-saturated proton density-weighted sagittal image depicts areas of hyperintensity (*) traducing bone marrow oedema and intraosseous cystic lesions (dashed arrow). (C) T2*-weighted gradient-echo coronal image depicting partial bone collapse (*) as well as fragmentation (arrow) on the radial side of the lunate bone. (D–F) Images on the same MRI sequences as A, B and C, respectively, after 77 (D) and 120 (E and F) sessions of HBOT, show a significant improvement of disease with remission of previous changes. There is a practically complete recovery of the signal and homogeneity of lunate bone (arrows). HBOT, hyperbaric oxygen therapy.

factors identified.[1 2] It is believed, as in the works published by Kienböck, that traumatic event associated with ligament and vascular injury may be implicated in the mechanism of the disease, and there are presently three coexisting theories: mechanical, vascular and traumatic theory.[3]

This condition has a higher predominance for males, especially in individuals with a strong impact on manual labour tasks. However, the incidence and prevalence of this disease in athletes is unknown.[3]

The vascularisation of the lunate occurs by dorsal and palmar branches of the radial and ulnar arteries. It is estimated that only in 20% of the cases the palm branches are found conferring an increased risk of osteonecrosis.[4] The vascularisation of the proximal pole is more exiguous which predisposes to avascular necrosis events with increased probability.[3]

The initial clinical manifestation may be subtle but may mimic a wrist sprain or any inflammatory condition affecting the limb.[3] Dorsal and palmar oedema and crepitation to palpation stand out. Joint mobility may be affected by the associated pain, aggravated by maximal flexion and extension, thus assuming pain as the main factor of dysfunction,[1] there may also be a decrease in grip strength.[2] This episodic pain may be associated with the period of fragmentation caused by fracture of the subchondral

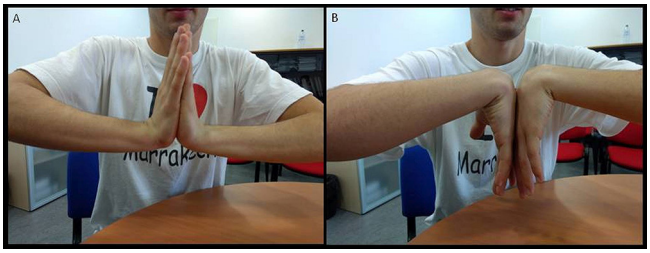


**Figure 3** Patient's range of movement.

bone, absence of restriction of joint mobility and concomitant synovitis.[1]

The clinical suspicion for the diagnosis, combined with an MRI allows for detecting the disease at an earlier stage. Thus, the T1 and T2 changes in the signal can detect changes in the vascularisation[2] with bone necrosis,[3] allowing for the establishment of an earlier therapeutic protocol.[1] The implementation of this protocol allows monitoring the evolution of the treatment conferred by the hypersignal in T2 that manifests bone revascularisation.[2]

The presumptive diagnosis presupposes the exclusion and control of several clinical conditions that may contribute to the aetiopathogenesis of this condition, such as coagulopathies, haemoglobinopathies, lupus, arterial disease, conditions that favour venous stasis (obesity), alcohol consumption and anabolic steroids.[1–3 5]

Regardless of the treatment selected, its primary goals should be to relieve pain, prevent bone collapse and prevent or delay osteoarthritis. There are currently several treatment modalities, varying from the conservative methodology with immobilisation and anti-inflammatory drugs and change in lifestyle,[3] to numerous surgical approaches, none of which is presently considered the gold standard in the treatment of this pathology.[1] Procedures such as lunate revascularisation, osteotomies or for the treatment of sequelae caused by the collapse of the lunate (graft, vascularised graft, prosthesis, proximal carpectomy, arthroplasty or arthrodesis) are well known.[4 6]

From the review of the initial imaging study, we can conclude that in the present case, we were faced with a stage IIIA lesion of the Lichtman classification. The most consensual therapeutic options for treatment of stage IIIA are radio-osteotomies, vascularised bone grafts, temporary scaphocapitate osteotaxia or scaphotrapeziotrapezoid, capitate shortening with or without capitohamate arthrodesis.[2]

Despite the surgical technique applied, the outcome regarding bone regeneration has not yet been established.[7] In this sense, other approaches were developed such as transfusion of bone marrow after microperfurations stabilised with osteotaxy and low-intensity pulsed ultrasound with a demonstration of non-inferiority results, postulating possible effect of the properties of mesenchymal stem cells and cytokines and growth factors with neovascularisation properties.[7]

Therefore, considering promising work already published on the effects of HBOT: (1) the biological potential of the mesenchymal cells, (2) the wound healing physiology, (3) the angiogenic potential effect associated with the treatment of osteonecrosis of the femoral head; in addition to the patient's family reluctance towards the surgical option, we opted for the conservative approach using HBOT.

The precise role of HBOT is not yet established. It is now known that HBOT allows for higher concentration and diffusion in the blood plasma and consequently the level of oxygenation in the tissues. In parallel, microvascular perfusion is stimulated by the synergistic effect of increased synthesis of nitric oxide.[8] HBOT seems to contribute to the reduction of bone and peripheral oedema, facilitating venous drainage and promoting angiogenesis by contributing to the cellular stimulus inducing the formation of fibroblasts and formation of collagen[9] through mechanisms that stimulate the production of antioxidant enzymes with the production of cytokines and growth factors.[8] In addition, angiogenesis may be enhanced by the recruitment of bone marrow stem cells under the effect of HBOT, with emphasis on the endothelial progenitor cells.[10–12]

Concomitantly, hyperoxia acts on bone metabolism by stimulating osteoclastic activity on the necrotic bone.[2 9] Animal studies have shown that areas with capillary proliferation and fibroblastic activity stimulate osteoblastic and osteoclastic activity promoting the replacement of necrotic bone with new bone.[6] The key factor here seems to be the balance between osteoblastic and osteoclastic activity. At the cellular and molecular levels, the beneficial effects of HBOT may be via regulating serum osteoprotegerin, accelerating osteoblast differentiation and suppressing osteoclasts genesis activation, shifting the balance between bone formation and bone resorption in a direction that promotes bone regeneration.[13]

The HBOT sessions, in this case, were held inside a hermetically sealed hyperbaric chamber classified as type IIb medical device (Directive 93/42 ECC of 14 June 1993, concerning medical devices). The patient performed our routine protocol (100% oxygen at 2.5 atm, for 70 min periods, once daily, five times per week) for general medical conditions. This treatment included a total of 120 sessions with two intervals coincident with the school holidays (2 weeks within the first 30 sessions and 8 weeks between 60 and 90 sessions). The first cycle of 30 sessions had a marked impact on the mobility gain, which we associate with the rapid resolution of oedema and pain management. During the treatment, clinical and radiological improvements were observed, triggering our decision to have the patient undergo more than 100 treatment sessions, surpassing the number established for other chronic conditions treated at our centre.

Despite this favourable clinical outcome, there are some unanswered questions, namely: in light of the current evidence related to the natural history of this disease, can we assume that an adolescent could have a stage IIIA spontaneous remission? This fact is known to be more frequent at the paediatric age[14–16]; however, while awaiting the start of HBOT there was no improvement in symptoms. Could interruption of our protocol have influenced the result obtained, even in the absence of cumulative side effects of hyperoxia (Lorrain-Smith and Paul Bert's effects)? Could the repetitive microtrauma associated with our patient's competition gym explain the occurrence of his illness? Is the patient fit in the medium to long term to resume his high-impact sports activities? On the objective clinical and functional evaluation of the patient's evolution during the HBOT period, there are some biases to disclose: the retrospective assessment of pain and functional scales, the absence of quantitative determination of the mobility and grip strength, the failure to perform arthroscopy for the initial in loco staging disease for improved characterisation of joint surfaces and eventual ligament dysfunction.

The indications for HBOT are well defined in the 10th European Consensus Conference on Hyperbaric Medicine held in 2016. Its use for the treatment of osteonecrosis of the femoral head is a type II indication and its application in Kienböck's disease continues to be off-label.[8 16] The success obtained with

## Learning points

- Kienböck's disease is a rare condition requiring a high index of suspicion.
- There is currently no gold-standard treatment.
- The current surgical treatments do not seem to totally change the natural history of the disease.
- Hyperbaric oxygen therapy (HBOT) may take place as a new successful therapeutic strategy.
- Can HBOT change the natural history of the disease in the long term?

this treatment could lead to HBOT implementation for Kienböck's disease, bringing great value added to the existing therapeutic arsenal. Therefore, this conservative treatment may be a valid option for this clinical context, requiring: (1) more cases to be treated with HBOT, (2) a specific HBOT protocol, (3) a longer follow-up period (possibly up to 30 years) and (4) a cost–benefit analysis versus other therapeutic approaches.

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**Patient consent** Obtained.

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