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Short Communication

Value of Ultrasound in Haemophilia. A Brief Review

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Haemophilia is an X-linked heritable coagulopathy with a prevalence of approximately 1 in 10,000 individuals [1]. The haemophilia A (factor VIII deficiency) comprises approximately 80% of cases and the haemophilia B (factor IX deficiency) comprises approximately 20% of cases [2].

Haemophilia is classified as "mild", "moderate" or "severe" according to the degree of clotting factor deficit, subjects with a severe deficit have a highest risk to develop soft-tissue haematomas, bruising, retroperitoneal bleeding, intracerebral hemorrhage and intraarticular blood (haemarthrosis), that is one of the most common problem in these patients.

The joint bleeding triggers and perpetuates synovitis and provokes cartilage damage, which may start even after the first exposure to blood through activation of cytokines and caspases, leading to a permanent and progressive joint damage [3], this phenomenon is denominate haemophilic arthropathy.

The replacement therapy of coagulation factors has reducing bleeding frequency [4] but it does not remove the risk of accidental traumatic conditions, furthermore subclinical haemarthrosis may developed at great joints as knee, ankle, and elbow when plasma levels of the factor are the lowest, previous to a new factor dose usually [5]. Typically joints that sustained several consecutive bleeding episodes (> 3) within a short time (< 6 months) are defined as 'target joints' and these are prone to develop bony and soft tissue changes, cartilage destruction and synovial hypertrophy with or without synovitis [6].

The magnetic resonance(MR) is the gold standard to identify early structural changes secondary to haemophilic arthropathy however it is expensive and it is not available at all hospitals. In the other hand, the ultrasound (US) is cheap, it do not need sedation in children and our experience in this diagnostic imaging modality has improved since 1987 when Wilson., *et al*. [7] described its use to control of bleeding in soft tissues.

The typical US findings of haemarthrosis are a dark, black (anechoic) area corresponding to the fluid pushing and bulging the tendon, this area modifies its size with the pressure of the transducer because the fluid is displacing when pushed by the probe. Fluid in the joint should not be confused with normal cartilage (Figure 1). Occasionally a greyish echogenic area can be seen (Figure 2), consistent with the reactive synovial hypertrophy (acute), if this persist during 15 days, it should be considered as chronic synovitis, both involves therapy because the highest bleeding risk [2]. In context of trauma, an intra-articular mixed echogenic material that correspond to haematoma can be see (Figure 3).

Poonnoose., *et al*. [8] developed a concordance study between the physical exam using the Hemophilia Joint Health Score (HJHS version 2.0) and the radiological evaluation. They used the Pettersson score for X-rays [9], the tool of the International Prophylaxis Study Group (IPSG score) for the MR and the corresponding US scale in 55 joints of subjects with severe haemophilia or Von Willebrand disease. In this cohort, subjects with normal physical exam had soft changes or osteochondral changes on US or MR evaluation, furthermore 33% of the joints had findings in US or MR but it did not have pathological changes on X-ray, so a normal x ray or physical exam do not exclude joint disease in haemophilia.

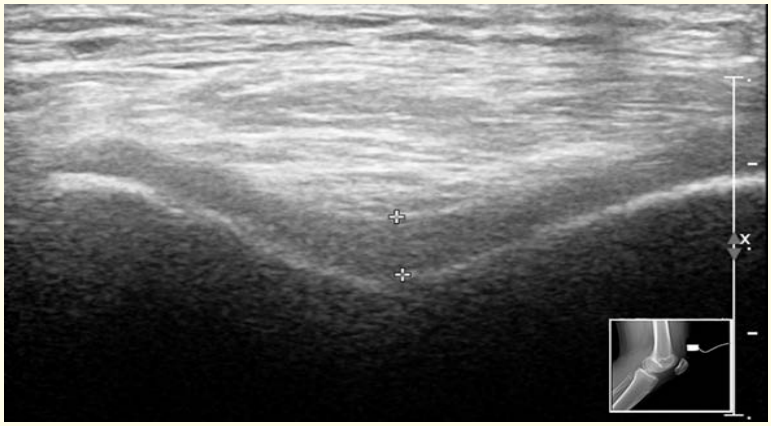

Figure 1: *Knee ultrasound. Axial scan over the knee. A hypoechoic band (between the calipers) over the tibia normal cartilage.*

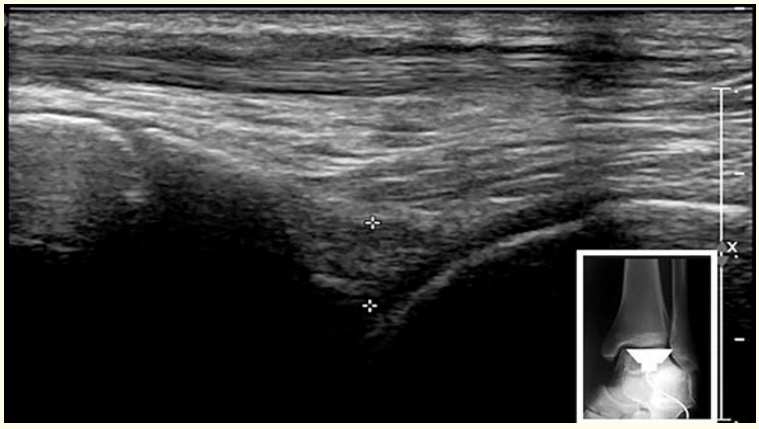

Figure 2: *Synovial hypertrophy. Transversal scan of ankle. The calipers indicate intra-articular isoechoic lesion, a typical image of reactive synovial hypertrophy.*

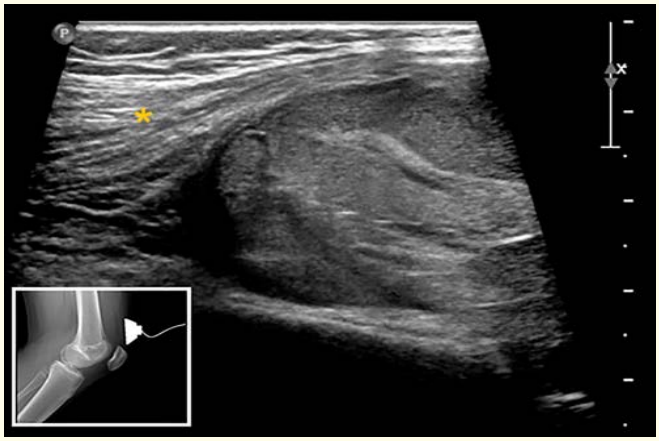

Figure 3: *Acute haematoma. Longitudinal scan of knee. An oval mixed echogenic lesion under the patellar tendon (asterisk) correspond to haematoma.*

Painful joints is not cause by bleeding always, for example, tendinosis and enthesopathy contributing to musculoskeletal pain and are present in 33% of haemophilic joints [10]. Tendinosis describes tendon swelling and enthesopathy are the structural abnormalities as tendon echogenicity changes, often caused by repetitive stress injury related with chronic degenerative changes. Other symptoms and findings such as loss of range of motion, warmth and swelling are non-specific and may occur with bleeding, sinovitis, arthritis or other musculoskeletal syndromes [11].

Other advantage of US is the possibility to optimize clotting factor replacement therapy with periodically joint evaluation during the treatment to determine the persistence of bloody effusions [10], however, the periodicity of US evaluation have not established yet.

Conclusion

In conclusion, the US is a fundamental tool in the current treatment of haemophilia and it is not limited to describe intraarticular bleeding only. The investigation road about haemophilia and US have begun until now.

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