Development and definition of a simplified scanning procedure and scoring method for Haemophilia Early Arthropathy Detection with Ultrasound (HEAD-US)

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Summary
The aim of this study was to develop a simplified ultrasound scanning procedure and scoring method, named Haemophilia Early Arthropathy Detection with UltraSound [HEAD-US], to evaluate joints of patients with haemophilic arthropathy. After an initial consensus-based process involving a multidisciplinary panel of experts, three comprehensive and evidence-based US scanning procedures to image the elbow, knee and ankle were established with the aim to increase sensitivity in detection of early signs of joint involvement while keeping the technique easy and quick to perform. Each procedure included systematic evaluation of synovial recesses and selection of a single osteochondral surface for damage analysis. Based on expert consensus, a simplified scoring system based on an additive scale was created to define the joint status and, in perspective, to offer a tool to evaluate disease progression and monitor the result of treatment in follow-up studies.

Keywords
Haemophilic arthropathy, joints, scoring systems, ultrasonography, synovitis, osteochondral damage

Introduction
Early signs of haemophilic arthropathy (HA) are known to be encountered in asymptomatic joints in which just one or a few bleeds have previously occurred (1). Diagnostic imaging has the potential to disclose subclinical disease and influence the specific treatment strategy to limit disease progression. Regardless of the technique, however, imaging at initial stages of arthropathy is often missed or underused in routine practice because of the lack of justification for submitting patients to imaging departments to scan multiple joints that appear nearly asymptomatic. Among the available modalities, ultrasonography (US) is intrinsically more flexible and time- and cost-effective than magnetic resonance imaging (MRI) as a screening tool. The main issue is how best to harness and quantify the information generated at US in a simplified and quick method for evaluating and scoring disease activity and damage in HA. Accordingly, the aim of this paper was to develop a simple US scanning procedure and scoring method for HA, able to shorten the examination and interpretation time with good intra- and inter-reader reliability. This method, named Haemophilia Early Arthropathy Detection with UltraSound [HEAD-US], was developed with the final aim to integrate US in the routine practice of haemophilia centres as a close complement of physical examination.

Materials and methods
Development of the scanning procedures and scoring method
We used a widely accepted consensus formation method, i.e. the nominal group technique (2), during four structured face-to-face meetings specifically designed to combine opinions from a group of nine experts with proven experience in haemophilia care and/or musculoskeletal US to facilitate the consensus on the field of study topics and the contents of the HEAD-US score. The following requisites for the HEAD-US scanning protocols were agreed: i) easiness in learning by non-expert musculoskeletal sonologists; ii) easiness of use by means of conventional US machines, including portable machines; iii) lack of any requirement for high-end and/or proprietary technology; iv) informativeness in the assessment of joint status compared with MRI; v) reliability and repeatability for being used to monitor treatment efficacy and damage evolution in longitudinal studies; vi) time efficiency to be implemented in daily
practice as part of the physical examination instead of being an independent diagnostic procedure; and vii) easiness to apply within a busy clinical practice. The agreed requisites for the contents of the HEAD-US scoring scale were: i) inclusion of indicators for disease activity and disease damage; ii) adequate weighing of these items to formulate a scale that most closely reflects the joint status; and iii) equal item list for the elbow, knee and ankle.

Within the nominal group technique meetings, live-demo interactive sessions examining joints of consenting adult patients allowed the board members to build-up a tailored methodology for HA evaluation. A total of sixty joints (including n=15 elbows, n=27 knees and n=28 ankles) from 30 patients with haemophilia A and B (mean age [SD] 31 ± 12, age range, 20-48 years), were examined during these meetings giving an acceptable coverage of the spectrum of severity of joint involvement. Ethic board permission and written informed consent were obtained. A fourth meeting resolved issues that did not previously achieve a consensus: the most relevant ones concerned the ability of US to recognise haemosiderin and the inclusion/exclusion of Doppler imaging in a comprehensive joint examination. With regard to haemosiderin detection, a further ad-hoc subset of eight patients with moderate to severe synovitis in the elbow, knee or ankle was examined with 12.5MHz US (iU22©, Philips ATL, Bothell, WA, USA) and correlative 1.5T (Avanto©, Siemens, Erlangen, Germany) gradient-echo (GRE) axial T2* MRI to obtain one-to-one comparison of findings. Seven of them presented with extensive haemosiderin deposits at GRE MRI; one did not exhibit the susceptibility artifact related to intra-articular haemosiderin deposits. Additional eight non-haemophilic patients affected by chronic joint disorders, such as juvenile idiopathic arthritis (n=2), rheumatoid arthritis (n=3) and osteoarthritis (n=3) who exhibited joint synovitis and absence of any detectable sign of haemosiderin deposition at MRI were also examined. Written informed consents of patients or parents and asests of minors were obtained. In these 16 patients, analysis of synovial echotexture was performed blindly by three readers with the DICOM viewer OsiriX© 4.1 software (OsiriX©, Geneva, Switzerland) in order to define whether US is able to i) recognise specific findings of haemosiderin deposition and ii) differentiate synovial proliferation in haemophilia compared to other chronic joint disorders. Readers were radiologists (C.M., A.T., G.R.) with different level of experience in musculoskeletal US and namely; reader 1 (C.M.), >15 years; reader 2 (A.T.), 7 years; reader 3 (G.R.), <3 months.

**Reading and reliability evaluation**

The next step included practical testing of the preliminary HEAD-US scoring system in the version agreed in the previous meetings. For this purpose, a fifth nominal group technique meeting consisting of a live-demo of US features followed by calibration exercises was arranged. Ten randomly selected haemophilia patients were scored by the same three readers separately using the version of the HEAD-US score agreed in the previous meetings. A preliminary atlas of US images was used in the reliability exercise to calibrate. Results and discrepancies were internally and critically discussed among the readers and no further suggestion that the scoring scale should have to be modified was made.

Lastly and following calibration, we performed the intra-reader and inter-reader reliability evaluation, the results of which are presented in this paper. The same three readers were asked to examine a sample of 96 joints (including 20 elbows, 34 knees and 42 ankles) from 49 haemophilic patients (Table 1). Two joints per patients, either symptomatic or asymptomatic having a prior history of at least one bleeding episode, were included. Blinded to clinical information and the other readers' scoring, they performed the US examination independently with a linear array 12.5MHz US transducer (iU22©, Philips Healthcare). After one-month wash-out period, each reader evaluated a reduced sample of 38 joints randomly selected from the previous patients' cohort to measure intra-observer variability. The intra- and inter-observer agreement of the HEAD-US score was then calculated. k statistics were used and k values were reported as weighed k with linear weights. 95% confidence intervals (CI) and standard error were also reported. Agreement was defined on the basis of Fleiss classification: <0.40, poor; 0.40-0.59, moderate; 0.60-0.75, good; >0.75, excellent (3). Cronbach’s alpha was used to assess the internal consistency of the method considering values of alpha useful for clinical purposes at least equal to 0.90 (4). Statistical analysis has been performed with statistical software (MedCalc - version 12.3.0).

**Results**

**The HEAD-US scanning procedures**

Based on the consensus work described above and literature analysis, the following scanning procedures to examine the elbow, knee and ankle in patients with HA were finalised.

**Elbow**

- **E1a.** The patient keeps the forearm on the examination bed palm up extending the elbow as much as possible. The probe is

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**Table 1: Main patients characteristics and clinical features.**

<table>
<thead>
<tr>
<th>Patients (n)</th>
<th>Mean age ± SD (range)</th>
<th>BMI</th>
<th>Haemophilia A/B</th>
<th>Disease severity based on factor levels (mild/moderate/severe)</th>
<th>Mean WFH score (elbow/knee/ankle)*</th>
<th>Mean HJHS 2.1 score (elbow/knee/ankle)*</th>
</tr>
</thead>
<tbody>
<tr>
<td>49</td>
<td>34±17(2-69)</td>
<td>24.5</td>
<td>41/8</td>
<td>17/5/27</td>
<td>3.15/3.38/3.63</td>
<td>6.42/5.35/6.0</td>
</tr>
</tbody>
</table>

* values referred to the examined joints. BMI, body mass index; HJHS 2.1, Hemophilia Joint Health Score 2.1; n, number; SD, standard deviation; WFH, World Federation of Hemophilia.
placed in an anterior transverse plane at the level of the mid-distal arm, 10 cm proximal to the elbow crease. While sweeping the transducer down toward the joint line, the humeral shaft is seen progressively enlarging and becoming flat. At the metaphyseal level, two adjacent bony grooves appear on the anterior aspect of the humerus, representing the radial (lateral) and the coronoid (medial) fossae (Figure 1A). The first one is shallow and houses the radial recess of the joint, the second is deeper and contains the coronoid recess. Both recesses are roofed by the anterior fat pad. In cases of abundant joint effusion, the radial and coronoid recesses coalesce to form a unique complex.

- **E1b.** Sweeping the transducer down from the level of the two joint recesses on transverse planes, the distal humeral epiphysis comes into view. It consists of the convex capitellum (lateral) and the concave trochlea (medial). This osteochondral surface can be identified based on its wavy appearance and bilayered structure formed by a superficial anechoic band of articular cartilage and a deep linear echo related to the subchondral bone (Figure 1B).

- **E1c.** After shifting the transducer slightly lateral to the midline, sweep it down on transverse planes to reach the rounded profile of the radial head. Forearm pronation and supination may help to check the status of the radial head as it rotates.

- **E2a.** The probe is then placed in an anterior sagittal plane aligned over the radiocapitellar joint. The main bony landmarks to identify the joint line are the convex humeral capitellum and the squared radial head (Figure 1C). Both are covered with a uniform band of cartilage. In normal states, the capitellum has a smooth rounded profile. Cranial to the capitellum, the radial recess is located on the floor of the radial fossa.

- **E2b.** Shifting the probe medially on sagittal planes over the joint line, the coronoid process appears as a pointed structure. Its shape looks different from the radial head that is more prominent and squared (Figure 1D). Cranial to the coronoid, the trochlea appears convex distally and concave proximally. The coronoid recess lies on the floor of the coronoid fossa.

- **E3.** The posterior elbow is examined while keeping the joint flexed 90° with the palm resting on a table. Cranial to the olecranon, the probe is placed in a mid-sagittal plane over the distal myotendinous junction of the triceps. Care should be taken to align the transducer to the long-axis of the forearm. The olecranon recess is located between the floor of the fossa and the posterior fat pad (Figure 1E).

**Knee**

- **K1.** The patient is asked to lie down on the examination bed keeping the knee at 30-40° flexion. The probe is placed in a mid-sagittal plane with its inferior edge on the upper pole of the patella. The suprapatellar recess can be found cranial to the patella and underneath the quadriceps tendon (Figure 2A). From the mid-sagittal plane, the probe is then shifted medially and laterally to cover the full width of the recess.
• K2a and K2b. Imaging is then extended over the lateral and medial sides of the patella to examine the parapatellar recesses. Keeping the knee at 30–40° flexion, the probe is placed over the middle third of the patella and then swept lateral and medial on transverse planes. The parapatellar recesses lie between the femoral condyles and the patellar retinacula (Figure 2B). Once they have been recognised, the probe is moved anteriorly and posteriorly to cover their full extension.

• K3. The V-shaped trochlea is examined on transverse planes by keeping the knee hyperflexed. With maximal knee flexion, the femoral trochlea emerges from underneath the patella and can be extensively evaluated with US. Patients with advanced knee arthritis may be unable to flex the joint maximally. In these cases, the examiner may push the patient’s leg gently towards the thigh to obtain the highest possible degree of flexion. The probe is placed cranial to the patella and shifted up and down to cover most of the extension of the trochlear cartilage. When sweeping the probe along the curved trochlea care should be taken to keep it perpendicular to its surface to avoid any artifact. The articular facets of the trochlea are carefully evaluated for symmetry, status of boundaries (osteoophytes) and thickness (Figure 2C).

• K4. The patient is finally asked to rotate the leg externally while maintaining 20–30° knee flexion. The transducer is placed in a coronal plane over the medial joint space in the long axis of the medial collateral ligament. In normal states, the external boundaries of the femur and tibia facing the joint space appear smooth and regular (Figure 2D). Osteophytes can be appreciated as discrete bony outgrowths extending superficially. Owing to the still cartilaginous surfaces of bones that makes osteophyte formation unfeasible in children, this scanning plane is not performed in this age group.

Ankle

• A1a. The patient is seated on the examination bed with the heel lying on a table and the forefoot slightly elevated above it. The probe is placed in a midsagittal plane over the dorsal aspect of the tibiotalar joint. With the ulnar side of the same hand holding the probe, gentle pressure is exerted on the dorsal forefoot to pull the talar dome as much as possible out of the tibial cover. The anterior recess of the tibiotalar joint extends from the joint line to the junction of the neck and head of the talus, underneath the intraarticular fat pad (Figure 3A). The probe is then shifted lateral to medial to cover the full extension of the osteochondral surface of the talar dome.

• A1b. The probe is placed on transverse planes over the anterior aspect of the distal tibia as a landmark and then swept down to examine the talus dome. Using these planes, the talus dome has a flat appearance and appears to be covered with a uniform layer of cartilage (Figure 3B). Because the talar dome is convex in a craniocaudal direction, the probe should be continuously tilted over it to maintain the US beam perpendicular to the osteochondral surfaces.

• A2. The foot is then placed flush on the table in an inversion position to examine the anterior recess of the subtalar joint with a lateral approach. The probe is placed horizontally over the sinus tarsi (Figure 3C). Any focal hypoechoic area partially filling this space should be regarded as a distended anterior recess of the subtalar joint.

• A3. The patient is invited to lie on the bed on their back with the toes pointing upwards to stretch the Achilles tendon. The probe is placed in a sagittal plane over the suprarcannlear portion of this tendon to examine the posterior recesses of the tibiotalar and subtalar joints (Figure 3D). These recesses are deep-seated and their examination may be better achieved by decreasing the transducer frequency, lowering the focal zones...
and increasing the gain in the deepest area of the field-of-view. As an alternative, the transducer may be placed alongside the medial aspect of the Achilles slightly converging towards the midline. In this latter position, the joint recesses are closer to the probe and can be examined with better image quality.

**The HEAD-US score**

The HEAD-US scoring method is reported in Table 2. It is based on an additive scale with assessment of one joint at any one time and includes indicators of disease activity (hypertrophic synovium) and structural osteochondral damage (articular cartilage and subchondral bone). The total score represents the sum of item scores for abnormalities detected. Its values range from 0 (minimum) to 8 (maximum). Hypertrophic synovium is graded in three steps (0: absent/minimal; 1: mild/moderate [score=1]; 2: severe [score =2]) based on the comprehensive evaluation of the joint recesses and the mean amount of synovial tissue contained in them. According to the technical guidelines described above, the relevant sweeps to score synovitis are: E1a, E1c, E2a, E2b, E3 for the elbow; K1, K2a, K2b for the knee; and A1a, A2, A3 for the ankle. In the ankle, the recesses of the

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**Table 2: HEAD-US scoring method.**

<table>
<thead>
<tr>
<th>Disease activity (synovitis)</th>
<th>Scale</th>
</tr>
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<tbody>
<tr>
<td>Hypertrophic synovium</td>
<td></td>
</tr>
<tr>
<td>0. Absent/Minimal</td>
<td>0</td>
</tr>
<tr>
<td>1. Mild/Moderate</td>
<td>1</td>
</tr>
<tr>
<td>2. Severe</td>
<td>2</td>
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<table>
<thead>
<tr>
<th>Disease damage (articular surfaces)</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Cartilage</td>
<td></td>
</tr>
<tr>
<td>0. Normal</td>
<td>0</td>
</tr>
<tr>
<td>1. Echotexture abnormalities, focal partial/full-thickness loss of the articular cartilage involving &lt;25% of the target surface*</td>
<td>1</td>
</tr>
<tr>
<td>2. Partial/full-thickness loss of the articular cartilage involving at least ≤50% of the target surface*</td>
<td>2</td>
</tr>
<tr>
<td>3. Partial/full-thickness loss of the articular cartilage involving &gt;50% of the target surface*</td>
<td>3</td>
</tr>
<tr>
<td>4. Complete cartilage destruction or absent visualization of the articular cartilage on the target bony surface*</td>
<td>4</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Bone</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>0. Normal</td>
<td>0</td>
</tr>
<tr>
<td>1. Mild irregularities of the subchondral bone with/without initial osteophytes around the joint</td>
<td>1</td>
</tr>
<tr>
<td>2. Deranged subchondral bone with/without erosions and presence of prominent osteophytes around the joint</td>
<td>2</td>
</tr>
</tbody>
</table>

**Note:** Elbow: anterior aspect of the distal humeral epiphysis; Knee: femoral trochlea; Ankle: anterior aspect of the talar dome.
tibiotalar and subtalar joints must be considered together as a single measurement. US criteria to identify synovial tissue and distinguish it from effusion are reported elsewhere (5). Based on the observation that the synovium appeared largely hypovascular at Doppler imaging in the series of 60 joints examined during the consensus meetings, we decided to exclude this parameter from the scale.

Evaluation of structural osteochondral damage is referred to specific target surfaces, namely: the anterior aspect of the distal humeral epiphysis in the elbow, the femoral trochlea in the knee, and the anterior aspect of the talar dome in the ankle. The relevant sweeps to demonstrate these surfaces are: E1b, E2a, E2b for the elbow; K3 for the knee; and A1a, A1b for the ankle. Concerning the articular cartilage investing these surfaces, the damage is graded in five steps (0: normal; 1: echotexture abnormalities and focal loss involving <25% of the target surface [score=1]; 2: partial/full thickness loss of the cartilage involving at least \( \leq 50\% \) of the target surface [score=2]; 3: partial/full thickness loss of the cartilage involving >50% of the target surface [score=3]; 4: complete cartilage destruction or absent visualisation of the articular cartilage on the target surface [score=4]). US criteria to identify the articular cartilage and the subchondral bone are reported in the literature (5). Scoring is assigned on the transverse plane of the E1b, K3 and A1b sweeps showing the most extended chondral abnormality. It measures the extension of the chondral loss in the selected plane regardless of whether the abnormality is a partial- or full-thickness defect (Figure 4). Grade-4 of chondral damage includes absent visualisation of the cartilage. This condition may occur in the ankle when a prominent anterior tibial osteophyte projects down to cover the talar dome completely. Similarly, the inability to flex the knee or to extend the elbow may impair visualisation of the osteochondral surfaces. Damage of subchondral bone is determined in the same reference plane used for scoring chondral abnormalities by means of a three-grade scale (0: normal; 1: mild irregularities of the subchondral bone with/without initial osteophytes around the joint [score=1]; 2: deranged subchondral bone with/without erosions and presence of prominent osteophytes around the joint [score=2]). For this last assessment, the osteophyte item is not limited to the selected target surface but is comprehensive of any finding encountered while performing the scanning protocols described above. Detection of large osteophytes around

Figure 4: Scoring osteochondral damage in the knee - spectrum of findings. A) Normal femoral trochlea. The concave osteochondral surface can be divided in two halves (medial and lateral facets) by the central groove. To estimate the amount of the damaged surface one should place the probe in the transverse plane over the area of maximum damage. As a reference, the distance between the deepest point of the groove and the medial or lateral end of the trochlea means 50% of the osteochondral surface, whereas half of the medial or lateral facet means 25%. The articular cartilage (1) appears as an band of uniform thickness below the quadriceps tendon (2) and the subchondral bone (3) as a smooth regular surface. B) Grade-1 cartilage, grade-0 bone score. A partial-thickness cartilage defect (arrowheads) involving <25% of one trochlear facet is seen. C) Grade-2 cartilage, grade-0 bone score. A combined partial- (narrow arrow) and full-thickness (large arrow) cartilage defect (arrowheads) involving more than half of a trocheal facet is found. The subchondral bone appears normal. D) Grade-3 cartilage, grade-1 bone score. A diffuse partial-thickness loss (ptl) of the cartilage (arrowrows) can be appreciated on both facets involving >50% of the osteochondral surface. The subchondral bone appears cobbled and irregular. On the right, an osteophyte (arrowheads) is seen. E) Grade-3 cartilage, grade-1 bone score. A full-thickness loss (ftl) of the articular cartilage is seen involving >50% of the osteochondral surface. Some residual cartilage (arrowheads) can be appreciated peripherally. The subchondral bone is irregular and focally depressed (arrow). F) Grade-4 cartilage, grade-1 bone score. No more articular cartilage is appreciated throughout the trochlea (curved arrows). The hypoechoic soft-tissue layer (void arrows) covering a slightly irregular subchondral bone refers to hypertrophic synovium.
the joint may, therefore, influence the score irrespective of the status of subchondral bone in the reference plane.

Observer agreements

The pre-test related to haemosiderin detectability resulted in full agreement between the readers that no differences existed between cases with haemosiderin-enriched and haemosiderin-free synovium. Similarly, there was no echotextural finding allowing the readers to differentiate proliferating synovium in haemophilic joints from other chronic disorders.

Tables 3 to 5 summarise the analysis of inter- and intra-observer tests. Good to excellent inter-observer agreement was found for the individual items (synovium, cartilage and bone) of the scale among the readers (κ = 0.61–0.93), with the lowest values found in the assessment of the osteochondral profile (Table 3). The overall inter-observer agreement was excellent (κ = 0.71–0.81). Intra-observer agreement proved to be moderate to excellent for the individual items of the scale (κ = 0.50–0.85), with the lowest values found in the assessment of the synovial profile (Table 4). The overall intra-observer agreement was good to excellent (κ = 0.69–0.78). Considering the three joints separately (Table 5), the global inter-observer and intra-observer agreement was excellent for the elbow and the knee (κ = 0.80–0.81) and good for the ankle (κ = 0.66–0.69). The Cronbach’s alpha values for all variables were excellent (α = 0.98).

Discussion

Synovial changes and destruction of the osteochondral surfaces are the most prominent histopathologic features in HA and represent two distinct domains of analysis included in the HEAD-US scor-
Martinoli et al. HEAD-US Scoring System

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Macrosopic inspection of joints structures during replacement procedures revealed that the worst affected areas are the load-bearing articular zones, whereas the peripheral cartilage is proportionally less affected (16). Regardless of its severity, however, the osteochondral damage is extensive in haemophilia involving areas that are usually spared in other degenerative conditions. It could be hypothesised that this diffuse pattern of involvement may be related to the direct harmful effect of iron on the joint surfaces with inhibitions of the mechanism underlying formation of the cartilage matrix (17). Following iron deposition, which may be ubiquitous in the joint cavity, damage may develop throughout the joint before, and possibly independent of, synovial mechanisms of cartilage destruction (14, 17). One of the most relevant drawbacks of joint US is the inability to access the central, load-bearing surfaces due to the inability of the beam to cross the bone boundaries (5).

Such a limitation invariably makes the assessment of the osteochondral surfaces incomplete and may preclude the evaluation of some relevant intra-articular structures. This limited access makes US unfeasible to reveal some focal osteochondral disorders (i.e. osteochondral fractures, osteochondritis dissecans) that are typically located in weight-bearing zones. Conversely, important information can be obtained in diffuse pathologies (e.g. chondrocalcinosis, inflammatory arthropathies), even if the evaluation of osteochondral surfaces remains partial. HA seems to belong to this latter category. In the HEAD-US score, a single osteochondral area has been selected in each joint as the key-surface for assessing and scoring the damage. These areas could be easily and reliably imaged after a short period of training. The good performance of the third reader, the less experienced one, in the inter-observer exercise basically confirms this observation. Appropriate positioning has been used to visualise these surfaces below the bone and maximally expose the load-bearing part of them to the US beam. Other smaller osteochondral areas, such as the posterior aspects of the humeral trochlea, femoral condyles and talar dome, could possibly have been screened, but they were excluded from the protocol to avoid more complexity than needed and to save examination time. In our preliminary series of patients we did not observe any abnormality in the excluded surfaces if the included surfaces appeared normal. Therefore, we can speculate that the diffuse osteochondral damage in haemophilic patients may warrant the policy of considering one surface representative of the overall status of the joint without significantly reducing the sensitivity of the method. Further experience on a large series of patients is needed to substantiate this statement.

Regardless of the modality, a reproducible scoring scheme is useful in evaluating disease progression and making therapeutic decisions and treatment evaluations. MRI scoring methods are widely used and approved as reference standards in haemophilia trials, although rarely applied in clinical practice for diagnosis and outcome because of their complexity and time commitment. In the large scale, the use of MRI is basically unfeasible at many institutions due to the high cost and the long waiting lists of patients to scan. US offers several advantages over MRI, such as lower cost, better accessibility, repeatability, a quicker examination, and the ability to screen multiple joints in a single study as well as to exam-
Ultrasound (US) is a useful modality for evaluation of joint status, being able to recognise joint effusion, synovial hypertrophy and abnormalities involving osteochondral surfaces.

US has proven to be able to detect joint abnormalities in haemophilic arthropathy (HA).

US has the potential to evaluate disease progression and monitor the result of treatment in follow-up studies.

Development of a new method for early HA detection with ultrasound (HEAD-US) with the aim to integrate this imaging technique in the routine practice of haemophilia centres as a complement of physical examination.

Definition of simplified ultrasound scanning procedures to image the elbow, knee and ankle that may be easy to learn from non-expert musculoskeletal sonologists.

Development of a scoring system based on an additive scale to define the joint status from disease activity and disease damage parameters.

What is known about this topic?

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