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The NEW ENGLAND JOURNAL of MEDICINE

ESTABLISHED IN 1812 AUGUST 9, 2007 VOL. 357 NO. 6

Prophylaxis versus Episodic Treatment to Prevent Joint Disease in Boys with Severe Hemophilia

- Sixty-five boys younger than 30 months of age were randomly assigned to prophylaxis (32 boys) or enhanced episodic therapy (33 boys).
- When the boys reached 6 years of age, 93% of those in the prophylaxis group and 55% of those in the episodic-therapy group were considered to have normal index-joint structure on MRI (P=0.006).

CLINICAL ASSESSMENT OF JOINTS

Clinical examination of joints, with assessment of swelling, strength, range of motion, pain, and gait, was performed semiannually, as previously described, and videotaped for central review at study entry, midpoint, and completion.[20,21]

- Surprisingly, the number of clinically evident hemarthroses correlated weakly with the outcome as determined by MRI.

fective. We propose that chronic microhemorrhage into the joints or subchondral bone in young boys with hemophilia causes deterioration of joints without clinical evidence of hemarthroses and that prophylaxis prevents this subclinical process.

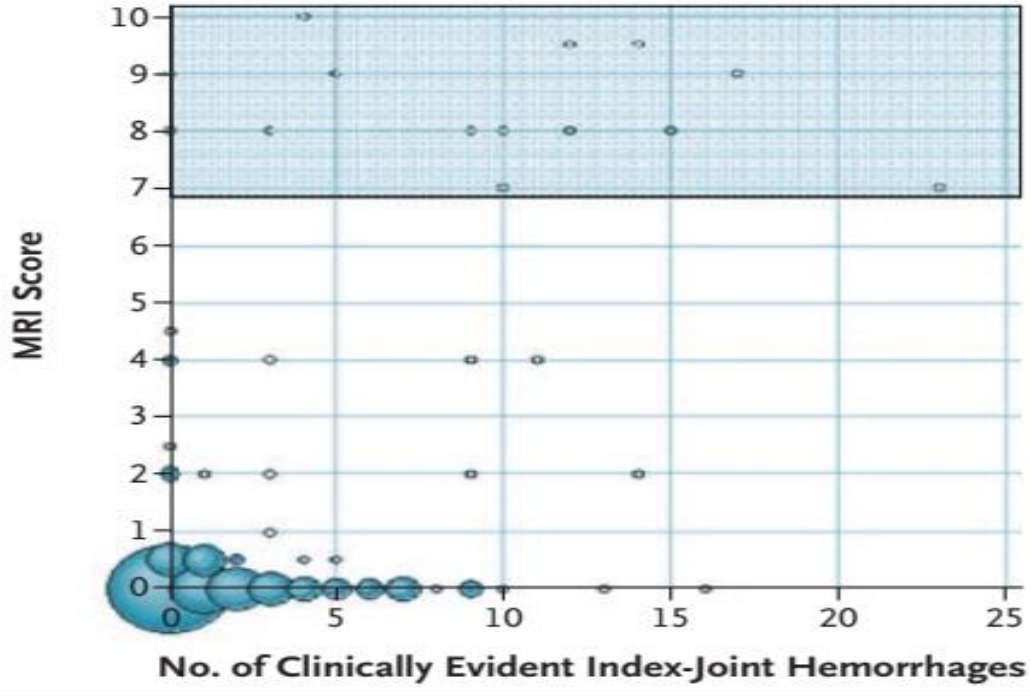

Figure 2. MRI Score for Index Joint According to the Number of Hemorrhages in That Joint for Both Treatment Groups.

The MRI score is the average of the two concordant readings; a score of 0 indicates a normal joint and a score of 7 to 10 indicates damage to joint bone or cartilage. The size of each circle corresponds to the number of cases. The largest number of index joints had no hemorrhage and an MRI score of 0. The shaded area of the graph shows the 14 joints with bone or cartilage damage on MRI. The larger bubbles represent more than one joint. Some joints with no hemorrhages had high MRI scores, whereas some joints with more than 10 hemorrhages did not show bone or cartilage damage on MRI.

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- It is possible that the joint score we used was insufficiently sensitive for the detection of early arthropathy, even though our physical-examination scoring system is more sensitive for the detection of mild abnormalities of gait, joint swelling, muscle strength, and atrophy than is that of the World Federation of Hemophilia.
- **Thus, the absence of overt hemarthroses and abnormalities of joints on physical examination *can lead to the erroneous assumption*.**

MSK Sonography in Haemophilia

1 | Introduction

Hemarthrosis, the most common haemorrhage in haemophilia, primarily affects the knees, elbows and ankles. Recurrent joint bleeds trigger synovial changes, including inflammation, proliferation, and neovascularization, making the synovial membrane prone to rebleeding. The bleeding-synovitis-bleeding cycle leads to chronic synovitis, cartilage and bone destruction, ultimately leading to irreversible arthropathy [1]. Imaging plays a vital role in detecting subclinical and symptomatic joint abnormalities to guide treatment decisions and prevent irreversible arthropathic changes [2, 3].

Haemophilia WFH

ORIGINAL ARTICLE

Evaluation of Point-of-Care Ultrasound in Paediatric Haemophilic Arthropathy: A Prospective Comparative Study With Comprehensive Ultrasound and MRI

Caroline Rutten[1,2] | Magdy Soliman[3,4] | Jyoti Panwar[3,5] | Arun Mohanta[1,2] | Alex Zhou[1,2] | Pamela Hilliard[6] | Tammy Rayner[1,2] | Ruth Weiss[1,2] | Carina Man[3] | Rahim Moineddin[7] | Manuel Carcao[2,8] | Vanessa Bouskill[8] | Jennifer Stimec[1,2] | Kevon Dindial[8,9] | Deidre Tetzlaff[6] | Victor S. Blanchette (in memoriam)[2,8,†] | Andrea S. Doria[1,2,3]

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Prophylaxis with factor VIII (FVIII) replacement therapy, or other non-factor agents, is currently the standard of care for the prevention of bleeding and preservation of long-term joint health in haemophilia A [3, 6, 7]. However, prophylactic treatment may not completely prevent joint bleeds; occasional clinical and subclinical bleeds may occur, resulting in progressive joint disease [3, 6].

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CME: Rheumatology

Applications of musculoskeletal ultrasound in inflammatory arthritis ☆

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Introduction

Musculoskeletal ultrasound is increasingly used in rheumatology as a diagnostic and disease-monitoring tool for patients with inflammatory arthropathies. The underpinning principle that drives this development is the ability of ultrasound to detect subtle joint and tendon inflammation that is not clinically apparent, thereby detecting inflammation earlier in the disease process. The main advantages of ultrasound include real-time visualisation, good patient tolerability, the ability to scan multiple joints in a short time, cost-effectiveness and the absence of ionising radiation.[1]

Haemophilia WFH

ORIGINAL ARTICLE OPEN ACCESS

Impact of Systematic Joint Examination (Ultrasound, Functional and Physical) on Treatment Management Decisions in Patients With Haemophilia A in France: Final Data From the Prospective, Observational A-MOVE Study

Nicolas Drillaud[1] | Virginie Barbay[2] | Jean Baptiste Valentin[3] | Romain Jailler[4] | Aurélien Lebreton[5] | Brigitte Pan-Petesch[6] | Sabine Marie Castet[7] | Birgit Frotscher[8] | Laurent Frenzel[9] | Sandrine Jousse-Joulin[10] | Hervé Chambost[11] | Mikaela Alenäs[12] | Markus Fusser[12] | Corinne Gandossi[13] | Meriem Zidi[13] | Oussama Mahdout[13] | Yohann Repessé[14] | on behalf of the A-MOVE study group

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ORIGINAL ARTICLE

Power Doppler sonography in the diagnosis of hemophilic synovitis – a promising tool

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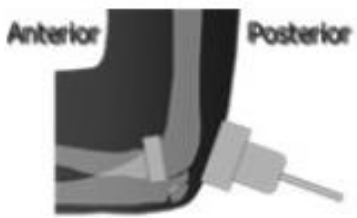

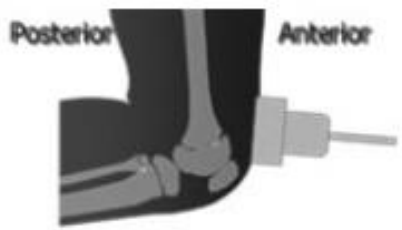

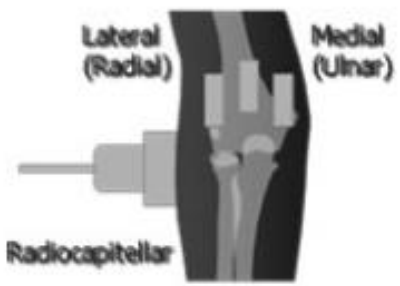

Elbow

- Posterior distal humeral and radiocapitellar regions were evaluated
- Lateral, middle, and medial aspects of posterior distal humeral region

Knee

- Anterior suprapatellar region was evaluated
- Lateral, middle, and medial aspects

Ankle

- Anterior tibiotalar region was evaluated
- Lateral, middle, and medial aspects

- Thirty-one subjects with hemophilia A/B (factor VIII/IX activity of <5 u dL)1) and other inherited bleeding disorders with a severe bleeding between the ages 6–60 years, with >2 lifetime hemorrhages into knees, ankles, and elbow joints.
- Subjects sustaining a median of 17 joint bleeds (range 4 to >20)

that locations for analyses were as similar as possible for comparison. Off-line analysis of ROIs was performed using QLAB software (Philips Medical), which calculates mean color pixel intensity (averaged over the ROI) expressing the data as decibels (dB) per mm^2 (Fig. 1B). Peak pixel intensities were averaged when cine loops were obtained. Prior to

Correlation of clinical bleeding and synovial pathology by imaging

- There was a significant correlation between Synovial thickness [pre-DCE-MRI ($P < 0.0001$), USG ($P = 0.001$)]
- and vascularity [PDS flow ($P = 0.013$)]
- in bleeders as compared with non-bleeders (Table 2).

Table 2 Synovial thickness and vascularity on magnetic resonance imaging (MRI) and ultrasonography (USG), power Doppler (PDS) flow (minimum, maximum, median) for bleeders and non-bleeders

	Non-bleeders			Bleeders		
	Synovial thickness pre DCE-MRI	Synovial thickness USG	PDS flow	Synovial thickness pre DCE-MRI	Synovial thickness USG	PDS flow
Median	3.65	5	3.075	9.15	13.45	8.53
Minimum	0	2	1.42	5.1	8	1.14
Maximum	13.3	18	31.75	20	18.3	81.89
P-value (2-tailed)				0.000	0.001	0.013

DCE-MRI, dynamic contrast enhanced MRI; USG, ultrasonography; PDS, power Doppler sonography. *P*-value indicates the medians for these variables which are being compared for bleeders vs. non-bleeders.

- There was a strong correlation for synovial thickness ($r = 0.77$, $P < 0.0001$) and vascularity ($r = 0.84$, $P < 0.0001$) in knee joints;
- Strong correlation in ankles for synovial thickness ($r = 0.68$, $P = 0.01$) but a (No) weak correlation for vascularity parameters($r = 0.25$, $P = 0.39$).
- For elbows (just 6 joints) there was no correlation between synovial thickness or vascularity using the two techniques.

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

Carlo Martinoli[1]; Ornella Della Casa Alberighi[3]; Giovanni Di Minno[5]; Ermelinda Graziano[6]; Angelo Claudio Molinari[4]; Gianluigi Pasta[7]; Giuseppe Russo[1]; Elena Santagostino[8]; Annarita Tagliaferri[9]; Alberto Tagliafico[2]; Massimo Morfini[10]

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- 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.
- 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.

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

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- 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.**

Haemophilia Early Arthropathy Detection with Ultrasound (HEAD-US)

a simplified scanning procedure and scoring method
Martolini et al., 2013

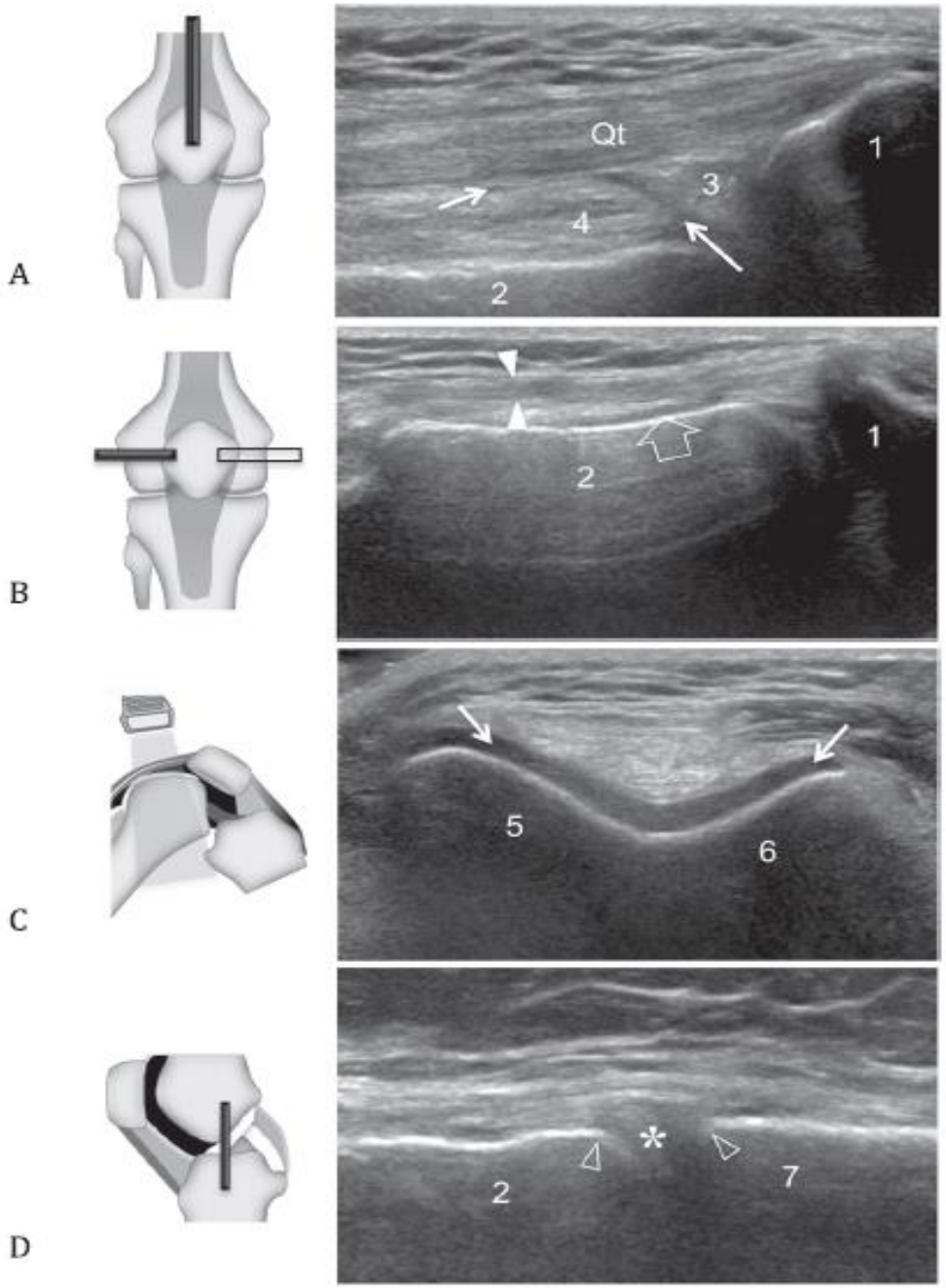

Figure 2: Knee protocol. A) Anterior mid-sagittal US image demonstrates the patella (1) and the distal femur (2). The suprapatellar recess (arrows) appears as a hypoechoic linear band located below the quadriceps tendon (Qt), between the suprapatellar (3) and the prefemoral (4) fat pads. B) Anterolateral transverse US image shows the patellofemoral joint and the lateral parapatellar recess (void arrow) located between the lateral patellar retinaculum (arrowheads) and the lateral femoral condyle. C) Transverse US image of the femoral trochlea at maximum degree of knee flexion. The medial (5) and lateral (6) facets of the trochlea are invested by a thick band of cartilage (arrows). D) Coronal US image over the medial femorotibial space shows the medial meniscus (asterisk). The boundaries (open arrowheads) of the femur (2) and tibia (7) are regular and flat up to the joint space.

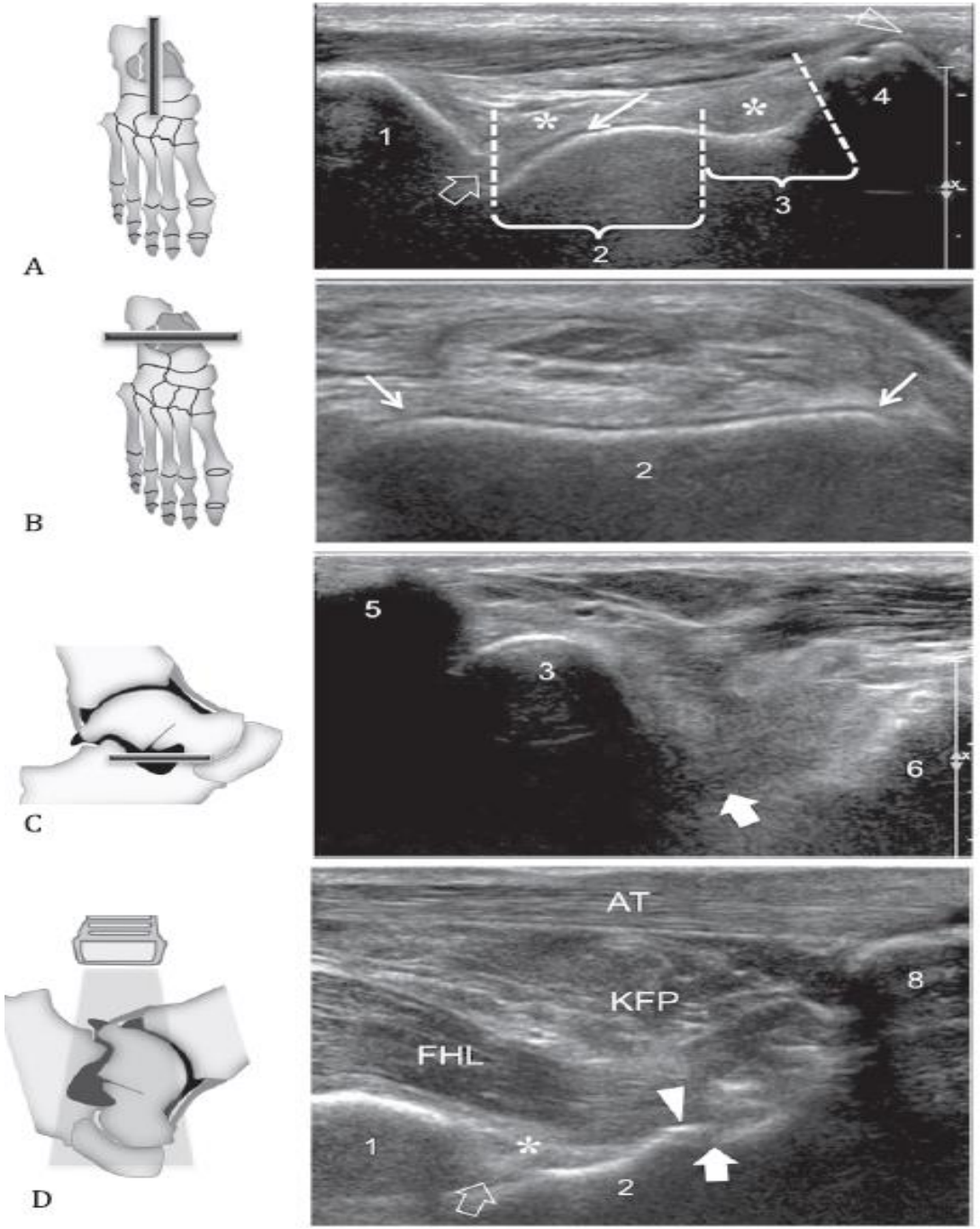

Figure 3: Ankle protocol. A) Anterior mid-sagittal US image over the tibiotalar joint (void arrow) demonstrates the distal tibial epiphysis and the talus, the latter consisting of three parts: the convex dome (2) covered with cartilage (narrow arrow), the concave neck (3) and the prominent head (4). More distally, the dorsal aspect of the talonavicular joint is seen (arrowheads). The anterior fat pad (asterisks) extends over the talar dome and neck. B) Anterior transverse US image over the talar dome reveals a flat bony surface (arrows) covered with a thin layer of cartilage. C) Lateral transverse US image over the sinus tarsi. The lateral malleolus (5), the talar neck posteriorly and the calcaneus (6) anteriorly are the main landmarks to identify the anterior recess of the subtalar joints. The sinus tarsi is a fat-filled space and looks hyperechoic normally. D) Posterior mid-sagittal US image over the Achilles tendon (AT). Below this tendon, the preachilles (Kager) fat pad (KFP) and the flexor hallucis longus muscle (FHL) are found. The posterior recess of the tibiotalar joint (void arrow) lies between the posterior malleolus (1) and the posterior aspect of the talar dome (2), underneath the posterior fat pad (asterisk). The posterior recess of the subtalar joint lies between the talus and calcaneus (8). A prominent posterior process of the talus (white arrowhead) may mimic an osteophyte.

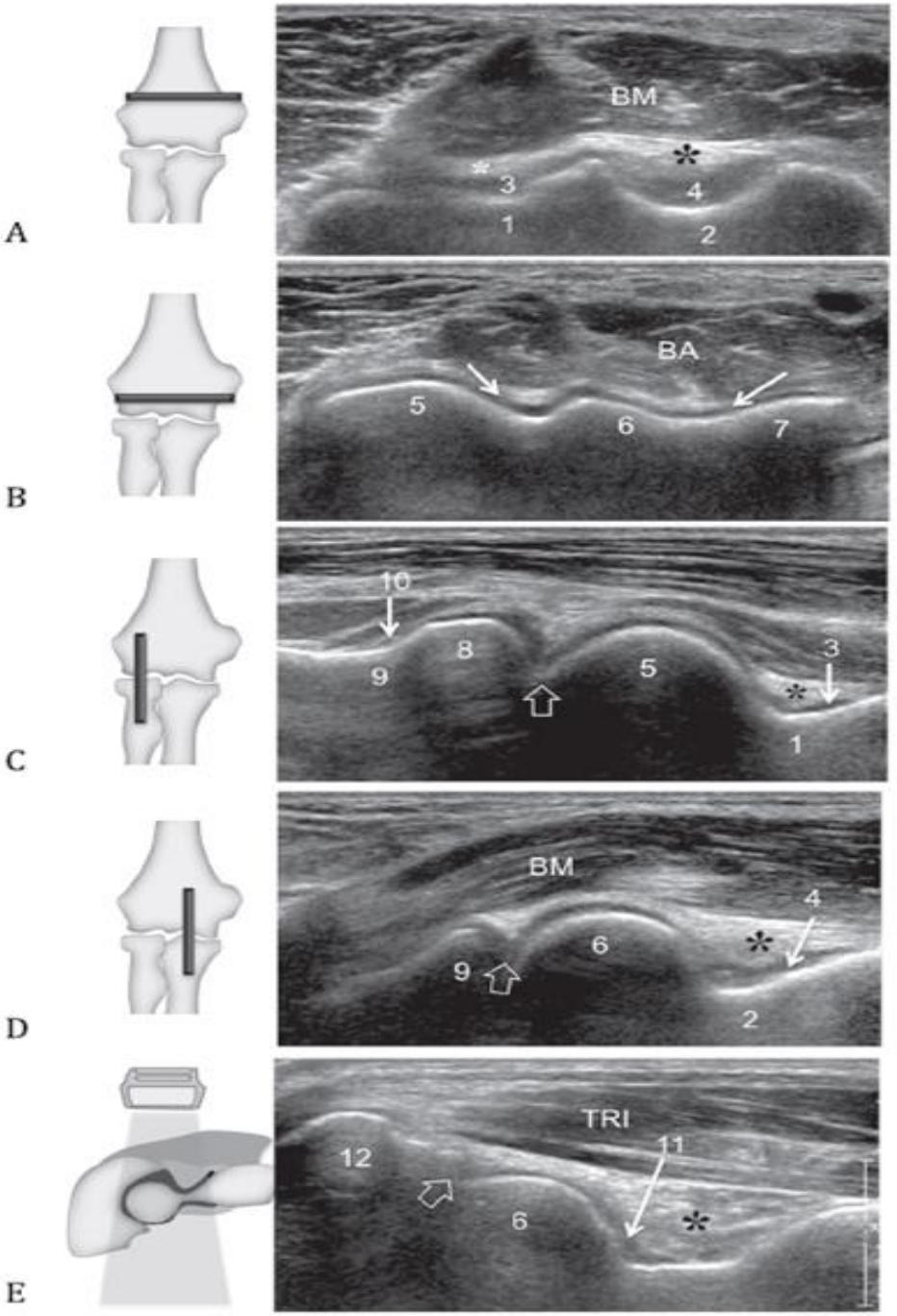

Figure 1: Elbow protocol. A) Anterior transverse US image over the distal humeral metaphysis demonstrates the radial (1) and coronoid (2) fossa with the respective radial (3) and coronoid (4) recesses. These structures lie underneath the anterior fat pad (asterisks) and the brachialis muscle (BM). B) Anterior transverse US image over the distal humeral epiphysis shows a wavy osteochondral surface consisting of the convex capitellum (5) and the concave trochlea that exhibits two facets, lateral (6) and medial (7). The articular cartilage appears as a uniform hypoechoic band (arrows) overlying the subcondral bone. C) Anterior sagittal US image over the radio-capitellar joint reveals the radial fossa that houses the radial recess (4). The joint line (void arrow) separates the humeral capitellum (5) from the radial head (8). Distal to it, the annular recess (10) surrounds the radial neck (9). D) Anterior sagittal US image over the trochlea-ulna joint demonstrates the coronoid fossa and recess in a cranial position. The joint line (open arrow) separates the trochlea from the coronoid process (9). E) Posterior sagittal US image over the olecranon fossa demonstrates the posterior fat pad (asterisk) and the olecranon recess (11) deep to the triceps muscle (TRI). The joint line (void arrow) lies between the posterior aspect of the trochlea and the olecranon process (12).

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Advanced Hemophilic Arthropathy: Sensitivity of Soft Tissue Discrimination with Musculoskeletal Ultrasound

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- Point-of-care (POC) musculoskeletal ultrasound (MSKUS) is increasingly used by hemophilia providers to guide management, however, pathologic tissue differentiation with US is uncertain.
- We sought to determine the extent to which POC MSKUS can identify and discriminate pathologic soft tissue changes in hemophilic arthropathy.

Materials and Methods—36 adult patients with hemophilia A/B were prospectively enrolled. POC MSKUS was performed on arthropathic joints (16 knees, 10 ankles, and 10 elbows) using standard views by a MSKUS-trained and certified hematologist, who recorded abnormal intra-articular soft tissue accumulation. Within three days, magnetic resonance imaging (MRI) was performed using conventional and multi-echo ultrashort echo time (UTE) sequences. Soft tissue identification (synovial proliferation with or without hemosiderin, fat, and/or blood products) was performed by a musculoskeletal radiologist. Findings obtained with both imaging modalities were compared and correlated in a blinded fashion.

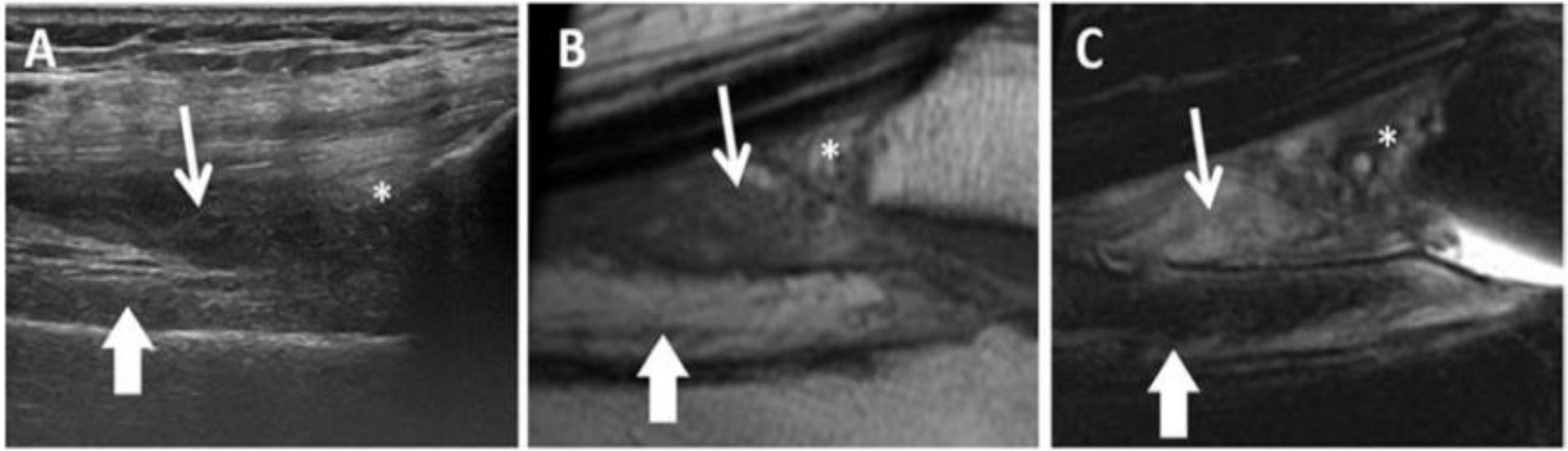


Figure 5. Soft tissue accumulation (blood clot)

(A) Long-axis US image shows hypoechoic tissue that was partially compressible on real time imaging in the suprapatellar recess (thin arrow). (B) Sagittal T1-weighted and (C) T2-weighted fat-suppressed MR images shows heterogeneously hyperintense signal in the suprapatellar recess on both sequences, consistent with coagulated subacute blood products. Prefemoral (thick arrows) and suprapatellar (asterisks) fat pads are marked. Note that the prefemoral fat pad is more hyperintense proximally compared with distally. US, ultrasound;

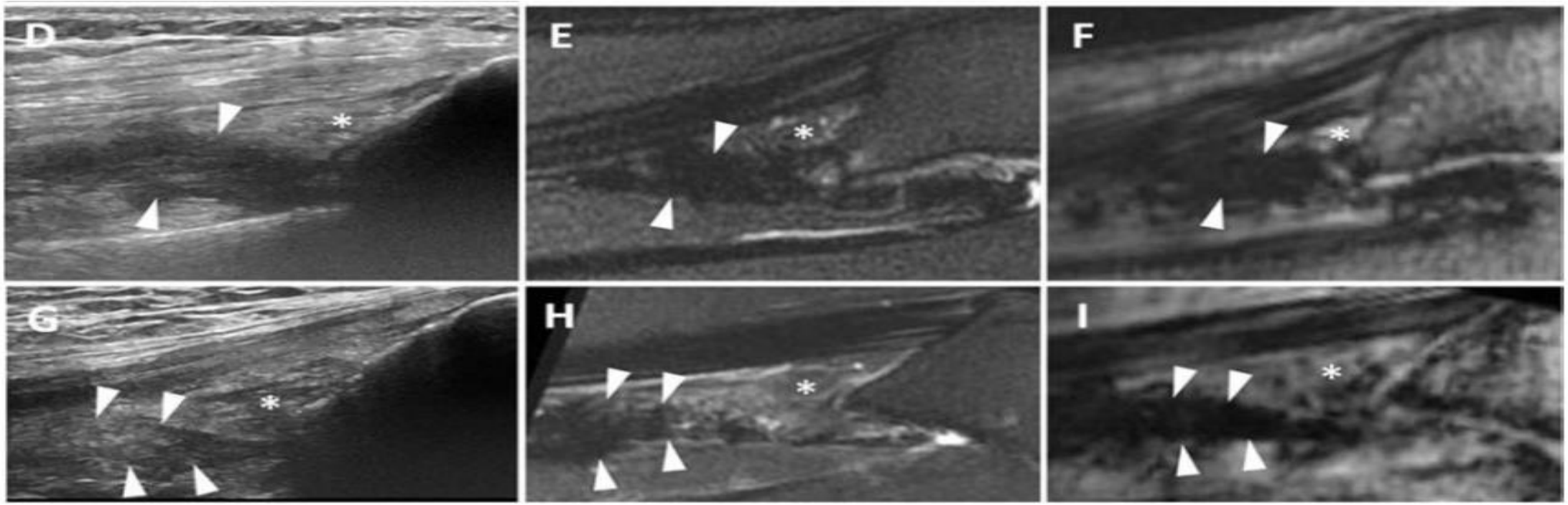
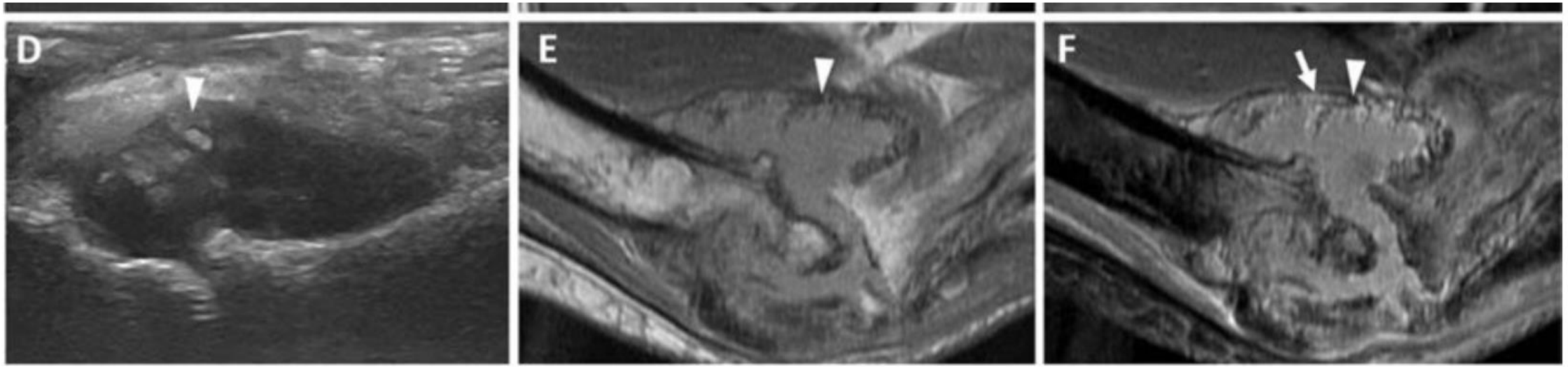


Figure 1. Various echotextures of hemosiderin-laden synovial proliferation

Figure 1. Various echotextures of hemosiderin-laden synovial proliferation

The annular recess in the elbow of a 39-year old (A-C) and suprapatellar recesses in the knees of a 23-year-old (D-F) and a 38-year-old (G-I) patient. (A, D and G) Longitudinal US images show thick, non-compressible tissue with increasing coarseness and heterogeneous echotexture (arrowheads) that differs between patients. (B,C,E,F,H,I) Sagittal T2-weighted and UTE gradient MR images (TE 6.0 ms) show hypointense tissue with blooming, consistent with hemosiderin-laden synovium. * = suprapatellar fat pad. US, ultrasound; MR, magnetic resonance; UTE, ultrashort time-to-echo.



- (D) Longitudinal US image shows mixed, predominantly hyperechoic, non-compressible tissue directly at the level of the joint line (arrowhead). (E) Sagittal T1-weighted MR image shows hypointense hemosiderin-laden synovium (arrowhead). (F) Sagittal T1-weighted fatsuppressed post-intravenous contrast MR image shows areas of avid enhancement (thin arrow) adjacent to hypointense areas (arrowhead), consistent with synoviu containing various amounts of hemosiderin.

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Results—There was perfect agreement between the modalities on the presence of abnormal soft tissue (34/36 cases). However, MSKUS was unable to discriminate between coagulated blood, synovium, intra- or extra-synovial fat tissue, or hemosiderin deposits due to wide variation in echogenicity.

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27
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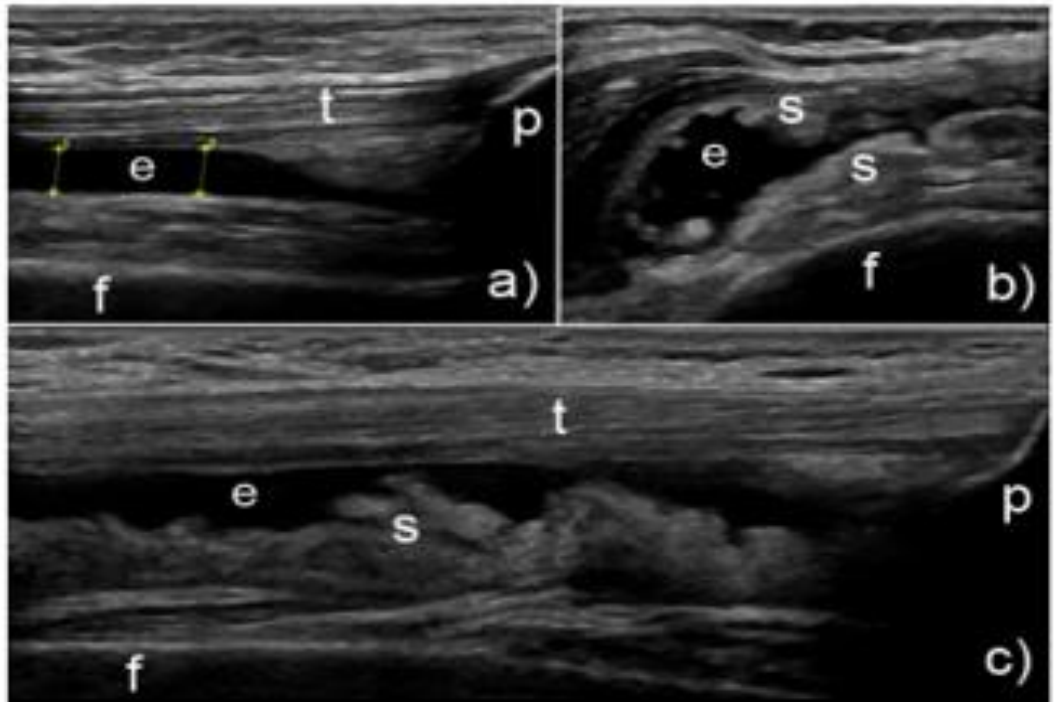
2016

Ultrasound of the knee in Rheumatology: Pitfalls, what is new?
Use of US in Rheumatology

Giasna Giokits-Kakavouli[1], Dimitrios Karokis[2], Ioannis Raftakis[3], Caterina Siagkri[4]

Katerini, Greece, Patras, Greece, Asklepeion Voulas General Hospital, Athens, Greece, Thessaloniki, Greece



- **Ultrasound is an operator-dependent imaging modality**

- **Training is crucial for development of image acquisition and interpretation skills**.

- Ultrasound performed by adequately trained specialists can be used to distinguish:
- Simple effusion (serous synovial fluid) from
- Complex effusion (joint bleeding) or
- Synovial hypertrophy.
- In the first hours from the start of a bleeding event, complex joint effusion appears as joint capsule distension due to isoechoic or hypoechoic compressible joint effusion with speckles (Figure) floating inside the joint during dynamic maneuvers with a characteristic pattern resembling a shaken snow globe (“snow globe sign”).

PRACTICE GUIDELINES FOR IMPLEMENTATION AND USE OF MUSCULOSKELETAL ULTRASOUND IN THE MANAGEMENT OF INDIVIDUALS WITH HEMOPHILIA AND OTHER BLEEDING DISORDERS

- Results of an International Prophylaxis Study Group Global Survey"
- Physiotherapist were identified as the clinical providers that perform POC-MSKUS the most in the surveyed HTC's **constituting over 50%** of the list of imaging providers,
- closely followed by **Hematologists at 41.5%.**
- Nurses and advanced care providers performed the least number of exams (5.7%)

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ORIGINAL ARTICLE

Ultrasound in addition to clinical assessment of acute musculoskeletal complaints in bleeding disorders: impact on patient management

Flora Hendrica Pieternella van Leeuwen[1] | Wouter Foppen[1] | Pim A. de Jong[1] | Wobke E. M. van Dijk[2] | Johan Blokzijl[2] | Kathelijn Fischer[2] | Merel A. Timmer[2]

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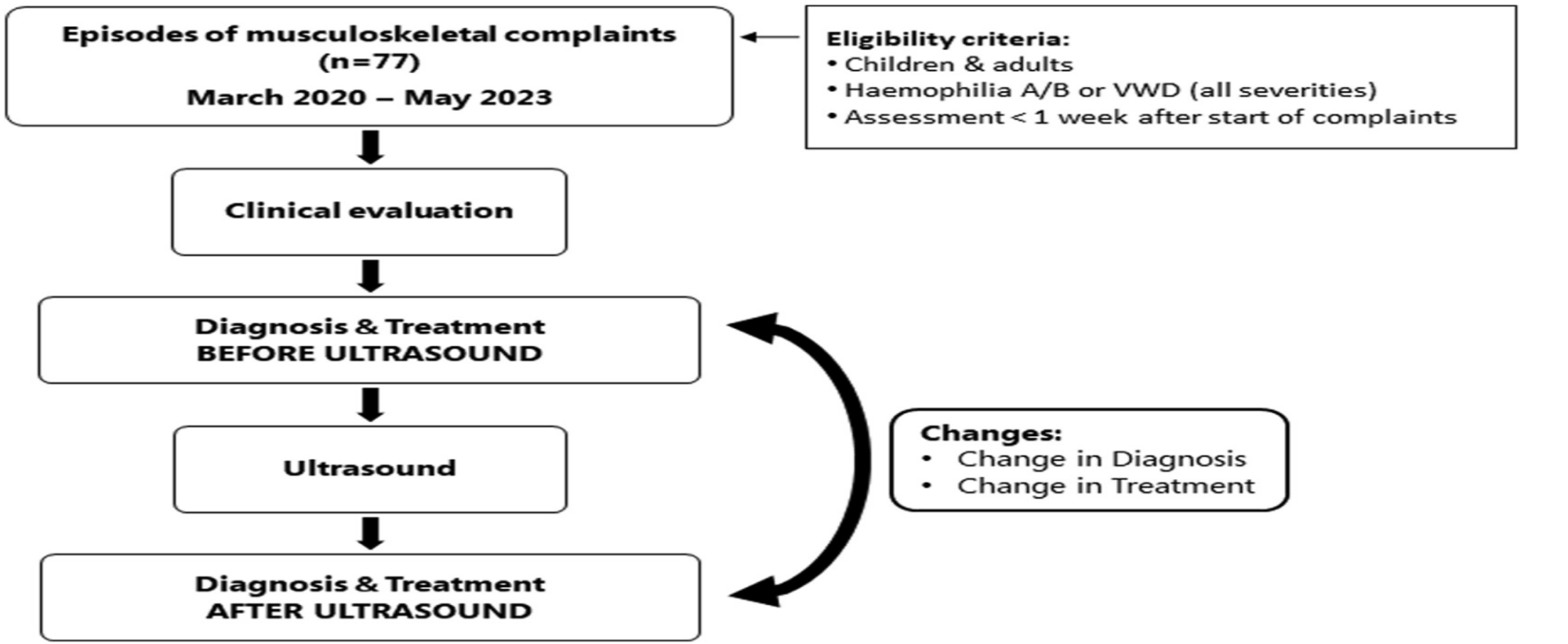

2.2 | Clinical assessment

The clinical assessment consisted of history and physical examination and was performed by physicians and physiotherapists (M.A.T and J.B.) of our hemophilia treatment center (HTC). Established symptoms to differentiate between joint bleeds and arthropathy flare-ups [27] were collected: the cause of the musculoskeletal complaints (trauma/overexertion/unknown), pain (yes/no), pain localization (local/diffuse), type of pain (stabbing/pressing), pain at rest (yes/no), sleep disruption due to pain (yes/no), response of pain to treatment with factor replacement therapy (yes/no), pain with active range of motion (AROM; yes/no), and the course of the pain (constant/increasing/increasing with motion/start pain decreasing with motion). For lower extremity episodes, participants were additionally asked about pain on weight-bearing (yes/no).

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- The median age at the time of assessment was 24 years **(IQR, 13-42 years**).
- Most patients had hemophilia (94%).
- A total of 25 of 26 people with severe hemophilia, 3 of 12 people with moderate hemophilia , and 3 of 25 people with mild hemophilia received prophylactic treatment..
- Approximately half of the musculoskeletal episodes (55%) were traumatic.
- Most episodes involved joints (79%),
- Followed by muscles (14%).
- **The majority of joint episodes (42/61, 69%) occurred in joints without signs of hemophilic arthropathy (HJHS, 0; HEAD-US cartilage and bone sum score, 0; Pettersson score , 0).**

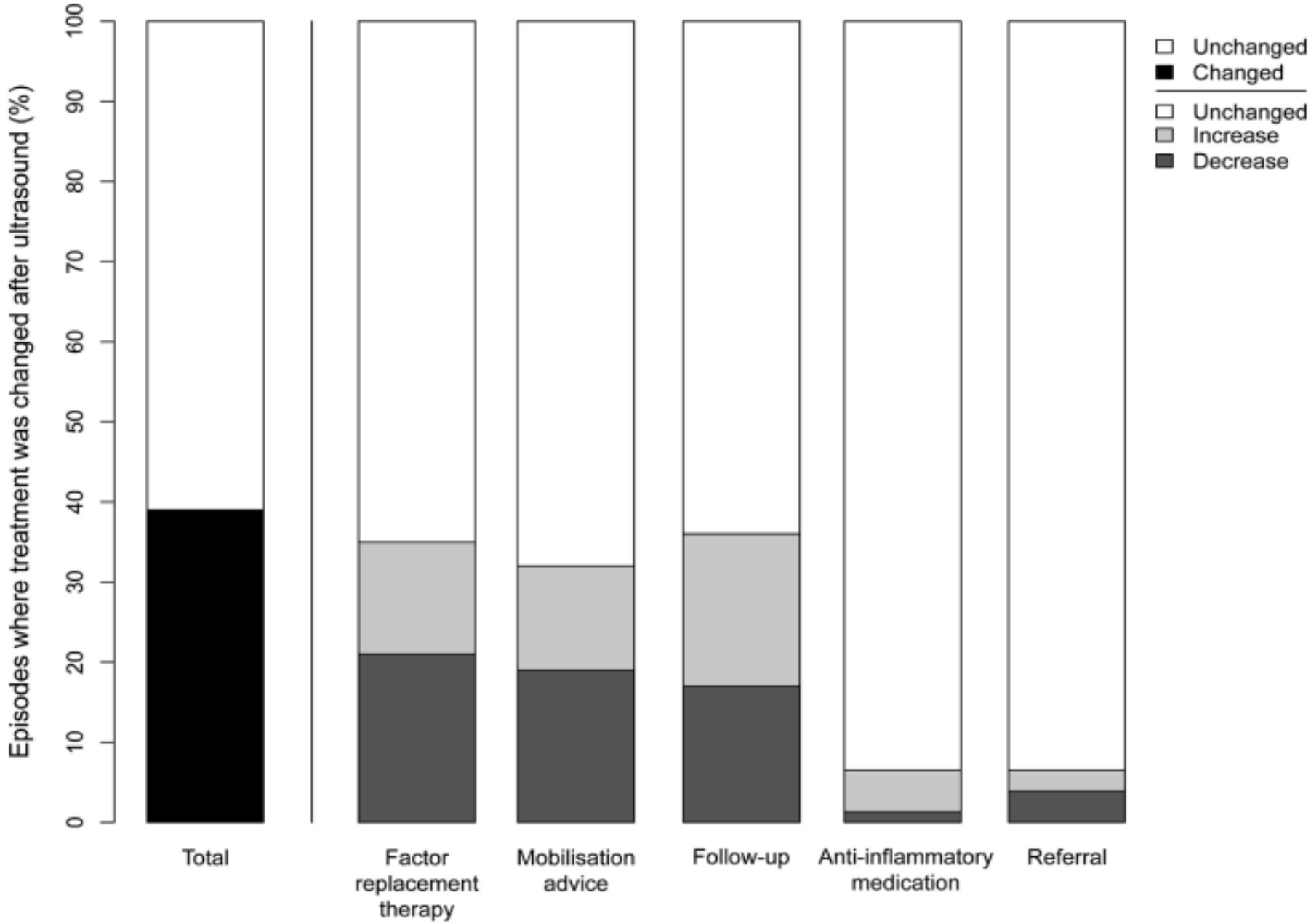

FIGURE 4 Stacked bar chart visualizing changes in treatment plans after ultrasound assessment.

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- .**We evaluated 77 episodes in 67 patients**
- Before ultrasound, 37 joint bleeds, 13 muscle bleeds, and 27 other diagnoses were diagnosed.
- After ultrasound, 33 joint bleeds, 11 muscle bleeds, and 33 other diagnoses were confirmed.
- **Nine 9 joint bleeds and 2 muscle bleeds were missed by clinical assessment.**
- **The ultrasound findings changed treatment strategy in 30 of 77 episodes (39%; 95% CI, 28%-51%).**
- **dose and/or duration of the factor replacement therapy was adjusted in 27 of 77 episodes (35%; 95% CI, 25%-47%).**
- **The net effect was a reduction (total -96,879 IU) of the total international units of factor prescribed in the study population.**

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ORIGINAL ARTICLE

Integrating musculoskeletal ultrasound as a shared decision-making tool in hemophilia care: observations from a 3-year study

Azusa Nagao[1] | Satoko Orita[2] | Katsuyuki Fukutake[1,3] | Hideyuki Takedani[4,5]

[1]Department of Blood Coagulation, Ogikubo Hospital, Tokyo, Japan

Abstract

- This study assessed the impact of incorporating musculoskeletal ultrasound (MSKUS) into shared decision-making processes on prophylaxis regimens for patients with hemophilia over a 3-year period between March 2018 and March 2022.

-**174 Participants** underwent 6 monthly MSKUS evaluations, generating **3582 images from 682 joints;** the findings guided adjustments of prophylaxis.

- Individuals **aged ≥6 years**, who had congenital moderate-to-severe hemophilia A or B (factor [F]VIII ≤ 2% or FIX ≤ 2%) and had undergone prophylaxis (regardless of factor product or regimen), were included.

- At the time of study enrollment, **their home infusion records for at least 6 months before enrollment and for >50 exposure days (EDs) to coagulation factors for children aged <12 years, or >150 EDs for adolescents aged ≥12 years and adults**, were required for enrollment.

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Abstract

- Two readers reviewed the imaging studies using standardized scoring systems. The inter-reader agreement was assessed with weighted kappa statistics.
- Diagnostic test performance of POCUS and comprehensive US was benchmarked against MRI findings using sensitivity, specificity, predictive values and accuracy.

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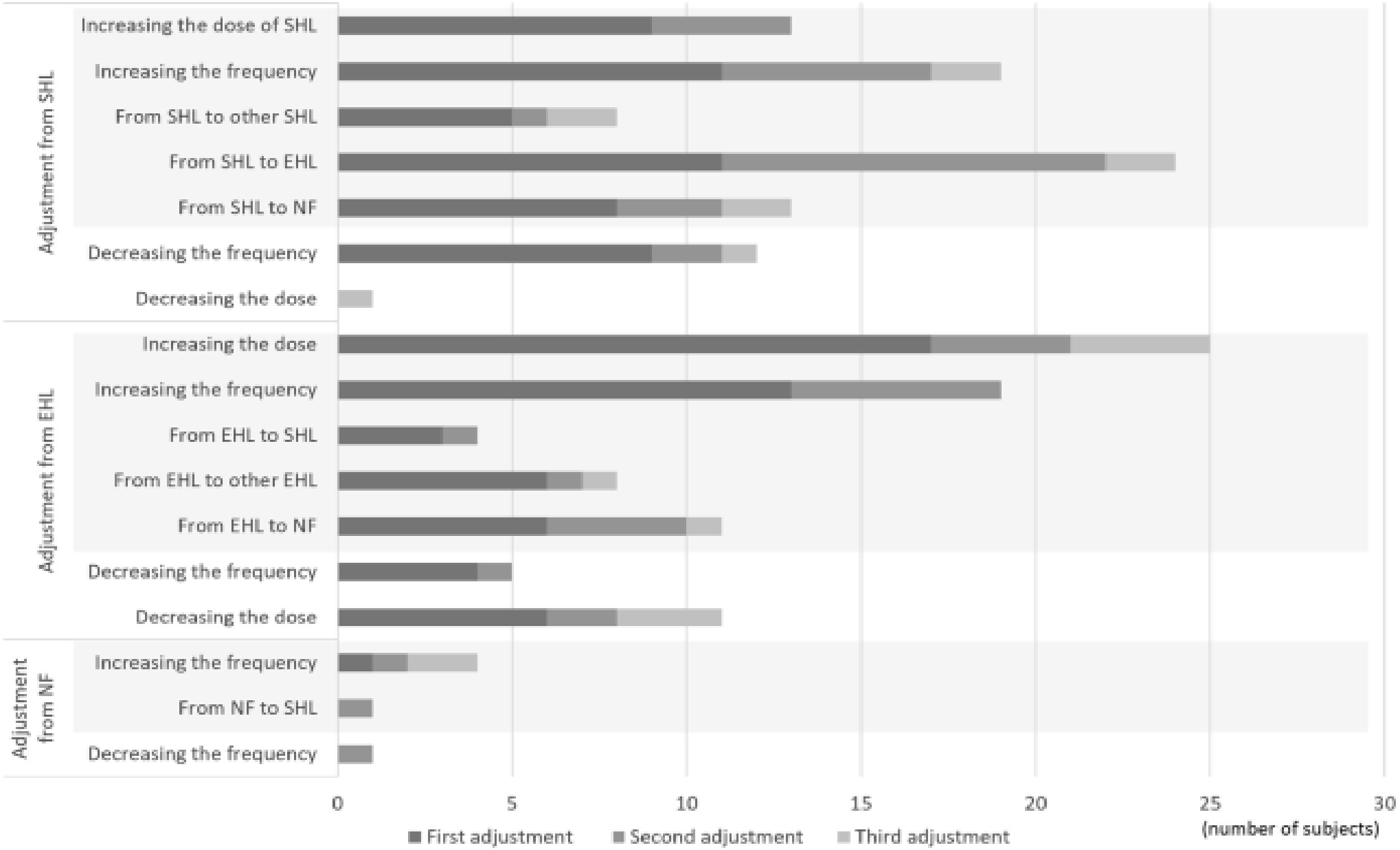
Abstract

MSKUS images were assessed blindly by another independent orthopedic surgeon with 35 years of experience in medical practice for hemophilia. This surgeon also received specific training from the HEAD-US group. The joint condition was classified by the severity of synovitis, cartilage damage, and bone degeneration, according to the HEAD-US scoring protocol [9], the primary scoring method for hemophilia in Japan in 2018. Although the protocol's suitability for pediatric assessments was not yet well established, its use was deemed provisional due to the absence of other pediatric-focused scoring methods.


Adjustment from SHL
Increasing the dose of SHL
Increasing the frequency
From SHL to other SHL
From SHL to EHL
From SHL to NF
Decreasing the frequency
Decreasing the dose
Adjustment from EHL
Increasing the dose
Increasing the frequency
From EHL to SHL
From EHL to other EHL
From EHL to NF
Decreasing the frequency
Decreasing the dose
Adjustment from NF
Increasing the frequency
From NF to SHL
Decreasing the frequency
0
5
10
15
20
25
30
First adjustment
Second adjustment
Third adjustment
(number of subjects)

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[1]Department of Blood Coagulation, Ogikubo Hospital, Tokyo, Japan

Abstract

- The HEAD-US scores for elbows, knees, and ankles significantly improved (all P < .0001).
- The spontaneous annual joint bleeding rate and Hemophilia Joint Health Scores also significantly improved (P = .001 and P = .004, respectively).
- **Synovitis detection decreased from 12.9% to 1.6%, with the majority of identified synovitis being subclinical (11.7%) and not associated with bleeding events in the 6 months preceding detection.**
- Conclusion: Integrating MSKUS into hemophilia care **as a shared decision-making tool** significantly facilitates the early detection of joint damage and supports personalized prophylaxis adjustments, markedly improvinn g patient outcomes.

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Abstract

2.3 | Definition of subclinical synovitis

There is no specific definition for subclinical synovitis. Therefore, we used definitions based on the sAjBR and HJHS. Based on the sAjBR, cases of synovitis detected by ultrasound but without bleeding events requiring treatment (including no "self-treatment for bleeding") within the last 6 months were considered subclinical synovitis. Based on the HJHS, cases of synovitis detected by ultrasound but with the HJHS examination showing no joint swelling or pain were considered subclinical synovitis.

Haemophilia

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Haemophilia WFH

ORIGINAL ARTICLE OPEN ACCESS

Impact of Systematic Joint Examination (Ultrasound, Functional and Physical) on Treatment Management Decisions in Patients With Haemophilia A in France: Final Data From the Prospective, Observational A-MOVE Study

Nicolas Drillaud[1] | Virginie Barbay[2] | Jean Baptiste Valentin[3] | Romain Jailler[4] | Aurélien Lebreton[5] | Brigitte Pan-Petesch[6] | Sabine Marie Castet[7] | Birgit Frotscher[8] | Laurent Frenzel[9] | Sandrine Jousse-Joulin[10] | Hervé Chambost[11] | Mikaela Alenäs[12] | Markus Fusser[12] | Corinne Gandossi[13] | Meriem Zidi[13] | Oussama Mahdout[13] | Yohann Repessé[14] | on behalf of the A-MOVE study group

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- **Aim:** To evaluate whether systematic joint examination with HJHS and/or **HEAD-US had an impact on treatment management decisions in France**, using final data from the A-MOVE study.
- **Methods:** A-MOVE (NCT04133883) was **a 12-month prospective**, multicentre study, which enrolled persons with haemophilia A (all severities, **aged 6–40 years**) treated prophylactically or on demand with standard/extended half-life FVIII replacement.
- At baseline, 6 and 12 months, HJHS/HEAD-US and changes in patients' management were assessed..

n (%) unless otherwise specified	Overall population (*N* = 86)	Subgroup analysis[a]		
		Patients with treatment changes due to joint assessments (*n* = 20)[b]	Patients with treatment changes due to other findings (*n* = 19)	Patients with no treatment changes (*n* = 42)
Age (years) at enrolment, median (range)	19.0 (7–40)	25.5 (12–39)	21.0 (7–40)	17.0 (7–35)
Age category (years) at enrolment				
<18	38 (44.2)	7 (35.0)	7 (36.8)	23 (54.8)
≥18	48 (55.8)	13 (65.0)	12 (63.2)	19 (45.2)
Severity of haemophilia				
Severe	55 (64.0)	13 (65.0)	11 (57.9)	27 (64.3)
Moderate	21 (24.4)	5 (25.0)	5 (26.3)	11 (26.2)
Mild	10 (11.6)	2 (10.0)	3 (15.8)	4 (9.5)

Results: Eighty-six patients from 20 sites were included in the final analysis; 68 had HJHS/HEAD-US assessments at 12 months. Over 12 months, 24.4% (n = 21/86) of patients experienced an impact on their haemophilia management due to HJHS/HEAD-US scores; these decisions were impacted by HJHS in about half of the patients (52.4%, n = 11/21) and HEAD-US in almost all patients (95.2%, n = 20/21). Both assessments contributed to a change in management decisions in about half of the patients

Evaluation of Point-of-Care Ultrasound in Paediatric Haemophilic Arthropathy: A Prospective Comparative Study With Comprehensive Ultrasound and MRI

Caroline Rutten[1,2] | Magdy Soliman[3,4] | Jyoti Panwar[3,5] | Arun Mohanta[1,2] | Alex Zhou[1,2] | Pamela Hilliard[6] | Tammy Rayner[1,2] | Ruth Weiss[1,2] | Carina Man[3] | Rahim Moineddin[7] | Manuel Carcao[2,8] | Vanessa Bouskill[8] | Jennifer Stimec[1,2] | Kevon Dindial[8,9] | Deidre Tetzlaff[6] | Victor S. Blanchette (in memoriam)[2,8,†] | Andrea S. Doria[1,2,3]

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- **To evaluate the diagnostic accuracy** of point-of-care (POC) ultrasound (US) in detecting joint changes in haemophilic arthropathy compared to comprehensive US, physical examination and MRI.
- Single-centre, prospective study examining 22 ankles, eight knees and nine elbows of **28 subjects** (median/range, 16/6– 27 years) with inherited bleeding disorders. **Same-day** physical and imaging joint assessment was performed using Haemophilia Joint Health Score (HJHS) 2.1, **POC-US**, **comprehensive US** and MRI.

Evaluation of Point-of-Care Ultrasound in Paediatric Haemophilic Arthropathy: A Prospective Comparative Study With Comprehensive Ultrasound and MRI

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Results: Moderate inter-reader agreement was observed for soft-tissue changes (kappa = 0.58), which was lower than comprehensive US (kappa = 0.76) and MRI (kappa = 0.77). Agreement for osteochondral lesions was poor (κ 0.09), trailing behind comprehensive US (kappa = 0.55) and MRI (kappa = 0.75). POC-US showed reduced accuracy across all joint parameters in the elbows and ankles compared to comprehensive US, but comparable diagnostic performance in the knees. A good correlation was noted between total POC-US scores and HJHS ($r = 0.68$, $p < 0.0001$), similar to comprehensive US ($r = 0.66$, $p < 0.0001$).

Conclusion: POC-US is valuable for assessing soft-tissue changes in haemophilic arthropathy, correlates satisfactorily with physical examination, but its accuracy is inferior to comprehensive US for ankles and elbows. Both POC-US and comprehensive US should be employed cautiously for assessing osteochondral joint abnormalities.

Haemophilia
The Official Journal of the World Federation of Hemophilia,
European Association for Haemophilia and Allied Disorders and
the Hemostasis & Thrombosis Research Society

Home-delivered ultrasound monitoring for home treatment of haemarthrosis in haemophilia A

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*Hemostasis and Thrombosis Unit; †Health Research Institute, La Fe University and Poly
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In 2010, we started a home-delivered ultrasound protocol to monitor the HT of haemarthrosis of elbows, knees and ankles applied to all patients with severe haemophilia A (SHA) undergoing secondary prophylaxis. The primary objective of this study was to minimize the incidence of JRB maintaining the length of the HT until the reabsorption of the intraarticular blood and the reduction in the synovial swelling as it was previous to the haemarthrosis.

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The home-delivered ultrasound programme was offered to all our patients who had SHA and were undergoing secondary prophylaxis (bi-or tri-weekly) at home. The dose and frequency of prophylaxis were monitored to keep the trough levels of factor VIII (FVIII) between 1.5 and 2 IU mL^{-1}. Only those who

These patients had a clinical and ultrasound joint examination when they had no bleeding episodes to obtain a baseline value. After the appearance of a haemarthrosis on elbows, knees or ankles, the patient would self-administer an initial FVIII dose of 25 IU kg^{-1}. Subsequently, the patient would inform the hospital, and a physiotherapist (with added qualification in technical imaging) of our unit would go to the patient's home to perform a clinical and ultrasound analysis within 24 h of the onset of the haemarthrosis. This home-delivered monitoring with ultrasound imaging and physical examinations covered an area of 38 500 km^2.

The diagnosis of haemarthrosis was based on clinical and ultrasound data compared to the basal state. The clinical criteria were the presence or increase in pain, an increased size of the joint perimeter or a reduced range of the useful mobility. The ultrasound criterion was the presence or increase in intra-articular liquid and synovial swelling in relation to the baseline image. Ultrasound examinations were performed using a portable ultrasonography device (Titan SonoSite; Sonosite Inc., Bothell, WA, USA) with a multi-frequency 5–10 MHz linear array-transducer without Power Doppler. The ultrasound examination protocol consisted of the following: (i) For elbows, we examined the posterior olecranon recess, by keeping the joint flexed 90°, placing the transducer in the mid-longitudinal plane using as reference the insertion of the triceps tendon. (ii) For the anterior knee, we examined the patient supine and a knee flexion of approximately 20–30°. The transducer was placed in the mid-longitudinal plane using as a reference the upper pole of the patellar insertion of the quadriceps tendon. Two longitudinal sections (one on each lateral recess) were used with the same reference to the patella with 30° of external inclination while maintaining a knee flexion of 20–30°. (iii) For ankles, we examined 10–20° of plantar flexion. We examined the anterior recess placing the transducer in the mid-longitudinal plane between extensor hallux longus tendon and extensor digitorum longus tendon, using as reference the tibial head and talus dome [4,6]. The haemarthroses were treated following the scheme showed in Fig. 1.

Table 1. Calls made by the patient to the hospital for an alleged haemarthrosis.

Call	Joint	Haemarthrosis confirmed by ultrasound	Spontaneous/ provoked	Joint rebleeding	Home visits
1	Elbow	Yes	Spontaneous	No	4
2	Knee	Yes	Spontaneous	No	2
3	Elbow	Yes	Spontaneous	Yes	6
4	Knee	Yes	Provoked	Yes	5
5	Elbow	Yes	Spontaneous	No	4
6	Ankle	Yes	Spontaneous	Yes	2
7	Knee	Yes	Provoked	No	2
8	Knee	Yes	Spontaneous	No	10
9	Elbow	Yes	Provoked	Yes	3
10	Knee	Yes	Provoked	No	6
11	Knee	Yes	Provoked	No	4
12	Elbow	Yes	Spontaneous	Yes	3
13	Elbow	Yes	Spontaneous	No	3
14	Knee	Yes	Spontaneous	No	2
15	Elbow	Yes	Provoked	Yes	4
16	Ankle	Yes	Provoked	No	3
17	Elbow	Yes	Spontaneous	No	4
18	Elbow	Yes	Spontaneous	Yes	4
19	Knee	Yes	Provoked	No	2
20	Elbow	Yes	Spontaneous	No	3
21	Elbow	Yes	Spontaneous	Yes	3
22	Elbow	Yes	Spontaneous	No	2
23	Elbow	Yes	Provoked	No	3
24	Knee	Yes	Provoked	No	3
25	Knee	Yes	Spontaneous	No	3
26	Knee	Yes	Provoked	No	3
27	Ankle	Yes	Spontaneous	No	3
28	Knee	Yes	Spontaneous	No	3
29	Elbow	Yes	Spontaneous	No	2
30	Elbow	Yes	Spontaneous	No	2
31	Elbow	Yes	Spontaneous	No	3
32	Ankle	No	–	–	2
33	Knee	No	–	–	1
34	Knee	No	–	–	1
35	Ankle	No	–	–	1
36	Knee	No	–	–	1
37	Ankle	No	–	–	1

Haemophilia
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Home-delivered ultrasound monitoring for home treatment of haemarthrosis in haemophilia A

J. A. AZNAR,*† S. PÉREZ-ALENDA,*‡ M. JACA,*† M. GARCÍA-DASÍ,*† C. VILA,*† A. MORET, *† F. QUEROL*‡ and S. BONANAD*
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Methods

38 patients with SHA Aged between 21 and 49 years (median 36 years) and undergoing home secondary prophylaxis.
These 38 patients had an average follow-up of 17.97 (range 7.17–28.17) months.

Numner of MSK Examination

These patients received at home a total of 113 physical examinations and 113 ultrasound examinations.

Results

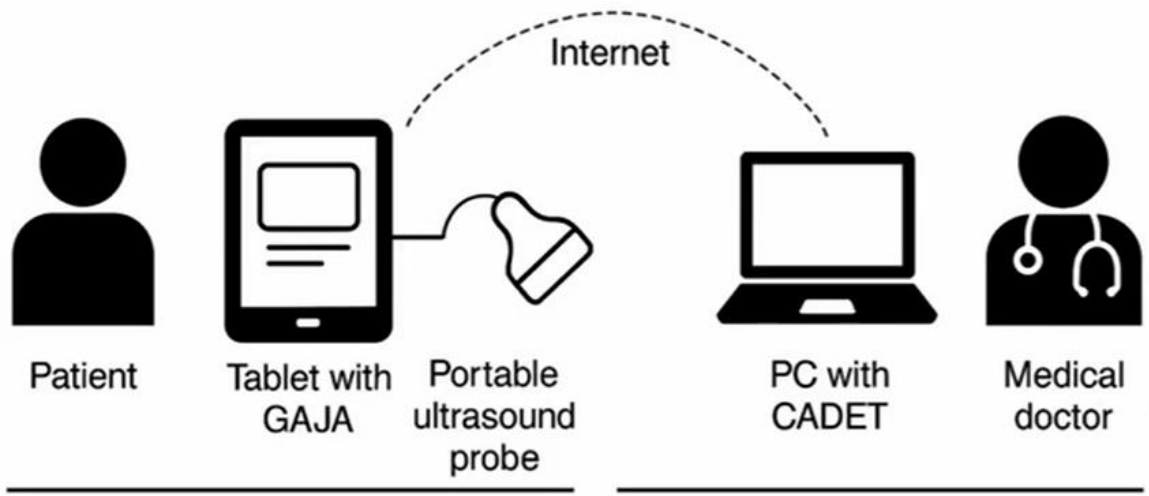
Of the 37 calls made to the hospital, in six cases (16.2%) the ultrasound image did not suggest the presence of intra-articular blood so the episode treatment was discontinued.

In the remaining 31 calls, the presence of haemarthrosis was confirmed.

Important

In 13 of these cases (41.9%), no pain was alleged despite observing intra-articular blood.

GAJA: Interactive guidance for self acquisition of ultrasounds



Teleguidance for Patient Self-Imaging of Hemophilic Joints Using Mobile Ultrasound Devices

A Pilot Study

Peter Aguero, PT, DPT, RMSK, Richard FW Barnes, PhD, MPH, Andres Flores, MS, ATC, Annette von Drygalski, MD, PharmD, RMSK

Received April 1, 2022, from the Department of Medicine, University of California San Diego, La Jolla, California, USA (P.A., R.F.B., A.F., A.D.). Manuscript accepted for publication July 31, 2022.

- A recent international survey of 121 Hemophilia Treatment Centers in 23 countries reported that the majority of hemophilia providers found that **the most important use of MSKUS was to rule out acute joint bleeds.**
- These findings highlight the potential of MSKUS to guide the administration of clotting factor products, **enabling their administration for confirmed bleeding episodes only.**
- **Not all patients have quick access to a specialized center where MSKUS can be performed in a timely manner.**

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- Tele-ultrasonography using tele-guidance **opens the door for ultrasound being performed by operators with limited experience**, which may include **patient self-imaging for remote monitoring and disease management.**
- Health care delivery may be advanced even more by outfitting patients with handheld ultrasound equipment to capture their own images with **real-time feedback and interpretation of findings.**
- Depends on the system's capabilities—but many **advanced portable tele-ultrasound platforms do support full voice and video interaction**, **making the experience much closer to an in-person exam**.


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GAIN: 69
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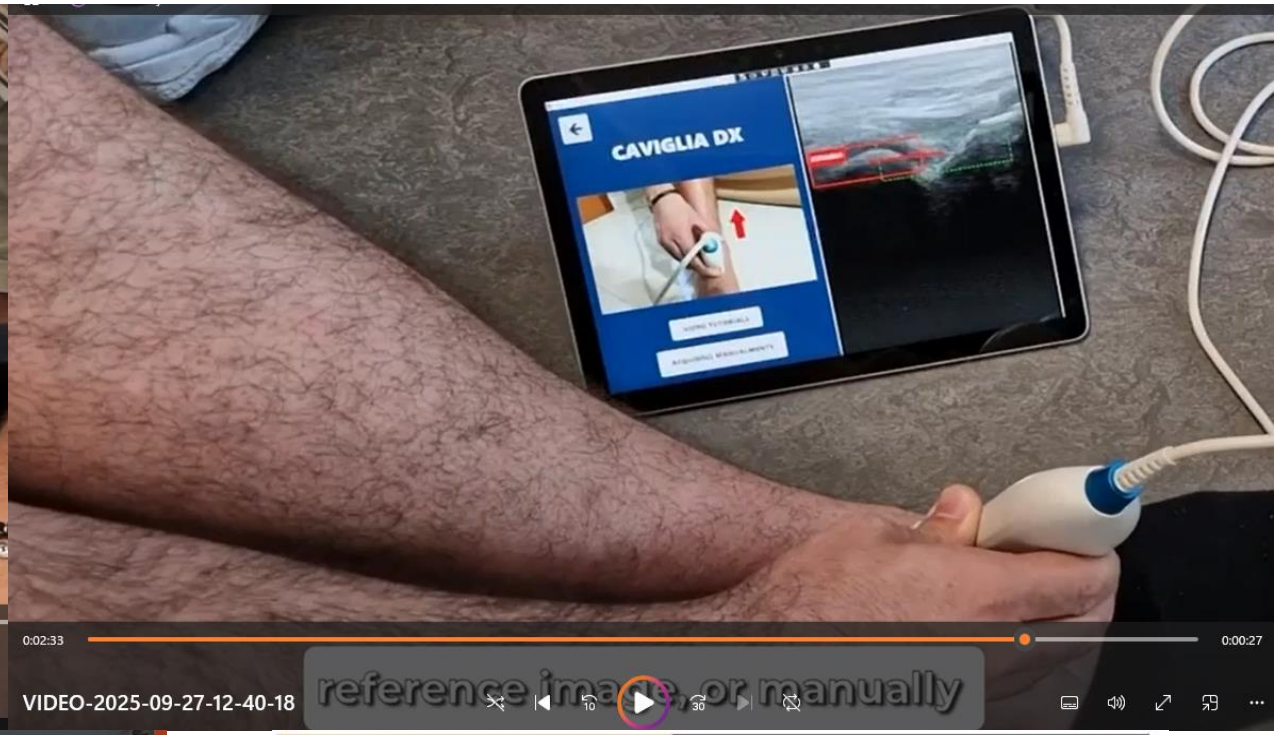
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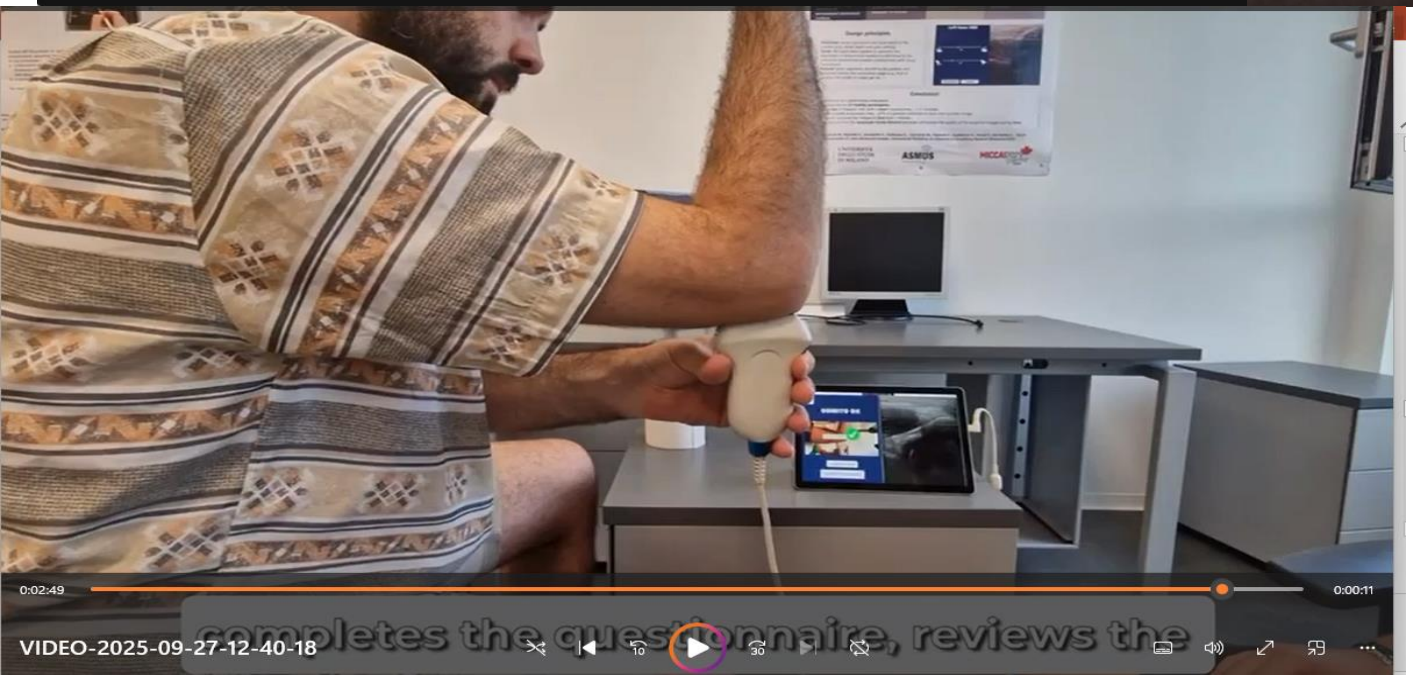
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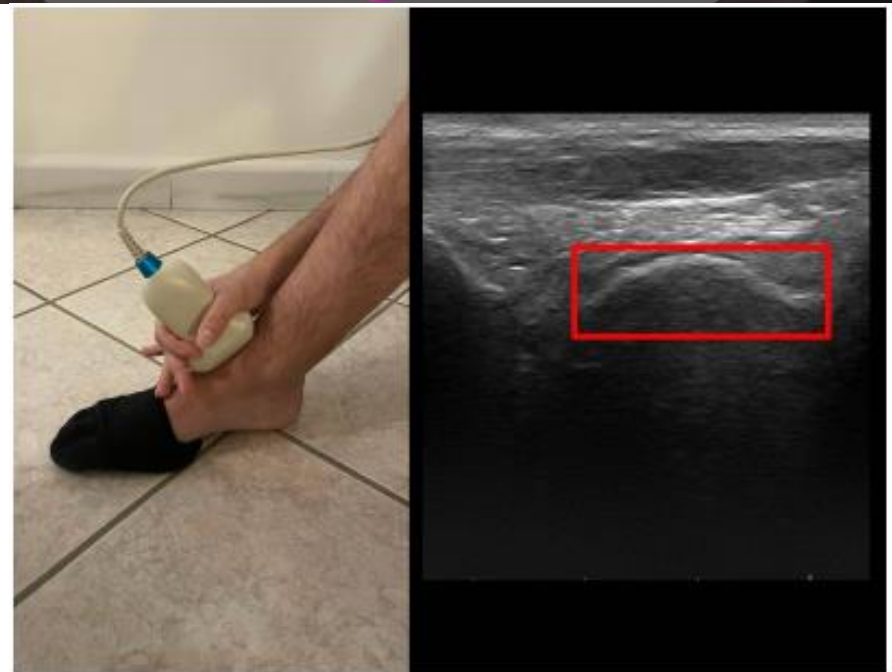

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- **Aims—Determine the feasibility of patient self-imaging with/without teleguidance.**

- The educational session was offered as a “free education” on joint health, joint bleeds, arthritis, and an introduction to MSKUS.
- Patients interested in attending the educational session were given the option to participate in the self-imaging study if they met the following inclusion criteria.

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- **Educational Session**
- The educational session lasted 4–5 hours and was taught in PowerPoint format by the Physical Therapist (PA) and Athletic Trainer (AT).
- It included general aspects of joint health and MSKUS, **explaining the basics of ultrasound physics, probe orientation, vocabulary, sono-anatomy, and hands-on practice.**
- **The hands-on practice included obtaining 11 standard images** from the Joint Tissue Activity and Damage Exam (JADE) protocol.

- These views are most pertinent for the detection of bleeds and soft tissue alterations in the ankle (anterior talocrural joint in short and long axis,
- Achilles tendon in long axis,
- subtalar joint in long axis),
- knee (suprapatellar bursa in long axis, infrapatellar fat pad in long axis, medial and lateral patellofemoral recess in short axis),
- elbow (anterior radio-humeral joint in short and long axis, posterior olecranon recess in long axis).
- Participants were given a handout that included photographs of the 11 standard JADE views, the title of the image (TOI), and photos of proper probe orientation (Figure S1)

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- he experts graded using semi-quantitative scoring on a 0–4 scale in comparison to the expert acquired view. They used the following definitions (Table S3):
- 0 = Incorrect area of the joint or incorrect joint.
- 1 = Not similar to standard image.
- 2 = Somewhat similar to standard image.
- 3 = Similar to standard image.
- 4 = Very similar to standard (expert) image

FIGURE 1. Illustration of teleguided ultrasound. These images show two examples of teleguided ultrasound to assess knee bleeding during a patient training session. The top two images illustrate imaging of the suprapatellar bursa performed by a patient caregiver (**A**) with remote guidance by the physician (AvD) (**B**). The bottom two images illustrate imaging of the lateral recess performed by a patient with remote guidance by the physical therapist (PA). Images: Courtesy of the Hemophilia & Thrombosis Treatment Center, University of California San Diego (with verbal patient consent).

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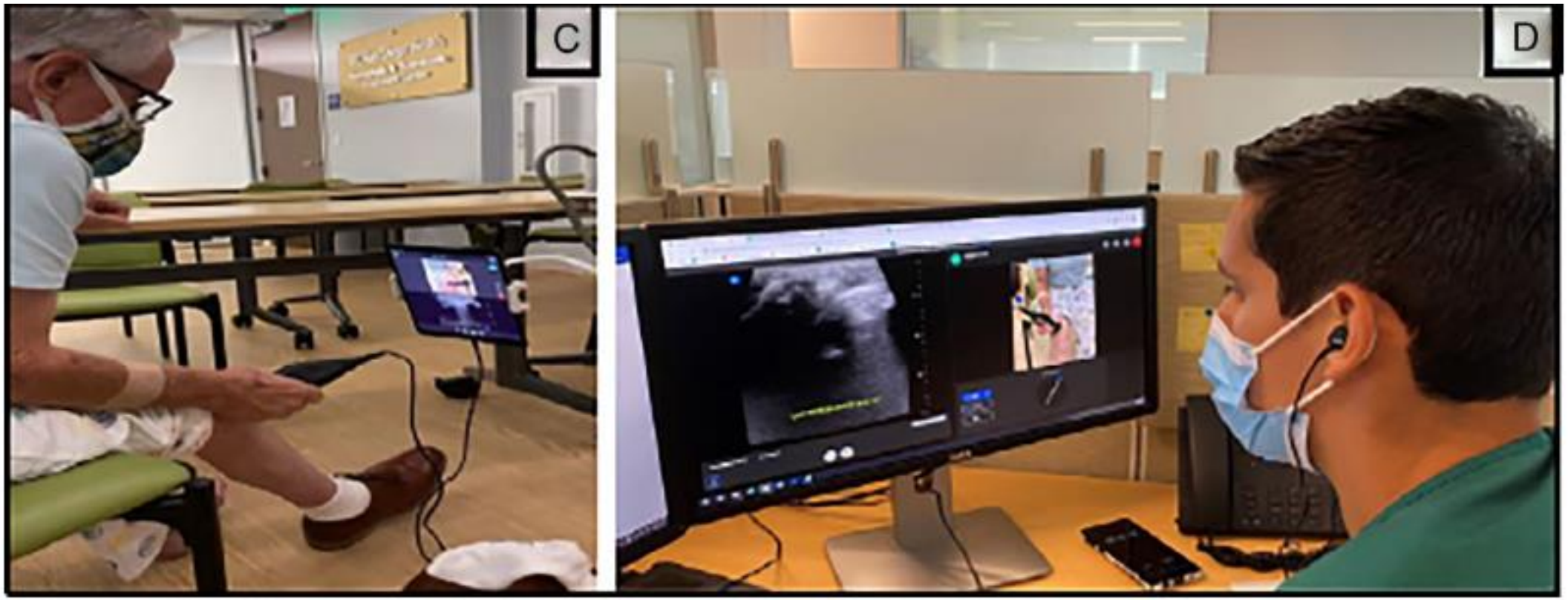

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- The patients acquired five randomly selected views with the Butterfly/IQ probe without assistance at 2, 6– 8 weeks, and 3–4 months later, followed by teleguidance.
- **There were three follow-up assessments after the initial Educational Session.**
- **The first and second followup assessments were conducted in-person 1–2 and 6– 8 weeks after the educational session, respectively.**
- **The final assessment was remote and took place at least 3–4 months after the educational session.**

- **Third (Final) Assessment With and Without Teleguidance**
- Images were acquired twice during the final assessment at least 3–4 months after the educational training session.
- The first session was performed without teleguidance (real-time feedback) by the expert instructor (PA) to assess unsupervised recall of imaging skills 3–4 months after the initial training analogous to the first two assessments (1–2 weeks and 6– 8 weeks post-training).
- The second session included real-time expert feedback during teleguidance to study feasibility and image improvements afforded by teleguidance.
- **Similar to the first two assessments, patients were assigned five randomly selected TOIs and were free to choose between their left or right joints.**

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- Results—Of 132 PWH, 10 (median age 52 years) opted for study inclusion.
- Most had severe Hemophilia A, were white/non-Hispanic, with at least a high school degree and, overall, similar to the other 122 PWH.
- **At 2 and 6 weeks after training, 80% images were acquired correctly compared with 53% at 12 weeks.**
- **Accuracy of landmark recognition was 55%.**
- **With teleguidance, all images were acquired correctly**, with near-perfect image quality ($P \leq .01$ compared with the 3–4 month time point).

TABLE 5. Percent correct joint structure annotation by participants using Musculoskeletal Ultrasound

	Median percent (IQR)		
	Initial	**6–8 Weeks**	**Wilcoxon matched-pairs signed-ranks test: *P***
Bony landmark	100 (100, 100)	80 (60, 100)	.063
Cartilage	60 (60, 90)	40 (30, 70)	.055
Joint space	40 (10, 70)	40 (20, 80)	1.000
Fat pad	40 (10, 70)	60 (30, 70)	.281
Tendons/ligaments	60 (30, 70)	60 (50, 80)	.594

RESEARCH ARTICLE

DOI: 10.4274/tjh.galenos.2021.2020.0717
Turk J Hematol 2021;38:101-110

Impact of the HEAD-US Scoring System for Observing the Protective Effect of Prophylaxis in Hemophilia Patients: A Prospective, Multicenter, Observational Study

Hemofili Hastalarında Profilaksinin Koruyucu Etkisinin Gözlenmesinde HEAD-US Skorlama Sisteminin Önemi: Prospektif, Çok Merkezli ve Gözlemsel Bir Çalışma

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- **HEAD-US Validates Prophylaxis in Hemophilia**
- Study Overview
  - Design: Prospective, multicenter, observational
  - Participants**: 147 hemophilia patients** study **(6-70 Yr)** included male patients aged ≥6 years with the diagnosis of moderate or severe hemophilia A or B from **8 centers across Turkey** between January **2017 and March 2019**. Patients were followed for 1 year with 5 visits (baseline and 3rd, 6th, 9th, and 12th month visits).
  - **Objective: Assess whether HEAD-US scoring reflects the protective effect of prophylaxis vs on-demand therapy**
- 📊 Key Results
  - **Lower HEAD-US scores in patients on prophylaxis → less joint damage**
  - **HEAD-US detected early structural changes, even in asymptomatic joints**
  - **Knees and ankles showed strongest contrast between prophylaxis vs. on-demand treatment**
- 🎓 Conclusion: HEAD-US is a sensitive tool for evaluating joint health and treatment impact

Table 4. Both scoring systems and relationship with frequency of prophylaxis.

	HJHS Total score Mean ± SD Median (min-max)					HEAD-US Total score Mean ± SD Median (min-max)				
	n	Receiving prophylaxis 1-2 times a week	n	Receiving prophylaxis ≥3 times a week	P	n	Receiving prophylaxis 1-2 times a week	n	Receiving prophylaxis ≥3 times a week	p
Baseline	35	11.74±13.32 8 (0-48)	15	5.87±9 2 (0-29)	0.517	39	10.05±10.05 7 (0-33)	13	6.69±6.17 5 (0-20)	0.172
3rd month	34	12.06±13.22 6.5 (0-48)	16	5.75±6.81 3 (0-25)	0.973	31	10.61±11.45 7 (0-36)	16	8.19±6.96 7 (0-22)	0.244
6th month	29	12.14±14.55 4 (0-48)	18	11.94±15.52 5.5 (0-54)	0.410	28	9.46±12 4 (0-40)	13	8.38±6.05 9 (0-23)	0.895
9th month	28	8.46±12.59 3 (0-48)	16	10.13±12.09 6 (0-38)	0.739	23	9.87±11.25 6 (0-40)	11	8.27±5.9 8 (0-22)	0.366
12th month	31	10.48±13.44 3 (0-48)	14	10.79±13.41 5 (0-41)	0.557	32	11.44±11.83 7 (0-40)	11	10.45±6.19 10 (0-22)	0.795

HJHS: Hemophilia Joint Health Score; HEAD-US: Hemophilia Early Arthropathy Detection with Ultrasound; SD: standard deviation; min-max: minimum-maximum.

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Table 5. Prophylaxis regimens and relationship with scores in subgroups of children and adults.

	Baseline		12th month	
Total scores	n	Mean ± SD Median (min-max)	Mean ± SD Median (min-max)	P
Children (6-17 years)				
HJHS	29	1.86±3.07 1 (0-13)	3.17±7.07 1 (0-36)	0.166
<3 years of age	17	1.41±2.67 0 (0-11)	3.24±8.55 1 (0-36)	0.158
≥3 years of age	10	2.7±3.86 2 (0-13)	3.6±4.86 2 (0-16)	0.438
HEAD-US	29	4.03±4.59 2 (0-16)	4.69±4.35 5 (0-14)	0.322
<3 years of age	17	4.53±5.57 1 (0-16)	4.94±4.6 5 (0-14)	0.623
≥3 years of age	9	3.44±2.96 3 (0-9)	4.67±4.58 5 (0-11)	0.395

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	Baseline		12th month	
Total scores	n	Mean ± SD Median (min-max)	Mean ± SD Median (min-max)	P
Adults (≥18 years)				
HJHS	23	17.96±12.55 17 (0-48)	22.43±15.4 22 (0-57)	0.017
Receiving <10 years of treatment	3	21.33±4.51 21 (17-26)	23.67±2.08 23 (22-26)	-
Receiving ≥10 years of treatment	20	17.45±13.35 13.5 (0-48)	22.25±16.55 20 (0-57)	0.031
HEAD-US	23	17.3±8.96 18 (0-33)	19.78±10.34 22 (0-40)	0.006
Receiving <10 years of treatment	4	21.25±6.29 22 (14-27)	26±4.08 24.5 (23-32)	-
Receiving ≥10 years of treatment	19	16.47±9.35 17 (0-33)	18.47±10.83 19 (0-40)	0.023

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- 📌 Recommendations
  - 🔑 Integrate HEAD-US into standard joint assessment protocols
  - 🔑 Use HEAD-US for long-term tracking of joint integrity
  - 🔑 Prioritize early prophylaxis to prevent irreversible damage
- 💥 Bottom Line: HEAD-US strengthens clinical decision-making and improves patient outcomes

Türkiye Klinikleri Tıp Bilimleri Dergisi
Turkiye Klinikleri Journal of Medical Sciences
Turkiye Klinikleri J Med Sci. 2024;44(3):97-104

ORIGINAL RESEARCH ORİJİNAL ARAŞTIRMA

DOI: 10.5336/medsci.2024-101178

Joint by Joint Correlations Between Ultrasound HEAD-US Scores and HJHS Joint Scores in Hemophilia A Patients: A National, Multi-Center, Prospective, and Cross-Sectional Study

Hemofili A Hastalarında Ultrason HEAD-US ve HJHS Eklem Skorları Arasındaki Korelasyon: Ulusal, Çok Merkezli, Prospektif ve Kesitsel Bir Çalışma

- **Study Type: National, multi-center, prospective, cross-sectional**
- **Population: A comprehensive national, multicenter, prospective, cross-sectional, non-interventional observational study was conducted across 19 medical centers in Türkiye, involving 192 hemophilia A patients over a two-year duration.**
- **Objective: Evaluate joint-by-joint correlation between:**
- **HEAD-US (ultrasound-based structural scoring)**
- **HJHS (Hemophilia Joint Health Score – clinical functional scoring)**

Key Findings:

- **Strong correlation between HEAD-US and HJHS scores, especially in ankles and knees**
- **Elbows showed weaker correlation, possibly due to early structural changes not reflected in function**
- **HEAD-US detected subclinical joint damage not captured by HJHS**

Türkiye Klinikleri Tıp Bilimleri Dergisi
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- Recommendations:
- **Integrate HEAD-US into standard joint assessment protocols**
- **Use both tools for comprehensive joint evaluation**
- Prioritize ankle and knee imaging for early intervention

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DOI: 10.1111/hae.15115

Haemophilia WFH WILEY

ORIGINAL ARTICLE

Musculoskeletal

Assessment of haemophilic joints in a low-resourced area using clinical tools: The effect of different types of prophylaxis

Ilham Youssry[1] | Rania El-Taweel[1] | Hadeel Seif[2] | Rania Sami[3] | Karim Okasha[4] | Ebtehal Taha[5] | Assad Haffar[6] | Marwa Abd Elhady[1]

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Abstract

Introduction: Haemophilic arthropathy (HA) is the most frequent complication in people with haemophilia (PWH). MRI is the gold standard to assess HA, however, there are limitations to its use in low-resourced areas.

Aim: Primary; to compare clinical-functional, laboratory, and ultrasonographic joint scores with MRI scores to determine a reasonable alternative to MRI. **Secondary**; to identify the effect of various replacement therapies on the degree of joint involvement.

Material and method: Fifty PWH with at least one affected joint, with or without inhibitors, and receiving either on-demand treatment or secondary prophylaxis, were included. All participants had a joint assessment by clinical HJHS 2.1, functional FISH, HEAD-US, and MRI DENVER scores. Also, serum COMP level was assessed by ELISA for the PWH and 50 healthy subjects as control.

- Their median age was **9 years, with a range of 3–19 years old** (Mean ± SD 9.80 ± 4.66 years).
- Their median age of diagnosis was 4 months, with IQR 6–12 months (Mean ± SD 4.12 ± 1.32 months).

- The included patients were on different treatment regimens based on the severity of their symptoms, and the availability of the products.
- Unfortunately, in our limited resources country, prophylaxis for all haemophilic patients is not feasible.
- Thus, 21 patients (42%) were on-demand treatment,
- 29 patients (58%) were on prophylaxis treatment by either plasma-derived factor-8 or recombinant factor-8, and emicizumab prophylaxis was given to haemophilic patients who developed inhibitors or patients with difficult venous access.
- There was no switch between different treatment regimens during the study period

TABLE 2 Correlation between all studied joint assessment scores.

	HJHS 2.1		FISH		MRI DENVER score		US HEAD Score	
	r	*p*	*r*	*p*	*r*	*p*	*r*	*p*
ABR	.668	>.001*	−.604	<.001*	.651	<.001*	.333	.018*
HJHS 2.1 value			−.877	<.001*	.886	<.001*	.533	<.001*
FISH score value	−.877	<.001*			−.878	<.001*	−.541	<.001*
MRI DENVER score value	.886	<.001*	−.878	<.001*			.452	<.001*
US HEAD Score value	.533	<.001*	−.541	<.001*	.452	<.001*		
Serum COMP level	.093	.469	−.104	.418	.173	.175	−.080	.534

Abbreviations: ABR, annual bleeding rate; COMP, cartilage oligomeric matrix protein; FISH, Functional Independence Score in Haemophilia; HJHS 2.1, Haemophilia Joint Health score version 2.1; MRI, magnetic resonance; US, ultrasound.

*p value significant ≤ .05.

TABLE 3 Homogeneity between the MRI DENVER score and the US HEAD score for all the studied joint changes in the studied joints ($N = 63$).

US HEAD score	MRI DENVER score								
The studied joint changes									
	Mild ($n = 31$)		Moderate and severe ($n = 32$)						
Synovial hypertrophy	No.	%	No.	%	Sensitivity	Specificity	PPV	NPV	Accuracy
Mild	30	96.8	2	6.3	93.750	96.774	96.774	93.750	95.238
Moderate and severe	1	3.2	30	93.8					
χ^2 (p)	51.626*(< .001*)								
	No + Mild ($n = 47$)		Moderate and severe ($n = 16$)						
Intra articular effusion	No.	%	No.	%					
No + Mild	41	87.2	0	.0	100.0	87.234	72.727	100	90.476
Moderate + Severe	6	12.8	16	100.0					
χ^2 (p)	39.969*(< .001*)								

	No ($n = 35$)		Yes ($n = 28$)						
Cartilage loss	**No.**	**%**	**No.**	**%**					
No	35	100.0	26	92.9	7.143	100.0	100.0	57.377	58.730
Yes	0	.0	2	7.1					
χ^2 (^{FE}p)	2.582 (.194)								
	No ($n = 32$		**Yes ($n = 31$)**						
Bony erosions	**No.**	**%**	**No.**	**%**					
No	32	100.0	30	96.8	3.226	100.0	100.0	51.613	52.381
Yes	0	.0	1	3.2					
χ^2 (^{FE}p)	1.049 (.492)								

Abbreviations: χ^2, Chi square test; FE, Fisher exact; NPV, negative predictive value; p, p value for association between different categories; PPV, positive predictive value.

*Statistically significant at $p \leq .05$.

TABLE 5 Joints scores and COMP level for patients on different treatment regimens.

	On recombinant factor 8 ($n = 11$)	On plasma derived factor 8 ($n = 10$)	On emicizumab ($n = 15$)	On demand ($n = 27$)	p
Age of patients(years)					
Min–Max	5–18	6–18	4–17	3–19	.700
Median	7	10	8.5	10	
Age of start of prophylaxis(years)					
Min–Max	4–17	5–17	3–16	_	.797
Median	6	9	8	_	
Prophylaxis duration (mon.)					
Min–Max	11–15	10–15	10–14	_	.024*
Median	14	12	12	_	
Sig. bet. Grps	$p_2 = .208, p_3 = .006^*, p_4 = .192$				
HJHS 2.1 values					
Min–Max	5.0–20.0	4.0–23.0	4.0–13.0	4.0–22.0	.001*
Median	14.0	12.50	6.0	13.0	
p_1	.857	.543	<.001*		

FISH score values					
Min–Max	14.0–30.0	15.0–28.0	21.0–30.0	14.0–30.0	.006*
Median	22.0	20.50	28.0	22.0	
p_1	.974	.292	.005*		
Sig.bet.Grps	$p_2 = .358, p_3 = .024^*, p_4 = .001^*$				
MRI DENVER score values					
Min–Max	4.0–10.0	4.0–10.0	4.0–6.0	4.0–10.0	.002*
Median	9.0	9.0	4.0	9.0	
p_1	.879	.741	.001*		
Sig.bet.Grps	$p_2 = .876, p_3 = .004^*, p_4 = .003^*$				
HEAD US score values					
Min–Max.	1.0–2.0	1.0–2.0	1.0–2.0	1.0–2.0	.044*
Median	1.0	1.0	1.0	1.0	
p_1	.041*	.105	.546		
Sig.bet.Grps	$p_2 = .766, p_3 = .020^*, p_4 = .051$				
Serum COMP level					
Min–Max	16.5–52	9.5–50	26–55	10–82.5	.879
Median	35.5	35.6	36	34	

Thank You

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ORIGINAL ARTICLE

Rapid musculoskeletal ultrasound for painful episodes in adult haemophilia patients

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Perception of pain etiology is unreliable

Many Patients reported inadequate pain relief or no response to prolonged factor replacement