

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

*The following practice guidelines were developed through the consensus of a diverse group of providers who work with bleeding disorder (BD) patients and edited by the National Hemophilia Foundation's Physical Therapy Working Group. The group of providers covered a range of disciplines (pediatric and adult hematologists/oncologists, musculoskeletal radiologists and physical therapists) who use musculoskeletal ultrasound (MSKUS) in their hemophilia treatment centers (HTC's). The information contained in these practice guidelines is not intended in any way to be used as primary medical advice or to replace medical advice. They are intended to guide the providers caring for individuals with BD and interested in implementing MSKUS in their comprehensive care model to the appropriate use of this technology, the need for formal training and establishing proficiency, available opportunities for credentialing and privileging and lastly MSKUS management, quality assurance and maintenance of competence. The group recommends that these practice guidelines are adapted to both scope of practice and local/institutional guidelines.*

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### ● **Introduction**

The musculoskeletal consequences of hemophilia constitute the largest cause of morbidity in persons with hemophilia (PwH)[1]. Hemophilic arthropathy, characterized by synovial hypertrophy and osteochondral damage, results from repeated bleeding episodes in the joint which continue to occur despite early initiation of prophylaxis regimens and a rapidly evolving hemophilia treatment landscape with the advent of novel factor, non-factor and gene therapies[2-5]. A single episode of hemarthrosis can impact long-term joint outcomes and HTC providers are seeking more accurate and objective assessments of painful musculoskeletal episodes and overall joint health. MSKUS has emerged as a promising tool to bridge the gap that continues to exist in our ability to detect and impact hemophilic arthropathy at the preclinical or asymptomatic phase when the disease process is early and potentially reversible [6-13]. MSKUS is widely available, accessible, safe, efficient and less costly, it also does not require sedation in pediatric patients. However, MSKUS is operator dependent and requires skills and knowledge to allow for accurate diagnosis and therapy.

- **The Role of Musculoskeletal Ultrasound in Bleeding Disorder Clinics and Hemophilia Treatment Centers**

MSKUS can provide a detailed and dynamic assessment of synovial joints and periarticular structures, including tendons, ligaments, muscles and soft tissues using high-resolution, high-frequency transducers (3.5–15 MHz) with power Doppler imaging.

In a BD clinic/HTC setting MSKUS can be divided based on scope of practice into:

1. Point-of-Care (POC-MSKUS)
2. Investigative MSKUS
3. Research MSKUS

**POC-MSKUS:** A focused and expedited evaluation, using limited scanning planes or an abbreviated scanning protocol, and is usually carried out by trained providers at the patient's bedside to answer specific yes–no questions. Examples of yes-no questions in the assessment of painful musculoskeletal episodes in patients with bleeding disorders would include presence or absence of hemarthrosis, synovial or intra-articular soft tissue proliferation with/without inflammation (synovitis), and sequelae of recurrent bleeding episodes including osteochondral derangement [11, 12]. Trained providers may also assess extra-articular structures including muscles and soft tissues for hematomas and in select cases perform ultrasound-guided joint aspiration with/without injection of steroids or viscosupplement [14-17]. Tendon and ligament evaluation requires additional imaging expertise.

It is imperative to emphasize that POC-MSKUS should not be used in lieu of clinical assessment but rather be performed as an adjunct or an extension of a detailed musculoskeletal physical examination completed by an experienced and trained health care provider. Proper use of POC-MSKUS will allow for expeditious, accurate diagnosis and treatment recommendations be made in real-time, at the patient's bedside, with the patient at the center of the care model. POC-MSKUS will then also allow for follow up of bleed and/or injury through resolution and inform decision-making around resumption of normal physical activity and return to school or work [18].

**Investigative MSKUS:** MSKUS can be integrated as part of the comprehensive or joint health clinic visit for longitudinal joint health assessment, early detection of hemophilic arthropathy and temporal evaluation of markers of disease activity: soft tissue proliferation and osteochondral derangement in index joints (bilateral elbows, knees and ankles, but shoulders and hips have been evaluated as well) [7, 8, 19-23]. With specialized training, providers can also evaluate for abnormal or disordered skeletal maturation and secondary ossification of pediatric joints. Multiple point-of-care (POC) and full diagnostic MSKUS scanning protocols and scoring systems have been proposed over the past two decades, with variable global adoption and implementation rates by HTC's [24]. Full diagnostic MSKUS is a 360° assessment of the joint, using all standard scanning planes, and is usually performed by expert radiologists, ultrasonographers or trained providers. POC-MSKUS protocols are 2-5 views per joints with ideal proposed scanning times of <5 minutes per joint. These image acquisition times would be after training and

significant experience. The two most commonly used POC-MSKUS protocols are the “Hemophilia Early Arthropathy Detection with Ultrasound” (**HEAD-US**) [25] and Joint Tissue Examination and Damage Exam (**JADE**) [26]. HEAD-US can be used for both pediatric and adult patients, is semi-quantitative, and relies on pattern recognition and little to no measurements; validation studies are ongoing. JADE on the other hand is a validated and provides an algorithm to quantify intraarticular soft tissue expansion, osteochondral changes, inflammation and vascular remodeling in mature hemophilic joints. POC-MSKUS has shown good correlation with joint MRI findings and functional joint outcome scores [8, 27-30]. Normal reference ranges are not currently available for either protocol

**Research MSKUS:** for accurate and objective comparative and longitudinal evaluation of annualized bleeding rates, bleed prevention and management strategies, and musculoskeletal outcomes.

- **Ultrasound Regulations and Qualified Providers in the United States of America: Accreditation and Privileging**

- 1. American Registry of Diagnostic Medical Sonography (ARDMS):**

The governing Board of Ultrasonography in the United States statement [31]: “Licensed allied health care professionals may perform ultrasonic imaging. An allied medical license is required to perform imaging ultrasound in the United States. A certification in imaging ultrasound is not required.”

BD clinics and HTC’s may establish ultrasound practice guidelines at an institutional level and subsequently implement this imaging modality into clinical care within the scope of their practice. Non-sonographers (physicians, physical therapists and advanced care providers) are eligible to earn the Registered in Musculoskeletal Sonography (RMSK) certification; the certifying examination requires applicants to be practicing, licensed, and meet clinical musculoskeletal ultrasound experience prerequisites. RMSK certification is regulated and administered through the Alliance for Physician Certification and Advancement (APCA), a physician-centric council spun out of the ARDMS.

- 2. Alliance for Physician Certification & Advancement (APCA):**

APCA oversees the testing of non-sonographers at the physician level but advanced care providers and other allied health care professionals can be certified at the level of a physician if they meet the qualifications set by the APCA and pass the RMSK exam [32]. In an HTC or BD clinic settings providers who may pursue certification through APCA include physicians, physical therapists, nurse practitioners and physician assistants. A full list of allied health care professionals for whom APCA oversees testing is referenced below and available online [33].

- 3. American Institute of Ultrasound in Medicine (AIUM):**

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AIUM is a multidisciplinary medical association and the leading scientific group in the area of medical ultrasound. The AIUM mission statement is: “Advancing the safe and effective use of ultrasound in medicine through professional and public education, research, development of guidelines, and accreditation” [34]. AIUM recognizes providers that can perform ultrasound examination as “licensed medical providers”, the list is similar to those specified by APCA and again in a BD clinic or HTC setting would include physicians, physical therapists, nurse practitioners and physician assistants.

- **Education, Training and Proficiency**

Ultrasound is an operator dependent imaging modality; training is crucial for development of image acquisition and interpretation skills. Training can be done using didactic modules, hands-on training, and supervised scans; variation in BD clinic and HTC staffing, finances, structure and regulations render it difficult to make an overall statement about “best practice” imaging training for BD clinic/HTC providers at this time. Ongoing education and practice are essential for achieving proficiency and subsequently maintaining imaging and interpretation competence.

Research studies evaluating feasibility, quality and inter/intra-reader variability of implementing POC-MSKUS in a BD clinic or HTC will support the development of training standards and guidelines. In one study by Volland et al. 20-24 hours of in-person, hands-on training were needed for a beginning level ultrasound imager to become effective and reliable in the acquisition and interpretation of the J.A.D.E. protocol images. The accuracy of these images was adjudicated by an MSKUS trained Radiologist [26].

Currently available opportunities for training and education are listed in appendix 1; courses use a combination of didactic modules which cover musculoskeletal/joint anatomy, ultrasound physics and controls, image acquisition, optimization, interpretation, and hemophilic arthropathy assessment and scoring. Additional training is required for providers interested in evaluating pediatric patients, extra-articular structures and muscles. Scope of practice determines the training required: POC-MSKUS for yes/no questions vs. investigative and serial MSKUS.

- **MSKUS Implementation, Process Management and Quality Assurance**

In the authors’ experience successful BD clinic/HTC ultrasound programs exhibited a collaborative working model where the hematologist, physical therapist and patient/parent worked closely to optimize joint outcomes and proper evaluation and management of musculoskeletal painful episodes. Web based collaboration with other HTC providers trained in POC-MSKUS is also encouraged (appendix 1).

It is recommended, but may not be always possible or feasible, to become acquainted with an experienced MSK radiologist who would then collaborate with BD clinic/HTC MSKUS trained

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providers for image interpretation and/or acquisition. Trained providers should seek every opportunity to maintain competence through self-directed active learning and hands-on practice.

Building a strong foundation for clinical implementation is key [18, 35]. MASAC MSKUS and when needed institutional guidelines should be established. Examination purpose and limitations must be disclosed to the patient/parent. This tool should only be used by the appropriate and trained BD/HTC providers. Studies evaluating parent/patient ability to safely use pocket hand-held ultrasound probes and interpret scans should be conducted. This is particularly important as these probes become widely available and affordable; preliminary data is promising [10, 36].

Indications for referral to the radiology department for a comprehensive/full diagnostic ultrasound or another imaging modality should be clearly disclosed to the patient and agreed upon by the treatment team. Reasons for referral may include, but are not limited to, pathology involving a structure outside the scope of practice or scanning expertise, POC-MSKUS findings do not explain the clinical presentation or are unclear as in cases where exam is limited by decreased range of motion or inability of the patient to tolerate the exam due to discomfort or pain.

MSKUS may be initiated upon request of the HTC/BD provider: physician, nurse, physician assistant or physical therapist. Given the musculoskeletal nature of the MSKUS imaging evaluation it has been shown to be very well suited to the physical therapist's skill set [37]. The POC-US/MSKUS examination can be a stand-alone evaluation or a valuable adjunct/extension of the physical therapists' exam and/or treatment. According to the American Physical Therapy Association information on diagnostic and procedural imaging in physical therapy (PT) practice "PT's have been imaging within their practice settings for several decades. Imaging instructional content is now foundational in PT educational programs and is mandated by accreditation standards, allowing for basic competencies in imaging use and decision making at entry level practice. Effective use of imaging in daily PT practice is validated by a multitude of entries in peer-reviewed literature" [38]. It is thus not surprising that in the recently published manuscript by Ignas et al. "Use of Ultrasound for Assessment of Musculoskeletal Disease in Persons with Hemophilia: Results of an International Prophylaxis Study Group Global Survey" PT's 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%) [39].

Images should be safely stored in DICOM format in a HIPAA compliant software and/or hardware. Images should be identifiable with patient information including name, date of birth, medical record number, location and date obtained. Image findings, especially when used to change management, should be clearly documented in the patient's electronic medical record. When a trained and experienced non-physician HTC provider is acquiring and interpreting the images, qualifiers such as, but not limited to, "has the hallmarks of...., is consistent with....., appears to be....." are recommended in describing the findings.

## Maintenance and Infection Control

Each BD/HTC should consult their equipment manufacturer for scheduled maintenance needs including US probes and attachments. This will maintain best working conditions and ensure consistency in optimal imaging. Furthermore, BD/HTC should follow their institutional infection control and equipment disinfection policies and procedures.

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## ● **Appendix 1**

### **1. The UC San Diego Musculoskeletal Ultrasound and Joint Injection Education Activities**

In-person and online didactic modules provide a comprehensive modular curriculum to teach point-of-care musculoskeletal ultrasound for the evaluation of ankles, knees, elbows, hips and spine, and to guide intra-articular needle placement for injections and aspirations.

<https://cme.ucsd.edu/httc/>

### **2. Musculoskeletal Ultrasound (MSKUS) Training For Hemophilia and Other Arthritic Conditions Rounds**

Web-based meeting every 2nd and 4th Tuesday of every month- Bring your case for discussion.

[mxzepeda@health.ucsd.edu](mailto:mxzepeda@health.ucsd.edu)

### **3. MSK Masters**

Dr. Randy Moore

<https://mskmasters.com>

### **4. Haemophilia Early Arthropathy Detection with Ultrasound System (HEAD-US system)**

Dr. Carlo Martinoli

<https://martinoli-ultrasoundcourses.com/carlo-martinoli/>

### **5. The Mohawk College Imaging Research Centre (MIRC)**

Provides an interactive and collaborative space for research and innovation in the area of allied health.

<https://www.mohawkcollege.ca/mohawk-college-imaging-research-centre>