

ORIGINAL ARTICLE

Musculoskeletal

Reliability and clinical features associated with the IPSG MRI tibiotalar and subtalar joint scores in children, adolescents and young adults with haemophilia

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Objectives: To assess the reliability of the IPSG MRI scale for tibiotalar (TTJ) and subtalar joint (STJ) changes in young haemophilic patients, correlating MRI findings with functional scores and 3D-rearfoot kinematics.

Methods: A total of 37 haemophilic patients underwent bilateral MRI of the foot/ankle, clinical evaluation and quantitative assessment of their 3D-rearfoot kinematics during walking. TTJ and STJ soft tissues were assessed twice along with osteochondral changes by two radiologists using the IPSG MRI scale. Inter- and intra-observer reproducibility of MRI scoring were tested by means of kappa statistics. Correlational analyses were performed between MRI findings and the Haemophilia Joint Health Score 2.1 (HJHS) and 3D-rearfoot kinematic data.

Results: The intra-reader reliability of MRI scoring was good to excellent (Kappa: 0.62-1), whereas the inter-reader reliability was moderate to good (Kappa: 0.54-0.79). Weak yet significant correlations were found between the frontal plane rearfoot range of motion (ROM) during loading response of gait and STJ score, as well as between frontal plane rearfoot ROM during the terminal stance phase and the rearfoot osteochondral lesions.

Conclusion: The IPSG score appears applicable to not only the TTJ but also the STJ. Contrary to TTJ lesions, those of the STJ do not correlate with the HJHS but do with 3D-rearfoot kinematic data.

KEYWORDS

Haemophilia Joint Health Score, haemophilic arthropathy, International Prophylaxis Study Group Score, magnetic resonance imaging, subtalar joint, tibiotalar joint

1 | INTRODUCTION

Haemophilic arthropathy is one of the main causes of morbidity in haemophilia.¹ Repeated occurrences of intra-articular bleedings cause

synovial hypertrophy along with cartilage and bone alterations that lead to joint destruction and abarticular soft tissue retraction, severely affecting joint function.² Although regular regular factor replacement can reduce the incidence of joint bleeds, these patients' ankle joints remain



particularly vulnerable, even in haemophilic children under primary or secondary prophylaxis, and this is now recognized as the primary affected joint^{3,4}. Clinical assessment scores such as the Haemophilia Joint Health Score 2.1 (HJHS)⁵⁻⁷ have been developed to estimate the functional status of ankle, knee and elbow joints. Until recently, no study had assessed the sensitivity of this score in detecting early changes related to pauci-symptomatic articular bleeding.^{6,8}

The impact of haemarthrosis on joint structure needs to be characterized, and imaging has become an important tool in haemophilic arthropathy follow-up.^{9,10} Radiography was initially used, yet it offers limited soft tissue assessment and secondary subchondral or bony abnormalities appear when the disease is already advanced. Compared to other modalities, radiography is less sensitive for depicting articular changes,^{10,11}. Ultrasound has a good potential for joint assessment,¹² being less costly and more accessible, easily performed and repeatable. It offers high sensitivity to synovial changes and effusion, although its osteochondral exploration capacities are restricted to the edge of the joints.^{9,13,14} Magnetic resonance imaging (MRI) appears a sensitive tool for detecting arthropathy changes,^{13,15} able to detect early changes not revealed by radiography.² The International Prophylaxis Study Group (IPSG) score was developed using MRI to assess both soft tissues and osteochondral changes in the elbow, knee and tibiotalar joint (TTJ).¹⁶ This scoring tool offers significant clinical potential, though critical appraisal has unveiled a number of elements that need to be addressed before its full clinical utility can be assessed. The first point of interest which should be further explored is the scale's reliability since these qualitative indicators have only been documented by a small expert group (IPSG). Second, the IPSG MRI scale only assesses the TTJ, thus includes no qualitative assessment of the subtalar joint (STJ). Third, early-stage STJ alterations are often well tolerated by patients who naturally adapt their gait, rendering them difficult to detect by clinical examination.¹⁷ Finally, new approaches in foot kinematics assessment using multisegment foot models¹⁸ now enable better comprehension of ankle and foot motion,¹⁹ though the degree of correlation between such measurements and medical imaging has as yet not been explored.

This study had three primary objectives: (i) to apply the IPSG MRI scoring system in the assessment of not only the TTJ but also the STJ, (ii) to determine intra- and inter-reader reliability of this scoring system in the assessment of both joints and (iii) to correlate the MRI findings to the HJHS assessed at the ankle and 3D-rearfoot kinematic data of young haemophilic patients.

2 | MATERIALS AND METHODS

2.1 | Patient selection

Thirty seven haemophilic children, adolescents and young adults (CAAwH), diagnosed with moderate (4/37) or severe (33/37) haemophilia A (31/37) or B (6/37), were recruited prospectively from January 2014 to January 2015 at the Haemophilia Treatment Centers of Cliniques Universitaires Saint-Luc (N = 15) and University Hospitals Leuven (N = 22) (Belgium). We intended to include CAAwH with previous episodes of ankle haemarthrosis with or without clinical evidence of

TABLE 1 Patient characteristics, presented as mean $\pm$ standard deviation [min-max]

Number of patients	37
Age (y)	13.6 $\pm$ 4.4 [6.0-20.3]
Height (meters)	1.58 $\pm$ 0.21 [1.13-1.90]
Weight (kilograms)	55.0 $\pm$ 22.8 [17.2-110.0]
BMI	20.9 $\pm$ 4.9 [13.5-35.5]
Haem A/Haem B	31/6
Severe form/Moderated form	33/4
Inhibitor	26 negative/11 positive
Prophylaxis (primary/secondary/no data)	9/17/11
IPSG MRI score in talocrural joint (/17)	1.4 $\pm$ 3.0 [0-12]
IPSG MRI score in posterior subtalar joint (/17)	0.5 $\pm$ 1.8 [0-10]
HJHS 2.1 score at the ankle (/20)	0.7 $\pm$ 1.6 [0-11]

BMI, body mass index; Haem A/B, haemophilia A/B; IPSG, International Prophylaxis Study Group; MRI, magnetic resonance imaging; HJHS, Haemophilia Joint Health Score.

arthropathy, as well as CAAwH with no ankle-related complaints. The population characteristics are presented in Table 1. The exclusion criteria included acute or unstable chronic disease and neurological or musculoskeletal impairment. Each participant or their parents/guardians completed and signed an informed consent document, which was approved by the two institutional review boards, who also approved the study design (protocol B403201317010).

2.2 | Clinical data

The HJHS 2.1⁵⁻⁷ was used for evaluating joint impairment in both ankles and knees. Each joint was assessed by a physiotherapist (S.L.) with over 15 years of experience in the field of haemophilia. The following items were scored, with a maximum joint score of 20: swelling, duration of swelling, muscle atrophy, crepitus on motion, flexion loss, extension loss, joint pain and joint strength.

2.3 | Imaging protocol

Magnetic resonance imaging examinations were performed using a specific protocol for ankle exploration in each centre's radiology department. Two 3T MRI scans were used (Magnetom Verio, Siemens Healthcare, Erlangen, Germany and Achieva, Philips Healthcare, Best, the Netherlands) equipped with dedicated foot-ankle 16-channel coils. Both ankles were analysed consecutively in a single examination using the following sequences: sagittal T1-weighted spin echo (SE), sagittal T2-weighted gradient echo, sagittal and coronal T2-weighted fast spin echo with fat saturation. The scanning time was approximately 30 minutes per ankle.

2.4 | Imaging analysis

Images were read twice at least 4 weeks apart by two radiologists (J-B.P. and T.B., with 15 and 3 years of experience, respectively), blinded

to the clinical and kinematic data, using the MRI IPSG scale¹⁶ to depict TTJ and STJ joint lesions. Final scoring was determined after consensus between the two radiologists.

2.5 | Ankle-foot kinematics assessment and data

Foot segmental mobility during walking was assessed by means of the Rizzoli Multi-Segment Foot Model (RMSFM).¹⁹ Each child was bilaterally mapped with the marker set of the RMSFM. A passive motion analysis system (Vicon Motion System Ltd, Oxford Metrics, UK) consisting of 10 T-10 cameras was used to track the kinematic data (100 Hz) of all the children while walking along a 10 m walkway. A custom-made force plate was placed in the middle of this walkway (100 Hz, Advanced Mechanical Technology, Watertown, NY, USA). This setup enabled detection of specific gait events. Initial contact and last foot contact of the stance phase was determined by the force platform, whereas the second heel strike associated with the gait cycle was manually determined using Nexus 1.8 software (Vicon Motion System). The RMSFM Plug-in (Aurion Srl, Milano, Italy) was used to calculate the sagittal and frontal plane motion between the shank and calcaneus, defined as the rearfoot. Eventually, the range of motion (ROM) was calculated during specific subphases of the stance phase, namely the initial contact phase (0-2% gait cycle), loading response (2-12% gait cycle), midstance (12-30% gait cycle), terminal stance (30-50% gait cycle), preswing (50-60% gait cycle), swing phase (60-100% gait cycle) and the full gait cycle (0-100% gait cycle).²⁰ This ROM was calculated by measuring the difference between the maximum and minimum values within each phase.

2.6 | Statistical analysis

Inter- and intra-reader reliability of the IPSG MRI scores were analyzed using Cohen's Kappa score for the TTJ and STJ, first separately then combined, giving the so-called rearfoot score. Soft tissue and

osteochondral changes were considered using IPSG additive score values (0-17). Agreements were considered poor with coefficient values < 0.40, moderate with values of 0.40-0.60, good for 0.60-0.80 and excellent for >0.80. Correlations between TTJ and STJ MRI IPSG scores, HJHS 2.1 score assessed at the ankle (HJHS 2.1 ankle score) and 3D-rearfoot kinematic data were evaluated using a nonparametric Spearman correlation test.

3 | RESULTS

We performed MRI scans on 73 ankles, with one patient refusing the second ankle MRI. Inter-reader reliability was moderate to good for TTJ, STJ, and rearfoot scores of soft tissue changes (Kappa: 0.54 to 0.79), osteochondral changes (Kappa: 0.59 to 0.62) and total IPSG MRI scores (Kappa: 0.58 to 0.66) (Table 2). Intra-reader reliability was good to excellent (Kappa: 0.62 to 1) for both the TTJ and STJ. After the radiologists reached a consensus, 19 patients were reported as presenting MRI anomalies, with three producing positive HJHS 2.1 ankle scores without MRI anomalies (Table 3). Unilateral damages were present in 12 patients, and seven had bilateral damages. Of the 26 ankles showing anomalies on MRI, 15 had isolated TTJ anomalies, five had both TTJ and STJ anomalies and six had isolated STJ anomalies (Table 3) (Figure 1).

Of the 73 ankles, 41 had normal IPSG MRI and HJHS 2.1 ankle scores and 14 had pathological results concerning both MRI and clinical evaluation (Table 3). MRI/clinical discordance was reported for 19 ankles (26%). Of these, 11 concerned small effusions (seven cases with small effusions on MRI and no clinical swelling, and four cases with clinical swelling and no effusions on MRI), four had TTJ lesions and three had STJ lesions on MRI with a negative HJHS 2.1 ankle score, while one was free of pathological MRI findings with a HJHS 2.1 score of 2.

Significant Spearman's correlations were found between HJHS 2.1 ankle score and total MRI score ($r = .59$, $P < .0001$) and subscores at the TTJ joint (ranging from $r = .69$, $P < .0001$ for haemosiderin to

TABLE 2 Inter- and intra-reader reliability of the IPSG MRI scale using Cohen's Kappa for the tibiotalar joint (TTJ) and subtalar joint (STJ), separately and combined (Rearfoot score)

	Effusion	Synovitis	Haemosiderin	Soft tissue score	Osteochondral score	IPSG MRI score
Inter-reader reliability						
Rearfoot score	0.75	0.60	0.72	0.60	0.60	0.61
STJ	0.69	0.70	0.79	0.65	0.62	0.66
TTJ	0.79	0.54	0.68	0.56	0.59	0.58
Reader 1 reliability						
Rearfoot score	0.80	0.82	0.77	0.70	0.76	0.66
STJ	0.88	0.65	0.70	0.72	0.85	0.72
TTJ	0.76	0.88	0.80	0.67	0.70	0.62
Reader 2 reliability						
Rearfoot score	0.84	0.88	0.92	0.82	0.83	0.76
STJ	0.85	1	1	0.86	0.85	0.80
TTJ	0.83	0.82	0.90	0.80	0.83	0.74

TTJ, tibiotalar joint; STJ, subtalar joint; IPSG, International Prophylaxis Study Group; MRI, magnetic resonance imaging.



Patient ID	Left TTJ MRI score	Left STJ MRI score	Right TTJ MRI score	Right STJ MRI score	Left ankle HJHS 2.1	Right ankle HJHS 2.1
1	0	0	7 (5 + 2)	0	0	0
2	0	3 (0 + 3)	1 (eff)	1 (eff)	0	0
3	6 (0 + 6)	0	0	0	0	0
4	0	0	0	10 (6 + 4)	0	0
5	1 (eff)	1 (eff)	1 (eff)	0	0	0
6	12 (4 + 8)	0	11 (3 + 8)	0	4	11
7	1 (0 + 1)	0	2 (2 + 0)	0	0	0
8	NA	NA	1 (eff)	0	0	0
9	0	2 (0 + 2)	0	0	1	0
10	0	0	10 (7 + 3)	0	0	2
11	9 (6 + 3)	1 (eff)	0	0	4	0
12	0	0	0	0	1	1
13	0	0	0	2 (1 + 1)	0	0
14	2 (2 + 0)	0	0	0	3	0
15	0	0	0	0	0	1
16	0	0	0	0	1	0
17	0	0	6 (3 + 3)	0	2	4
18	0	1 (eff)	0	0	0	0
19	8 (8 + 0)	0	7 (6 + 1)	0	4	2
20	2 (1 + 1)	10 (3 + 7)	3 (1 + 2)	6 (2 + 4)	2	2
21	0	0	6 (0 + 6)	0	0	2
22	0	1 (eff)	1 (eff)	0	0	0

TTJ, tibiotalar joint; STJ, subtalar joint; MRI, magnetic resonance imaging; HJHS, Haemophilia Joint Health Score.

TABLE 3 Patient scores with HJHS 2.1 and/or inter-reader consensus IPSPG MRI score > 0 are presented for left/right tibiotalar joint (TTJ) and subtalar joint (STJ). Soft tissue and osteochondral subscores are presented in brackets, respectively. When the only finding was a small effusion, (eff) was reported

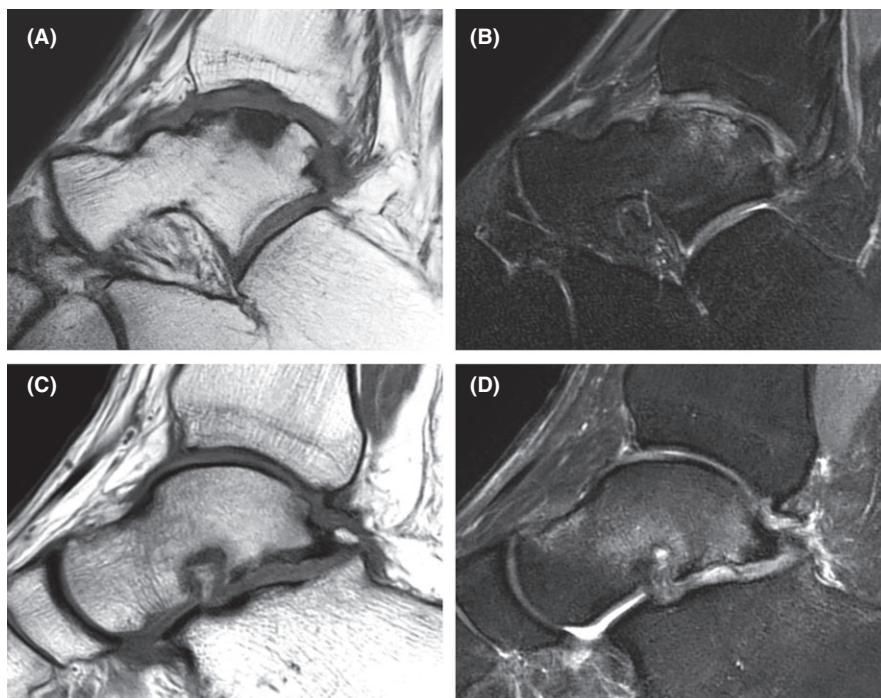


FIGURE 1 T1-weighted (left-sided) and T2 fat-suppressed weighted (right-sided) sagittal images from two patients with tibiotalar joint (A and B) and subtalar joint (C and D) lesions

TABLE 4 Spearman's correlation between IPSPG MRI score and subscores for tibiotalar joint (TTJ) or subtalar joint (STJ) and HJHS 2.1 clinical score evaluated for the same ankle

	TTJ		STJ	
	Spearman's ρ	P	Spearman's ρ	P
Effusion	0.45	<.0001	0.12	ns
Haemosiderin	0.69	<.0001	0.05	ns
Synovial hypertrophy	0.61	<.0001	0.20	ns
Soft tissues total score	0.57	<.0001	0.11	ns
Bone surface erosion	0.48	<.0001	0.16	ns
Half surface eroded	0.44	.0001	0.30	.011
Subchondral cyst	0.48	<.0001	0.15	ns
Any loss of joint cartilage	0.58	<.0001	0.21	ns
Full-thickness loss of cartilage	0.42	.0002	0.10	ns
Total osteochondral score	0.60	<.0001	0.17	ns
Total IPSPG MRI score	0.59	<.0001	0.14	ns

Bone surface erosion: any surface erosion. Subchondral cyst: at least one subchondral cyst. Half surface eroded: half or more of the articular surface eroded in at least one bone.

TABLE 5 Summary of rearfoot sagittal and frontal plane ROM occurring in the different phases of the gait cycle (mean $\pm$ standard deviation)

	Rearfoot frontal plane ROM ($^{\circ}$)	Rearfoot sagittal plane ROM ($^{\circ}$)
Loading response (0%-12%)	3.4 $\pm$ 1.7	6.4 $\pm$ 3.1
Midstance (12%-30%)	1.7 $\pm$ 1.1	5.9 $\pm$ 2.2
Terminal stance (30%-50%)	3.7 $\pm$ 1.4	4.5 $\pm$ 2.1
Preswing (50%-60%)	2.3 $\pm$ 1.5	15.3 $\pm$ 4.0
Swing (60%-100%)	3.9 $\pm$ 1.6	19.3 $\pm$ 4.8
Full gait cycle (0%-100%)	7.6 $\pm$ 1.9	21.9 $\pm$ 4.9

$r = .42$, $P = .011$ for full-thickness loss of cartilage) (Table 4). In contrast, no correlation was found between HJHS 2.1 ankle score and either STJ total score or subscores, except for the "half or more of the articular surface eroded in at least one bone full-thickness loss of cartilage" subscore ($r = .30$, $P = .011$).

Rearfoot sagittal and coronal plane ROM data are presented in Table 5. Significant correlations between these data and the IPSPG MRI score are presented in Table 6. Rearfoot sagittal plane ROM showed poor inverse correlations with TTJ bone erosion or osteochondral lesion scores or subscores for all phases except the midstance phase (r value from $-.25$ to $-.35$, P value from .035 to .003). The presence of haemosiderin was inversely correlated with the loading response and terminal stance phase ($r = -.25$ and $-.26$, $P = .0035$ and .032, respectively). No correlation was observed between sagittal ROM and STJ MRI score. Rearfoot frontal plane ROM during loading response phase data revealed poor yet significant inverse correlation with rearfoot MRI score ($r = -.25$, $P = .039$), with the presence of STJ lesions on MRI ($r = -.28$, $P = .018$) and particularly osteochondral lesions of the STJ joint ($r = -.29$,

$P = .015$). Interestingly, rearfoot frontal plane ROM during the terminal stance phase was positively correlated with the rearfoot osteochondral score on MRI ($r = .26$, $P = .028$), with the presence of osteochondral lesion more specifically of the TTJ joint ($r = .27$, $P = .024$).

4 | DISCUSSION

4.1 | STJ changes on MRI

The primary aim of our study was to conduct the first-ever assessment of arthropathic changes related to haemophilia in the STJ. To our knowledge, no systematic evaluation of STJ had ever before been reported using MRI with the IPSPG score. The IPSPG MRI scale originally enabled the structural changes of the TTJ to be described in the context of haemophilic arthropathy management, even if the involvement of STJ in haemophilic arthropathy of the ankle is well established.^{17,21} Very few studies had even addressed this field for other imaging modalities.^{22,23}

The assessment of STJ on MRI was possible in all ankles, with the same criteria as those for TTJ. The STJ has thick enough cartilage and the orientation of the joint space is primarily in the horizontal plane, thus easy to explore on sagittal images. Thus, subtle changes are easily depicted in the STJ. Soft tissue changes were analyzed in the posterior recess of the joint, along with the medial and anterolateral recess. The posterior recess, however, frequently communicates with the posterior recess of the TTJ, thus STJ effusion was considered only when fluid was also present at the anterior aspect of the joint.²⁴

Most of our patients had severe haemophilia, yet the number of altered ankles was moderate, with IPSPG MRI score or positive HJHS 2.1 ankle score found in 22 patients (34 ankles). Less abnormalities were observed in the STJ compared to the TTJ, as well as a lower mean IPSPG score. MRI-detected changes were found in 11 ankles, six with osteochondral lesions $\pm$ soft tissue changes and five with small effusions only. These STJ changes were associated with TTJ changes in only 45%



TABLE 6 Summary of significant Spearman's correlations between rearfoot sagittal and coronal plane range of motion (ROM) and IPSPG MRI scores and subscores for subtalar (STJ) and tibiotalar (TTJ) joints. The *P* value is shown in parentheses for each Spearman's ρ coefficient. Given that correlations with rearfoot sagittal ROM were found for TTJ only, STJ is not showed

	Coronal		Sagittal		
	TTJ	STJ	TTJ		
Loading response (0%-12%)		IPSG positive	-0.28 (.021)	Bone surface erosion	-0.27 (.021)
		IPSG total score	-0.25 (.039)	Full cartilage half bone lesion	-0.27 (.023)
		Osteochondral positive score	-0.29 (.015)	Osteochondral lesion	-0.26 (.026)
				Haemosiderin	-0.25 (.035)
Midstance (12%-30%)					
Terminal stance (30%-50%)	IPSG positive	0.27 (.023)		Haemosiderin	-0.26 (.032)
	Osteochondral lesion	0.27 (.28)			
Preswing (50%-60%)				Full cartilage half bone lesion	-0.29 (.017)
				Subchondral cyst	-0.25 (.032)
Swing (60%-100%)				Subchondral cyst	-0.35 (.003)
				Subchondral cysts in two bones or a third	-0.26 (.034)
Full gait cycle (0%-100%)				Subchondral cyst	-0.34 (.004)
				Subchondral cysts in two bones or a third	-0.29 (.016)
				Full cartilage half bone lesion	-0.26 (.034)
				Half surface eroded	-0.25 (.035)

Bone surface erosion: any surface erosion. Half surface eroded: half or more of the articular surface eroded in at least one bone. Subchondral cyst: at least one subchondral cyst. Subchondral cysts in two bones or a third: subchondral cysts in at least two bones, or cystic changes involving a third or more of the articular surface eroded in at least one bone. Full cartilage half bone lesion: full-thickness loss of joint cartilage including at least one half of the joint surface in at least one bone.

of cases (Table 3). Of the six ankles with isolated STJ changes, only one was symptomatic. In contrast, one ankle with an IPSPG MRI score of 10 was asymptomatic. This is in accordance with the lack of correlation that was found between HJHS 2.1 ankle score and IPSPG MRI score at the ankle, supporting the need for STJ assessment in imaging screening of haemophilic patients.

Ultrasound and MRI have been compared for joint analysis, producing strong correlations for soft tissue changes and most osteochondral lesions for several joints in TTJ assessment at the ankle site.¹⁴ The STJ is more difficult to access, and assessing chondral lesions is impossible using ultrasound. In contrast, the synovial recess is easy to evaluate with a posteromedial approach, and effusion can be visualized in the tarsal sinus in the anterolateral recess of the joint. To date, no specific study has yet evaluated the respective sensitivity or specificity of these two techniques for STJ changes.

4.2 | Reliability of the IPSPG MRI scale for TTJ and STJ assessment

We found moderate to excellent reliability indices for inter- and intra-reader reliability when assessing the TTJ using this scale. Interestingly, its reliability for STJ and TTJ joint lesion depiction was

almost in the same range. The same scientific credibility awarded to this scale with respect to TTJ assessment¹⁶ thus appears to be also merited for its assessment capacities of the STJ joint. We chose to use Kappa to calculate this reliability based on the ordinal nature of the variables contained in the IPSPG MRI score. The results are still in accordance with previous reports from Lundin et al¹⁶ despite being reported in terms of intra-class correlation scores. Our results appear more moderate to good compared to their good to excellent scores, however, which could be partially explained by the statistical tests we used. Finally, the difference in reliability between the two readers could be partly related to their differing experience in musculoskeletal imaging.

4.3 | Correlation between MRI consensus score and functional assessments

Tibiotalar MRI changes were found to be significantly correlated with clinical alterations, as already reported.^{6,8,9,16,20} Nevertheless, studies including both HJHS and MRI findings have only recently been published.^{16,25,26} Doria et al²⁵ reported high correlations between ankle erosion scores and HJHS. Oymak et al⁶ reported moderate correlations between HJHS and MRI additive or progressive score, though

their study included knee and elbows and only 10 ankles. Poonnoose et al⁸ reported on 22 knees and 33 ankles in a population of 51 patients with a distribution based on the Petterson radiographic score. Their patients had more severe TTJ lesions and most were positive for HJHS and joint changes on MRI. We found moderate correlations between osteochondral changes on MRI and HJHS ($r = .60$ for TTJ in our study compared to $r = .51$ for Poonnoose et al though their results included both knee and ankle assessment). In contrast, the correlation between MRI scoring and HJHS clinical score was stronger in our study for soft tissue changes ($r = .57$ vs $.19$ for Poonnoose et al). Nine ankles showed discordance with negative HJHS while producing positive IPSPG MRI scores: Two patients had osteochondral lesions, one had both osteochondral and soft tissue changes, one small effusion and small synovial hypertrophy, and the remaining five showed only small effusions. This raises the question of the pathological nature of these effusions. Foppen et al²⁷ found similar scores for effusion in the ankles of both haemophilic and control patients, notably for small effusions (12% and 10%, respectively), concluding that including joint effusions in the MRI scale can be expected to reduce its specificity for haemophilic arthropathy. A similar number of small effusions without any clinical findings were found in the STJ than in the TTJ (four and five, respectively), raising the question for this joint as well. Finally, the four cases showing small effusion without fluid found on MRI illustrate the difficulties to differentiate between false positive clinical assessment and false negative MRI scoring related to limited reliability.¹³

Contrary to TTJ, STJ joint changes on MRI have very few correlations with clinical scoring. Indeed, the HJHS 2.1 score does not tend to detect STJ lesions because clinical tests are not focused on this particular joint. This limitation underlines the need to add STJ-related scoring items to the clinical scoring system. In our study, we incorporated in vivo 3D motion analysis in order to assess the dynamic behaviour of the rearfoot, it being the rotation between the shank and calcaneus. We considered discrete variables representing the range of sagittal and frontal plane mobility between the shank and calcaneus during the stance phase of gait. The observed correlations between the sagittal plane rearfoot ROM and IPSPG MRI score highlight that the progression of gait over the support foot, also known as the three functional rockers, appears to be affected in the arthropathy patients. There seems to be evidence for a weekend fulcrum function of the heel during loading response (heel rocker), followed by a reduced ankle dorsiflexion during terminal stance (ankle rocker). Finally, a reduced ankle plantar flexion during propulsion seems to be associated to the degree of arthropathy, potentially illustrating decreased forward propulsion (third rocker). Despite being weak, a negative correlation was evidenced between the frontal plane rearfoot ROM at loading response and the STJ MRI score. During the loading response, STJ pronation provides shock absorption and participates in distributing ground reaction forces over the complete forefoot region. The reduction of rearfoot frontal plane ROM as a consequence of recurrent bleeding episodes and arthropathic lesions may affect this natural shock absorption function and, in turn, have a negative effect on loading distribution of the STJ and TTJ. In contrast, we found a positive correlation between the rearfoot

frontal plane ROM during the terminal stance phase of walking and the rearfoot osteochondral score, and more specifically the TTJ osteochondral lesions. During this phase, a second rocker function involves considerable TTJ dorsiflexion ROM, yet with increasing osteochondral lesions, and the limited sagittal motion may lead to compensatory ROM in the frontal plane. These correlations need further investigation to corroborate the value of kinematics assessment in the monitoring of patients under prophylaxis.

Our study had some limitations. The number of patients included in this study and the large number among them without clinical or MRI changes in the ankle limited the number of pathological joints that could be assessed. Our difficulty in recruiting patients can be explained by the rarity of this disease and the difficulty for some participants to travel to two investigation sites. The heterogeneity of the sample in terms of age is also a point of interest. Our results for the TTJ are in line with others from the literature, whereas for the STJ evaluation, our findings will have to be confirmed in a larger sample of patients. The clinical evaluation was based on the HJHS 2.1 score, which was not originally designed to detect STJ lesions. Specific tests, such as palpation of the tarsal sinus recess or evaluation of pain triggering with varus and valgus test applied at the calcaneus in ventral decubitus position, could be added, although the sensitivity and specificity of these signs are not well established. Eventually, the number of bleedings and therapeutic regimens were not precisely reported either, we thus could not test their correlation with clinical and MRI findings or the impact of our STJ change assessment on prophylactic treatment. While this was not the exact aim of our study, a validation of the IPSPG scale added to optimized clinical scoring for STJ assessment would enable future studies to address this question and to better evaluate the clinical impact of depicting pathological findings in asymptomatic joints.

5 | CONCLUSION

The IPSPG MRI score enables a reproducible depiction of structural changes of the TTJ and STJ in haemophilic ankle arthropathy follow-up. TTJ lesions are significantly associated with clinical dysfunctions. The STJ joint can be independently affected without immediate repercussions in articular function detected using the HJHS 2.1 score, yet correlations are seen with kinematics assessment.

DISCLOSURE

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