

Diagnosi precoce e monitoraggio dell' artropatia nel giovane

L' approccio 2.0 all' emofilia

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La valutazione clinica/radiografica classica

- I sanguinamenti articolari nei pazienti emofilici (target joints: caviglie, ginocchia, gomiti) determinano ipertrofia sinoviale e alterazioni osteocondrali

Valutazione:

- **Petterson Score** (radiografico)
- **Gilbert Score** (clinico)
- **HJHS** (clinico)

Cosa è cambiato?

- Con la terapia sostitutiva profilattica è aumentata l' enfasi sul concetto di **PREVENZIONE**
- Entrambi gli scores (Gillbert e Pettersson) non sono adatti a rilevare alterazioni articolari precoci

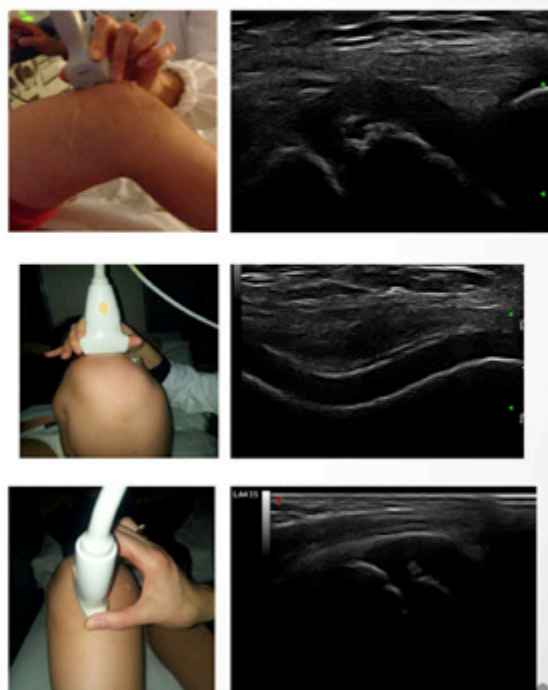
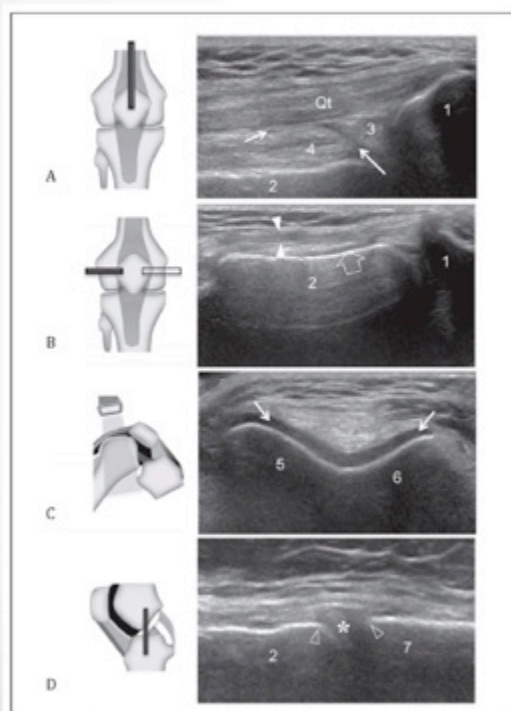
Problematiche

- Discrepanza tra clinica e imaging nel paziente emofilico
- **RM**: elevata sensibilità, rileva ipertrofia sinoviale, depositi di emosiderina, alterazioni cartilaginee e ossee precoci
- Non adatta per un monitoraggio routinario

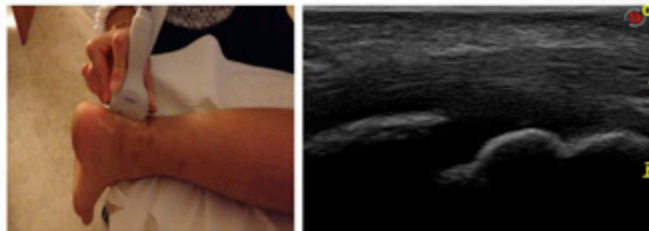
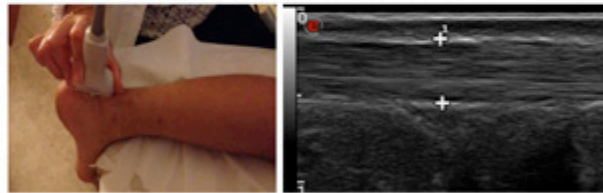
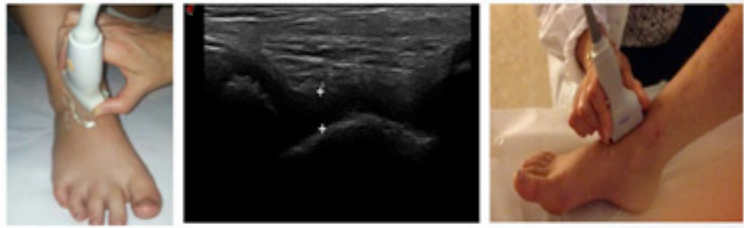
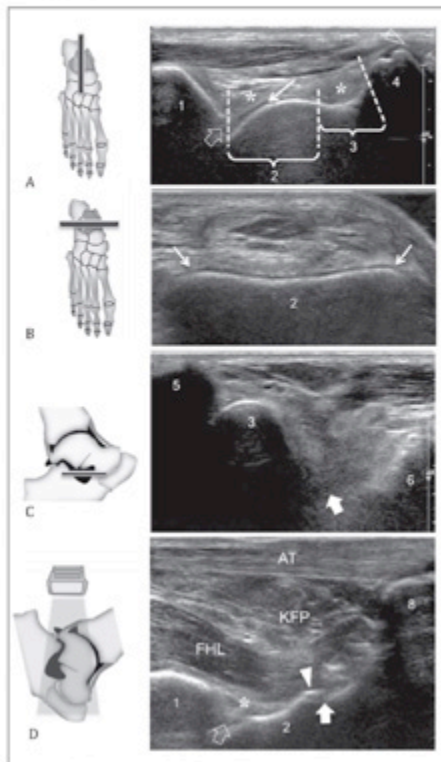
Gli ultrasuoni

- L' **ecografia** per la valutazione di routine delle articolazioni nei giovani emofilici (Emofilia moderata/severa) in profilassi mostra alterazioni precoci, anche con HJHS score basso
- Utile per il follow-up di alterazioni già note
- Utile per la valutazione di episodi acuti (emartro, sinovite)
- Facile e veloce da eseguire, non invasiva

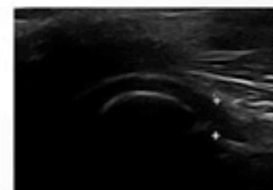
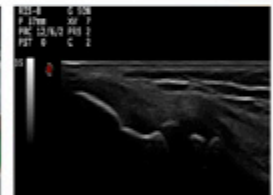
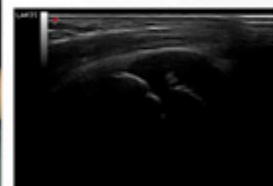
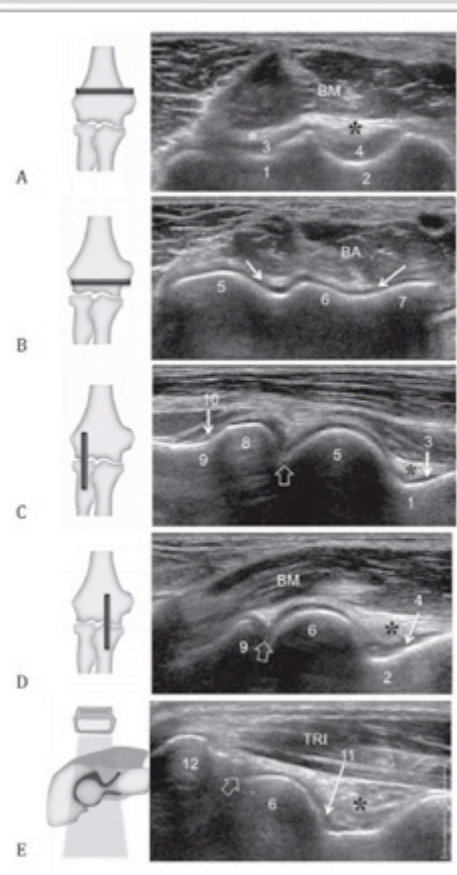
Ginocchio



Caviglia



Gomito



Protocolli

- **HEAD-US** (Haemophilia Early Arthropathy Detection with UltraSound): protocollo sviluppato per medici non-radiologi

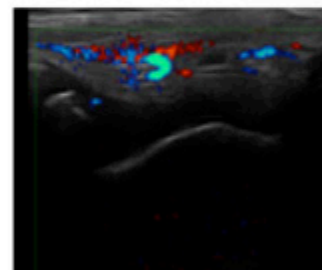
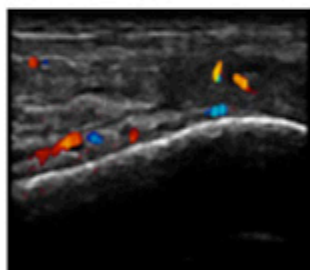
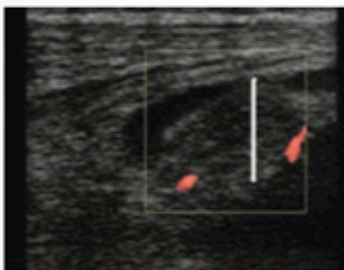
Table 2: HEAD-US scoring method.

Disease activity (synovitis)	Scale
Hypertrophic synovium	
0. Absent/Minimal	0
1. Mild/Moderate	1
2. Severe	2
Disease damage (articular surfaces)	
Cartilage	
0. Normal	0
1. Echotexture abnormalities, focal partial/full-thickness loss of the articular cartilage involving <25% of the target surface*	1
2. Partial/full-thickness loss of the articular cartilage involving at least \leq 50% of the target surface*	2
3. Partial/full-thickness loss of the articular cartilage involving >50% of the target surface*	3
4. Complete cartilage destruction or absent visualization of the articular cartilage on the target bony surface*	4
Bone	
0. Normal	0
1. Mild irregularities of the subchondral bone with/without initial osteophytes around the joint	1
2. Deranged subchondral bone with/without erosions and presence of prominent osteophytes around the joint	2

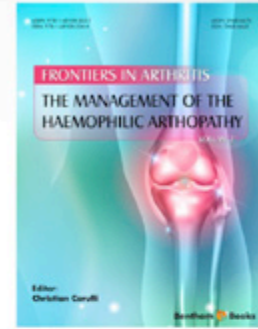
Note: Elbow: anterior aspect of the distal humeral epiphysis, Knee: femoral trochlea; Ankle: anterior aspect of the talar dome.

Valutazione PDUS

- **PDUS**: neoangiogenesi sinoviale



Protocolli



This scoring method is applied to each target joint with a range from 0 to 21 and with cut off ≤ 5 or >5 useful to define the early stage of arthropathy [11].

Item	Concept	Evaluation	Score
1	Effusion: hydropsyrosis or haemarthrosis	Absent = 0; small = 1; moderate = 2; Absent = 0; small = 1, moderate = 2;	
2	Fibrotic septa	Absent = 0; present = 1	
3	Synovial hypertrophy with flags on PDUS	1 = <3 flags, 2 = >3 flags	
4	Synovial hypertrophy without flags on PDUS	Thickness <1.5 mm = 1 Thickness 1.5-2.5 mm = 2 Thickness >2.5 mm = 3	
5	Haemosiderin deposition	Absent = 0; small = 1; moderate = 2; large = 3	
6	Bone erosion	Absent = 0; present = 1	
7	Osteophytes	Absent = 0; present = 1	
8	Bone remodelling	Absent = 0; present = 1	
9	Cartilage modifications	Hyperechoogenicity = 1; irregular profile = 2; calcification = 3	

Table 1. Personal scoring system for haemophilic arthropathy.

Haemophilia

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



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Letter to the Editor

Comparing findings of routine Haemophilia Joint Health Score and Haemophilia Early Arthropathy Detection with UltraSound assessments in adults with haemophilia

M. A. Timmer , W. Foppen, R. E. G. Schutgens, M. F. Pisters, K. Fischer

First published: 16 March 2017 [Full publication history](#)


DOI: 10.1111/hae.13147 [View/save citation](#)

- In conclusion, in adults with haemophilia, HEAD-US may have **additional value** to HJHS by detecting the origin of slightly impaired function, or in revealing of early osteochondral changes in apparently healthy joints.



Review Article

A systematic review of ultrasound imaging as a tool for evaluating haemophilic arthropathy in children and adults

C. C. Ligocki, A. Abadeh, K. C. Wang, T. Adams-Webber,
V. S. Blanchette, A. S. Doria 

First published: 21 April 2017 [Full publication history](#)

DOI: 10.1111/hae.13163 [View/save citation](#)

- The association between frequent monitoring in asymptomatic patients in early stages of the disease with the high cost of prophylaxis and treatment can make US a **very promising** tool for assessment of soft tissue changes in haemophilic arthropathy once standardized scoring system and protocols are available leading to reliable US conclusions.

(spesso in presenza dell'ematologo)

- Raccolta dell'anamnesi
- Esame obiettivo
- Visualizzazione di esami strumentali/ consulenze/ cartella del centro emofilici
- **Ecografia** (articolazioni target, comparativa)

Cosa fare dopo la diagnosi

- Prescrizione di attività fisica costante
- Attività sportiva nei giovani a basso impatto
- Fisioterapia dopo emartri
- Infiltrazioni (viscosupplementazione, sinoviortesi)
- **Monitoraggio periodico delle articolazioni bersaglio**

GRAZIE PER L'ATTENZIONE

