REHABILITACIÓN EN PERSONAS CON HEMOFILIA

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Hemofilia

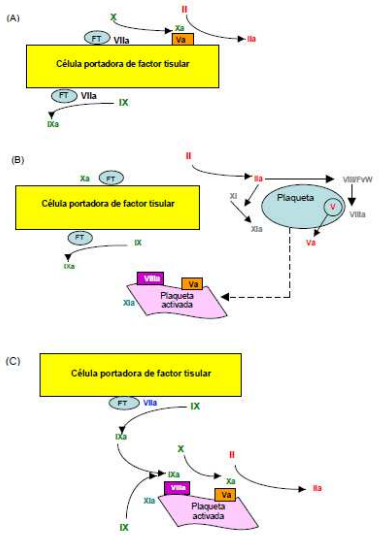


Figura 1. Modelo celular de la hemostasia: A) iniciación, B) ampliación, C) propagación.

Base hematológica

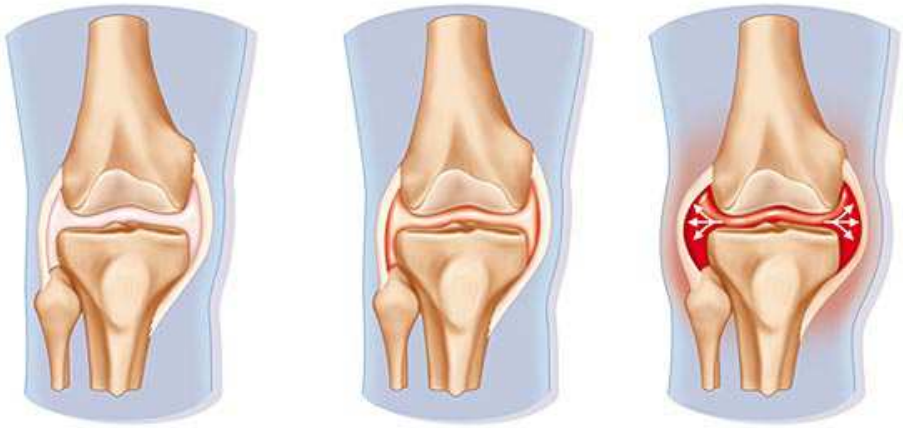
Clínica ortopédica

Manifestación más frecuente :

HEMARTROSIS

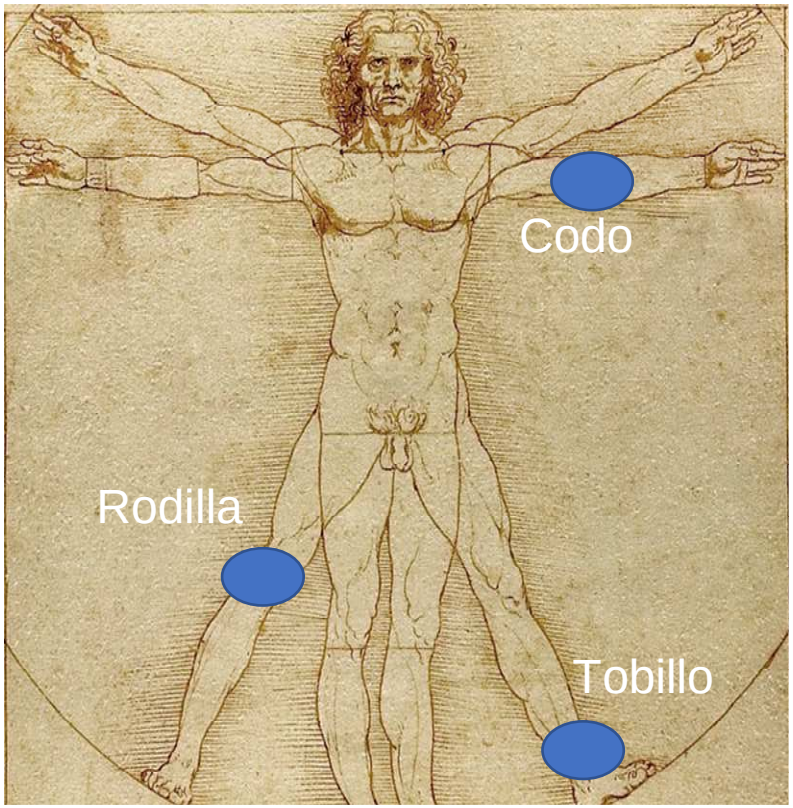
Hemartros

- El sangrado intra-articular procede de la membrana sinovial, originándose en el plexo subsinovial
- Suponen el 75-85% de todos los sangrados

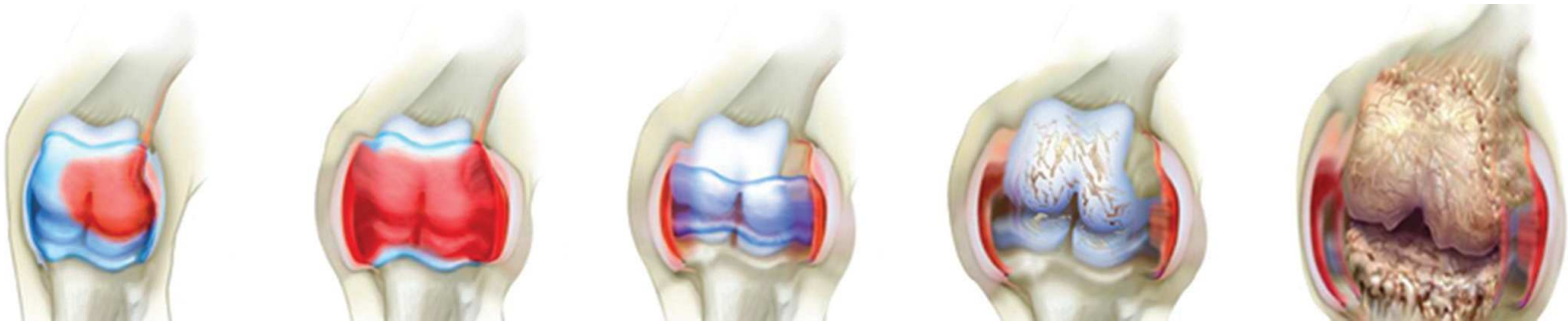


Rodríguez-Merchán EC et al. Haemophilia 2007; 13:613-9.
Roosendaal G *The Haemophilic Joints: New Perspectives. Wiley-Blackwell, Oxford, 2003: 12-16.*
Berntorp E. Baillieres Clin Haematol 1996; 9:259-71.

- El 80% de los hemartros en pacientes con H grave ocurren en estas 3 articulaciones



Betsy M, Gilbert MS. En: *The Haemophilic Joints: New Perspectives. Wiley*- Blackwell, Oxford, 2003:17-19.
Rodríguez-Merchán EC. En: *The Haemophilic Joints: New Perspectives, Wiley-Blackwell, 2003:73-9.*



- La hemorragia recidivante sobre una misma articulación si no recibe el tratamiento adecuado hematológico y rehabilitador, dará lugar a la **artropatía hemofílica** y sus secuelas altamente invalidantes

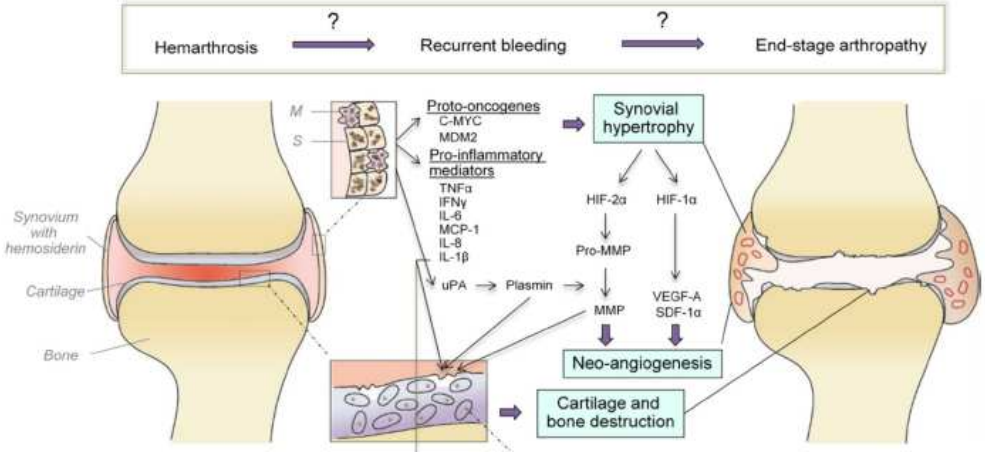
- Principal causa de morbilidad y disminución de la **calidad de vida** de los pacientes con Hemofilia grave A o B

Artropatía hemofílica

- Es la consecuencia de la extravasación repetida de sangre en las cavidades articulares y se caracteriza por dos rasgos fundamentales:

Sinovitis crónica (proliferativas)

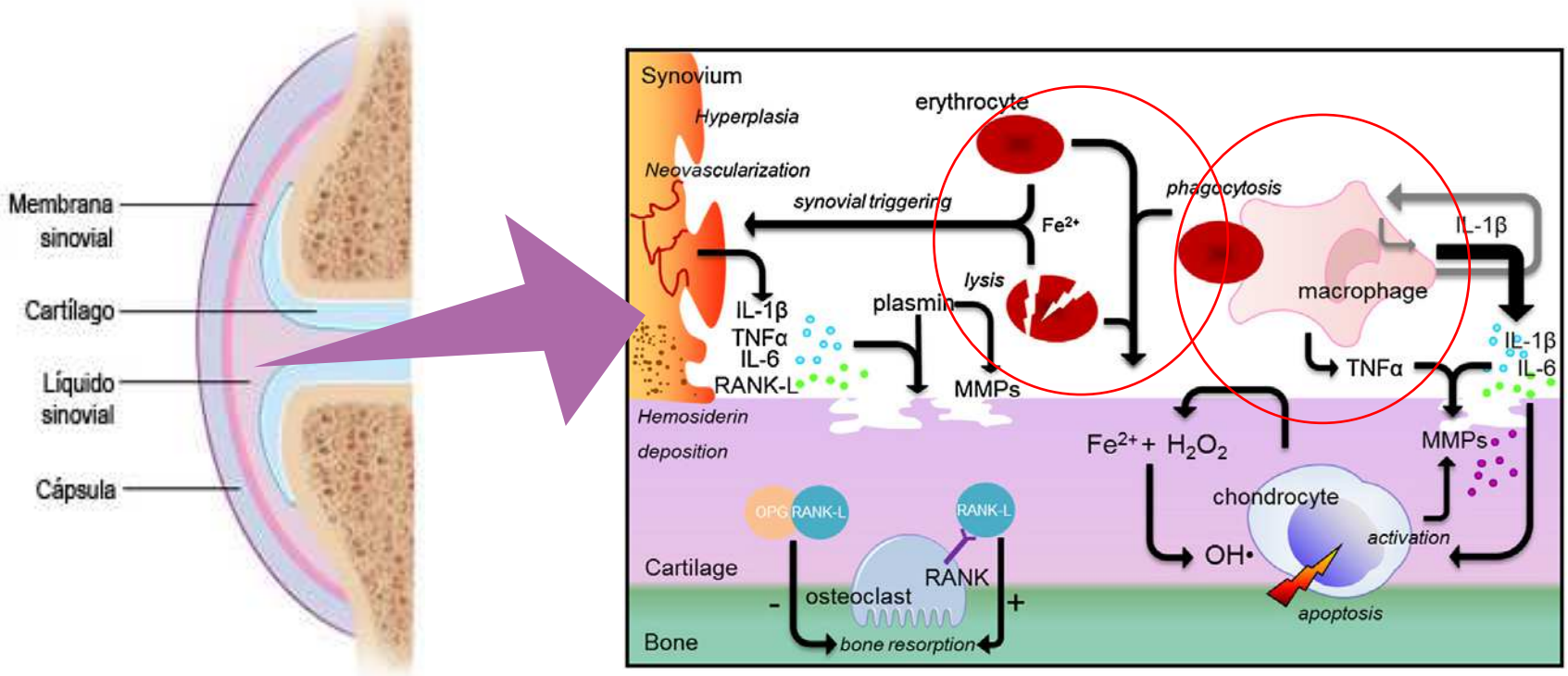
Daño osteocondral (degenerativas)



Procesos paralelos

Lafeber G et al. Haemophilia 2008; 14 (Suppl 4):3-9.
Wyseure T et al. Semin hematol 2016; 51-1:10-19

Depósitos de hierro y proceso inflamatorio!!!



Pulles AE et al. Pharmacol Res. 2017 Jan;115:192-199

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

Haemophilia (2016), 1–14 DOI: 10.1111/hae.13088

REVIEW ARTICLE

Choosing outcome assessment tools in haemophilia care and research: a multidisciplinary perspective

K. FISCHER,* P. POONNOOSE,† A. L. DUNN,‡ P. BABYN,§ M. J. MANCO-JOHNSON,¶ J. A. DAVID,** J. VAN DER NET,†† B. FELDMAN,‡‡ K. BERGER,§§ M. CARCAO,¶¶ P. DE KLEIJN,*** M. SILVA,††† P. HILLIARD,‡‡‡ A. DORIA,§§§ A. SRIVASTAVA¶¶¶ and V. BLANCHETTE¶¶ ON BEHALF OF THE PARTICIPANTS OF THE INTERNATIONAL SYMPOSIUM ON OUTCOME MEASURES IN HEMOPHILIC ARTHROPATHY[a]

Table 5. Recommended and/or mandatory outcome parameters in haemophilia according to field of use and ICF domain.

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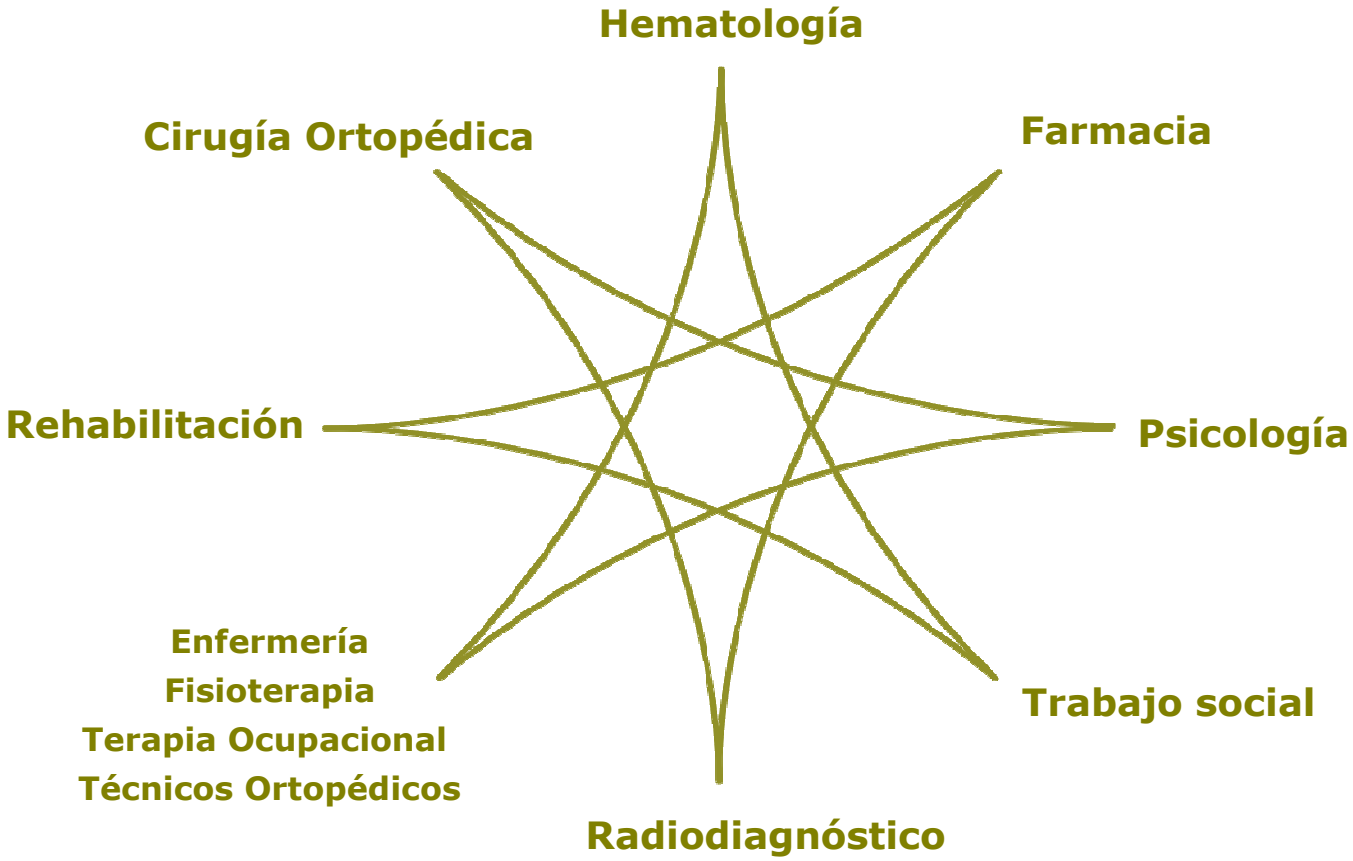
	Self-reported activities	R	R (adults) R (children)	– HAL, from age 18 upwards – pedHAL, from age 4 onwards
Participation	Days lost from school/work	M	M	
	Paid employment	M	M	Include information on full-time yes/no
Economic	Clotting factor consumption	M	M	Combine with body weight and treatment regimen
	Haemophilia-related surgeries	M	L	Not for short term studies
	Hospital visits	M	O/R	
	Days in hospital.	M	O/R	
	Utility assessment	O	R/M	Not for short term studies Choice of tariff (calculation method) affects results

M, mandatory; O, optional; R, recommended; L, limited value; U, unknown; NR, not recommended.

Results and conclusion: Recommendations for choice of outcome tools were made according to the ICF domains, economic setting, and reason for use (clinical or research). The next step will be to identify a 'core' set of outcome measures for use in clinical care or studies evaluating treatment.

Equipo multidisciplinar



Medicina Física y Rehabilitación

Especialidad MÉDICA a la que concierne la **evaluación**, **prevención, diagnóstico y tratamiento de la discapacidad**, encaminados a mantener o devolver el mayor grado de capacidad funcional e independencia posible.

Haemophilia (2008), **14** (Suppl. 3), 153–161

The role of the physiatrist in the haemophilia comprehensive care team in different parts of the world

L. HEIJNEN,*† G. DIRAT,‡ L. CHEN,§ A. B. M. TULAAR,¶ L. MOYSISYAN,** N. M. M. NASSAR†† and L. R. BATISTELLA‡‡

**Van Creveldkliniek, University Medical Centre, Utrecht, The Netherlands; †Rehabilitation Centre De Trappenberg, Huizen, The Netherlands; ‡Department of Haemophilia Physical Therapy, La Perle Cerdane, Osséja, France; §Peking Union Medical College Hospital, Beijing, China; ¶Faculty of Medicine, University of Indonesia, Jakarta, Indonesia; **Republican Pediatric Rehabilitation Centre, Yerevan, Armenia; ††Department of Physical Medicine and Rheumatology Ain Shams University, Cairo, Egypt; and ‡‡School of Medicine, University of Sao Paulo, Sao Paulo, Brazil*

The Utilization of Rehabilitation in Patients with Hemophilia A in Taiwan: A Nationwide Population-Based Study

Chien-Min Chen[1,2,3], Yao-Hsu Yang[4,5,6,7], Chia-Hao Chang[8], Chih-Cheng Chen[2,3,9]*, Pau-Chung Chen[5,10]

Citation: Chen C-M, Yang Y-H, Chang C-H, Chen C-C, Chen P-C (2016) The Utilization of Rehabilitation in Patients with Hemophilia A in Taiwan: A Nationwide Population-Based Study. PLoS ONE 11 (9): e0164009. doi:10.1371/journal.pone.0164009

The role of physical medicine and rehabilitation in haemophiliac patients

Hortensia De la Corte-Rodriguez[a] and E. Carlos Rodriguez-Merchan[b]

Physical medicine and rehabilitation aim to evaluate, diagnose and treat disability in haemophiliac patients, while preventing injury or deterioration. They also aim to maintain the greatest degree of functional capacity and independence in patients with haemophilia, or to return them to that state. Rehabilitation, together with clotting factor replacement therapy, has revolutionized the management of these patients in developed countries and reduced their morbidity/mortality rates. A knowledge of the musculoskeletal signs and symptoms of haemophilia is essential for providing a treatment which is suitable and customized. Physical medicine and rehabilitation techniques, which are based on physical means, are intended to reduce the impact which these injuries and their consequences or sequelae can have on the quality of life of patients with haemophilia. Under ideal haemostatic control conditions (primary prophylaxis), people with haemophilia could achieve good physical condition which will allow them to enjoy both physical activity and a daily life without limitations. Currently, children undergoing primary prophylaxis are quite close to this ideal situation. For these physical activities to be carried out, the safest possible situations must be sought. *Blood Coagul Fibrinolysis* 23:000–000 © 2012 Wolters Kluwer Health | Lippincott Williams & Wilkins.

Blood Coagulation and Fibrinolysis 2012, 23:000–000

Keywords: haemophilia, physical medicine, rehabilitation, treatment

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E-mail: ecrmerchan@gmx.es

Received 24 July 2012 Accepted 14 September 2012

Objetivos

Enfoque hematológico

Prevenir y tratar la hemorragia

Enfoque rehabilitador

Prevenir y tratar las consecuencias MSK de los sangrados

NO reversibilidad

Sangrado 0

Haemophilia
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Haemophilia (2014), 20, 459–463 DOI: 10.1111/hae.12375

REVIEW ARTICLE

The burden of bleeding in haemophilia: is one bleed too many?

A. GRINGERI,* B. EWENSTEIN† and A. REININGER*
*Baxter Innovations GmbH, Vienna, Austria; and †Baxter Healthcare Corporation, Westlake Village, CA, USA

Conclusion

The aspirational goal of zero bleeding episodes is conceivably attainable and undeniably critical to optimizing joint health and HRQoL in children and adults with haemophilia. Achieving this objective requires individualized, outcome-based, multidisciplinary care to maximize the effectiveness of the prophylactic regimen without increasing overall health care costs.

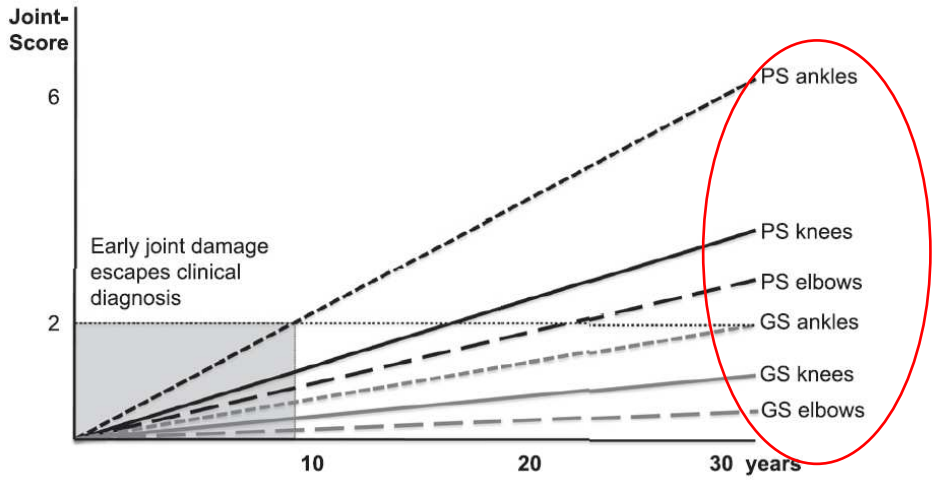
The massive proliferation of smaller blood vessels that occurs after an episode of joint bleeding may provide one explanation for subclinical bleeding. The walls of these new vessels are often defective, predisposing to the continual leakage of blood into the joint cavity [33]. Alternatively, intra-articular blood has been shown to adversely affect cartilage in the absence of inflammation [14] – in other words, without clinically evident pain and swelling.

INHERITED BLEEDING DISORDERS

Optimal treatment strategies for hemophilia: achievements and limitations of current prophylactic regimens

Johannes Oldenburg

Institute of Experimental Haematology and Transfusion Medicine, University Clinic Bonn, Bonn, Germany



prophylactic group. This study impressively demonstrated that prophylaxis is largely protective for joint disease over a 4-year period of observation. However, joint disease still occurred during this time in about 7% of the patients treated with an intensive prophylactic regimen (6000 IU/kg BW at the age of 6 years). Projected to a lifetime treatment, this means that at the age of 30 to 40 years, most hemophilia patients will suffer from some joint arthropathy.

Oldenburg J. Blood. 2015 Mar 26;125(13):2038-44.

- En la era moderna de la terapia profiláctica de reemplazo del factor de coagulación, la artropatía hemofílica no es totalmente prevenible (h objetivables y subclínicos)

- Para poder prevenir sus consecuencias es necesario un **diagnostico** precoz y certero

Aparisi F et al. Rev Iberoam Tromb Hemostasia 2001; 14:220-7.

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the Hemostasis & Thrombosis Research Society

Haemophilia (2016), **22**, 760–764 DOI: 10.1111/hae.12936

ORIGINAL ARTICLE *Musculoskeletal*

Potential biomarkers of haemophilic arthropathy: correlations with compatible additive magnetic resonance imaging scores

J. OLDENBURG,* R. ZIMMERMANN,† O. KATSAROU,‡ E. ZANON,§ E. KELLERMANN,¶ B. LUNDIN** and P. ELLINGHAUS††

Conclusions: Compatible additive MRI scores showed no clear correlations with any of the potential biomarkers for haemophilic arthropathy in the overall population. CS846 levels were significantly correlated with MRI scores in patients treated on demand.

Table 1. Biomarker levels* and correlation with compatible additive MRI score (per-protocol population).

Biomarker	Marker for	Normal range†	Biomarker levels (n = 117) Mean ± SD	Median (range)	Correlation with MRI score‡
COMP, μg mL^{-1}	Cartilage degradation	0.99–2.54	1.5 ± 0.3	1.6 (0.0–2.0)	0.100
CS846, ng mL^{-1}	Cartilage formation	§	255.3 ± 213.9	200.3 (0.0–1722.2)	0.053
CTX-I, ng mL^{-1}	Bone degradation	0.115–0.748	0.7 ± 0.6	0.6 (0.0–3.5)	–0.203
MMP3, ng mL^{-1}	Joint cartilage destruction	2.1–64.0	17.4 ± 9.4	16.6 (0.0–74.4)	–0.121
MMP9, ng mL^{-1}	Joint cartilage destruction	169–705	244.5 ± 161.8	219.1 (0.0–939.7)	0.156
TIMP-1, ng mL^{-1}	Periarticular bone loss	87–524	121.3 ± 41.2	121.8 (0.0–242.9)	0.005
VEGF, pg mL^{-1}	Inflammation and angiogenesis	62–707	226.4 ± 177.0	191.3 (0.0–845.5)	–0.084

COMP, cartilage oligomeric matrix protein; CS846, chondroitin-sulphate aggrecan turnover 846 epitope; CTX-I, C-terminal telopeptides of type I collagen; MMP, matrix metalloproteinase; MRI, magnetic resonance imaging; TIMP-1, tissue inhibitor of metalloproteinase 1; VEGF, vascular endothelial growth factor.

*Serum levels were measured for all the biomarkers except VEGF, MMP3, and MMP9, which were measured in platelet-poor plasma samples.

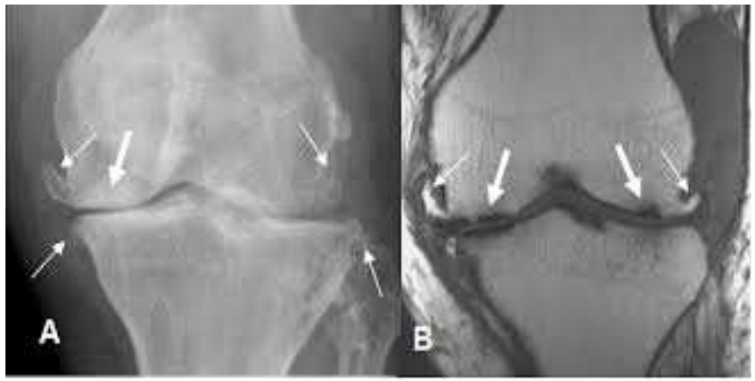
†Normal ranges for healthy patients as provided by supplier of test kits.

‡Spearman rank correlation coefficient for the correlation of biomarker and compatible additive MRI score.

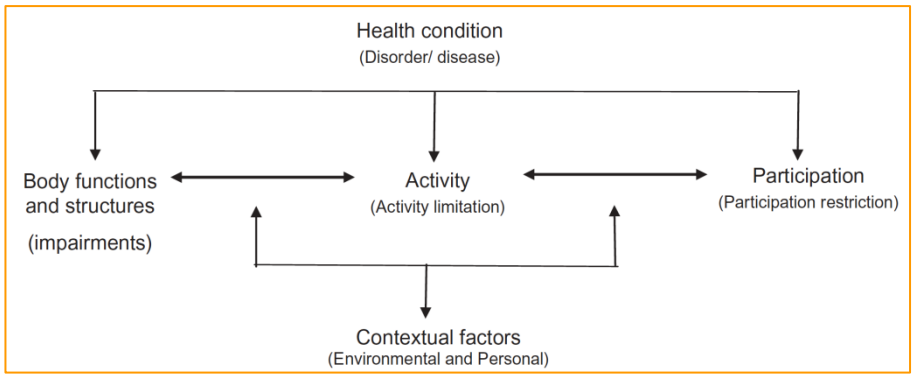
§No normal range for healthy patients was provided by supplier of test kits. Serum CS846 levels in patients with haemophilic arthropathy who had not experienced any joint bleeds in the previous 3 months have been reported [5].

EF

Imagen



Funcional



De la Corte-Rodriguez H et al. Haemophilia. 2015 Nov;21(6):723-30.

Exploración clínica

EXPLORACIÓN FÍSICA detallada a fin de conocer el estado basal y detectar posibles lesiones.

- **Análisis de 4EE :**

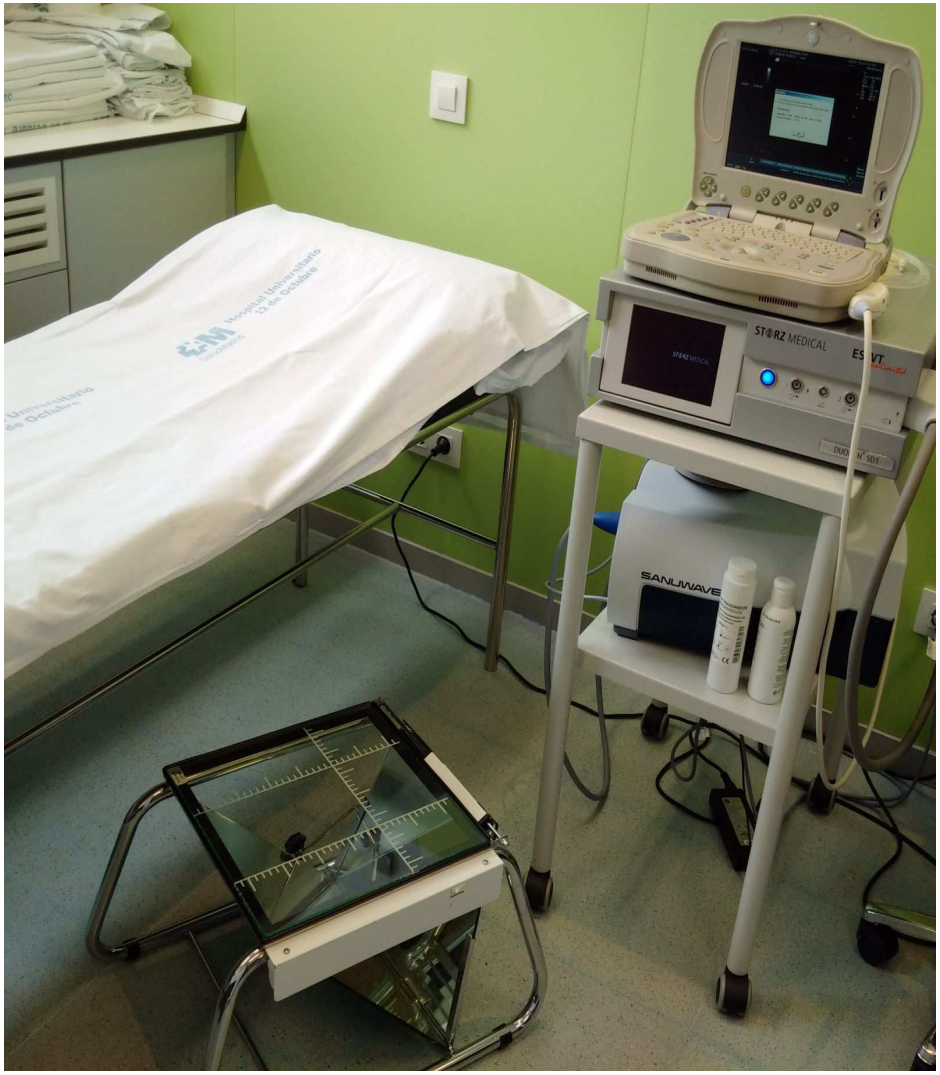
  - Dolor
  - Hinchazón
  - Balance articular (BA)
  - Trofismo muscular
  - Balance muscular (BM)
  - Estabilidad articular
  - Alineación de ejes
  - Propiocepción
  - Pisada

- **Columna vertebral:**

  - Estudio de las desviaciones axiales
  - Valoración de discrepancia MMII

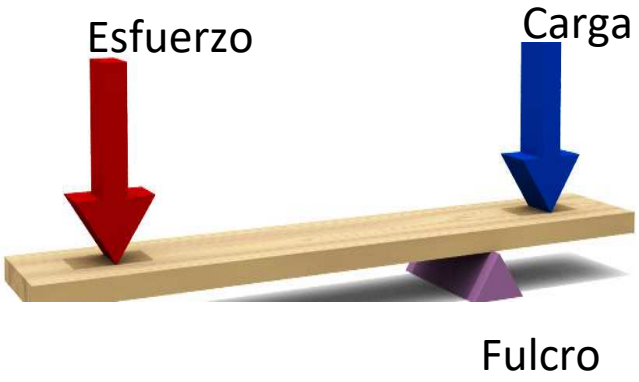
- **Análisis de la marcha**

Valoración clínica del estado clínico



Articulación = fulcro

Sistema de palancas: permite amplificar la fuerza mecánica que se aplica a un objeto



LIMITACIÓN FUNCIONAL Y DE AVD

Dolor

Balance articular

Propiocepción

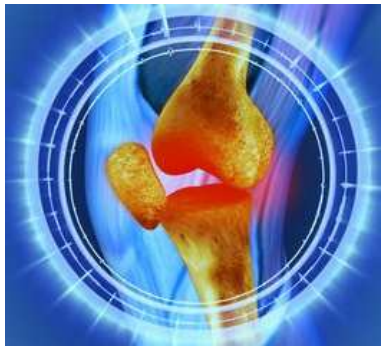
Balance muscular

Estabilidad articular

Crepitación

Pisada

Alineación de ejes



- En Hemofilia, las lesiones suelen ser **poliarticulares** e implicar graves trastornos biomecánicos que provocan impotencia funcional

- En pacientes que comenzaron a utilizar la profilaxis, la incidencia es mucho menor

- En pacientes que usaron ttos a demanda o con inhibidor, la artropatía es mucho más frecuente

ESCALAS DE VALORACIÓN FÍSICA

- Las más utilizadas:

- WORLD FEDERATION HEMOPHILIA O GILBERT (1980)

- HEMOPHILIA JOINT HEALTH SCORE (2008)

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Participation	Days lost from school/work	M	M	
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	Haemophilia-related surgeries	M	L	Not for short term studies
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	Days in hospital.	M	O/R	
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Results and conclusion: Recommendations for choice of outcome tools were made according to the ICF domains, economic setting, and reason for use (clinical or research). The next step will be to identify a 'core' set of outcome measures for use in clinical care or studies evaluating treatment.

Escala de la WFH

Escala acumulativa

Desarrollada por Gilbert

Refleja mejor el estado articular en pacientes >20 a

Puntúa de 0-1 ó de 0-2 cada uno de los hallazgos

Puntuación máxima de 12 para el tobillo y la rodilla, y de 10 para el codo

Tabla 5. Escala de evaluación clínica osteomuscular de la hemofilia de Gilbert

Inflamación	0	No
	2	Presente (añadir S en caso de sinovitis)
Atrofia muscular	0	<1 cm
	1	Presente
Deformidad axial (rodilla y tobillo)	0	0 a 7º de valgo
	1	8 a 15º de valgo ó 0 a 5º de varo
	2	>15º de valgo o >5º de varo
Crepitación	0	No
	1	Presente
Movilidad articular	0	Pérdida <10% del total del recorrido
	1	Pérdida del 10-33% del recorrido
	2	Pérdida >33% del recorrido
Contractura en flexión (irreducible)	0	<15%
	1	>15%
Inestabilidad	0	No
	1	Presente. No interfiere con la función. No precisa férula
	2	Si interfiere con la función y/o precisa férula

Gilbert MS. Semin Hematol1993; 30:(2):3-6

Escala HJHS

Hemophilia Joint Health Score 2.1 - Summary Score Sheet

	Left Elbow		Right Elbow		Left Knee		Right Knee		Left Ankle		Right Ankle	
Swelling		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE
Duration (swelling)		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE
Muscle Atrophy		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE
Crepitus on motion		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE
Flexion Loss		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE
Extension Loss		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE
Joint Pain		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE
Strength		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE		☐ NE
Joint Total												

Sum of Joint Totals ☐

+

Global Gait Score ☐

(☐ NE included in Gait items)

HJHS Total Score **=** ☐

NE = Non-Evaluable

Parece haber una buena correlación entre ambas

Escala acumulativa

Desarrollada para niños 4-18 a

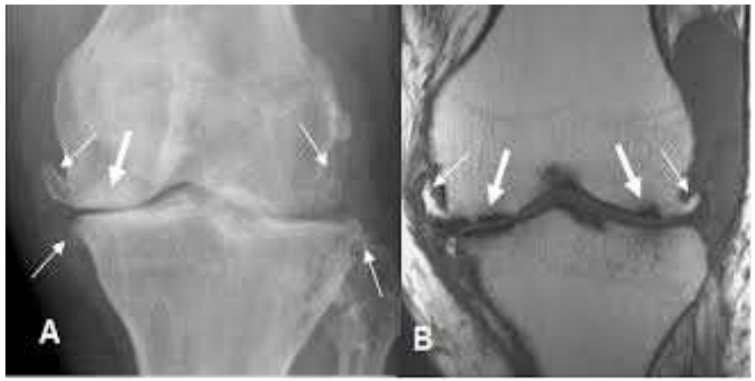
Puntúa de 0-1,2,3,4 hallazgos

Puntuación máxima de 20 por articulación y 4 para la marcha

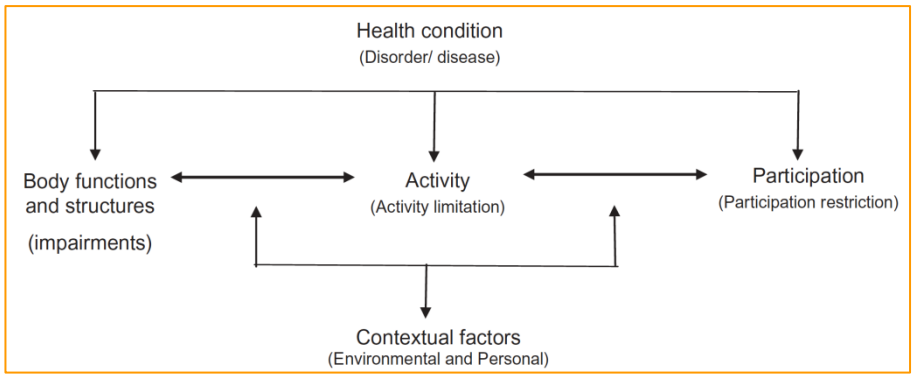
Feldman B et al. Arthritis Care & Research 2011;63:223–30

EF

Imagen



Funcional



De la Corte-Rodriguez H et al. Haemophilia. 2015 Nov;21(6):723-30.

Pruebas de imagen

- Importantes para apoyar la sospecha clínica

- Objetivo
  - Alta sensibilidad para detectar lesiones
  - Técnica sencilla
  - Permita monitorizar cambios
  - Protocolo para construir escalas

Goddard NJ et al. Haemophilia 2007; 13 (Suppl 3):14-9

- Las estrategias terapéuticas frecuentemente se dirigen al uso del factor
- Se ha demostrado que la valoración subjetiva de paciente y médico no son suficientes
- El diagnóstico por imagen permite hacer un uso racional de las terapias

Berntorp E, Shapiro AD. Lancet 2012; 379: 1447–56..
Ceponis A, Wong-Sefidan I, Glass CS, von Drygalski A. Haemophilia 2013; 19: 790–8.

- Las estrategias terapéuticas frecuentemente se dirigen al uso del factor
- Se ha demostrado que la valoración subjetiva de paciente y médico no son suficientes
- El diagnóstico por imagen permite hacer un uso racional de las terapias

- **Necesidad de complementar la valoración con pruebas de imagen !!!!**

Berntorp E, Shapiro AD. Lancet 2012; 379: 1447–56..
Ceponis A, Wong-Sefidan I, Glass CS, von Drygalski A. Haemophilia 2013; 19: 790–8.

Haemophilia
The Official Journal of the World Federation of Hemophilia,
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the Hemostasis & Thrombosis Research Society

Haemophilia (2016), 1–14 DOI: 10.1111/hae.13088

REVIEW ARTICLE

Choosing outcome assessment tools in haemophilia care and research: a multidisciplinary perspective

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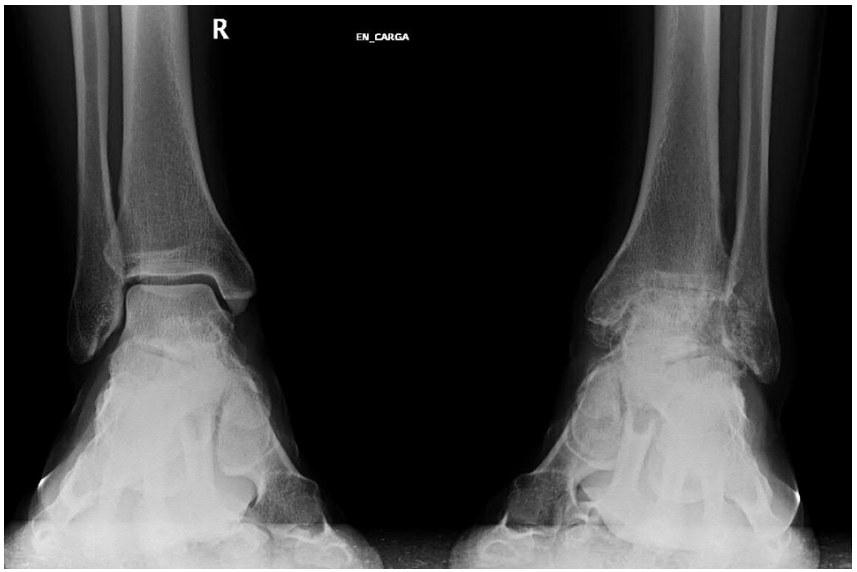
Table 5. Recommended and/or mandatory outcome parameters in haemophilia according to field of use and ICF domain.

ICF domain	Tool	Clinical	Research	Comments
Joint function and structure	Bleeding	M	M	– At least an annual review of bleeding – Reporting on periods of no less than 12 months – Use recommended definitions
	Physical examination	M	M	– HJHS v2.1 when including patients with early joint changes (all paediatric studies) – If using HJHS is impossible, collect AROM
	Imaging	O	R	– US or MRI for evaluation of early changes – Pettersson score (X-ray) for advanced osteochondral changes (interval no shorter than 3 years)
Activities	Observed activities	R	O/R	– FISH in populations with more advanced joint disease
	Self-reported activities	R	R (adults) R (children)	– HAL, from age 18 upwards – pedHAL, from age 4 onwards
Participation	Days lost from school/work	M	M	
	Paid employment	M	M	Include information on full-time yes/no
Economic	Clotting factor consumption	M	M	Combine with body weight and treatment regimen
	Haemophilia-related surgeries	M	L	Not for short term studies
	Hospital visits	M	O/R	
	Days in hospital.	M	O/R	
	Utility assessment	O	R/M	Not for short term studies Choice of tariff (calculation method) affects results

M, mandatory; O, optional; R, recommended; L, limited value; U, unknown; NR, not recommended.

Results and conclusion: Recommendations for choice of outcome tools were made according to the ICF domains, economic setting, and reason for use (clinical or research). The next step will be to identify a 'core' set of outcome measures for use in clinical care or studies evaluating treatment.

Radiografía



Radiología simples

Ventajas:

- Fácilmente disponibles, proyecciones estándar y sus resultados son prácticamente instantáneos

- Radiografía digital es más sensible que la radiología convencional

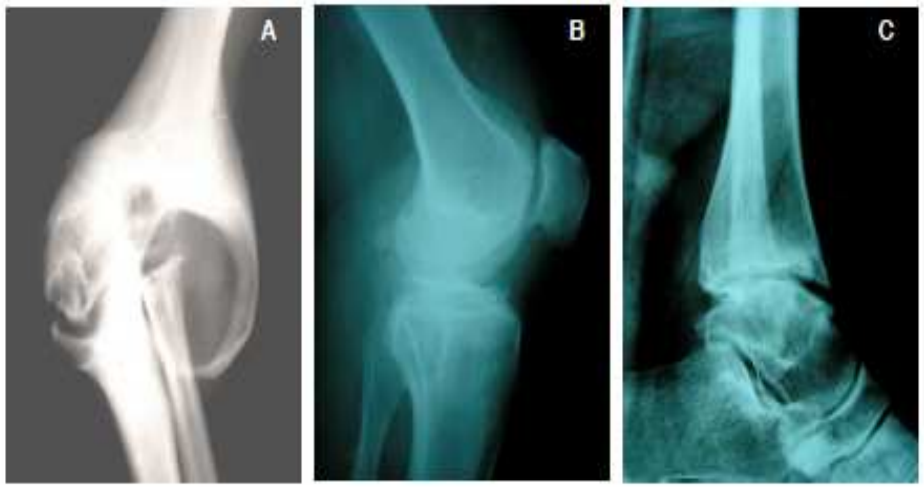
Arnold WD, Hilgartner MW. J Bone Joint Surg Am 1977; 59:287-305.

Pettersson H, Ahlberg A, Nilsson IM. Clin Orthop Relat Res 1980; 149:153-9.

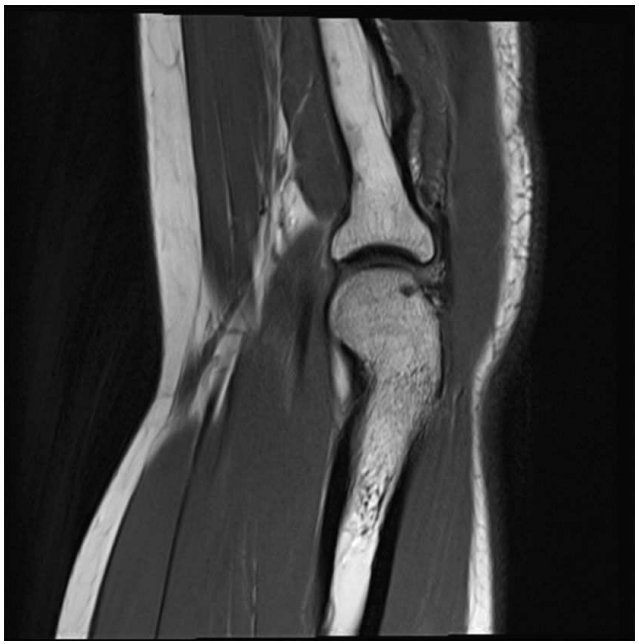
Radiología simples

Limitaciones:

- Subestima el grado de patología articular real en fases iniciales y ppbb
- Su uso más apropiado es en pacientes adultos con enfermedad articular avanzada o sospecha de lesión ósea



Resonancia Magnética



Resonancia Magnética

Ventajas:

- Tiene una alta resolución espacial y capacidad para distinguir los elementos intra y extraarticulares
- RM es útil para detectar microhemorragias
- Grandes avances por mejoras en antenas y secuencias, lo que permite una mayor resolución y un mejor contraste tisular

Nuss R et al. Haemophilia 2000; 6:162–9 (Denver)
Funk MB et al. Haemophilia 2002; 8:98-103
Soler R et al. Eur Radiol 2002; 12: 836–43
Dobón M et al. Haemophilia 2003; 9:76–85
Lundin B et al. Haemophilia 2004; 10:383–9 (Europea)
Lundin B et al. Haemophilia 2005; 11:109–15

Doria AS et al. Haemophilia 2010;16 (5):107–14
Pergantou H et al. Haemophilia 2006;12:241-7

Resonancia Magnética

Denver (10p) Europea (20p)

Table 3. The compatible scales for progressive and additive MRI assessments.

	Progressive scale (P)	Additive scale (A)
Effusion/haemarthrosis (IPSG)		
Small	(1)___	
Moderate	(2)___	
Large	(3)___	
Synovial Hypertrophy		
Small	(4)___	(1)___
Moderate	(5)___	(2)___
Large	(6)___	(3)___
Haemosiderin		(1)___
Small	(4)___	
Moderate	(5)___	
Large	(6)___	
Changes of subchondral bone or joint margins		
Any surface erosion	(7)___	(1)___
Any surface erosion in at least two bones		(1)___
Half or more of the articular surface eroded in at least one bone	(8)___	(1)___
Half or more of the articular surface eroded in at least two bones		(1)___
At least one subchondral cyst	(7)___	(1)___
More than one subchondral cyst	(8)___	(1)___
Subchondral cysts in at least two bones		(1)___
Multiple subchondral cysts in each of at least two bones		(1)___
Cartilage loss		
Any loss of joint cartilage height	(9)___	(1)___
Any loss of joint cartilage height in at least two bones		(1)___
Any loss of joint cartilage height involving more than one third of the joint surface in at least one bone		(1)___
Any loss of joint cartilage height involving more than one third of the joint surface in at least two bones		(1)___
Full-thickness loss of joint cartilage in at least some area in at least one bone	(10)___	(1)___
Full-thickness loss of joint cartilage in at least some area in at least two bones		(1)___
Full-thickness loss of joint cartilage involves at least one third of the joint surface in at least one bone		(1)___
Full-thickness loss of joint cartilage involves at least one third of the joint surface in at least two bones		(1)___
	Highest number (max value = 10)	Add numbers (max value = 20)
Scores	(P)___	(A)___

Nuss R et al. Haemophilia 2000; 6:162–9
Lundin B et al. Haemophilia 2005; 11:109–15

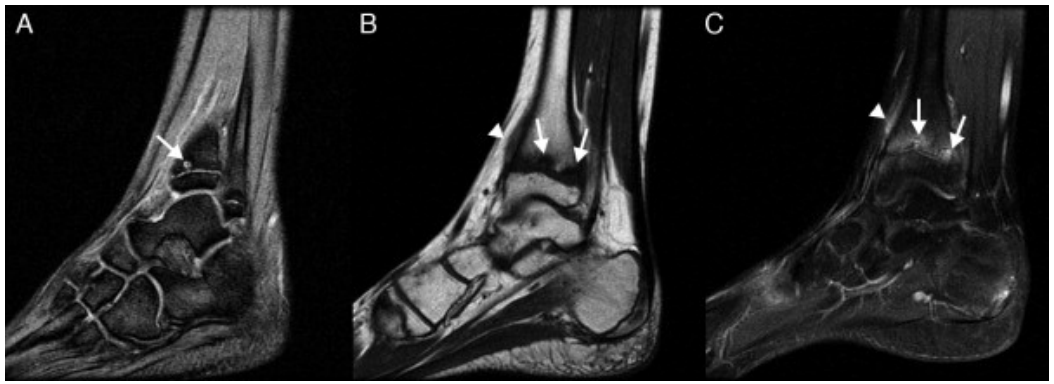
Resonancia Magnética

Limitaciones:

- Sin embargo, la RM es más costosa, menos accesible y requiere más tpo

- En niños suele requerir sedación

- Aunque la objetividad es extraordinaria, resulta utópico poder realizar estudios seriados de RM, por el coste económico



Doria AS et al. Haemophilia 2010;16 (5):107–14
Pergantou H et al. Haemophilia 2006;12:241-7

Ecografía

Ecografía

- El US MSK, ha demostrado su utilidad como herramienta para el diagnóstico MSK en la hemofilia
- En cuanto a la detección de hipertrofia sinovial y daño del cartílago, así como los episodios de hemartrosis

Klukowska A et al. Haemophilia 2001; 7: 286-92,
Doria AS et al. AJR Am J Roentgenol 2015; 204: W336-47.
Martinoli C et al. Thromb Haemost 2013; 109: 1170-9.
Melchiorre D et al. Haemophilia 2011; 17: 112-7.
Muça-Perja M et al. Haemophilia 2012; 18: 364-8.
Zukotynski K et al. Haemophilia 2007; 13: 293-304.
Querol F, Rodríguez-Merchán EC. Haemophilia 2012; 18, e215–e226

Klukowska A et al. Haemophilia 2001; 7: 286-92,

Recuerdo histórico

- En el año 1992, nuestra Unidad publica el primer estudio comparativo, concluyendo la utilidad del US en los tres aspectos

Table 1. References evidencing the use of ultrasonography for the diagnosis and/or treatment of musculo-skeletal lesions in haemophilia.

				USG	Identifies / points at value for:						
Year	Authors	Objective	Cases	Available, fast, effective, safe, cheap technique	Effusion	Synovial	Arthrosic disease	Haematoma	Pseudotumour	Therapy control	Remarks/conclusions
1992	Daly BD [17]	Diagnosis of psoas haematoma	Yes	-	Yes	-	-	Yes	-	-	USG choice in children. CT and MRI complete the study.
1992	Hermann G [18]	Describe techniques	Yes	Yes	-	-	-	Yes	Yes	-	USG method of choice, diagnosis and
1992	Merchan ECR *et al.* [19]	Diagnosis of arthrosic disease	Yes	Yes	Yes	Yes	Yes	-	-	-	USG simple, effective technique diagnosis and/or control of effusion, synovitis and early stages of arthrosic disease.
	Merchan EC [20]	pseudotumour									of pseudotumour.
2001	Klukowska A *et al.* [14]	Protocol description	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	USG useful for effusion, synovial, and cartilage. Synovitis involves therapy with factor. MRI best technique, but not useful for routine. Good USG-MRI correlation.
2005	Balkan C *et al.* [21]	Diagnosis of psoas haematoma	Yes	Yes	-	-	-	Yes	-	-	USG useful for psoas haematoma. CT and MRI best techniques, but practical disadvantages.
2007	Goddard NJ *et al.* [22]	Diagnosis of synovitis	-	Yes	Yes	Yes	Yes	-	-	-	USG particularly useful in effusion and synovitis. MRI best technique for diagnosis of arthrosic disease.
2007	Zukotynski K *et al.* [15]	Protocol description	-	Yes	Yes	Yes	Yes	-	-	Yes	USG useful for diagnosis and assessment of therapeutic effects. Very complete protocol.
2008	Acharya SS *et al.* [23]	Comparative efficacy study of images	Yes	Yes	Yes	Yes	Yes	-	-	Yes	USG particularly important for diagnosis of synovitis.
2008	Acharya SS [7]	Study of diagnostic approaches	-	Yes	-	Yes	Yes	-	-	Yes	USG useful in synovitis. MRI best technique, but practical disadvantages.
2008	Antunes SV *et al.* [24]	Evaluate USG for diagnosis of arthrosic disease	Yes	Yes	Yes	Yes	Yes	-	-	-	USG useful in diagnosis of arthrosic disease.
2008	Mauser-Bunschoten EP *et al.* [25]	Describe techniques	-	Yes	Yes	Yes	-	-	-	-	USG useful in synovitis. MRI best technique, but practical disadvantages.
2008	Querol F *et al.* [26]	Description of exploratory protocol	Yes	-	Yes	Yes	-	-	-	Yes	USG useful for diagnosis and control of haemarthrosis.
2009	Jelbert A *et al.* [8]	Description of imaging techniques for arthrosic disease	-	Yes	Yes	Yes	Yes	-	-	Yes	USG useful in synovitis. Limitations for arthrosic disease. MRI best technique.
2009	Keshava S *et al.* [9]	Protocol description	-	-		Yes	Yes	-	-		USG proposes protocol standardisation.
2009	Robertson JD [27]	Description of case report	Yes	Yes	Yes	-	Yes	-	-	Yes	USG useful for evaluating joint effusion and/or suction guide.

Merchan EC, De Orbe A, Gago J. Acta Orthop Belg. 1992;58(2):122-5.
Querol F, Rodriguez-Merchan EC. Haemophilia ;2012:18, e215–e226

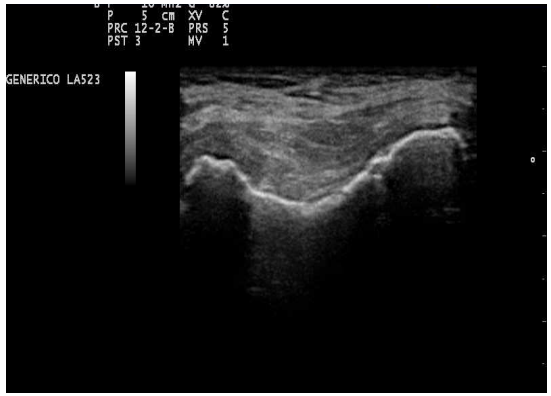
HEMOFILIA

Desde entonces, y especialmente en los últimos años, se han multiplicado las publicaciones sobre el uso de la ecográfia MSK en hemofilia

Ecografía

Ventajas:

- Fácilmente disponible, económica y no invasiva
- Pertinente en los estadios precoces de la enfermedad ya que puede detectar cambios incipientes
- Permite determinar la necesidad de otras pruebas de imagen.



Strike KL et. Haemophilia. 2015 Nov;21(6):828-31
Martinoli C et al. Haemophilia. 2016 Jan;22(1):20-1
Kidder W et al. Haemophilia. 2015 Jul;21(4):530-7

Ecografía

Limitaciones:

- Imposibilidad de valorar simultáneamente el hueso
- Es necesaria una metodología bien establecida
- Precisa un evaluador experimentado
- Variabilidad interobservador

Diferentes protocolos

Se han publicado múltiples sistemas de puntuación estandarizados con el objetivo de estandarizar mediciones y minimizar en gran medida la dependencia del operador.

Klukowska et al. Haemophilia 2001;7:286-92
Zukotynski K et al Haemophilia 2007;13:293–304
Querol F et al. Haemophilia 2008; 14 (6):36-44
Keshava S et al. Haemophilia 2009;1: 1159–79
Melchiorre et al. Haemophilia 2011,17:112-7
Xavier F et al. Rheumatol Curr Res 2012, S2-S9
Martinoli C et al. Thromb Haemost 2013;109(6):1170-9
Doria As et al. AJR AM J Roentgenol 2015;204(3):W336-47.

Clasificados como :

1.Protocolos de US de art. periférica completa (**FPJ-US**), que son más exhaustivos y realizados por expertos en radiología.

2.Protocolos de US de punto de atención (**POC-US**), traída al paciente y realizada por el médico en tiempo real, para proporcionar una respuesta rápida en la práctica clínica y facilitar la toma de decisiones.

No son comparables

Foppen W et al. Haemophilia. 2017 Sep;23(5):645-647

HEAD-US

Haemophilia Early Arthropathy Detection with UltraSound

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]

Thrombosis and Haemostasis 109.6/2013

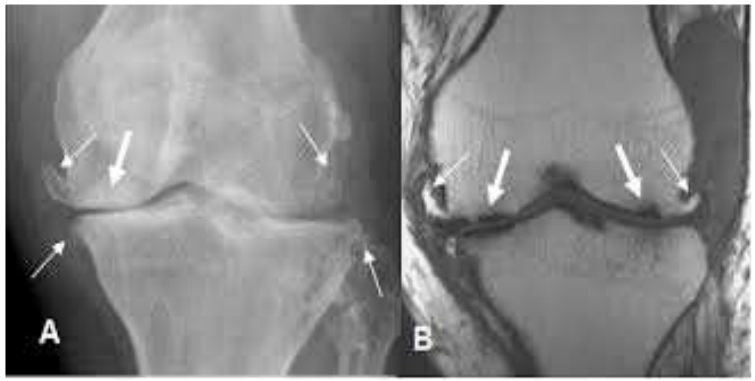
- Único desarrollado para POC-US
- Basado en un consenso de expertos multidisciplinar
- Describe la sistemática de exploración de codo, rodilla y tobillo

Recesos articulares	→	**Hemartros, Sinovitis**
Superficie articular	→	**Daño condral y subcondral**

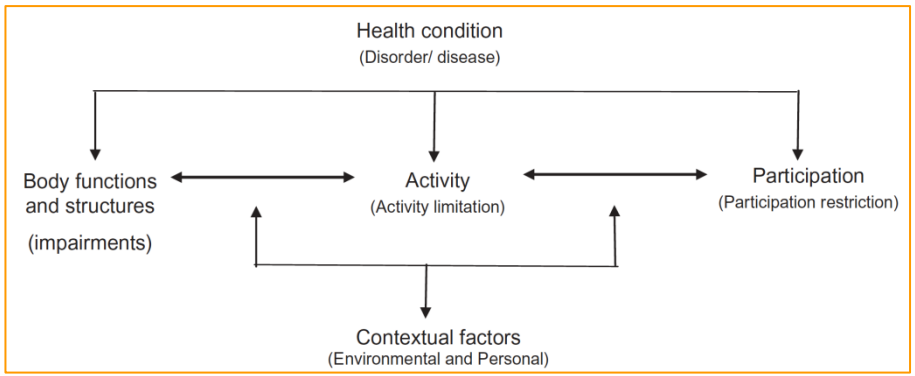
Martinoli C et al. Thromb Haemost. 2013;4:109(6)

EF

Imagen



Funcional



De la Corte-Rodriguez H et al. Haemophilia. 2015 Nov;21(6):723-30.

Valoración funcional

FISH

Eating and grooming
Bathing
Dressing
Transfers-sitting
Transfers-squatting
Going up a step
Walking
Running

HAL

Lying/sitting/kneeling/standing
Functions of the legs
Functions of the arms
Use of transportation
Self-care
Household tasks
Leisure activities and sports

Se recomienda usar los dos instrumentos complementarios.
Medir diferentes dominios

Functional Independence Score in Hemophilia (FISH)

FUNCTIONAL INDEPENDENCE SCORE IN HEMOPHILIA (FISH)

Performance based instrument

Patient Name:	Patient Code:
	Today (dd/mm/yyyy):___/___/____.
A.Self Care	
1. Eating and grooming	○ 1 ○ 2 ○ 3 ○ 4
2. Bathing	○ 1 ○ 2 ○ 3 ○ 4
3. Dressing	○ 1 ○ 2 ○ 3 ○ 4
B. Transfers	
4. Chair	○ 1 ○ 2 ○ 3 ○ 4
5. Squatting	○ 1 ○ 2 ○ 3 ○ 4
C. Locomotion	
6. Walking	○ 1 ○ 2 ○ 3 ○ 4
7. Stairs (12 - 14 steps)	○ 1 ○ 2 ○ 3 ○ 4
8. Running	○ 1 ○ 2 ○ 3 ○ 4
Total Score	

Scores range from 1 - 4 depending on the degree of independence (see scoring key)

Comments:

4. The subject is able to perform the activity without any difficulty like other healthy peers.
3. The subject is able to perform the activity without aids or assistance, but with slight discomfort. He is unable to perform the activity like his healthy peers.
2. The subject needs partial assistance/ aids/ modified instruments/ modified environment to perform the activity.
1. The subject is unable to perform the activity, or needs complete assistance to perform the activity.

No requiere validación transcultural porque no es autoadministrable.

Poonnoose PM et al. Haemophilia. 2005 Nov;11(6):598-602.

Haemophilia Activities List (HAL)

Requiere a validación transcultural

Lying down/ sitting / kneeling / standing

In the previous month, did you have any

Sitting down
(e.g. on a chair or couch)

Rising from a chair with armrests

Rising from a chair without armrests

Kneeling / squatting

Bending forward

Kneeling for a longer period of time

Squatting for a longer period of time

Standing for a longer period of time

Functions of the legs

In the previous month, did you have any difficulty, due to hemophilia, with:

	n/a	Impossible	Always	Mostly	Sometimes	Rarely	Never
Walking short distances (less than 1 kilometer / 15 minutes)	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Walking long distances (more than 1 kilometer / 15 minutes)	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Walking on a soft surface (e.g. on the beach or through the woods)	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Walking on an uneven surface (e.g. cobblestones, high sidewalks)	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Strolling / (window-)shopping	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Climbing up the stairs	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Climbing down the stairs	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Running (e.g. in order to catch the bus)	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Jumping	☐8	☐1	☐2	☐3	☐4	☐5	☐6

Functions of the arms

In the previous month, did you have a

Lifting heavy objects

Carrying heavy objects in the arms

Fine hand movements
(e.g. closing buttons)

Reaching above your head (to pick something up from a high shelf)

Use of transportation

In the previous month, did you have

Riding a bicycle

Getting in and out of a car

Using public transportation
(bus, train, subway)

Self care

In the previous month, did you

Drying your whole body

Putting on a shirt, sweater etc.

Putting on sock and shoes

Putting on a tie or closing the top button of a shirt

Going to the toilet

Household tasks

In the previous month, did you have any difficulty, due to hemophilia, with:

	n/a	Impossible	Always	Mostly	Sometimes	Rarely	Never
Going out shopping (for food, drink etc.)	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Washing the dishes, cleaning the sink	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Cleaning the house	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Other household tasks (ironing, making the beds)	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Doing odd jobs (both in and around the house)	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Gardening	☐8	☐1	☐2	☐3	☐4	☐5	☐6

Leisure activities and sports

In the previous month, did you have any difficulty, due to hemophilia, with:

	n/a	Impossible	Always	Mostly	Sometimes	Rarely	Never
Playing games (outdoors, e.g. with your children)	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Sports	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Going out (theatre / museum / movie theatre / bar)	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Hobbies	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Dancing	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Going on a holiday (active)	☐8	☐1	☐2	☐3	☐4	☐5	☐6
Going on a holiday ("passive"; beach-/hotel holiday)	☐8	☐1	☐2	☐3	☐4	☐5	☐6

van Genderen FR et al. Haemophilia. 2006 Jan;12(1):36-46.

Pediatric HAL (PedHAL)

PedHAL

Ssitting/kneeling/standing
Functions of the legs
Functions of the arms
Use of transportation
Self-care
Household tasks
Leisure activities and sports

En proceso de validación

3. Valoración funcional

*

Health Assessment Questionaire (HAQ)

	Durante la **última semana**, ¿ha sido usted capaz de…	Sin dificultad	Con alguna dificultad	Con mucha dificultad	Incapaz de hacerlo
Vestirse y asearse	1) Vestirse solo, incluyendo abrocharse los botones y atarse los cordones de los zapatos?	☐	☐	☐	☐
	2) Enjabonarse la cabeza?	☐	☐	☐	☐
Levantarse	3) Levantarse de una silla sin brazos?	☐	☐	☐	☐
	4) Acostarse y levantarse de la cama?	☐	☐	☐	☐
Comer	5) Cortar un filete de carne?	☐	☐	☐	☐
	6) Abrir un cartón de leche nuevo?	☐	☐	☐	☐
	7) Servirse la bebida?	☐	☐	☐	☐
Caminar	8) Caminar fuera de casa por un terreno llano?	☐	☐	☐	☐
	9) Subir cinco escalones?	☐	☐	☐	☐
Higiene	10) Lavarse y secarse todo el cuerpo?	☐	☐	☐	☐
	11) Sentarse y levantarse del retrete?	☐	☐	☐	☐
	12) Ducharse?	☐	☐	☐	☐
Alcanzar	13) Coger un paquete de azúcar de 1 Kg de una estantería colocada por encima de su cabeza?	☐	☐	☐	☐
	14) Agacharse y recoger ropa del suelo?	☐	☐	☐	☐
Prensión	15) Abrir la puerta de un coche?	☐	☐	☐	☐
	16) Abrir tarros cerrados que ya antes habían sido abiertos?	☐	☐	☐	☐
	17) Abrir y cerrar los grifos?	☐	☐	☐	☐
Otras	18) Hacer los recados y las compras?	☐	☐	☐	☐
	19) Entrar y salir de un coche?	☐	☐	☐	☐
	20) Hacer tareas de casa como barrer o lavar los platos?	☐	☐	☐	☐

Otras genéricas: HAQ, I Katz, I. Barthel, FIM

De la Corte-Rodriguez H et al. Thromb Res. 2017;153:51-56.

Calidad de vida

Es una construcción multidimensional que incluye el bienestar físico, emocional y social

Table 1. Overview of QoL instruments for use in patients with haemophilia.

Type of instrument*	Target age group	
	Children	Adults
Generic	KINDL [19]	Nottingham Health Profile (NHP) [20]
	CHQ [21]	Quality of Well-being Index (QWB) [22]
	TACQOL [23]	Sickness Impact Profile (SIP) [24]
	PedsQL [25]	SF-36 Health Survey (SF-36) [26]
	–	EQ-5D [27]
	–	WHOQoL Questionnaire [28]
Haemophilia-specific*	Haemo-QoL [29]	Mediap [30]
	CHO-KLAT [31]	Haem-A-QoL [32]
	HemDux [33]	Hemofilia-QoL [34]
	QoL for young children [35]	Hemolatin-QoL [36]
	QUAL-Hemo [37]	QUAL-Hemo [37]

*Only two of the haemophilia-specific quality of life (QoL) instruments include a dimension to assess sports activities: the paediatric instrument Haemo-QoL and the adult instrument Haem-A-QoL.

Table 11.1 Studies assessing HRQoL with disease-specific measures in hemophilia

Year	Authors	Country	Participants	Sample (*n*)	Measure
2000	Bullinger et al. [6]	6 European countries	Children/adolescents and parents	58*	Haemo-QoL
2004	v. Mackensen et al. [7]	6 European countries	Children/adolescents and parents	339#	Haemo-QoL
2004	Young et al. [8]	Canada	Children/adolescents and parents	50**	CHO-KLAT
2004	Arranz et al. [9]	Spain	Adults	73**	Hemofilia-QoL
				35*	(version 1.0)
2005	Remor et al. [5]	8 Latin-American countries	Adults	50**	Hemolatin-QoL
2005	Remor et al. [10]	Spain	Adults	121#	A36 Hemofilia-QoL
2005	v. Mackensen et al. [11]	Italy	Adults	233#	Hemo-A-QoL
2006	Gueróis et al. [12]	France	Children/adolescents and parents, adults	70**	QUAL HEMO
2008	Rentz et al. [13]	Germany, Spain, USA, Canada	Adults	221#	Haemo-QoL-A

*Pilot testing, psychometric; **Development study; #Psychometric field study. CHO-KLAT (Canadian Haemophilia Outcomes – Kids Life Assessment Tool); Haemo-QoL (Children's Haemophilia Quality-of-life questionnaire); Haemo-QoL-A (Adults Haemophilia Quality-of-life questionnaire) A36 Hemofilia-QoL (Adults Haemophilia Quality of Life questionnaire); HemoLatin-QoL (Adults Haemophilia Quality of Life questionnaire); Hemo-A-QoL (Hemophilia QoL questionnaire for adults).

Table 11.2 HRQOL instruments for hemophilia available

Children (C/P version)	Age groups	Adults	Age focus
Cho-KLAT	4–7 (C/P), 8–12 (C/P), 13–17 (C/P)	A36 Hemofilia-QoL	> 17 years old
Haemo-QoL	4–7 (C/P), 8–12 (C/P), 13–17 (C/P)	Haemo-QoL-A	> 18 years old
QUAL HEMO	2–12 (C/P), 13–17	QUAL HEMO	> 18 years old
		Haem-A-QoL	> 18 years old
		HemoLatin-QoL	> 17 years old

C/P, Child/parents.

Quality of Life in Hemophilia. En: *Current and Future Issues in Hemophilia Care*, First Edition. Edited by EC Rodrıguez-Merchan and LA Valentino. 2011 JohnWiley & Sons, Ltd. Published 2011 by Blackwell Publishing Ltd.

Tabla 3. Instrumentos utilizados en investigación para medir la CV en pacientes con hemofilias adultos

Instrumento	Autor(es)	Versiones y año de publicación	Objetivo	Población	Estructura	Específico para hemofilia	Confiabilidad y/o validez	Referencia o acceso
SF-36 (The Short Form Health Survey)	Ware y Sherbourne (1992)	Versión 1: 1992	Evaluar CV relacionada con la salud	Desde los 14 años en adelante	36 ítem y 8 dimensiones: 1) Función física; 2) rol físico; 3) dolor corporal; 4) salud general; 5) vitalidad; 6) funcionamiento social; 7) rol emocional y 8) salud mental	No	Consistencia interna= 0,76 para la dimensión de funcionamiento social y 0,8 para el resto de las dimensiones.	Ware, J.E., & Sherbourne, C.D. (1992). The MOS 36-Item Short-Form Health Survey (SF-36®): I. conceptual framework and item selection. *Med Care*, 30(6), 473-83.
		Versión 2: 1996						http://www.sf-36.org/tools/sf36.shtml#MODEL
	Alonso, Prieto & Antó (1995)	Adaptación española: 1995						Alonso, J., Prieto, L., y Antó, J.M. (1995). La versión española del 'SF-36® Health Survey' (Cuestionairo de Salud SF-36®): un instrumento para la medida de los resultados clínicos. *Med Clin (Barc)*, 104(20), 771-6.
A36 Hemofilia-QoL	Arranz y Remor	Única versión: 2005	Evaluar la CV en adultos con hemofilia	Mayores de 17 años	36 ítems que cubren 9 dominios: salud física, actividades diarias, daño articular, dolor, satisfacción con el tratamiento, dificultades con el tratamiento, funcionamiento emocional, salud mental, relaciones y actividad social.	Sí	confiabilidad de 0,95 y validez con un rango entre 0.49 y 0.77 según la sub-escala	**Referencias del instrumento original:** Remor, E., Arranz, P., Quintana, M., Villar, A., Jiménez-Yuste, V., Díaz, J.L., … Hemofilia-QoL Project Group. (2005). Psychometric field study of the new haemophilia quality of life questionnaire for adults: the Hemofilia-QoL. *Haemophilia*, 11(6), 603-10. Arranz, P., Remor, E., Quintana, M., Villar, A., Díaz, J.L., Moreno, M., … Hemofilia-QoL Group. (2004). Development of a new disease-specific quality-of-life questionnaire to adults living with haemophilia. *Haemophilia*,10(4), 376-82
Hemolatin qol - Cuestionario Latinoamericano Para la Evaluacion de la CV en Adultos con Hemofilia	Remor	2002	Evaluar la CV en adultos con hemofilia	Mayores de 18 años	47 ítems que evalúan: funcionamiento físico; funcionamiento emocional; apoyo social percibido; actividades y funcionamiento social; tratamiento médico; salud mental; satisfacción con las condiciones del entorno; bienestar general; salud general.	Sí	No hay datos	Remor, E. (2005). Desarrollo de una medida específica para la Evaluación de la CV en Pacientes Adultos Viviendo con Hemofilia en América-Latina: el Hemolatin-QoL. *Revista Interamericana de Psicología*, 39(2), 211-220. Proyecto de investigación en desarrollo. http://hemolatin-qol.info/

Table 2. Common domains of haemophilia-specific quality of life (QoL) questionnaires for adult patients.

	Specific domains		
Aspects	Medtap	Haem-A-QoL	Hemofilia-QoL
Physical	Physical functioning	Physical health	Physical health
	–	Sports and leisure	Joint damage
	–	–	Pain
Functional	Role functioning	Work and school	Daily functioning
Social	Psychosocial-related	Family planning	Relationship and social activities
	–	Partnership and sexuality	–
Emotional	Fear/worry	Feeling	Emotional functioning
	Positive effect	View	–
	–	Future	–
Mental	–	–	Mental health
Treatment and disease	Treatment worry	Treatment	Treatment satisfaction
	–	Dealing	Treatment difficulties
No. of domains	6	10	9
No. of items	46	46	36

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

Haemophilia (2015), **21**, 430–435 DOI: 10.1111/hae.12731

REVIEW ARTICLE

Assessments of outcome in haemophilia – what is the added value of QoL tools?

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In conclusion, prospective systematic assessment of outcome of therapeutic interventions in haemophilia and related bleeding disorders is important. HTCs need to allocate resources to develop capacities to use appropriate tools that will provide relevant data. The usefulness or additional contribution of HRQoL data to those obtained by the more direct assessment of joint health, activities and participation is unclear at this time.

TRAS LA VALORACIÓN…

No lesiones / situaciones de riesgo → Prevención 1ª

→ Revaloración

TRAS LA VALORACIÓN…

No lesiones / situaciones de riesgo → Prevención 1ª

→ Revaloración

Detección de lesiones MSK → Prevención 2ª

→ Tratamiento

Prevención de lesiones

LA PREVENCIÓN

de las alteraciones del aparato locomotor en el niño durante la etapa del desarrollo osteo-articular

Mejora la calidad de vida niño / familia

Prevención de lesiones

Uso profiláctico de factor deficitario

Evitar sobrepeso

Higiene postural

Reparto de cargas

Alineación de ejes

Consejos sobre calzado

Desarrollo psicomotor

Acondicionamiento físico

Actividad física y deporte

- En las últimas décadas, el incremento en la disponibilidad y seguridad de los concentrados de factor de la coagulación, ha permitido disminuir las restricciones físicas.

MODALIDAD PROFILÁCTICA CON FACTOR

Beneficios

- Existe un amplio reconocimiento de los beneficios que trae aparejada la actividad física y el ejercicio desde varios puntos de vista

Físico

Socioafectivo

Psíquico

Tratamiento de lesiones

El equipo multidisciplinar aplicará el tratamiento más conveniente en cada caso:

- Tto hematológico
- Tto rehabilitador
- Tto quirúrgico

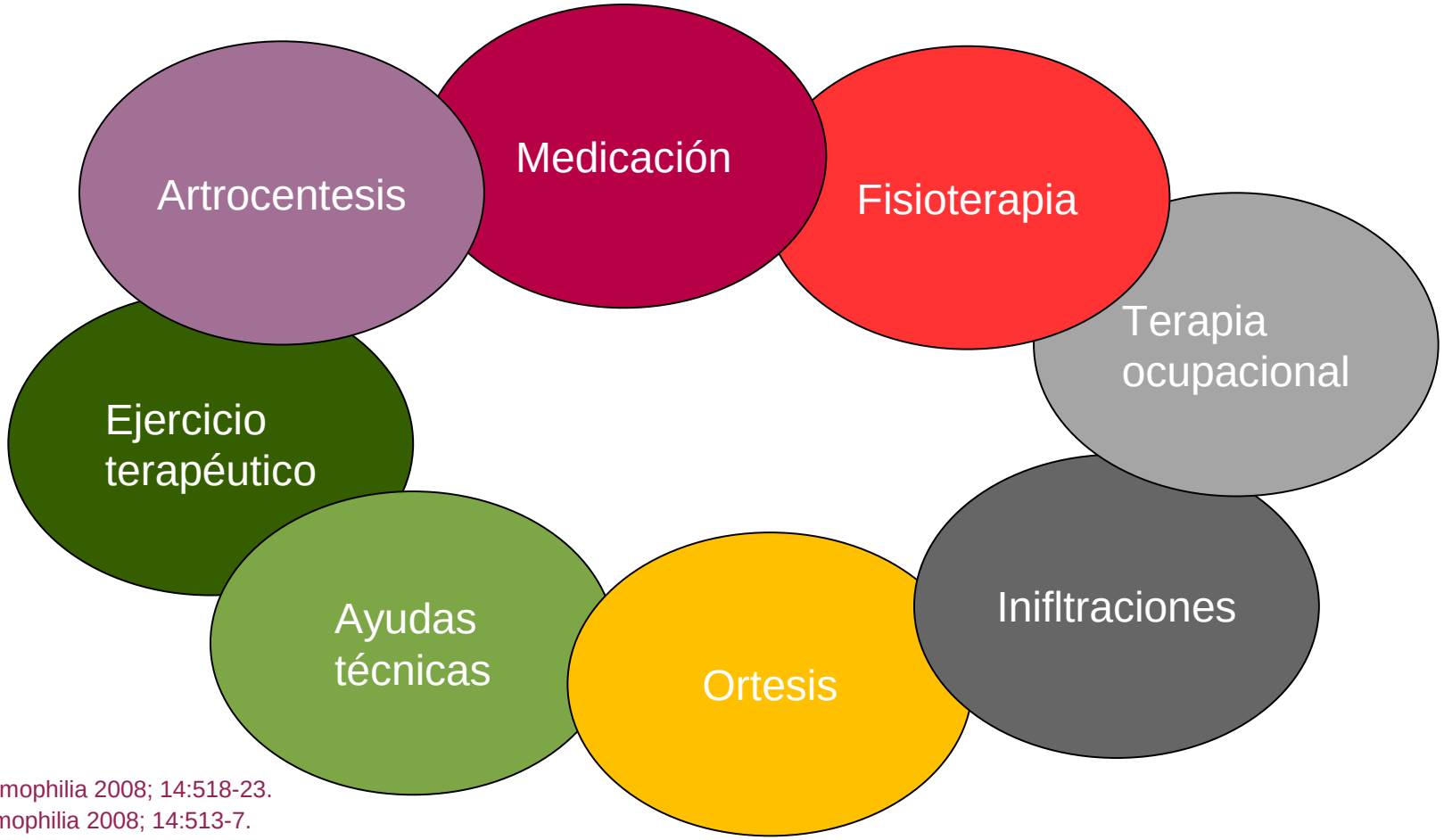
Tratamiento rehabilitador

Tabla 4. Objetivos del tratamiento rehabilitador de las lesiones músculo-esqueléticas de los pacientes con hemofilia.

Aliviar el dolor
Recuperar el rango de movimiento
Prevenir la atrofia muscular
Mejorar la potencia y resistencia musculares
Recuperar la propiocepción
Prevenir las secuelas y deformidades
Mejorar las habilidades funcionales
Mantener un patrón de marcha adecuado
Reducir la frecuencia de sangrados articulares
En general, mejorar la calidad de vida

De la Corte-Rodriguez H, et al. Blood Coagul Fibrinolisys.2013;24:1-19

Tratamiento rehabilitador



Kavakli K et al. Haemophilia 2008; 14:518-23.
Brecelj J et al. Haemophilia 2008; 14:513-7.
De la Corte-Rodriguez H, et al. Blood Coagul Fibrinolisys.2013;24:1-19
Rodríguez-Merchán EC, Wiedel JD. Haemophilia 2001; 7 (Supp 2):6-10.
López-Cabarcos C et al. En: Recomendaciones sobre Rehabilitación en Hemofilia y otras Coagulopatías. 2009

Tratamiento rehabilitador

Kavakli K et al. Haemophilia 2008; 14:518-23.
Brecelj J et al. Haemophilia 2008; 14:513-7.
De la Corte-Rodriguez H, et al. Blood Coagul Fibrinolisys.2013;24:1-19
Rodríguez-Merchán EC, Wiedel JD. Haemophilia 2001; 7 (Supp 2):6-10.
López-Cabarcos C et al. En: Recomendaciones sobre Rehabilitación en Hemofilia y otras Coagulopatías. 2009

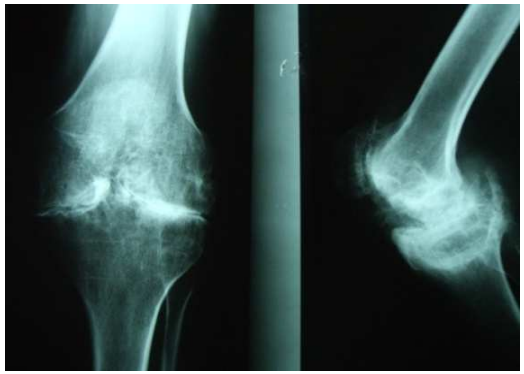
Tratamiento postquirúrgico

Sinovitis hipertrófica → Sinovectomía artroscópica

Artropatías →
- Desbridamientos articulares
- Osteotomías
- Artroplastias (prótesis)
- Artrodesis.

Deformidades articulares →
- Alargamientos tendinosos
- Osteotomías
- Distracciones progresivas con fijadores externos

Tratamiento postquirúrgico



Establecer un programa rehabilitador postquirúrgico inmediato, con el objetivo de alcanzar la máxima recuperación funcional

Es precisa la cobertura hemostática

CONCLUSIONES

1.Las lesiones del aparato locomotor son las que producen mayor morbilidad en las personas con hemofilia

2.La anamnesis, la exploración y las pruebas de imagen apoyan el diagnóstico y la toma de decisiones terapéuticas por parte del equipo multidisciplinar

3.El abordaje **rehabilitador** es complejo, debe hacerse de forma individualizada y con objetivos establecidos

4.El fin último es disminuir el impacto que las lesiones producen sobre la calidad de vida