

How to evaluate the state and the progress of haemophilic joints with no or only mild haemarthropathy in routine and research and in countries with limited resources

Axel Seuser,¹ Claude Negrier,² Lily Heijnen,³ Claudia Djambas Khayat,⁴

1. Private Practice for Prevention, Rehabilitation and Orthopaedics, Bonn, Germany; 2. Centre Régional de Traitement de l'Hémophilie, Lyon, France; 3. Van CreveldKliniek, UMC, Utrecht, The Netherlands; 4. Hôtel Dieu de France Hospital, Beirut, Lebanon.

Introduction

- Children with haemophilia have silent bleeds that cause immediate biochemical and biomechanical changes
- The young heavy bleeder and others who bleed less show good scores for years, but when scores deteriorate it is too late to intervene and change the evolution of the joints.
- Very often they are recognized too late with the usual clinical tools. Daily routine leaves the haemophilia team, especially in countries with limited factor resources, with unsatisfactory results of scores and QoL's.
- Tools or combination of tools that are useful in detecting early alterations as well are required. They should :
 - Help to describe the joint status in all periods of the illness
 - Measure outcome of present treatment
 - Give additional information to help to propose a better adapted treatment for patient at an individual level
 - Give additional information to help to propose new strategies of treatment and evaluate the new treatment
 - Give additional information to help to promote the importance of rehabilitation and physiotherapy
 - Give additional information to help to discuss with payers to allow, maintain or increase the standard of care

Methods

- A literature review of clinimetric instruments and evaluation tools used for joint assessment in haemophilia was performed . Out of 685 papers, 32 were selected for final evaluation using the Oxford Evidence Scale and expert decisions.
- Assessment of clinimetric tools was done by clinical relevance (CR), content validity (Cont), sensitivity to change (Sen), disease specificity (DS) and feasibility (Fea).
- To enable use in children (C > 3) early age data was integrated in the evaluation.
- The integrative model of joint function (Fig. 1) served as basis to describe specific and therapy-focused impact. The power of the tools was evaluated on Form Closure (Structure), Force Closure (FC), Motor Control (MC) and Neural Control (NC) (Fig.1).
- Need for a more functional insight according to the ICF model (International classification of functioning, disability and health) lead to integrating activity and participation (A+P) as a criterion.
- How much do the tools contribute to Clinic (C), Quality control (Q) and Research (R).
- Scores: 0 for none, 1 for low, 2 for medium and 3 for high impact. (Fig.2)

Conclusion

- The first assessment needed is clinical examination. It is the basis of interpretation of the status of the patient with haemophilia and his joints. There is need to add the search for silent symptoms. This ensures detecting early joint problems clinically
- Even in countries with the highest treatment standards deterioration of joints especially the ankle joint is recognised in the beginning of adulthood . Joint evolution cannot be changed if intervention is not started early enough, still in subclinical phases. This can only be done with functional measurements like 3D MA and sEMG
- In the experts opinion, countries with poor resources encounter the same issues much earlier. As physiotherapy is not as expensive as factor replacement, rehabilitation and muscle reinforcement are a very interesting way to try to compensate for less factor
- The proposed recommendations, based on the assessment tools analysis table will help to assemble individual tools for individual purposes. And it will help to judge what the single tool can contribute to analyse the impact musculoskeletal care has on haemophilia and support the mobilisation of resources for comprehensive care

Assessment tools - recommendations

Clinical examination (must)

- Clinical examination should be performed in all cases and include measuring circumference, joint angle, pain with VAS and inspection of silent symptoms

Scores at structure/function level (must)

- One of the three structural scores (HJHS or Colorado) is the minimum recommended

Scores at activity/participation level (must)

- Quality of life instrument or the FISH as a disease specific tool to measure activity and participation as a very important part of the ICF

Imaging (recommended)

- Ultrasound should be performed, especially in children
- MRI if ultrasound leaves open questions or in preparation for surgery
- Pettersson score in PWH with medium or manifest haemarthropathy

Function measuring techniques (recommended)

- 3D motion analysis and superficial kinetic EMG scored highest. Sensitive to all stages of haemarthropathy and all age groups. Early detection of problems and early individual therapy planning

Assessment tools - analysis table

Criteria/Tools	CR a b c	Cont a b c	Sen a b c	DS	Fea	C>3	Structure	FC a b c	MC a b c	NC a b c	A+P	Aims C Q R	Sum a b c	Sum Total
Circ	0 1 0	x 1 x	x 1 x	n	3	3	y	x 0 x	x 0 x	x 0 x	0	2 0 0	0 3 0	11
Pain VAS	0 2 2	0 1 1	1 2 2	n	3	1	y	0 0 0	0 0 0	0 0 0	0	2 2 1	1 5 5	20
Gon	1 2 2	0 3 3	0 2 2	n	3	3	y	0 0 0	0 0 0	0 0 0	0	2 1 0	1 7 7	24
SS	2 2 2	3 3 3	3 3 2	n	3	3	y	0 0 0	0 0 0	0 0 0	0	3 2 1	8 8 7	35
GS	0 1 2	0 2 2	0 2 2	y	3	1	y	0 0 0	0 0 0	0 0 0	0	2 1 2	0 5 6	20
HJHS	0 2 1	0 2 2	0 2 2	y	3	2	y	0 0 0	0 0 0	0 0 0	0	2 1 2	0 6 5	21
Col	0 2 1	0 2 2	0 2 2	y	3	2	y	0 0 0	0 0 0	0 0 0	0	2 1 2	0 6 5	21
Pett	0 1 2	0 1 1	0 2 2	n	2	2	y	0 0 0	0 0 0	0 0 0	0	1 1 2	0 4 5	17
MRI	3 3 2	2 2 2	3 3 3	n	0	2	y	0 0 0	0 0 0	0 0 0	0	3 3 3	8 8 7	34
Ther	3 3 2	2 2 2	3 3 1	n	2	3	y	0 0 0	0 0 0	0 0 0	0	3 3 3	8 8 5	35
US	3 3 2	2 2 2	2 2 1	n	2	3	y	2 2 2	0 0 0	0 0 0	0	3 3 3	9 9 7	39
SF36	1 2 2	0 2 2	0 2 2	n	3	1	n	0 0 0	0 0 0	0 2 2	2	2 2 2	1 8 8	29
HAL	0 2 3	0 2 2	0 2 2	y	3	2c	n	1 1 1	1 1 1	0 0 0	2	1 1 1	2 8 9	29
QoL	1 2 2	0 2 2	0 2 2	y	3	1	n	0 0 0	0 0 0	0 2 2	3	2 2 2	1 8 8	30
FISH	0 1 3	0 2 2	0 2 2	y	3	1	n	1 2 2	1 2 2	1 1 1	3	3 2 2	3 10 12	39
MA	3 3 2	2 2 2	3 3 3	n	0	3	n	2 2 2	3 3 3	3 3 3	0	3 3 3	16 16 15	59
EMG	3 3 2	2 2 2	3 3 3	n	2	3	n	3 3 3	3 3 3	2 2 2	0	3 3 3	16 16 15	61

Fig.2 Table summarising analysis of assessment tools according to parameters above described

Legend

- Yellow: Physical Examination (Circumference, Pain/VAS, Goniometer, silent symptoms)
- Light Green: tools assessing structure and function (Gilbert Score, HJHS, Colorado)
- Dark Green: Imaging techniques (X-Ray/Pettersson, MRI, Sznitigraphy, Thermography, Ultrasound)
- Light blue: tools assessing activities and participation and QoL (SF36, HAL, QoL, FISH)
- Dark Blue: Functional measurements (3D Motion Analyses, superficial kinetic Electromyography)

Discussion

- Our aim is to give new impulses to a topic that is of growing concern in haemophilia. The tools that are available right now do not give an early enough feedback concerning joint deterioration. To the contrary they give a false positive result in early haemarthropathy. When scores deteriorate its often too late to change the evolution of the joints
- To change this we found necessary to have another view on the assessment of joints. This is why we looked at all available tools from a different angle, using criteria that are specifically focused on discovering early functional or structural deficits like Ultrasound or kinetic superficial EMG
- We added the integrative model of joint function as a criterion for tools. It has been developed over the last 12 years to describe joint function in all its aspects. Most of the traditional scores do not fulfil the specific joint needs in early stages of haemarthropathy concerning this model
- We all know what we can expect from the traditional scores but we should also accept their limits
- The table (see above) indicates the usage for every known measuring tool as mirrored in literature and in our expert opinion. It is a way to combine new approaches like Ultrasound and functional measuring tools with the often used measuring tools
- From the Score Table arises a new standard of measurement tools to be used: Clinical examination including measuring circumference, joint angle, pain with VAS and looking for silent symptoms (like tender points on ligaments only aching when pressed and not reported by the patient), HJHS or Colorado score as a disease specific score, Ultrasound in children or MRI if ultrasound leaves open questions or for preparation for surgery. Pettersson score only in the group with medium or manifest haemarthropathy, Quality of life instrument or the FISH as a disease specific tool to figure out activity and participation as an important part of the ICF; functional measurements with superficial kinetic EMG or 3D motion analysis if the therapy options after EMG are not enough to solve the functional problems
- Especially in countries with restricted resources early detection of symptoms can help preserving the joint by individual therapy

Integrative Model of Joint Function



Fig. 1 Integrative Model of Joint Function

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