

**WORLD FEDERATION OF HEMOPHILIA**



# **11<sup>TH</sup> INTERNATIONAL MUSCULOSKELETAL CONGRESS**

**Cartagena de Indias, Colombia  
April 26–29, 2009**

## **FINAL PROGRAM AND ABSTRACTS**

**ORGANIZED BY**



**IN COLLABORATION WITH**



**LIGA COLOMBIANA DE HEMOFÍlicos  
Y OTRAS DEFICIENCIAS SANGUÍNEAS**



# Greetings

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Dear Friends,

Welcome to Latin-America, to Colombia and to Cartagena. It is intrinsic to our culture to feel infinite gratitude for your coming to our region of the world, and to make it our privilege to turn this meeting into an unforgettable experience.

Each of you has a pivotal role in creating the critical mass of challenge and debate, required to generate change in our countries of origin. The task is formidable and best framed in Santayana's phrase, "we must welcome the future, remembering that soon it will be the past; and we must respect the past, remembering that it was once all that was humanly possible". Past, present and future happen at different times around the world. During this Congress, we will have the unique opportunity to work towards achieving synchrony between the three, and thus contribute to the improvement of the quality of care for persons with musculoskeletal discomfort due to hemophilia.

In exchange for your efforts, Cartagena, a World Heritage Site on the beautiful Caribbean Ocean, will welcome you within its walls and share its history, romanticism and magic. The relationship of the visitor with Cartagena has been masterfully characterized by its iconic poet Luis Carlos López in his masterpiece, "to my native city".

Noble rincón de mis abuelos:  
nada como evocar cruzando callejuelas,  
los tiempos de la cruz y de la espada  
del ahumado candil y de las pajuelas  
pues ya paso, ciudad amurallada, tu edad de folletín  
Las carabelas se fueron para siempre de tu rada  
¡ya no viene el aceite en botijuelas!  
Fuiste heroica en tus años coloniales  
cuando tus hijos, águilas caudales,  
no eran caterva de vancejos.  
Mas hoy, plena de rancio desaliño,  
bien pueden inspirar ese cariño  
que uno le tiene a sus zapatos viejos.

On behalf of the World Federation of Hemophilia and of the *Liga Colombiana de Hemofilia* I welcome you to the **World Federation of Hemophilia's 11th International Musculoskeletal Congress** in Cartagena de Indias.



Adolfo Llinás  
Chair, WFH Musculoskeletal Committee



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## Acknowledgements

The World Federation of Hemophilia, organizer of the 11<sup>th</sup> International Musculoskeletal Congress, would like to thank the following companies for their significant contributions:

Novo Nordisk

Wyeth

Talecris

Baxter

## Committees

### **WFH Musculoskeletal Executive Committee**

Adolfo Llinas (Colombia): *Chair*

Luigi Solimeno (Italy): *Senior Vice Chair*

Angela Forsyth (USA ): *Junior Vice Chair*

Rachel Tiktinsky (Israel): *Secretary*

Pamela Narayan (India): *Member at Large*

### **WFH Musculoskeletal Abstract Review Committee**

Adolfo Llinas (Colombia)

Luigi Solimeno (Italy)

Angela Forsyth (USA )

Pamela Narayan (India)

### **Musculoskeletal Congress Organizing Committee**

Adolfo Llinás, (Colombia) Chair

Luigi Solimeno, (Italy) Vice-Chair

Angela Forsyth, (USA) Junior Vice-Chair

Rachel Tiktinsky, (Israel) Secretary

Sergio Robledo, (Colombia) Liga Colombiana de hemofílicos y otras deficiencias sanguíneas

Assad Haffar, (Canada) WFH, Program Regional Manager

Maria Milagros Salas, (Canada) WFH, MSK Project Manager—Congress and Meetings



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## Congress Information

### Congress Venue

The WFH 11<sup>th</sup> International MSK Congress is being held at the Hilton Cartagena.

### Badges

Delegates, speakers, and exhibitors must wear their name badges. Entrance to sessions and the social event on Tuesday night is limited to badge holders only.

### Duplication/Recording

Photography, audio-taping, video recording, digital taping, or any other form of duplication is strictly prohibited in the session's halls.

### Cellular Phones and Pagers

As a courtesy to all congress attendees and speakers, cellular phones, pagers, and other electronic devices must be operated in the silent/vibrate mode during educational sessions. Devices that beep and ring are strictly prohibited. No cellular phone conversations are permitted within the meeting rooms.

### Smoking Policy

Please note that smoking is not allowed inside the Hilton Cartagena.

### Registration

Congress registration is located above the lobby area of the Hilton Cartagena, in front of the Bolivar meeting room.

### Registration/Information Desk Schedule:

Sunday, April 26:	11:30 – 16:00
Monday, April 27:	07:00 – 16:00
Tuesday, April 28:	07:45 – 16:00
Wednesday, April 29:	07:45 – 11:00

### Coffee & Tea Breaks:

For registered participants wearing their name badges, coffee and tea during the breaks are included in the registration fee.

### Dress Code

Dress code during the congress is smart casual

(Please avoid denims)

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## About Cartagena de Indias

Cartagena is a port city on the northern coast of Colombia and the capital of the Bolivar Department, named after Venezuelan Simon Bolivar (one of the most important leaders of Spanish America's successful struggle for independence).

The metropolitan area has a population of 1,240,000 and the city proper, 1,090,000 (according to the 2005 Census). It is the fifth largest urban area in Colombia.

Founded in 1533 by Spaniard Don Pedro de Heredia and named after the port of Cartagena in Spain's Murcia region, the city was a major center of early Spanish settlement in the Americas.

### Climate & Clothing

Daytime temperature in April is around 30 degrees Celsius. Evenings are about 21 degrees so it will be handy to have a light jacket.

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## Social Program

### Opening Ceremony

Date: Sunday April 26, 2009

Time: 17:30-18:00

Venue: Salon Bolivar A/F

Dress: Smart Casual

Cost: Complimentary

### Welcome Reception – White night on the beach

Date: Sunday, April 26, 2009

Time: 18:20-20:00

Venue: Playa Oasis (Oasis Beach) – Hilton Cartagena

Dress: Smart Casual

(make sure to wear sandals—we will be in the sand)

Cost: Complimentary

### Farewell Dinner

Date: Tuesday April 28, 2009

Time: 18:00-22:00

Venue: Club de Pesca

A 40 minute sightseeing tour, followed by dinner at the Club de Pesca, located at the Fuerte Pastelillo (Pastelillo Fort).

Dress: Casual Chic

Cost: Complimentary for full registered delegates

Meeting point: Hotel lobby, buses will depart at 18:00, please make sure to be at the Hotel lobby by 17:45.

## Program at a Glance

## WFH 11<sup>th</sup> International Musculoskeletal Congress

Sunday April 26		Monday April 27	Tuesday April 28	Wednesday April 29
Pre-Congress		DAY 1	DAY 2	DAY 3
		<b>08:00– 09:40</b> Setting the Tone for Musculoskeletal Care	<b>08:00– 11:00</b> Repetitive Psoas Bleeds, Secondary Knee Damage, and the Development of Pelvic Pseudotumors	<b>08:00–10:00</b> Superior Extremity Involvement in the Acute, the Chronic, and Sequelae
		<b>09:40-10:00 HEALTH BREAK</b>	<b>9:35-9:55 HEALTH BREAK</b>	<b>10:00-10:20 HEALTH BREAK</b>
		<b>10:00–11:00</b> Regional State of Affairs: Chal- lenges & Creative Solutions	<b>11:00–12:00</b> FREE PAPERS III: Factor Treatment and Synovectomy	<b>10:20–12:00</b> Formidable Tasksand Future Outlook
		<b>11:00–12:00</b> FREE PAPERS I: General Regional Issues		
<b>11:30–15:30</b>  Registration		<b>12:20–13:35</b> Lunch / Baxter Symposium Goals of Prophylaxis with Re- combinant FVIII in Muscu- loskeletal Care	<b>12:20–13:35</b> Lunch / Novo Nordisk Symposium Modern Management for an Opti- mal Outcome of Elective Orthope- dic Surgery in Hemophilia Patients with Inhibitors	<b>12:00–13:00</b> Annual General Meeting of Muscu- loskeletal Committee of The World Federation of Hemophilia
<b>13:00–16:00</b>  Pre-congress Physio 1) PT standards IHTC 2) Physiotherapy mod- ule of the IHTC 3) Train the Trainers Program Update	<b>13:00–16:00</b>  Pre-congress Ortho 1) Total Knee Replace- ment Registry 2)Orthopedic Treatment Guidelines 3)Development of Re- gional Study	<b>13:45–14:45</b>  Wound Site Hemostatic Comple- ments: Hematologists Appreciate the Help	<b>13:45–15:05</b>  Monitoring, Anesthesia, Post- operative Pain Management, and Nerve Blocks in Hemophilia Surgery	<b>14:00-15:00</b>  SCCOT: Everything you Wanted to Know About Hemophilia and Were Afraid to Ask
		<b>14:45-15:00 HEALTH BREAK</b>		
		<b>15:00–16:10</b> Exercise Programs and Sports in Hemophilia	<b>15:05–16:20</b> FREE PAPERS IV: Surgical and Invasive Procedures	
		<b>16:10–17:30</b> FREE PAPERS II: Non-Surgical Issues		
<b>17:30</b>  Opening Ceremony			<b>18:00-22:00</b>  Social Program & Farewell Dinner	

## Sunday, April 26, 2009

13:00–16:00

### Concurrent Pre-Congress Sessions

13:00–16:00

#### Physiotherapy

*Chairs: A. Forsyth (USA) and K. Mulder (Canada)*

PT Standards IHTC  
*K. Mulder (Canada)*

Physiotherapy Module of the IHTC  
*K. Mulder (Canada)*

Train the Trainers Program Update:  
Egypt and Russia  
*P. DeKleijn (Netherlands), A. Forsyth (USA), and  
A. Sabbour (Egypt)*

Workshop in Nepal  
*P. Narayan (India)*

13:00–16:00

#### Orthopedics

*Chairs: A. Llinas (Colombia) and L-P. Solimeno (Italy)*

Total Knee Registry  
*L-P. Solimeno (Italy) and N. Goddard (USA)*

Orthopedic Treatment Guidelines  
*G. Pasta (Italy)*

Development of Regional Descriptive Study  
*H. Caviglia (Argentina) and A. Llinas (Colombia)*

17:30-18:00

#### Opening Ceremony

Venue: Salon Bolivar A/F

Dress: Smart Casual

Cost: Complimentary

#### Welcoming Remarks

*M. Skinner (USA)*  
*President of the World Federation of Hemophilia*

18:20-20:00

#### Welcome Reception – White night on the beach

Venue: Playa Oasis (Oasis Beach) – Hilton Cartagena

Dress: Smart Casual

(make sure to wear sandals—we will be in the sand)

Cost: Complimentary

## Monday, April 27, 2009

8:00–9:40

### Session A1

#### Setting the Tone for Musculoskeletal Care

*Chairs: A. Llinas (Colombia) and R. Tiktinsky (Israel)*

8:00 Introduction  
*A. Llinas (Colombia)*

8:05 The Spectrum of Prophylaxis  
*C. Kasper (USA)*

8:20 Treatment for All...Implications for Future  
Orthopedic and Rehabilitation Services  
World Wide  
*B. Evatt, USA*

8:35 Cartilage Deterioration as a Function of the  
Speed of Blood Clearance from the Joint  
*N. Jansen (Netherlands)*

8:50 Practical Applications of Gait Analysis in  
Hemophilia: Surgical Decision-Making  
*A. Seuser (Germany)*

9:05 The Natural History of the Development of  
Inhibitors: What to Expect in Countries  
Approaching Access to Concentrates  
*P. Mannucci (Italy)*

9:20 Nature of Scientific Truth  
*H. Winet (USA)*

9:35 Discussion

9:40–10:00

#### Health Break

10:00–11:00

### Session A2

#### Regional State of Affairs: Challenges and Creative Solutions South of the Border

*Chairs: H. Caviglia (Argentina) and S. Toledo (Uruguay)*

10:00 Is Innovation Possible with Limited  
Resources? Let Me Give You a Few Examples!  
*F. Fernandez-Palazzi (Venezuela)*

10:10 Articular Cartilage Response to Chemical  
Synovectomy  
*O. Lazala (Colombia)*

10:20 Embolization of Geniculate Arteries for the  
Treatment of Synovitis of the Knee  
*G. Capetillo (Mexico)*

10:30 Implementation of a Successful  
Synoviorthesis Program in Matto Grosso:  
How Did We Achieve It?  
*S. Thomas (Brazil)*

10:40 Extension-Desubluxation Hinges for Flexion  
Contracture of the Knee  
*PJ Llinas H. (Colombia)*

10:50 Total Knee Arthroplasty in Angular  
Deformities: How Do We Do It in Santiago?  
*A. Oyarzun (Chile)*

**Monday, April 27, 2009**

**11:00–12:00**

**Session A3**

**Free Papers I: General Regional Issues**

*Chairs: N. Jansen (Netherlands) and G. Pasta (Italy)*

- 11:00 Evaluation of QoL in Hemophilic Children through the HaemoQoL Scale (Bogota)  
*D. Valencia (Colombia)*
- 11:07 Quality of Life in Mexican Children with Hemophilia  
*A. Tlacuilo-Parra (Mexico)*
- 11:14 Orthopedic Interventions in Patients with Hemophilia: Experience in Hospital de San Jose  
*MH. Solano (Colombia)*
- 11:21 Orthopedic Management of Musculoskeletal Complications of Hemophilia Patients in Hungary  
*L. Nemes (Hungary)*
- 11:28 Patterns of Orthopedic Complication of Hemophilia at Khartoum Haemophilia Clinic  
*S. Shaheen (Sudan)*
- 11:35 Frequency and Risk Factors of Reduced Bone Density among Iranian Patients with Hemophilia  
*MR. Managhchi (Iran)*
- 11:42 Osteoporosis/Osteopenia Prevalence in Severe Hemophilic Patients at Kerman City in Southeast of Iran  
*A. Naderi (Iran)*

**12:00–12:20**

**Lunch Box Pick-Up**

**12:20–13:35**

**Industry Symposium: Baxter**

**Goals of Prophylaxis with Recombinant FVIII in Musculoskeletal Care**

*Chair: Silverio Castano*

- 12:20 Introduction  
*S. Castano (Colombia)*
- 12:30 Goals of Prophylaxis with Recombinant FVIII in Musculoskeletal Care  
*L. Valentino (USA)*
- 13:00 Colombia Prophylaxis Experience  
*S. Castano (Colombia)*
- 13:20 Panel Q & A

**13:45–14:45**

**Session A4**

**Wound Site Hemostatic Complements: Hematologists Appreciate the Help**

*Chairs: A. Ruiz-Saez (Venezuela) and L-P. Solimeno (Italy)*

- 13:45 Fibrin Glue: Mechanism of Action and Surgical Applications  
*T. Sohail (Pakistan)*
- 14:00 Collagen Sponges, Local Hemostats, Drains, Hemovacs, and VACs  
*H. Caviglia (Argentina)*
- 14:15 Physiotherapy: Procedures that Hinder or Enhance Coagulation  
*K. Mulder (Canada)*
- 14:30 Discussion

**14:45–15:00**

**Health Break**

**15:00–16:10**

**Session A5**

**Exercise Programs and Sports in Hemophilia**

*Chairs: G. Blamey (Canada) and PJ Llinas H. (Colombia)*

- 15:00 Believe it or Not! Zero Contact in Argentinean Soccer  
*N. Moretti (Argentina)*
- 15:12 Upper Extremity Conditioning in the Athletic Severe Hemophiliac  
*N. Zourikian (Canada)*
- 15:24 Conceptual Framework and Indications and Outcomes of Hydrotherapy in Hemophilia  
*R. Tiktinsky (Israel)*
- 15:36 Hydrotherapy at *Tabasqueña de Hemofilia*  
*R. Coliaza (Mexico)*
- 15:48 The Recreational Exercise Program of the *Liga Colombiana de Hemofilia*  
*E. Benavides (Colombia)*
- 16:00 Discussion

**16:10–17:30**

**Session A6**

**Free Papers II: Non-Surgical Issues**

*Chair: J. Weidel (USA)*

- 16:10 Interactive CD-ROM for Physiotherapists in Hemophilia Care  
*G. Blamey (Canada)*

## Monday, April 27, 2009

16:10–17:30

### Session A6

#### Free Papers II: Non-Surgical Issues (CONT'D)

- 16:17 Role of Physiotherapy in Management of Hemophilic Joints: A Critical Review  
*T. Amalnerkar (Malaysia)*
- 16:24 Sports for Health in Hemophilia: Comparing Dates  
*L. Beltrame (Brazil)*
- 16:31 Continuous Femoral Nerve Block for Physiotherapy of the Knee Joint in Hemophilic Patients: A Preliminary Report  
*R. Espandar (Iran)*
- 16:38 Impact of Hemophilic Ankle Arthropathy on Gait Disability: Analysis of Energetic and Mechanical Variables  
*S. Lobet (Belgium)*
- 16:45 Ankle Hemarthropathy: Improved Outcomes in Pain and Disability  
*L. Short (UK)*
- 16:52 Intra-Articular Single Injection of Very High Molecular Weight Hyaluronic Acid in Ankle Hemophilic Arthropathy: Clinical and MRI  
*C. Ruosi (Italy)*
- 16:59 Variations of the Articular Mobility of Elbows, Knees, and Ankles  
*M. Garcia (Brazil)*
- 17:06 Follow-up of Progression of Hemophilic Arthropathy in Children  
*H. Pergantou (Greece)*
- 17:13 The Natural History of Muscle Hematoma: Diagnosis, Treatment, Rehabilitation, Prevention, and Impact for Patients with Hemophilia  
*R. Beyer (Denmark)*
- 17:20 Osteoid Osteoma in a Child with Severe von Willebrand Disease  
*D. Petratos (Greece)*

## Tuesday, April 28, 2009

8:00–11:00

### Session B1

#### Repetitive Psoas Bleeds, Secondary Knee Damage & the Development of Pelvic Pseudotumors

Chair: *N. Moretti (Argentina)*

- 8:00 Introduction
- 8:05 3D Anatomy of the Psoasiliacus Muscle  
*J. Ramirez (Colombia)*
- 8:20 Clinical Evaluation of the Normal Psoasiliacus Muscle  
*C. Briceno (Colombia)*
- 8:35 Clinical Manifestation of a Psoasiliacus Hemorrhage & Differential Diagnosis  
*N. Zourikian (Canada)*
- 8:50 Diagnostic Imaging of the Psoasiliacus: Radiology, Echography, and MRI  
*G. Pasta (Italy)*
- 9:05 Gait Analysis of the Extremity Devoid of Psoasiliacus Function  
*A. Seuser (Germany)*
- 9:20 The Knee as a Target Joint Due to Femoral Nerve Insufficiency Following a Psoasiliacus Bleed  
*M. Silva (USA)*

### 9:35–9:55 Health Break

- 9:55 Sex and the Psoas: Some Bleeds Are Worthwhile!  
*G. Blamey (Canada)*
- 10:10 Rehabilitation of the Psoasiliacus Muscle: How to Get the Strength, the ROM, and the Speed Back  
*S. Lobet (Belgium)*
- 10:20 How to Deal with Quadriceps Fibrosis or Paralysis: Does Anything Help?  
*F. Fernandez-Palazzi (Venezuela)*
- 10:35 When does a Repetitive Bleed Become a Pseudotumor? Formation and Management of Pelvic Pseudotumors  
*H. Caviglia (Argentina)*
- 10:50 Discussion

**Tuesday, April 28, 2009**

**11:00-12:00**

**Session B2**

**Free Papers III: Factor Treatment/Synovectomy**

*Chairs: K. Mulder (Canada) and M. Silva (USA)*

- 11:00 Clinical Dosing and Efficacy of Recombinant Factor VIIa (rFVIIa) in Specific Target and Non-Target Joint Hemorrhages  
*L. Valentino (USA)*
- 11:07 Validation of a Novel Efficacy Endpoint in Treatment of Hemophilia Bleeding Episodes: the Global Response to Treatment Algorithm  
*M. Christensen (Denmark)*
- 11:14 Scientific Rationales Behind Physiotherapy Recommendations for Chronic Hemophilic Synovitis and Pre/Post-Medical Synovectomy  
*N. Zourikian (Canada)*
- 11:21 Short-Term Anti-TNF Co-Therapy with Coagulation Factor Replacement Improves Prevention of Hemophilic Synovitis in Hemophilia A Mice  
*P. Monahan (USA)*
- 11:28 Cartilage Growth Alteration in Pigs Treated with Colloid Chromic Phosphate P-32  
*A. Douglas Price (Argentina)*
- 11:35 P32 Enlarged Colloidal (P32EC) in Knee and Elbow Synovitis  
*M. Martin (Argentina)*
- 11:42 Radioisotope Synovectomy in Children with Hemophilia Below 10 Years of Age  
*S. Aydogdu (Turkey)*
- 11:49 Radiosynoviorthesis with Yttrium-90 Hydroxyapatite in Hemophilic Patients: A New Challenge in a Developing Region of Brazil  
*E. Eulalio (Brazil)*
- 11:56 The Use of <sup>153</sup>Samarium Hydroxyapatite Synovectomy in Hemophilic Knees  
*M. Sayago (Brazil)*

**12:05–12:20**

**Lunch Box Pick-Up**

**12:20-13:35**

**Industry Symposium: Novo Nordisk  
Modern Management for an Optimal Outcome  
of Elective Orthopedic Surgery in Hemophilia  
Patients with Inhibitors**

*Chair: A. Llinas (Colombia)*

- 12:20 Welcome and Introduction  
*A. Llinas (Colombia)*
- 12:25 Optimizing Elective Orthopedic Surgery in Hemophilia: the Multi-Disciplinary Approach  
*A. Ruiz-Saez (Venezuela)*
- 12:45 A Consensus Protocol for the Use of rFVIIa in Pre-, Peri-, and Post-Operative Management of EOS in Inhibitor Patients  
*P. Giangrande (UK)*
- 13:05 Challenges in Post-Operative Care of EOS in Inhibitor Patients (including a case study)  
*A. Llinas (Colombia)*
- 13:25 Questions and Closing Remarks  
*A. Llinas (Colombia)*

**13:45-15:05**

**Session B3**

**Monitoring, Anesthesia, Post-operative Pain  
Management and Nerve Blocks in Hemophilia  
Surgery**

*Chairs: M. Heim (Israel) and P. Narayan (India)*

- 13:45 Venous Access and Monitoring for Anesthesia in Major Orthopedic Surgery: How to Approach Zero Morbidity  
*F. Raffan (Colombia)*
- 14:00 Pre-operative Evaluation: Risk Assessment  
*P-L. Solimeno (Italy)*
- 14:15 Intra-operative Thromboelastography as a Coadjuvant to Monitoring Coagulation Status During Major Orthopedic Surgery  
*F. Raffan (Colombia)*
- 14:30 Post-Operative Pain Management for Musculoskeletal Surgery: Conceptual Framework and Practical Approaches  
*G. Blamey (Canada)*
- 14:45 Peripheral, Post-Operative Nerve Blocks  
*S. Martinez (Colombia)*
- 15:00 Discussion

## Tuesday, April 28, 2009

15:05-16:20

### Session B4

#### Free Papers IV: Surgical & Invasive Procedures

*Chairs: P. De Kleijn (Netherlands) and  
T. Sohail (Pakistan)*

- 15:05 Arthrodesis of the Ankle and Subtalar Joint in Patients with Hemophilia  
*J. Weidel (USA)*
- 15:12 Ankle Prosthesis in Bleeding Patients  
*JG. Ascencio (France)*
- 15:19 Upper Ankle Joint Prostheses in Hemophilia Patients  
*D. Schott (Germany)*
- 15:26 Total Knee Replacement in a Patient with a Below-Knee Amputation with Hemophilia  
*N. Goddard (UK)*
- 15:33 Follow-Up of Primary Hinge Total Knee Arthroplasty in Patients with Hemophilia  
*S. Claeysens-Donadel (France)*
- 15:40 Incidence of Total Knee Replacement Infection in Hemophilic Patients with Negative HIV and HCV Positive Serology  
*H. Caviglia (Argentina)*
- 15:47 The Use of Plasma-Derived Biomaterial in the Treatment of Hemophilic Bone Cyst: Case Report  
*M. El Ekiaby (Egypt)*
- 15:54 Hip Surgery in Hemophilic Patients Under Low-Dose Thromboprophylaxis  
*TBD (Macedonia)*
- 16:03 Percutaneous Treatment of Discogenic Pain in Hemophilic Patients  
*F. Querol (Spain)*
- 16:10 Catastrophic E. Coli-Induced Necrotizing Fasciitis of the Lower Extremities in Hemophilic Patient with HIV  
*M. Shaheen (Saudi Arabia)*

18:00-22:00

### Cultural Event & Farewell Dinner

A 40 minutes sightseeing tour, followed by dinner at the Club de Pesca, located at the Fuerte Pastelillo (Pastelillo Fort).

Meeting point: Hotel lobby, buses will depart at 18:00, please make sure to be at the Hotel lobby by 17:45.

## Wednesday, April 29, 2009

8:00-10:00

### Session C1

#### Superior Extremity Involvement in the Acute, the Chronic, and Sequelae

*Chairs: F. Fernandez-Palazzi (Venezuela) and  
N. Zourikian (Canada)*

- 8:00 Physiotherapy for Recurrent Bleeding of the Elbow: Tips, Tricks, and Milestones  
*P. De Kleijn (Netherlands)*
- 8:10 Diagnosis and Treatment of Synovitis of the Elbow  
*M. Heim (Israel)*
- 8:20 Role of Radial Head Resection in the Recuperation of Prono-Supination of the Forearm  
*L. Pacheco (Brazil)*
- 8:30 What Can Surgery Do for Established Flexion Contractures of the Elbow?  
*M. Silva (USA)*
- 8:40 Diagnosis and Treatment of the Acute Compartment Syndrome of the Forearm  
*R. Bernal-Lagunes (Mexico)*
- 8:50 What Can Be Done about Volkmann's Ischaemic Contracture of the Forearm?  
*C. Moreno (Colombia)*
- 9:00 Conservative Management of the Acute Shoulder Bleed  
*P. Narayan (India)*
- 9:10 Arthroscopic Management of Shoulder Hemophilic Arthropathy  
*OS Perfetto (Italy)*
- 9:20 Management of Grade IV Arthropathy of the Shoulder  
*G. Pasta (Italy)*
- 9:30 Total Shoulder Arthroplasty  
*G. Pasta (Italy)*
- 9:40 Rehabilitation of Total Shoulder Arthroplasty  
*A. Forsyth (USA)*
- 9:50 Discussion

10:00-10:20

### Health Break



## Wednesday, April 29, 2009

10:20-11:45

### Session C2

#### Formidable Tasks and Future Outlook

*Chairs: A. Forsyth (USA) and A. Llinas (Colombia)*

- 10:20 Pharmacological Protection of Cartilage from Blood Induced Damage: State of the Art  
*N. Jansen (Netherlands)*
- 10:30 Total Knee Arthroplasty and Infections, and Joint Salvage  
*J. Weidel (USA)*
- 10:40 Arthroplasty in Patients with High-Titer Inhibitor and Prosthetic Infection: Are the Rates Acceptable?  
*N. Goddard (UK)*
- 10:50 Is the Bleeding Tendency in Hemophilia B Less Severe than in Hemophilia A?  
*P. Giangrande (UK)*
- 11:05 Revision of Infected Total Joint Arthroplasty in the HIV Positive Patient  
*P-L. Solimeno (Italy)*
- 11:15 Idealized Physiatry Model for the 21st Century: Where Should We Aim?  
*L. Chen (China)*
- 11:25 The Future of Hematological Management of Hemophilia: What is Brewing?  
*M. El Ekiaby (Egypt)*
- 11:35 Unintended Consequences of Modern Treatment  
*C. Kasper (USA)*

12:00-13:00

Lunch / Annual General Assembly

14:00-15:10

### SCCOT Session

#### Everything You Wanted to Know about Hemophilia and Were Afraid to Ask

*Chairs: O. Lazala and A. Llinas (Colombia)*

- 14:00 Hemophilia: Past, Present & Future  
*P. Mannucci (Italy)*
- 14:07 Clinical Manifestations and Management of the Acute Articular Bleed  
*P-L. Solimeno (Italy)*
- 14:14 Diagnosis and Management of Chronic Synovitis  
*M. Heim (Israel)*
- 14:21 Muscular Bleeds and Their Consequences  
*K. Mulder (Canada)*
- 14:28 Articular Manifestations of Hemophilia  
*M. Silva (USA)*
- 14:35 Gait Analysis in Hemophilia  
*A. Seuser (Germany)*
- 14:42 Total Joint Arthroplasty in Hemophilia  
*TBD*
- 14:49 Diagnosis and Treatment of Pseudotumors  
*F. Fernandez-Palazzi (Venezuela)*
- 14:56 No Lab Test Echoes Clinical Response  
*C. Kasper (USA)*
- 15:03 Closing Remarks  
*A. Llinas (Colombia)*



## ABSTRACTS

### Sunday, April 26, 2009

#### Pre-Congress—Physiotherapy

##### PT Standards IHTC

K MULDER

*Department of Physiotherapy, Children's Hospital,  
Winnipeg, Canada*

Building on the work done at the 10th MSK Congress in Stresa, this working session will continue to develop standards of physiotherapy care that can be used in hemophilia treatment facilities around the world.

##### Physiotherapy Module for IHTC

K MULDER

*Department of Physiotherapy, Children's Hospital,  
Winnipeg, Canada*

Over the past few years, it has been demonstrated that the IHTC fellowship model, where fellows from less developed countries travel to the developed world for training, is not effective for physiotherapists and physiatrists. In the developed world, most persons with hemophilia are on home care and factor replacement prophylaxis regimes, and therefore they have mild, if any, joint disease. This means that IHTC fellows do not get to assess and treat patients that could help them learn the skills they need when they return home. We are recommending that IHTC trainers go to the developing centres to teach the fellows in their own environment. In order to ensure consistent training, a standardized training module, based on learning objectives, is being developed by a working group of WFH physiotherapists. This working session will provide an opportunity for trainers and fellows to share their experiences, and to guide the formulation of the final objectives.

##### Train the Trainers Program Update: Global Physiotherapy Initiative (GPI) – Future plans and involvement of the MSC based on experiences of two pilot workshops

P DE KLEIJN<sup>1</sup>, K MULDER<sup>2</sup>, A FORSYTH<sup>3</sup>, AND A SABBOUR<sup>4</sup>

<sup>1</sup>Department of Rehabilitation and Sports Medicine, University Medical Center, Utrecht, The Netherlands; <sup>2</sup>Department of Physiotherapy, Children's Hospital, Winnipeg, MB, Canada; <sup>3</sup>Penn Hemophilia and Thrombosis Program, Philadelphia, PA, USA; <sup>4</sup>Faculty of Physical Therapy, Cairo University, Egypt

The GPI is based on the 'train the trainers' principle. Its final goal is to implement basic physiotherapy in HTCs worldwide, on a permanent and organized basis. To begin this project, two pilot workshops were held. The first was in Cairo, Egypt, in December 2007 and the second was in St. Petersburg, Russia, in October 2008. Both consisted of a one-day symposium and 3-4 workshop days. The pilot sites had similarities including: initiation, organizing parties, and overall set-up, and differences, including: lan-

guage spoken, involvement of local physiotherapists, participating professionals, and PWH volunteering in practical sessions. To develop future GPI plans, the following items have to be addressed: The total set up of the workshop, including time schedule, and parties involved; the content of the symposium and workshops; the role of the WFH (including coordinators) and NMOs; the role of the PWH; the coordination between the pool of physiotherapists (future trainers) of the MSK Committee and WFH, including NMOs; the expansion of the project to include rehabilitation specialists and orthopedic surgeons of the MSK. The evaluation of the two pilots is to promote the **chance to success** of this particular process by looking carefully at each item separately. This project will be discussed, and physiotherapists attending the Pre-Congress Day will be encouraged to participate in order to gather information on recommendations for the future of the GPI project. Furthermore, a discussion on the involvement of rehabilitation specialists and orthopedic surgeons will be included during the MSK general meeting.

##### Train the Trainers Program Update: Nepal, September 2008

P NARAYAN

*Lakshmi Neuro Centre, Hyderabad, India*

Physiotherapy is an important part of hemophilia care in the developing world. Nepal is a landlocked country in South Asia and is the world's youngest republic. It has a population of approximately thirty million. Katmandu is the capital of Nepal. Nepal has, within it, eight of the highest mountains in the world. Although the majority of its people are Hindus, the country has strong Buddhist traditions. The Nepal Hemophilia Society has 50 members and three full-time staff. A total of 195 persons with hemophilia have been registered with the society thus far. As part of the WFH's global initiative, a multidisciplinary symposium was organized in Katmandu in September 2008. All professionals involved with hemophilia care were invited to attend the symposium. The morning was spent addressing all the professionals together through a series of lectures. In the afternoon, concurrent workshops were conducted for doctors, laboratory technicians, and physiotherapists. This presentation will provide an update on the physiotherapy workshop conducted as part of this symposium. The presentation will detail physiotherapy concerns in Nepal and how these might be addressed by training local therapists. Details of the contents of the physiotherapy workshop will also be highlighted during this talk.

**ABSTRACTS**  
**Sunday, April 26, 2009**  
**Pre-Congress—Orthopedics**

**A Registry for Total Knee Replacement in Hemophilic Patients**

LP SOLIMENO<sup>1</sup>, ME MANCUSO<sup>2</sup>, E SANTAGOSTINO<sup>2</sup>, OS PERFETTO<sup>1</sup> AND G PASTA<sup>1</sup>

<sup>1</sup>Orthopaedics and Traumatology Department and Angelo Bianchi Bonomi Hemophilia Center, IRCCS Maggiore Hospital Foundation, Milan; <sup>2</sup>Angelo Bianchi Bonomi Hemophilia and Thrombosis Center, IRCCS Maggiore Hospital Foundation, Milan

The knee is the most common joint affected in hemophilia. Despite prophylactic treatment that has improved the lifestyle of hemophiliacs over the last years, there are still patients who have a severe degree of joint destruction as a result of repeated articular bleeding episodes. Even though replacement surgery on hemophiliacs started in the 70s, some questions remain cause for concern. The early literature was pessimistic, reporting poor results and in particular, a high incidence of infection. In recent years, significant changes in hematological management, patient selection, and surgical devices have led to better results. Moreover, an increased number of challenging revision surgery has been registered. The lack of adequate information on indications, implants, surgical technique, and clinical results could be improved by the collection of data in an International Registry. This could be a useful tool to fill the gap between clinical data and clinical practice, in preparation of suitable guidelines for surgical treatment of hemophilic arthropathy of the knee.

**Developing a Centralized Registry for Hemophilic Arthropathy Management**

N GODDARD

*Royal Free Hospital, London, UK*

I believe that there is an overwhelming need for a centralized registry for patients with haemophilia undergoing joint replacement. Arthroplasty registries are now well established and allow patients and health care professionals to establish and monitor standards and to identify potential problems at an early stage. The haemophilia community is relatively small by comparison to the larger orthopaedic community and the patient numbers limited. It therefore should be possible to collaborate and to share information and outcomes. It is difficult however to put this into practice as I found out in attempting to collect data from just the UK centres. The reasons for this are not clear. As the Musculoskeletal Committee of the WFH we should be responsible for the collection of data so that we can provide information on the use and effectiveness of the implants that we use. We should be able to report accurate outcome measures, audit our results, especially the incidence of failures and infection with the objectives of identifying and potential risk factors. We should establish guidelines for the optimal peri-operative

haemostatic management and post-operative care of our patients. The establishment of such a registry would provide a valuable resource for future research and support evidence-based decision making to improve the quality of care for our future patients. It is now time to act.

**Developing Clinical Guidelines for Hemophilic Arthropathy Management**

G PASTA, E CRISTINI, OS PERFETTO, AND LP SOLIMENO  
*Orthopaedics and Traumatology Department and Angelo Bianchi Bonomi Hemophilia Center, IRCCS Maggiore Hospital Foundation, Milan*

With the increased emphasis on evidence-based medicine in orthopedics, the surgeon is faced with the challenge of evaluating the effectiveness of various treatment interventions. Furthermore, the growing number of technologies and interventions has led to large variations in practice. Consequently, clinical guidelines are being increasingly promoted to guide individual decision-makers in their choices regarding orthopedic interventions while ensuring a standard of high-quality evidence-based health care. In this lecture, we aim to describe and promote the methods for guidelines development and quality assessment and to identify clinical guidelines currently available for orthopedic surgeons in the hemophilic arthropathy setting.

## ABSTRACTS

### Monday, April 27, 2009

#### Session A1 Setting the Tone for Musculoskeletal Care

##### A1.1: The Spectrum of Prophylaxis

C KASPER

*Orthopaedic Hospital, Los Angeles, USA*

The aims of prophylaxis are the prevention of damage to the musculoskeletal system and the optimization of well-being. The term is used most often to describe regular scheduled clotting factor replacement, which is very effective at preventing joint damage. Such programs overshadow other effective preventive measures. Muscle strengthening and fitness programs have an outstanding prophylactic utility by themselves, and can potentiate clotting factor replacement. Early guidance of a child towards a positive attitude, a good education, and a physically-appropriate occupation will help prevent physical damage in adulthood.

##### A1.2: Treatment for All...Implications for Future Orthopedic and Rehabilitation Services World Wide

BL EVATT

*Atlanta, GA, USA*

During the past 50 years, observations of the management of hemophilia have documented the evolution of clinical outcomes that strongly support the comprehensive and preventive care model for hemophilia patients. Today, the use of this model in developed countries continues to prove itself superior for survival and effective utilization of resources. As the economies of many other countries continued to emerge, these countries have implemented this model, limited by the funds available for treatment products. Initial results normally appear rapidly in the form of reduced mortality and morbidity. A series of issues are then imposed by economic realities in each individual country that produce a pause in the progress. First the need for orthopedic and rehabilitation interventions rise rapidly, due to a backlog of severely affected joints produced by years of suboptimal treatment. This produces a secondary rise in the need for treatment products necessary to manage patients during surgery. Second, if insufficient treatment products are available to insure the long term optimal therapy, orthopedic and rehabilitation services will be needed to treat less damaged joints on a continuing basis. An increase in the number of patients with inhibitors may produce priority issues for countries with limited resources. In the future, relocation of orthopedic and rehabilitation training programs are expected to shift from the most developed countries to countries with good medical resources and numerous patients needing these services. In the long term, optimal regimens of treatment with new cost effective products tailored to reduce inhibitors should have dramatic effects on the ability of most countries to obtain optimal care.

##### A1.3: Cartilage Deterioration as a Function of the Speed of Blood Clearance from the Joint

NWD JANSEN<sup>1,2</sup>, G ROOSENDAAL<sup>2</sup>, FPJG LAFEVER<sup>1</sup>

<sup>1</sup>Rheumatology & Clinical Immunology, <sup>2</sup>Haematology/  
*Van Creveldkliniek, UMC Utrecht, The Netherlands*

**Introduction:** It is apparent that recurrent joint bleeding leads to joint deterioration. We have performed both in vitro and animal in vivo studies on the effect of the speed of blood clearance from the joint on the development of joint damage. **Methods:** Cartilage tissue explants were cultured in the presence or absence of 50% v/v blood for 1, 2, 3 or 4 days or for 4 days in the presence of 0, 5, 10, 20, 30 or 50% v/v blood. Blood was injected into the knee joint of Beagle dogs, 48 hours, 24 hours or 15 minutes before termination. The effects on cartilage matrix turnover and synovial tissue destructive properties were determined. **Results:** In vitro exposure of cartilage to blood leads to a concentration- and time-dependent deterioration of the cartilage matrix turnover. The effects of hemarthroses in the canine knee have a time-dependent effect on both cartilage and synovial tissue and hence appear to be dependent on the speed of clearance of blood from the joint. **Discussion:** Taken together, it is apparent that joint damage occurs very rapidly after a joint bleed and that the magnitude of the effect is, at least in part, dependent on the blood load (i.e. the amount of blood, and the duration of the blood exposure). This information may be particularly useful in the discussion of whether aspiration of blood from a joint after a hemarthros is indicated or not to prevent joint damage later in life.

##### A1.4: Practical Applications of Gait Analysis in Hemophilia: Emphasis on Surgical Decision-Making

A SEUSER<sup>1</sup>, G SCHUMPE<sup>2</sup>, P BERDEL<sup>3</sup>

<sup>1</sup>Orthopedic Department, Kaiser-Karl-Klinik Bonn; <sup>2</sup>Institute for Motion Analysis and Quality Control of the Locomotive System (IBQ) Bonn; <sup>3</sup>Orthopedic Department, University Hospital Bonn

In more than 20 years we have performed over 2000 motion analyses in hemophilic adults and children. We have experience with ankles, knees, hips, spines, and elbows using Ultrasound Topometry we do online measurement of motion. We measure the elapsed time of an ultrasound impulse from transmitter to receiver. The transmitters are fixed above and below the patient's joint, the four receivers are positioned in a fixed frame. We calculate the joint angle, angular velocity, and angular acceleration during walking on a treadmill and a squat. In addition, we can calculate the role/glide mechanism during the squat for the knee joint and the distribution of the force vector on the femoral head during gait. Criterion for a physiological function is a regular, rhythmic, and sinusoid curve characteristic and a good distribution of the load onto the joint surface. This is how we can tell physiological from pathological function. Even if the functional pathology is partly caused by structural changes, we are able to work out a muscle-based therapy program with the aim of improving individual functional losses. This has

been proven to work out for all joint systems. If the joint function and the correlated clinical symptoms do not improve, indication for surgery is given. With this procedure, we can make sure that the functional capacity of the joint is fully used before surgery takes place and guarantee a better surgical result. Motion analysis after surgery will help to restore optimal function.

### **A1.5: The Natural History of the Development of Inhibitors: What to Expect in Countries Approaching Access to Concentrates**

PM MANNUCCI

*Angelo Bianchi Bonomi Hemophilia and Thrombosis Center, Department of Medicine and Medical Specialties, IRCCS Maggiore Hospital and University of Milan, Italy*

Inhibitors following treatment with factor VIII (FVIII) are the most challenging complication of hemophilia and the highest economic burden for a chronic disease. For countries approaching access to replacement therapy, it is important to know whether or not plasma-derived and recombinant products are associated with different risks of inhibitors in previously untreated patients with severe hemophilia (PUPs). In PUPs treated with a single plasma-derived concentrate, the cumulative risk of inhibitor was reported to range from 0 to 12.4%. For patients treated with a single recombinant product, this risk is much higher, ranging from 36.0 to 38.7%. These contradictory findings on the natural history of inhibitor development emphasize the need of a randomized trial in order to provide a definite answer on the different immunogenicity of plasma-derived vs recombinant FVIII. The SIPPET study is a randomized international study that has the goal to test the hypothesis that plasma-derived FVIII products containing von Willebrand factor (VWF) are less immunogenic than rFVIII products devoid of VWF. Patients will be randomized to be treated exclusively with a single FVIII product, either plasma-derived VWF-containing or recombinant, and followed up until inhibitor development or until 50 exposure days have elapsed. We will compare the two classes of products and not specific products belonging to these classes. This approach will allow us to generalize the findings of the study to all patients who are going to be treated with any product belonging to the class of rFVIII or to that of plasma-derived VWF/FVIII.

### **A1.6: THE NATURE OF SCIENTIFIC TRUTH**

H WINET

*Departments of Orthopaedic Surgery and Bioengineering, University of California at Los Angeles, USA*

Medicine and engineering are “truth” professions, their histories covering thousands of years of applying empirical reasoning to problems to obtain “truths”. Science is an “understanding” profession, covering less than 500 years of applying analytical reasoning to understanding the natural world. Using Francis Bacon’s inductive logic and Galileo Galilei’s experimentation as their methodology, scientists (nee Natural Philosophers) took advantage of the Enlightenment to initiate a quest that moved them from the constraints of Scholasticism to the brink of be-

coming professors of truth. Engineers and physicians began to incorporate results of this quest and launched a revolution in technology and disease treatment. The quest lost its link with truth in 1927 when Niels Bohr and Werner Heisenberg transformed scientific analysis into a statistical methodology. The social consequences of scientific thinking have been profound. It has produced an understanding of the place of humans in the universe that shifts our species from its center (pre-Copernican) to just another organism (Darwinian). Some, unable to cope with the shift, have declared war on science.

### **Session A2**

### **Regional State of Affairs: Challenges and Creative Solutions South of the Border**

#### **A2.1: Is Innovation Possible with Limited Resources? Let Me Give You a Few Examples!**

F FERNÁNDEZ-PALAZZI

*Orthopaedic Unit, National Haemophilia Center, Caracas, Venezuela*

Is the treatment of children with hemophilia different from developed countries? Definitely the answer to this question is YES. Though the professional knowledge and training of the pediatric orthopedic surgeon is mainly similar to that in developed countries, there are many causes, due to the state of underdevelopment, that make the practice of orthopedics in hemophilia different. In these special circumstances, the surgeon is compelled to improvise the techniques to apply and use non-conventional implants to solve fractures. We must realize that 33% of the world population is living in these circumstances in developing countries. Aside from financial constraints, these limitations are caused by difficulty in obtaining updated equipment or material, religious, cultural, and social considerations, issues with hospital structure, people’s idiosyncrasies and corruption. This work presents cases of the most frequent orthopedic problems that, due to their severity, and in some cases that are not so severe, require the surgeon to solve them, analyzing causes and consequences. Examples are given of critical situations during surgery that need to be solved by unconventional means: for example, wrapping non-sterile instruments with sterile gauze; treating fractures without X-ray control because of the lack of the apparatus; being unable to follow the patient post-op due to social and economical constraints; working in hospital-deficient situations due to the infrastructure of the hospital; needing to use an incorrect tool for a surgical procedure because the hospital does not have the proper one; using an inappropriate implant because that was the only one available; or being limited by religious ideas. Furthermore, all these situations mean that the treatment of hemophilic patients in developing countries is done, sometimes, with methods not completely accepted by all surgeons. Examples of these are given.

## **A2.2: Articular Cartilage Response to Chemical Synovectomy**

O LAZALA

*Ortopedia y Traumatología, Universidad Nacional de Colombia, Bogotá, Colombia*

Chemical synovectomy is a well-known and researched procedure useful in hemophilic patients, in order to obtain synovial membrane ablation. We are doing chemical synovectomy with oxytetracycline (OT) using Fernandez-Palazzi's modified protocol. Until now, results have been very satisfactory and we think this is a feasible method for developing countries, as we showed in Vancouver at the WFH's 2006 meeting. Because of concern about the effect of OT upon articular cartilage, in our orthopedic unit (*Colombia National University and Fundacion Hospital la Misericordia in Bogotá*) we designed an experimental trial applying five different substances, including OT, inside the knees of White Australian rabbits. After sacrifice, the width of articular cartilage was measured and the number of chondrocytes per microscopic camp was counted. OT was responsible for middle decrease of cartilage's thickness and cell count. Besides these anatomical changes, to investigate the joint tissue turnover, in the future, we need to take a look and analyze more variables in our patients after OT: clinical, serum and imaging urinary markers, immunohistology studies, and arthroscopic views. We move about these items throughout the presentation.

## **A2.3: Mid Genicular Artery Embolization Indications or Angiosynoviortesis**

G CAPETILLO, F LOPEZ, E ZURITA, E RAMON, S CASTRO, M HERNANDEZ, F FERNANDEZ, F MAGDALENO, S GÓMEZ, R SOLIS, C ALEJANDRO

Embolization is different from other techniques acting in an extrinsic way. Its action is intrinsic, directly affecting the nutritious artery of the bleeding synovial tissue and producing the cessation of the bleeding events. Without irrigation, the synovial tissue atrophies and ends up losing its hemorrhagic action. With this procedure, all the synovial tissue gets affected, which is not the case with other procedures. The accurate indications are frequent events of acute synovitis, chronic synovitis, and second-grade arthropathy. In third-grade arthropathy, we have proved clinically and by magnetic resonance the presence of active sacs of synovial tissue even in presence of subchondral cyst of diminution of patellar space and angular deformities. The procedure is not indicated in grade 4 and grade 5 arthropathy. The grade 3 patients benefit from being able to start physiotherapy without bleeding complications and end up giving up orthopedic devices, crutches or wheelchair. **CONCLUSION:** Embolization is a procedure that effectively controls the hemorrhagic events and the outcome is better for preserving the knee articulation if performed in early stages.

## **A2.4: Implementation of a Successful Synoviorthesis Program in Mato Grosso: How Did We Achieve it ?**

S THOMAS

*Hemocentro de Mato Grosso, Cuiaba, Brazil*

Brazil has 8,600 patients with hemophilia (PWH) registered. The federal constitution grants universal rights to health care that includes factor concentrates, usually on demand; there is little access to prophylaxis and orthopedic surgery. In April 2003 we started the first study with Yttrium<sup>90</sup> Citrate (C-<sup>90</sup>Y) in PWH in Brazil, through a partnership including IPEN - *Instituto de Pesquisas Energeticas Nucleares, Mato Grosso HTC*, and local nuclear medicine clinic, IMN, after approval by the Board of Ethics. Initially we used C-<sup>90</sup>Y imported from France. PWH directed by 22 HTC, we treated without charge: 366 joints (knees, elbows, ankles, and shoulders); 264 with C-<sup>90</sup>Y, 72 with <sup>90</sup>Y-hydroxiapatite (HA-<sup>90</sup>Y), produced by IPEN; 30 with <sup>153</sup>samarium-hidroxiapatite (HA-<sup>153</sup>Sm). Results were a global decrease in mean number of bleedings: from 19.21 one year before to 2.75 (n=264) one year after treatment with C-<sup>90</sup>Y, and from 18.17 to 6.75 with HA-<sup>90</sup>Y (n=72). No leakage to skin occurred. Presently IPEN delivers HA-<sup>90</sup>Y and HA-<sup>153</sup>Sm to four other HTCs and there is growing interest in spreading the access. Yttrium produced in Brazil - economically feasible - could be comparable to C-<sup>90</sup>Y; a longer follow-up period and comparable number of patients treated with HA is needed for further conclusions. A double blind, randomized clinical trial comparing both radionuclides is underway and will probably add more information. It is crucial that RSO is performed under controlled protocols and all steps must follow the comprehensive care concept.

## **A2.5: Treatment of Knee Flexion Contractures Using Casts and Hinges**

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A knee flexion contracture is an articular complication frequently found in severe hemophilia patients. Since such facilities offer specialized care to hemophilia patients, their finding suggests that even despite moderate access to expert medical care, factor concentrates, and rehabilitation, contractures are difficult to prevent. The deformity produces an evident shortening of the limb requiring the patient to use external support devices, which induce both bleeding and articular impairment of elbows and shoulders. Thus, besides restricting ambulation, lower limb flexion contractures may contribute to upper limb skeletal impairment; consequently, correcting them is essential. There are several treatment options, which include physical therapy, corrective devices, and surgical procedures. When choosing the best therapeutic option, some key parameters should be considered including the patient's age, contracture severity, presence of surrounding contractures, duration of the condition, muscular development, gait pattern, presence of an articular subluxation, radiological findings, willingness of the patient to cooperate, and availability of health care coverage. Treatment options may be included in three groups: (1) physi-

cal therapy; (2) orthotic therapy and corrective devices; and (3) surgical procedures. We will focus on the second category, and will describe the management of knee flexion deformities by using corrective casts with hinges. We suggest their use for managing chronic knee flexion contractures greater than 30° resulting from intra- or extra-articular bleeding. We also suggest using hinges in the post-operative treatment of acute flexion deformities in patients who have undergone either total knee replacement or osteotomies close to the knee.

## Session A3

### Free Papers I: General Regional Issues

#### A3.1: Evaluation of Quality of Life in Hemophilic Children Through the HaemoQoL Scale in the Misericordia Hospital in Bogota 2006 - 2008

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The measurement of quality of life (QoL) in patients with hemophilia has become a standard measurement of treatment outcome. Using the HaemoQoL questionnaire version II we evaluated quality of life in 30 hemophilic children, aged 8-12 years, in the Misericordia Hospital between 2006 and 2008. We also used the FISH questionnaire as a functionality measurement. We used Epi-Info software version 3.5.1 and SPSS version 15. Univariate and bivariate analysis was performed (statistical non-parametric) to relate clinical variables (severity, pain, alteration in walking, among others) and QoL. **Results:** The average age was 11 years. The commitment joints were ankle in 57% of patients, knee in 47%, and elbow in 23%. Point average of HaemoQoL was 53 and FISH was 25. Variables of QoL with greater commitment were friends, perception of support, and adaptation. There was no significant relationship between QoL score and age, severity, pain (EVA), or number of commitment joints. A weak correlation ( $p=0.190$ ) between FISH and HaemoQoL existed. A significant relationship ( $p=0.02$ ) between perception of general health and QoL was observed. **Conclusions:** The evaluated group showed moderate commitment in the total QoL score, especially in the variables friends and perception of support. No relationship was observed between the QoL score and the evaluated clinical variables. A relationship existed between the perception of general health and QoL. This basal measurement will be useful in the follow-up of this cohort of patients.

#### A3.2: Quality of Life in Mexican Children with Hemophilia

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**Introduction:** People with hemophilia have poorer health-related quality of life (HRQL) than the general population. There are no studies in Mexico. **Objective:** To assess the

quality of life of Mexican children with hemophilia. **Patients and methods:** The study involved a national sample of 102 patients with severe, moderate, and mild hemophilia A and B, aged 4-16 years. Quality of life was assessed using the Haemo-QoL long version questionnaires translated into Spanish (scale 0 to 100, 0 = best, 100 = worst QoL). **Results:** We included 85 type A and 17 type B patients from 18 states of the country. In terms of severity this included 24 mild, 56 moderate, and 22 severe cases. In group I, ages 4-7, we included 34 patients, mean age  $5 \pm 1$  year. The mean transformed scale score (TSS) was  $42.44 \pm 16.9$  (range 9.52 - 69.05). Group II, ages 8-12 included 40 patients, mean age  $10 \pm 1$ , mean TSS  $38.29 \pm 13.7$  (range 3.51 - 69.92). Finally group III, ages 13-16, included 28 patients, mean age  $14 \pm 1$ , mean TSS  $33.67 \pm 13.9$  (range 10.38-56.81). The comparison of TSS values between groups was statistically significant in group I vs. III ( $p=0.015$ ). The scores above 50 in groups I and II were items family and friends (55.51 and 58.82; 56.15 and 51.28, respectively), in group III there were no domains above 50. **Conclusion:** The QoL of hemophilic patients in Mexico needs improvement especially in the areas of family and friends.

#### A3.3: Orthopedic Interventions in Patients with Hemophilia: Experience in Hospital de San Jose

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Historically, surgical interventions in patients with hemophilia have been always considered a challenge to the medical group, because of the great risk of bleeding, high cost of the replacement therapy, postoperative infections, transmission of bloodborne infectious agents and need of a multidisciplinary team. Years ago, in Colombia was very difficult to perform these types of procedures, however, nowadays there are many institutions that can perform them with success. The purpose of this report was to describe major orthopedic surgeries in patients with deficiency of coagulation factors, with and without inhibitors, in Hospital de San Jose in Bogota, Colombia. Sixteen surgeries were reviewed in 13 patients. The clinical characteristics included: nine patients with hemophilia A, three with hemophilia B, and one with factor VII deficiency; four of them presented inhibitors and the average age at the time of surgery was 26 years. The orthopedic surgeries included: eight synovectomies, two chondroplasties, one resection of pseudohemophilic tumor, one total hip replacement, and five knee replacements. Sometimes two or more interventions were performed at the same time. Pre-operative evaluation included screening for hepatitis B, C, and HIV, and use of vaccine for hepatitis B when needed. Hemostatic control was achieved in all cases. The agents used included factor VIII, factor IX, FEIBA, and PROTOPLEX T. There were few complications: two infections, one catheter-related and one of the surgical wound, three minor bleedings, and two reinterventions; all initiated physical rehabilitation during the hospitalization. Hospital de San Jose has a special program for hemophilic patients, including trained surgeons and hematology

gists, laboratory services, and factor availability, that have made it possible to perform these high risk procedures with minimal complications and excellent results.

### **A3.4: Orthopedic Management of Musculoskeletal Complications of Hemophilia Patients in Hungary**

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**Patients and methods:** 45% of hemophilia patients in Hungary are treated in the National Hemophilia Center (NHC), Budapest. For the past 15 years, musculoskeletal complications of the disease were treated in close cooperation between the Orthopedic Department of the Semmelweis Medical University and the NHC. The treatment courses of 65 patients with severe congenital bleeding disorders are presented by the authors. The diagnostic distribution of the patients: hemophilia A: 55; hemophilia B: 4; von Willebrand disease: 6, with a mean age of 30 at the time of the surgical intervention. **Results:** 43 patients went through an invasive orthopedic procedure. Twenty-two patients were managed exclusively by conservative methods. The indications for arthroscopy and open surgery included severe bleeding episodes unresponsive to factor replacement and articular blockage. For the total prosthesis implantations, the main indications were resistant bleeding and resting pain. The distribution of the different orthopedic procedures was the following: total knee replacement: 12; total hip replacement: 7; arthroscopic synovectomy: 2; open synovectomy: 1. The average FVIII consumption was 116,500 IU during the prosthesis operations and 49,190 IU during the arthroscopic procedures. As a result of the operations, the number of recurrent bleeds has significantly decreased and the quality of life improved with the cessation of resting pain. The range of motion of the joints did not decrease significantly.

### **A3.5: Patterns of Orthopedic Complication of Hemophilia at Khartoum Haemophilia Clinic**

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Hemophilia A and B are X-linked recessive bleeding disorders. They can occur sporadically due to mutation or are very rarely acquired following an immune abnormality. This study reviews patients who presented to the Khartoum Haemophilia Clinic between March 2004 and June 2005. Objectives were to study the clinical and radiological patterns of musculoskeletal disorder in hemophilic patients. Of the 78 patients, 64 (82.1%) had hemophilia A and 14 (17.9%) had hemophilia B. Their ages ranged between 1½ and 50 years. Sixty-nine patients (91%) were under 30 years of age, while only six patients (9%) were over 30 years. None of them received prophylactic treatment. Only one forth of patients (17) showed good work performance, while 50% (34) had average performance. Swelling, pain, and reduced range of movement were the

commonest presenting features. Joints were affected in 83.3% of patients, and in more than 50%, the knee joint was the target joint. In two thirds of patients, the condition was polyarticular. In 14.1% the bleed was intramuscular. Radiological findings were less severe in patients with hemophilia B than with hemophilia A. In 85% of patients under 30 years of age, severity was in its early stages (I, II, or III), whereas 82% of patients older than 30 years showed stage IV or V. **Recommendations:** All patients with severe hemophilia should be covered with early prophylactic factor VIII to prevent complications and improve lifestyle. Hospitals need to be equipped with facilities for safe and adequate surgical intervention in hemophilic patients to help early medical care.

### **A3.6: Frequency and Risk Factors of Reduced Bone Density Among Iranian Patients with Hemophilia**

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**Introduction:** Hemophilia places affected individuals at an increased risk of reduced bone mineral density (BMD) and even pathologic fracture. We aimed to assess BMD among Iranian patients with hemophilia A and the associated risk factors. **Method:** We performed a cross-sectional survey of forty-two male patients with moderate to severe hemophilia A. All patients underwent a standard food frequency questionnaire to estimate calcium intake and the International Physical Activity Questionnaire (IPAQ) to estimate physical activity. Dual energy X-ray absorptiometry (DEXA) was carried out to define real bone density among the study group. **Results:** Forty-two patients with a mean age of 31.04 ±12.45 years and mean body mass index of 23.06 ±5.1 kg/m<sup>2</sup> were included. A reduced BMD defined as osteopenia and osteoporosis by World Health Organization criteria (T-score <-1 to -2.5 and ≤-2.5, respectively) was found in 16/42 (38.1%) and 5/42 (11.9%) patients, respectively. We also found one pathologic fracture in the hip region of a 21-year-old hemophiliac. There was no difference between groups with regard to calcium intake; however patients with vigorous-intensity activity were less likely to have severely reduced BMD (T-score <-2.5) compared to patients with moderate or low activity (odds ratio: 0.80, 95% confidence interval: 0.68-0.94). **Conclusion:** Our results, along with other studies, emphasize the high frequency of reduced BMD among patients with hemophilia and the role of physical activity in achieving normal bone density. This achievement would potentially avoid skeleton fragility and fractures in this group of patients.

### **A3.7: Osteoporosis/Osteopenia Prevalence in Severe Hemophilic Patients at Kerman City in Southeast of Iran**

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Background: Osteoporosis is a skeletal condition charac-

terized by decreased density (mass/volume) of normally mineralized bone. The reduced bone density leads to decreased mechanical strength, thus making the skeleton more likely to fracture. In view of the current high prevalence of hemophilia and osteoporosis, the aim of this study was to determine the frequency of osteoporosis in severe hemophilia patients (A, B) over 20 years of age in Kerman. Methods: We performed a cross-sectional survey of bone density among 40 men with severe hemophilia. Lumbar spine and hip bone mineral densities were measured using dual energy X-ray absorptiometry. A T-score of -2.5 to -1 was considered as osteopenia and values less than -2.5 were considered as osteoporosis. Data of variables were collected through a form and analysed by SPSS-15. Results: Bone mineral density (BMD) was normal in 37.5% of patients, 55% were osteopenic, and the others (7.5%) were osteoporotic. The pattern of joint involvement was significantly different between hip and spine (p value= 0.046). Mean factor intake and number of hemorrhages in the past year were significantly higher in patients with reduced BMD than those with normal BMD (p value = 0.018 and 0.004 respectively). Binary logistic regression analysis demonstrated that mean factor intake and number of hemorrhages in the past year were significant independent predictors of both spinal and femoral BMD (OR= 1.29 and 1.17 respectively). Discussion: Our results suggest that men with severe hemophilia have reduced BMD. Patients at risk are those with signs of hemophilic arthropathy. Because osteoporosis may complicate the future treatment of patients with hemophilia, screening of patients for reduced bone density and prevention therapies are recommended.

## Session A4

### Wound Site Hemostatic Complements: Hematologists Appreciate the Help

#### A4.1: Fibrin Glue: Mechanism of Action and Surgical Applications

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Hemophilic patients are equally exposed to various kinds of injuries during their lifetime. Range of injuries varies from superficial skin lacerations to mucosal injuries, while during adulthood arthropathies and other deformities need various kinds of surgeries including joint replacement. Factor requirement and procurement is a tedious, formidable, and expensive issue. To minimize the factor requirement other options are exercised, especially use of fibrin glue - tissue sealant. The majority of bleeding points are taken care of and the factor requirement is reduced to minimum. In developed countries it is available as a ready-to-use commercial product but in developing countries it can be made in the outpatient department or operating theater as and when required, thus minimizing factor need and saving a lot of money. Fibrin tissue sealant is also used in various other surgical procedures for hemostasis in non-hemophilic patients as well. This review will discuss the basic hematology of fibrin glue, and its indications in hemophilic and also in non-hemophilic patients.

#### A4.2: Collagen Sponges, Local Hemostats, Drains, Hemovacs and VACs

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Bleeding in patients with hemophilia is a great problem that the surgeon has to act in prompt and proper way to solve this situation. There has been progress in surgical techniques that reduce the possibility of bleeding post-operatively. In orthopedic surgeries when the tissue planes are exposed using standard surgical techniques only immediate bleeding vessels are ligated, but a lot are sealed by primary haemostasis. Diathermy knives or laser knives are used to open tissue planes, will seal off those small vessels and reduce bleeding. Several studies have been done on the use of the fibrin glue to reduce operative and post-operative blood loss. Fibrin sealant (FS) Surgery A liquid is a commercial product composed of purified fibrinogen and thrombin used to seal operative wounds by partially re-enacting the final stage of the coagulation cascade, in which fibrinogen is converted to fibrin in the presence of thrombin, factor XIII, fibronectin and calcium ions. FS are mixed at the 'table', and used to seal complex surgical wounds, such as, anorectal fistulas. However home made product with cryoprecipitate, thrombin and, traxenamic acid can be used through syringes with a Y type connector. Other products can be used to achieve the hemostatic action. Spongostan®/Surgifoam® is a gelatin-based absorbable hemostat. It absorbs 45-50 times its own weight in blood and within 4-6 weeks it is completely absorbed in the human body. The gelatine sponge is available in different sizes, addressing specific surgical needs and furthermore it can be cut into desired size or shape without fragmentation. The sponge can be applied dry or moistened with sterile saline or thrombin. The surgicel is a dense knit material that provides high tensile strength and excellent coverage in the presence of heavy bleeding. Unlike gelatin sponges, SURGICEL NU-KNIT Hemostat won't disintegrate during surgery. It is a plant-based formula that eliminates the possibility of animal or human-borne contaminants and does not need a reconstitutive mix or soak. Arista™AH is a sterile, absorbable hemostatic powder consisting of Microporous Polysaccharide Hemospheres (MPH®), Medafor's patented clotting technology derived from purified plant starch. Arista™AH is a natural composition synthesized from a purified plant polymer and free of all biological components. As such, it is non-toxic, non-irritating, non-hemolytic and non-mutagenic. The flowable particles provide complete coverage on both rough surfaces and in small, hard-to-reach spaces.

#### A4.3: Physiotherapy: Procedures that Hinder or Enhance Coagulation

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Physiotherapists use many electrical and physical agents in their work. This paper will examine what we know and what we don't know about how common physiotherapy modalities affect coagulation.



## **Session A5: Exercise Programs and Sports in Hemophilia**

### **A5.1: Believe it or Not! Zero Contact Argentinean Soccer**

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Soccer is undoubtedly the most popular sport in Argentina, and every Argentine boy has dreamt at some time in his life of becoming a professional soccer player and representing his country in an international competition. Even in spite of the fact that the National Hemophilia Foundation has repeatedly warned hemophiliacs against the dangers of soccer practice, it became evident that hemophilic boys did play soccer on their own, and with other players who were not physically disabled and with the inherent aggressive characteristic of this activity. With this state of affairs in mind, a sports program was devised with a selected group of patients and with the following characteristics: a) Alteration of the sport's regulations with a view to avoiding physical contact; b) Search for similarity of handicap; c) Supervision of the game by extremely active and strict umpires, who prevented physical contact among the players and the breakage of the rules. Acceptance as members of the team of players depended on the fulfilment of certain conditions: a) Absence of Inhibitors; b) Absence of synovitis; c) Lack of aggressive personal characteristics and commitment to team work. The amount of time each player is admitted and the position he occupied on the field is proportional to his degree of disability. Six is the number of patients per team. They played on lawn, as a synthetic field would have caused too much friction. Everybody received anti-hemophilic factor concentrates before the competitions. No major complications were observed during the activity (neither fractured bones, nor hemarthrosis, nor big hematomas). The two main advances of this undertaking are, in the first place, that the patients are protected by safety regulations, and in the second, that the majority of the other hemophiliacs who attend the centre have joined the pre-sport rehabilitation program to be able to fulfil the requirements needed.

### **A5.2: Upper Extremity Conditioning in the Athletic Severe Hemophiliac**

N ZOURIKIAN

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Many people with hemophilia (PWH) recognize the overall joint health improvements obtained by maintaining or increasing joint strength. These benefits are echoed in numerous hemophilia-related literature and recommendations. They include but are not limited to improved joint support, proprioception, bone density, and ultimately, decreased hemarthroses. Athletic individuals are likewise familiar with the value of strengthening exercises. Competitive athletes will typically train regularly and intensely to help attain, maintain, or improve optimum strength and physical condition, whilst aiming to be equal to or better

than their peers. This drive, characteristically present in many athletes, can potentially be a cause of delayed tissue healing in an acute lesion. Similarly, it may also contribute to developing or perpetuating chronic, overuse types of injuries as a result of prematurely returning to their training programs, following musculoskeletal (MSK) injuries. PWH tend to bleed into their joints or muscles following major, minor, or possibly even non-observable trauma, depending in part on the severity and phenotype of their disease, the overall condition of their MSK system, as well as whether factor prophylaxis is available or not. Repeated bleeds or significant bleeds can eventually lead to crippling morbidity. Therefore, the challenge for athletic individuals with severe hemophilia interested in improving their overall physical strength and condition by regular training is to do so while minimizing or preventing the occurrence of hemarthroses or muscle bleeds. This presentation will explain why certain, commonly performed athletic strengthening and conditioning techniques specifically for the upper extremities, when used by PW(severe)H, may theoretically result in an increased risk of joint or muscle bleeds. Subsequently, relatively less risky alternatives for strengthening / conditioning will be introduced along with their rationales.

### **A5.3: Conceptual Framework and Indications and Outcomes of Hydrotherapy in Hemophilia**

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The advantages and benefits of hydrotherapy are well known and recommended in the treatment of hemophilia. The properties of water when used wisely can be very instrumental during treatment. The warm water provides elasticity to the tissue. Buoyancy, which eliminates stress on the joints, helps strengthen muscles with ease of movement. Specific gravity, hydrostatic pressure, density, and refraction are also characteristics that enable treatment in the water to be very advantageous. In our centre, there is a children's group treatment and there is one-on-one for older patients with specific problems. The treatment goals are to increase muscle strength and endurance, mobilization of joints, increase range of motion, relaxation, pain reduction, improved balance, symmetry, and co-ordination, and functional activity and recreation. Additionally, for the youngest children the aims are to help the child acclimate to water easily and cheerfully. Indications for treatment may include a painful or stiff joint after hemarthrosis, joint synovitis, chronic arthritic pain, mobilization after long periods of bed rest, pre- and post-surgery, and joint contractures. There are many treatment techniques that can be used such as hold-relax, breathing exercises, repeated contractions, and gait training. Parents of our youngest patients report that when the child regularly exercises with the use of hydrotherapy, he has 40% fewer bleeds than when he does not attend therapy. Patients who receive hydrotherapy treatment post-surgery have a 50% shorter rehabilitation period than other patients who do not receive hydrotherapy treatment.

#### **A5.5: Program of Functional Rehabilitation Developed by the Colombian League of Hemophiliacs: Playful Presentation of Clinical Cases and Factories**

E BENAVIDES

Colombia

At present, musculoskeletal disorders are the most frequent, severe, and disabling complications of hemophilia and limit the important activities of daily living. Their prevention and treatment are the main cause of lyophilized factor consumption and the implementation of rehabilitation guidelines according to individual patient characteristics. In the context of Latin America, economic conditions for the supply and procurement of treatment are limited and it is usually not easy to access therapy for rehabilitation. In addition, the development of inhibitors remains one of the major challenges in treatment. The main purpose of this article is to present the results of a physiotherapy program developed by *Colhemofilicos* in patients with a significant degree of disability or the presence of inhibitors, and without replacement therapy. The program includes simple, practical, and functional techniques applied according to patient response, without trying to force it, avoiding the risk of new bleeding and pain and simultaneously with the implementation of recreational activities including workshops and education programs of selfcare and involving them in the game. Integration and recreation is an important part of promoting, physical activity around the hemophilic is harmless under the monitoring, collaboration and intervention of a multidisciplinary team. The knowledge and application of a hemophilia physiotherapy program is important in the development of medical skills for health professionals throughout the country and improves care in orthopedics and physical therapy, resulting in a decline of economic costs, and promotes a decent quality of life.

#### **Session A6: Free Papers II: Non-Surgical Issues**

##### **A6.1: Interactive CD-ROM for Physiotherapists in Hemophilia Care**

G BLAMEY

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This presentation will detail the development and production of an interactive CD-ROM designed for use by Canadian Physiotherapists with minimal experience in the musculoskeletal aspects of hemophilia treatment. Four case studies and a series of follow up questions for each study were designed by a group of eight experienced members of the Canadian Physiotherapists in Hemophilia Care (CPHC). The case studies address the many of the major MSK manifestations of hemophilia such as acute or chronic synovitis, arthropathy, and acute joint or muscle bleeding. The user is given the necessary information to answer the series of follow up questions, and given feedback on both correct as well as incorrect responses. The intention of the CD-ROM is to act in part as an outreach tool increase the breadth and depth of clinicians in Can-

ada working with hemophilia.

##### **A6.2: Role of Physiotherapy in Management of Hemophilic Joints: A Critical Review**

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**Objective:** This review is an attempt to determine the effectiveness of exercises and other physiotherapeutic interventions for treatment of joints affected with hemophilia and the subsequent improvement in activities of daily living. **Methodology:** A systemic review of the literature was conducted using PUBMED, CINHALL, COCHRANE LIBRARY, and MEDLINE. **Results:** In acute attacks, significant pain reduction and improvement have been observed with immobilization and cryotherapy. In the chronic stage, when symptoms subside, aggressive techniques like soft tissue therapy, manual traction, muscle guarding using proprioceptive neuromuscular techniques, and strength and balance training for the muscles surrounding the affected joints have been successful. Gradual stretching and regular active exercises with gait modulation, pace walking, treadmill practice, measured weight-bearing, limited resistance training, bicycling, and brisk walking have been incorporated. These interventions maintain and improve the function of neuromuscular junctions and the strength / flexibility ratio of soft tissue. Electrotherapy equipment such as functional electrical stimulation, iontophoresis, I.F.T., T.E.N.S., etc., also have been used clinically and have been well documented. In addition, lifestyle modification and the psychotherapeutic benefits of exercises reduce stress, nervousness and rationally confer a dynamic approach towards life. **Conclusion:** Physiotherapy alone can not treat the condition but can play a vital role in modulating and moderating the disease process. An aptly supervised blend of physiotherapy methods and anti-hemophilic drugs certainly can preserve regular joint range of motion, mobility, and quality of life. An inexpensive approach with proper supervision can be extremely helpful and may show benefits for patients.

##### **A6.3: Sports for Health in Hemophilia: Years in Comparison**

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IHTC - Brazil, Brasilia

Physical inactivity is a serious, nationwide problem. A healthy diet and regular, adequate physical activity are major factors in the promotion and maintenance of good health throughout the entire life course. This is a comparative study between sports / physical activities most recommended for persons with hemophilia through programs launched in 1998 (Go for It), in 2002 (Fit for Life), and in 2005 (Playing It Safe). The aim of this study is to identify the safest and healthiest sport or activity for hemophiliacs. The study compared 14 sports or activities most recommended in 1998 (Go for It), with the same sports reported in 2002 (Fit for Life) and in 2005 (Playing

It Safe). The classification methods were different within each study analysed. However almost every study showed the risk level for each sport/physical activity. We found, as an exercise, cycling, fishing, frisbee, sailing, hiking, snorkeling, table tennis, and tai chi all represented no risk in two of the studies analysed; and walking, swimming, and golf as sports / physical activities most recommended for hemophiliacs in all the samples analysed; badminton, yoga, and dance were safe in one study and represented a moderate risk in another study. In conclusion, swimming, golf, and walking are still safe activities for hemophiliacs according to the samples analysed. We need more studies in the field of sports including general guidelines to evaluate physical activity risk level in sports for hemophilia, and to compare the evolution of treatment and sports.

#### **A6.4: Continuous Femoral Nerve Block for Physiotherapy in Fixed Contracture of the Knee Joint in Haemophilic Patients: A Preliminary Report**

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**Background:** Contractures are usual components of hemophilic arthropathy. Physiotherapy has a limited role in the management of established contractures as patients are impaired by pain, which limits their ability to perform active exercises to regain range of motion (ROM) and muscle strength. **Aim:** Should pain during physiotherapy be alleviated, a higher ROM could be regained and muscle strength could be restored. **Materials and Methods:** Five patients with severe hemophilia and severe unilateral fixed flexed knee were included. Under adequate factor replacement and using electrical nerve stimulation, the anterior cutaneous branch of the femoral nerve was located and a Polyplex catheter was lodged beside the nerve. A solution of bupivacaine 0.125% and lidocaine 2% was used to confirm proper positioning of the catheter. The catheter was connected to a disposable pump (Autofuser®) containing bupivacaine 0.1%. The patients underwent an intensive physiotherapy program for two weeks afterwards. They were evaluated for ROM, pain (visual analogue scale, VAS), adverse effects/complications (questionnaire), and their satisfaction of pain control. The physiotherapist's satisfaction of the patients' cooperation and progress was also recorded. **Results:** All but one of the patients had increased ROM and the contracture was reduced to <5%. VAS diminished to nearly zero in all of the patients. The most common reported adverse reaction was anterior thigh numbness followed by fatigue of the quadratus femoris muscle. **Conclusion:** Should adequate factor levels be secured during catheter placement and physiotherapy, using infusion pumps is a safe procedure for reduction of pain during physiotherapy. Analgesia resulted in good compliance of the patients with painful physiotherapy, which resulted in improvement of knee biomechanics.

#### **A6.5: Impact of Hemophilic Ankle Arthropathy on Gait Disability: Analysis of Energetic and Mechanical Variables**

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The ankle is a common site for spontaneous hemarthrosis in patients with hemophilia. Bleeding episodes affecting the tibiotalar and/or the subtalar joints may lead to severe degenerative changes. Although ankle arthropathy is commonly evaluated by radiological (Pettersson, Arnold and Hilgartner) and clinical (WFH Gilbert, HJHS, Colorado) scores, the functional impairment of hemophilic ankle arthropathy on gait has so far not been studied. The aim of the present study was to evaluate the impact of ankle arthropathy on gait by analysing biomechanical parameters and energetic variables and to explore the relationship between functional gait impairment and the radiological degenerative changes. An observational clinical gait analysis was undertaken in patients with hemophilic ankle arthropathy. The severity of the arthropathy was evaluated using the Gilbert clinical score, the Pettersson radiological score, and the Arnold and Hilgartner radiological score. Foot function was evaluated with the foot function index FFI-R. The study involved 25 hemophilic patients [A (n=21), B (n=4); severe (n=22) and moderate (n=3) factor deficiency] with bilateral (n=22) or unilateral (n=3) ankle arthropathy. The mean age was 40 ±10 (range 21-60). An instrumented motion analysis was used to assess spatiotemporal parameters, ankle kinematics, and mechanical work. Energy expenditure was analysed using an ergospirometer. Electromyographic muscle activity timing of the tibialis anterior, peroneus longus and brevis, and external and internal gastrocnemius were simultaneously recorded. The volunteers walked on a force-measuring treadmill at preferred speed. Results were compared to those obtained in a large group of healthy subjects matched for age. Our results show that the kinematic evaluation (ankle dorsal and plantar flexion), the kinetic variables (maximum ankle muscle torque and power), and the muscle activity are permanently impaired in patients with hemophilia and ankle arthropathy. Normal values of external and internal mechanical work were observed to the detriment of a higher energy cost. Interestingly, no alteration in the pendulum-like mechanism of walking could be observed, even in patients with dramatically decreased range of ankle motion. This study provides new insight into the functional impact on gait of hemophilic ankle arthropathy.

#### **A6.6: Ankle Hemarthropathy: Improvement in Pain and Disability Using Functional Foot Orthoses and Rocker Sole Shoe Adaptations Prescribed with the Aid of an In-shoe Pressure Analysis System to Inform Outcome**

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Functional foot orthoses have been shown to have a therapeutic effect on varying degrees of haemarthropathy of the ankle joint (Slattery and Tinley 2001). However little has been written about the prescription of such devices in the haemophilia population. As part of a clinical audit and service review of 31 haemophilia patients, we identified a sub-group with advanced ankle haemarthropathy. These patients were deemed as being at medical end point for further management of their ankle pain. They also had varying degrees of disability including back pain. A series of 3 case studies are presented, demonstrating the local and global effects of a combination of modified rocker soled shoes and therapeutic functional foot orthoses in this sub group of patients. We demonstrate the benefits of using in-shoe pressure analysis modalities to enhance prescription choice and improve outcomes. The case studies also highlight the potential dangers of treating this group without objective force or pressure/time data.

#### **A6.7: Intra-Articular Single Injection of Very High Molecular Weight Hyaluronic Acid in Ankle Haemophilic Arthropathy: Clinical and MRI**

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**Background:** Intra-articular injections of hyaluronic acid (HA) have been successfully used in the conservative treatment of osteoarthritis of large joints since 1989. Several studies have documented their safety and the efficacy in hemophilic arthropathy, in particular with respect to relief of pain. **Methods:** We investigated the effects of a single very high molecular weight (HMW) intra-articular HA (10,000 kd, Durolane, Q-Med AB, Sweden) injection on symptoms and functional outcome in patients with hemophilic ankle arthropathy. Patients were evaluated prior to the injection, four weeks, and three months later. **Results:** Fifteen ankles in 11 severely affected hemophilic patients (age range 28-43 years) were treated with a single very HMW HA injection, after 40 IU/kg factor concentrate replacement. All joints had grade II or III arthropathy, characterized by subchondral cysts, irregularity or erosion of subchondral cortex, partial chondral destruction, effusion, hypertrophic synovitis, and presence of hemosiderin. Four weeks after the injection of HMW HA, all patients reported a significant subjective improvement, particularly in longer walking distance, stair-climbing, or

initial pain. Mean orthopedic scores of the treated joints were 7.5, 5.8, and 6.9 at baseline, one, and three months after injection, respectively. On MRI evaluation a significant improvement in the ankle joint cartilage quality after treatment was shown. Mean MRI score of the 15 ankles decreased from 19.6 at baseline to 13.4 three months later. **Conclusions:** Our short-term results suggest that a single intra-articular injection of very HMW HA is an effective approach for the conservative treatment of patients with hemophilic ankle arthropathy, with clinical results comparable to those from previous studies with multi-injection protocols.

#### **A6.8: Variations of the Articular Mobility of Elbows Knees and Ankles**

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**Objective:** To analyse and demonstrate the improvement of articular range of motion in patients with hemophilia submitted to free active movement in a pool with warm water. To investigate how this improvement occurs looking at the physiology of immersion, specifically the hydrostatic pressure, and to compare the results with a control group. **Methods:** The arch of active articular movement on the ankle, knee, and elbow was evaluated by the same observer in both groups at the beginning and end of each session using a goniometer. Anatomical references were indicated on the patient's body at the beginning of the session, enabling the exact positioning of the goniometer right after immersion. **Results:** In the hemophilic group, there was a mobility gain in the ankle and knee articulations and higher amplitudes were observed in the ankle, especially during the first sessions. No significant mobility gain in the elbow articulation was observed. In the group without articular anomaly, there was no significant gain in any of the assessed joints. **Conclusion:** The authors suggest that the buoyancy and hydrostatic pressure, as well as the thermodynamic properties of the water heated at 34°C, are responsible for the gain in articular mobility in patients with hemophilia. The authors believe that exercises of free active movement in a pool with warm water represents an approach of great importance in order to gain amplitude of movement in the articulations, prevent dysfunctions that result from hemorrhage, predispose the articulation for functional activities, and facilitate muscular strengthening.

#### **A6.9: Follow-up of Progression of Hemophilic Arthropathy in Children, with Clinical, X-ray, and MRI Scores**

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Arthropathy is considered an irreversible or progressive complication in patients with hemophilia, even in children on prophylaxis. To evaluate the progression of hemophilic arthropathy, 84 joints in 24 boys with severe (n=18) and moderate hemophilia (A= 22, B= 2) were investigated using clinical examination, X-rays, and MRI at two time periods (Time 0 and 1). Patients' age at time 0 was 10.5 ±3.6 years and time elapsed was 3.8 ±1.4 years. At time 0: All investigated joints had a history of more than three bleeds. Sixteen boys were on secondary prophylaxis for 5.4 ±2.8 years. Clinical score: 2.0 ±3.6, Pettersson score: 2.1 ±2.8, Denver score: 4.5 ±3.8. After the first evaluation, prophylaxis was intensified in 11 children and initiated in four. At time 1: Clinical score: 1.5 ±3.1, Pettersson score: 1.7 ±2.7, Denver score: 5.1 ±4.1. On average, the clinical and X-ray scores improved (26% and 40% respectively) while the MRI score deteriorated (34%). The number of hemarthroses per year was reduced (0.7 ±1.76 vs. 2.0 ±1.8). Comparing the findings, deterioration was found in 15.3%, 15.3%, and 34.1%, and improvement in 25.9%, 40%, and 16.5% of the joints with clinical, X-ray, and MRI scores, respectively. Initiation or intensification of prophylaxis resulted in a three-fold reduction of hemarthroses (p<0.01) and significant improvement of clinical and Pettersson scores (p<0.01). MRI findings were only reversible in joints with mild or moderate synovitis without cartilage degradation (14 joints, 16.5%). **Conclusion:** Prophylaxis seems to improve hemophilic arthropathy. It is worthwhile for physicians to persist even in patients with severely affected joints.

#### **A6.10: The Natural History of Muscle Hematoma - Diagnosis, Treatment, Rehabilitation, Prevention, and Impact for Patients with Hemophilia**

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**Introduction:** Muscle hematomas represent 10% of bleeds in patients with severe hemophilia. There is limited consensus on diagnostic or treatment strategies and little knowledge about the natural history of muscle hematoma and optimal treatment goals. **Aim:** Perform a systematic description of the natural history of muscle hematoma in healthy athletes, focusing on diagnosis, treatment, rehabilitation, and prevention. **Methods:** Publications and

educational textbooks on management of sports injuries were used as data source. **Results:** Muscle hematomas occur following contusion, strain, or laceration and can be categorized as mild, moderate, or severe. Muscle hematoma may be inter- or intramuscular. In healthy athletes, the healing process takes 21-30 days. Optimal diagnosis includes history, physical examination (observation, palpation, active and passive range of motion test, muscle length test, isometric strength test, biomechanical examination, full spinal examination, peripheral nerve test, and slump test), ultrasounds, MRI or CT. Treatment is conducted based on: 1) super-acute stage: control of the bleeding and minimization of the size of the hematoma; 2) acute stage: restoration of pain-free range of motion; 3) subacute stage: functional rehabilitation; and 4) gradual return to normal activity. Treatment and preventive strategies include: RICE, protected mobilization, stretching and strengthening exercises, manual therapy (articular, neural, and soft tissue mobilization and massage), correction of movement dysfunction, functional rehabilitation, and electrotherapeutic interventions. **Future perspectives:** The study describes the natural history of muscle hematoma and state-of-the-art diagnosis and treatment in healthy athletes. Results seem useful to optimize diagnosis and treatment in patients with hemophilia.

#### **A6.11: Osteoid Osteoma in a Child with Severe von Willebrand Disease**

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A six-year-old boy with type III von Willebrand disease was admitted with a two-month history of right leg pain that was relieved with painkillers. On physical examination there was tender swelling on the medial aspect of the middle third of the right leg. Hematological and biochemical tests were within normal limits. Plain radiographs showed reactive sclerosis with periosteal elevation at the midshaft of the right tibia, CT scan was positive for bone cortex thickening, bone scan revealed increased uptake, and MRI revealed periosteal reaction with concomitant edema of the bone marrow and surrounding soft tissues. The differential diagnosis included benign and malignant tumors, subacute osteomyelitis, as well as the usual complications secondary to recurrent hemorrhage. We hypothesized that we were dealing with a subperiosteal hematoma and the treatment included bed rest and regular administration of VWF+FVIII concentrates. The patient was discharged 25 days later, free of symptoms. One month later he came back with the same pain. New radiographs showed fresh periosteal reaction and a nidus, indicative of osteoid osteoma. The child was readmitted for surgery. En bloc excision of the lesion was performed and histology confirmed the suspected diagnosis. Seven months post-operatively, the patient is free of symptoms and has never complained of pain since then. Although an osseous lesion in a severely affected child with von Willebrand disease is usually related to the main bleeding disorder, the differential diagnosis should include common tumors like the osteoid osteoma, a benign tumor with various clinical and radiological features.

**Tuesday, April 28, 2009**

**Session B1**

**Repetitive Psoas Bleeds, Secondary Knee Damage, and the Development of Pelvic Pseudotumors**

**B1.2: Clinical Evaluation of the Normal Psoasiliacus Muscle**

C BRICENO

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Psoasiliacus bleeding episodes are frequent in patients with hemophilia. They can be crippling in the short-term and lead to serious complications in the long-term due to their size, origin, and trajectory through the inguinal channel until insertion. Hence the importance of total knowledge of these structures, their anatomy and mechanics in order to facilitate the confirmation of the structural and functional integrity of each and every one of the elements and their joint action. The psoasiliacus muscle is the main and most powerful hip flexor. It is the only muscle that originates on the lumbar spine and pelvis and inserts in the femur; as a result of its localization it does not only produce movement but it is also a stabilizer. During the walking process, it acts on the following phases: pre-balance, initial balance, and half balance. The psoasiliacus is a posture muscle, active in the upright position for the lumbar lordosis maintenance. The relevance of this muscle is due to its involvement in the majority of movements of everyday life. The clinical evaluation for the biomechanical properties is performed through evaluations of the active and passive performance of the muscle. The passive maneuvers determine the muscle tone and extensibility, and the active ones determine the excitability and contraction. Of the passive tests, the "extension muscle passive test" is done in the supine position with the inferior members extended; the anterior basculation of the pelvis and the increase of the lumbar lordosis are valid criteria to suspect shortening of the hip flexors, which is confirmed with other specific tests. With active muscular tests, we can identify the power of the psoasiliacus to promote voluntary joint movements. With the origins fixed, the psoasiliacus bends the hip joint. With the insertion fixed and acting bilaterally, it bends the trunk on the femur in order to move from the supine to the sitting position. When acting unilaterally, it performs the lateral bending of the trunk towards the same side. Psoasiliacus' retraction leads to loss of hip extension, modifies the inferior member posture and lumbar region into a flexion contracture, and modifies normal posture.

**B1.3: Clinical Manifestations of Psoasiliacus Hemorrhage and Differential Diagnosis**

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Muscle bleeds account for approximately 10-30% of all musculoskeletal bleeds in people with hemophilia (PWH).

Compared to hemarthroses, muscle bleeds occur less frequently. Therefore PWH, as well as new HTC team members, may sometimes be less familiar with the early recognition, treatment, and potential complications related to these bleeds. If unrecognized or not addressed promptly and appropriately, a single, significant muscle bleed such as in the iliopsoas (psoas, psoas-iliacus) can potentially lead to serious functional morbidity or even result in life-threatening consequences, in a relatively short interval. Conversely, a single, significant joint bleed will generally not result in severe morbidity—at least not immediately, nor be life-threatening in the PWH. Depending on the cited source, episodes of iliopsoas muscle hemorrhages occur in 0.3% – 13% of people with severe or moderate hemophilia. Knowledge of the clinical manifestations and early identification of iliopsoas bleeds, as well as their appropriate treatment, is therefore essential in helping to ensure a very good or excellent outcome in the PWH. This presentation will describe the commonly observed clinical presentations of iliopsoas bleeds in PWH as well as possible differential diagnoses. Certain relevant anatomical, theoretical, and practical relations, as well as implications of the clinical manifestations, will also be explained.

**B1.4: Diagnostic Imaging of the Iliopsoas: Radiology, Echography, and MRI**

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Spontaneous bleedings occurring into joints and muscles represent the more common manifestations of hemophilia A and B. Clinical diagnosis of iliopsoas hematoma could be a difficult problem to solve. Hip flexion contracture and a positive iliopsoas sign may indicate hemorrhage into the hip joint, the rectus femoral muscle, or even the anterior abdominal wall. It is important to distinguish between the different causes as the treatment differs considerably. The authors reviewed the results of imaging in hemophilic patients with iliopsoas bleeding to assess the role of ultrasound, computed tomography, and magnetic resonance in the investigation of this deep-seated and often clinically undetectable hematoma.

**B1.5: Gait Analysis of the Extremity Devoid of Iliopsoas Function**

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In more than 20 years we have performed over 2000 motion analyses in hemophilic adults and children. We have experience with ankles, knees, hips, spines, and elbows using Ultrasound Topometry we do online measurement of motion. We measure the elapsed time of an ultrasound impulse from transmitter to receiver. The

transmitters are fixed above and below the patient's joint, the four receivers are positioned in a fixed frame. We calculate the joint angle, angular velocity, and angular acceleration during walking on a treadmill and a squat. In addition, we can calculate the role/glide mechanism during the squat for the knee joint and the distribution of the force vector on the femoral head during gait. Criterion for a physiological function is a regular, rhythmic, and sinusoid curve characteristic and a good distribution of the load onto the joint surface. This is how we can tell physiological from pathological function. Especially for this lecture, we analysed five patients with psoas bleeds in their history. We examined the spine, hip, and knee joints, as the psoas has an influence on all those joint systems. Unfortunately we do not have the motion data from before the bleeding to directly compare the bleeding influence. From theory, we know that the psoas has an impact on the spine, the hip, and the knee joint. It flexes the hip and doing it exerts load to the facet joints of the lumbar spine. If it is shortened (like after a bleed), it prevents hip extension and thus extension of the knee joint before toe off during gait. It is a multifunctional and multisegmental muscle and not easy to compensate when dysfunctioning. The reaction to dysfunction after a bleed is very individual, as the motion analysis shows, but always increases gait load in all connected joints and has a negative impact on load distribution of hip and knee joints. Individual conservative therapy is needed to restore function and prevent hip and knee joints from overloading and bleeding.

### **B1.7: Sex and the Psoas : Some Bleeds are Worthwhile!**

BLAMEY G

*Canadian Physiotherapists in Hemophilia Care, Winnipeg, MB, Canada*

This presentation is a follow-up to a case presentation done at the WFH Congress in Vancouver in 2006 regarding sexual intercourse-related psoas bleeding. This topic has sparked much interest and attention and a patient-focused educational resource is in development to address musculoskeletal issues related to sexual intercourse. This presentation will focus on how sexual intercourse impacts the musculoskeletal system and may lead to new or repeat bleeding episodes in individuals who have either a history of psoas bleeding, a recent bleed that needs ongoing management, or established permanent musculoskeletal dysfunction from past bleeding episodes. Retrospective studies looking at psoas bleeds will be examined to bring a new perspective to what has historically been labeled spontaneous psoas bleeding. Specific positions for sexual intercourse that minimize risk to the individual with hemophilia will be discussed, as well as the importance to view sexual health as equal to other domains in the framework of comprehensive care.

### **B1.8: Rehabilitation of the Iliopsoas Muscle: How to Get the Strength, the ROM, and the Speed Back?**

S LOBET<sup>1</sup>, B HIDALGO<sup>2</sup>

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Iliopsoas hematoma is a well-recognized complication of hemophilia. It can be responsible for severe disabilities, among which femoral nerve compression is the most common. Iliopsoas bleeds can be difficult to diagnose clinically because they are not necessarily accompanied by significant bruising or swelling. Therefore, they have a high rate of recurrence often in relation to poor early recognition of the initial symptoms and with insufficient duration of treatment. In the acute phase, medical treatment with replacement factors and postural relaxation have priority. Rehabilitation has to start only on termination of the acute period. In countries where adequate replacement therapy is not available, extra caution should prevail before starting physiotherapy. A skilled physiotherapist has to evaluate the muscle function and identify the possible causes of the iliopsoas bleed in order to provide advice and reduce recurrence of the bleeding. Iliopsoas retraction is often a neglected complication. This retraction leads to a hip flexion contracture, which not only limits the physical activities of the patient but also distorts his posture and gait. Tightness in the iliopsoas causes downward rotation of the pelvis, and this position in turn causes exaggeration of the normal lumbar curvature. The patient either extends the trunk and has excessive lumbar lordosis, or flexes the knees and reduces the forward pelvis tilt. Often a combination of slight leaning forward and slight knee flexion is the result. Careful stretching exercises of the iliopsoas should be therefore performed in order to restore the mobility in hip extension. Because almost all hip flexion contractures affect the sartorius, rectus femoris, tensor fascia latae, and all the soft tissues of the anterior compartment of the hip, specific attention should also prevail on these structures. When the pain has disappeared and hip flexion has diminished, muscular strengthening of the iliopsoas can be carefully and progressively performed in order to restore the limitation in active flexion. In case of femoral nerve dysfunction, physiotherapy may be helpful to maintain muscle strength of the quadriceps, pectineus, and sartorius.

### **B1.9: How to Deal with Quadricipital Fibrosis or Paralysis: Does Anything Help?**

F FERNANDEZ-PALAZZI AND N FERNANDEZ

*Orthopaedic Unit, National Haemophilia Center, Caracas, Venezuela*

The quadriceps muscle is fundamental for knee kinematics. It not only moves the knee, but also gives support and stability to the joint. Therefore it is most important for the hemophiliac to have and maintain correct strength and force in this muscle, thus the importance of a continuous plan of exercises in order to always be fit. The quadriceps can be involved in hemophilia either directly or indirectly: directly when a muscular hematoma occurs in any one of

its components, and indirectly either after a psoas hematoma or after a fixed knee contracture. It is easy to understand that a local hematoma will involve the muscular fibers, shortening them, and then producing an enlargement of the muscle and a proximal and distal contracture. Immediate treatment, not only with coverage with anti-hemophilia factor (AHF), but local means and rehabilitation, must be started. On the affection after a psoas hematoma, strong rehabilitation must be started early, especially if the femoral nerve is involved. When it is involved after a knee contracture, this contracture should be treated before we can start the rehabilitation of the quadriceps. We must keep in mind that even a very small flexion contracture interferes with the quadriceps muscle function. Unfortunately, operations developed in the poliomyelitis era to strengthen the force of the quadriceps, such as flexors to patella, can not be performed in hemophiliacs. If the contracture is fixed, we must straighten the knee by means of a bone femoral surgery, such as a Moore type recurvatum osteotomy.

### **B1.10: Formation and Management of Pelvic Pseudotumors**

H CAVIGLIA

*Fundacion de la Hemofilia, Buenos Aires, Argentina*

Bleeding episodes in the iliopsoas muscle can be very incapacitating in the short-term and may have serious complications in the long-term. The most serious complication is the development of lumbar or pelvic pseudotumors. A pseudotumor is an encapsulated hematoma, which has a tendency to progress and produce clinical symptoms in relation to its anatomical location. Therefore it is more a clinical entity than a pathological lesion. The incidence of this complication is 1 to 2% in patients with severe hemophilia. The anatomic characteristics the muscle insertions and bone circulation of the iliopsoas muscle can explain the high incidence of pseudotumors because a possibility of developing a subperiosteal hematoma exists. The elevation of the periosteum along the cortex produces necrosis by putting pressure on the bone. The progression of hematoma is through muscle compartment. In the lumbar region, the hematoma goes out from the same place as a lumbar hernia. The pressure of the hematoma produces necrosis of the skin and opens the door to infection. The pathogenesis of the pseudotumor is simple. Untreated bleeding can produce the pseudotumor, and the progressive increase of erythrocyte LDH increases the intracystic osmolarity that attracts liquid from the interstitial space, leading to progressive growth of the pseudotumor. The treatment consists of location of the pseudotumor and aspiration of the content guided with the laparoscopy cannula. The cavity is refilled with OH coralline apatite or lyophilized bone graft and spongostane.

## **Session B2**

### **Free Papers III: Factor Treatment and Synovectomy**

#### **B2.1: Clinical Dosing and Efficacy of Recombinant Factor VIIa (rFVIIa) in Specific Target and Non-Target Joint Hemorrhages**

LA VALENTINO <sup>1</sup> AND DL COOPER <sup>2</sup> ON BEHALF OF THE HTRS INVESTIGATORS

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**Background:** Joint bleeding (hemarthrosis) in patients with congenital hemophilia with inhibitors (CHwI) can result in target (T) joints and functional disability. Clinical trials have assessed the efficacy of rFVIIa for the treatment of hemarthrosis. However, such studies have not been designed to address whether specific joints are more difficult to treat. **Methods:** We analysed data from 2,041 rFVIIa-treated hemorrhages in the HTRS registry (01/04-11/08) to identify hemarthroses in patients with CHwI. **Results:** There were 1,163 hemarthroses treated with rFVIIa (initial treatment: 1,065, salvage treatment: 98). Median age at time of hemorrhage was 9 yrs (range: 0-60). The 798 non-target (NT) and 344 target (T) joint bleeds included (NT/T): ankle (279/169); elbow (202/90); knee (143/76); shoulder (46/5); foot (45/1); wrist (43/1); hand (43/0); and hip (24/4). Median (range) total rFVIIa dose per treatment episode was lower for NT joints at 400mcg/kg (46-18,130mcg/kg) than for T joints at 630mcg/kg (50-21,392mcg/kg) (p<0.0001). Total dosing varied across joints and was generally higher for T joints (NT/T): ankle (300 mcg/kg /650mcg/kg); knee (360mcg/kg /558mcg/kg); elbow (490mcg/kg /556mcg/kg); hip (494mcg/kg /884mcg/kg); and shoulder (720mcg/kg /1080mcg/kg). Overall efficacy for all hemarthrosis was 90%, and for target joints was 90%. **Conclusions:** The HTRS registry provides a larger dataset on outcome of rFVIIa-treated hemarthrosis than trials. Median total dose of rFVIIa for NT joints was lower than for target joints. The data suggest load-bearing joints (ankle, knee) were treated with lower doses than non-load-bearing joints (elbow). Efficacy was comparable between T and NT joints.

#### **B2.2: Validation of a Novel Efficacy Endpoint in Treatment of Hemophilia Bleeding Episodes: The Global Response to Treatment Algorithm**

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*Novo Nordisk A/S*

**Objective:** A novel global response algorithm incorporating pain and joint mobility assessment has been developed for hemophilia to allow for more detailed and scaled grading of treatment efficacy. The objective of this study was to investigate the degree of agreement between the algorithm and the commonly used alternative efficacy endpoint: need for rescue medication within 48 hours after joint bleeding. **Methods:** A meta-analysis of the efficacy results reported for F7HAEM-1510 and F7HAEM-2068



(studies investigating the efficacy of 270 µg/kg rFVIIa and 3x90 µg/kg rFVIIa administered within one hour of joint bleeding onset) was performed. Cohen's kappa statistic was derived for each treatment in each study to assess agreement between algorithm results and use of rescue medication. A formal test for the treatments having equal value of kappa was calculated along with an estimate of the common value of kappa. **Results:** Moderate to substantial agreement (Cohen's kappa  $\geq 0.4$ ) was observed for 270µg/kg in the F7HAEM-2068 trial. However, no consistently high degree of agreement was observed in the two trials and the two dose regimens between the use of rescue medication within 48 hours and the global response algorithm as measures of treatment success. **Conclusion:** In this first validation of the global response algorithm, our results suggest that in its current version the metric may not be optimal for efficacy assessment. Nevertheless, efficacy endpoints that incorporate pain and joint mobility assessments in hemophilia are urgently needed, and further research that aims to refine or modify the current algorithm is strongly encouraged.

### B2.3: Scientific Rationales Behind Physiotherapy Recommendations for: Chronic Hemophilic Synovitis and Pre/Post-Medical Synovectomy

N ZOURIKIAN

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Acute hemophilic synovitis can result following joint bleeds in people with hemophilia (PWH). If allowed to progress to a chronic synovitic state, the involved articulation risks developing early joint arthropathy. Treatment options for chronic hemophilic synovitis may include prolonged factor prophylaxis, COX-2 inhibitor NSAIDs, activity modification or cessation, education, temporary use of braces, compression, intra-articular (steroidal) infiltration, and medical (or surgical) synovectomy. Medical synovectomies have been performed in PWH since the 1970s and may be indicated when other treatment approaches are either unsuccessful or unavailable. This presentation will use published references to help explain the rationale of certain physiotherapy recommendations and protocols provided to PWH who present with chronic joint synovitis leading up to, as well as immediately prior to and following, medical synovectomy.

### B2.4: Short-term Anti-TNF Co-therapy with Coagulation Factor Replacement Improves Prevention of Hemophilic Synovitis in Hemophilia A Mice

PE MONAHAN<sup>1,2</sup> AND J SUN<sup>1</sup>

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**Objective:** Inflammatory pathologic changes in joints persist after intra-articular bleeding and may become chronic. Therapy to antagonize inflammatory cytokines is efficacious in inflammatory arthritides including rheumatoid arthritis and could augment hemostatic replacement in hemophilic arthritis. **Methods:** Factor VIII knockout

mice develop bleeding-induced joint pathology that resembles human hemophilic arthropathy. A standardized mouse knee puncture injury results in hemarthrosis and reproducible synovitis; changes can be scored using a validated mouse synovitis scale that awards 0-10 points for increasing pathology. We induced three joint bleeding episodes in the same knee on days 0, 5, and 10, with or without I.V. FVIII and anti-tumor necrosis factor alpha therapy (etanercept, Enbrel®, Amgen/Wyeth); joint pathology was scored at day 35 following the first injury. **Results:** Without FVIII replacement, mice developed severe synovitis (mean score  $6.6 \pm 0.6$ ). Human factor VIII replacement (150 Units/kg) given at the time of each injury reduced the pathologic grade to  $2.4 \pm 1.6$ ; another group of mice receiving the same injury schedule and FVIII replacement but treated with etanercept (100 microgram I.P. Q.O.D.) following the development of synovitis (ten total doses, days 15-35) scored  $1.8 \pm 1.4$ . Mice that in addition to factor VIII replacement began etanercept at the time of the first injury and continued until after the last injury (ten total doses, days 0-20) developed minimal pathology, scoring  $0.8 \pm 1.0$  (significantly less than FVIII therapy alone,  $p=0.03$ ). **Relevance:** Short-course etanercept may augment hemostatic support to prevent hemarthropathic changes; however it may need to be initiated prior to established synovitis.

### B2.5: Cartilage Growth Alteration in Pigs Treated with Colloid Chromic Phosphate P-32

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**Objective:** To assess the cartilage growth alteration in two-month-old pigs treated with intra-articular FMC in their knees. **Materials and methods:** Twenty two-month-old Yorkshire male pigs, weighing between 35 and 40 kilograms, were included in the study. All of them received, under sterile conditions, 2.2 mCi of chromic phosphate P-32 colloid in their right knees. As control case, all of the pigs received, under the same conditions, 1 mL of saline sterile solution in their left knee. There was no evidence of any case of bleeding complications, septic arthritis, or skin injuries. Seven animals were slaughtered six months after having received the intra-articular injection, seven after one year, and six after two years. Lower limb X-rays and CTs were performed post-mortem on all the animals. Measurement of the lower limbs was carried out with a digimatic caliper Mitutoyo®. Measurements lines of the femur and tibia were made in the front and side X-rays (eight lines in the femur and four lines in the tibia); the epiphyses were measured in the CT. **Results:** The results were comparative with the control lower extremity for each line belonging to one anatomic region. It was expressed in millimeters to make the statistic study easier. No significant differences were found in the studies. **Conclusion:** We believe that it is a safe procedure to use in growing people.

## **B2.6: P32 Enlarged Colloidal (P32EC) in Knee and Elbow Synovitis**

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**Aim:** To assess the outcome of radiosynovectomy with P32EC preparation, predominantly in inflamed synovitic knees and elbows of hemophilic (H) patients, shown in a three-phase bone scan and to evaluate the therapeutic cost/benefit ratio. **Material & Methods:** 65 male hemophilic patients, ages: 4-51 years, 62 knee and three elbow, radiosynovectomies with P32EC. Comparison was drawn with intra-articular synovitic treatment with rifamycin (n=15). Informed consent was obtained. Routine blood tests, X-ray, ultrasound, and a three-phase bone scan were performed, as well as three- and six-month methylene diphosphonate (MDP) controls. Exclusion criteria: Bone destruction as by Larsens' classification. This was a single blind comparison study to the patient. Records were kept in follow-up charts on the state of joint involvement, pain, motility, and requirements of anti-hemophilic factors (AHF), corticoids, or analgesics. Saline flushing followed intra-articular instillation. P-32 Bremsstrahlung emission was used in the gamma camera for imaging and in the gamma well counter. Percentage of leakage was investigated counting blood and urine samples in the well counter. Patients were immobilized for 72-h following the procedures. **Results:** Absence of local or systemic effects. Leakage with the 200 nm colloid was less than 5-7%. Traditional intra-articular rifamycin and corticoids procedures required several injections. Comparison of regions of interest (Rols) in treated knees during soft tissue scintigraphies in pre- and post-third MDP controls and Severity Index (SI) showed knee improvement when SI was higher than 1. An increase in joint motion, diminished volume, and less requirement and frequency in the use of AHF was observed in 86% of the radiosynovectomies in the follow-up. **Conclusions:** Radiosynoviorthesis in H patients in early stages of osteoarticular compromise (Larsen 2-3), provides 4-6 months of relief of symptoms. It is a safe, economic procedure in emerging nations, where availability of AHF is difficult and expensive.

## **B2.7: Radioisotope Synovectomy in Children with Hemophilia Below 10 Years of Age**

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During a seven-year period, we treated 41 joints in young (below 10 years of age) children by radioisotope synovectomy (RS) for refractory chronic hemophilic synovitis. These patients represented 23% of all our RS cases (n=221). The youngest age at the time of treatment was three. All had severe factor deficiency and established target joints. Of 41 joints treated, 19 were knees, 11 el-

bows, and 11 ankles. We used Yttrium-90 as the radioisotopic agent in all knees, eight ankles, and five elbows, dosed according to the patient's age and the amount of synovial tissue (3 to 4 mCi for knees; 2 to 3 mCi for others). Additionally, we used Rhenium-186 in six elbows and three ankles. RS was performed in joints with grade II and III synovitis according to the Fernandez-Palazzi's four-grade scale. Mean follow-up period after the procedure was two years (range: 6 months to 3 years). Evaluation was based on the decreasing rate of bleeding episodes. We did not observe any serious complication. We only encountered transitory inflammatory changes around the ankle joint treated by Yttrium-90 in a five-year-old boy. The grading of synovitis seemed to be more important for success than the age of the patient. Even in patients below 10 years old, outcomes were less satisfactory if grade III synovitis was established. All inhibitor patients had satisfactory outcomes. RS seems to be a safe and effective modality for treatment of chronic synovitis related to recurrent joint bleedings. Young age does not seem to be a success-guaranteeing factor. Once synovitis has progressed and articular cartilage damage has occurred, response to treatment is less satisfactory, even in young patients.

## **B2.8: Radiosynoviorthesis with Yttrium-90 Hydroxyapatite in the Treatment of Recurrent Hemarthrosis in Hemophilic Patients: A New Challenge in a Developing Region of Brazil**

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**Objective:** Brazil is a developing country with almost eight thousand hemophilic patients. Only three specialized centres provide radiosynoviorthesis (RSO) as effective treatment of recurrent joint bleeds and all of them are located in the southwestern region of the country, raising difficulties in treating patients from other regions such as the northeast. We decided to evaluate the effectiveness of RSO with yttrium-90 (90Y) hydroxyapatite, a new radioactive compound in this form of therapy, in the treatment of recurrent knee hemarthrosis in hemophiliacs of Ceará, Brazil, and to assess the patients' satisfaction with RSO. **Methods:** In December 2008, RSO was performed on two patients with recurrent knee hemarthrosis. After intra-articular injection of local anesthetic (2% xylocaine) and direct injection of 0.4 mL 90Y-hydroxyapatite (5 mCi) and 0.1 mL betamethasone, 20-minute and 48-hour anterior and posterior knee images were taken, for 10 minutes each, with 80 KeV (20% window) for acquisition of Bremsstrahlung energy of 90Y, VXGP collimator. **Results:** The subjectively estimated success rate for this procedure was excellent. No patient suffered extra-capsular extravasations of 90Y-hydroxyapatite. The bleeding frequency decreased and no additional knee hemarthrosis had occurred almost two months after the procedure. Both patients reported that the treated joint was much better or better than before the RSO. **Conclusions:** RSO with 90Y-hydroxyapatite is a feasible and effective treatment for

recurrent knee hemarthrosis in hemophilic patients in a developing region of Brazil. Nowadays, our hemophilic patients have one more therapy option and they don't need to travel long distances anymore.

### **B2.9: The Use of 153-Samarium Hydroxyapatite Synovectomy in Hemophilic Knees**

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Synoviorthesis has to be considered the initial procedure of choice for the treatment of patients with hemarthrosis in hemophilia. The penetration of beta energy of 153-samarium is appropriate for the synovectomy of articulations. The objective of this study was to evaluate the efficacy of 153-samarium in 31 patients with hemophilia A or B and a total of 36 knee replacements (ks), with different evolution of arthropathy. We analysed the evaluation before and one year after synovectomy, using the following criteria: reduction in the number of hemarthroses (NH), use of coagulation factor (CF), and improvement in articular motility (IM). The occurrence of adverse events was also considered. The results were a reduction in hemarthroses in 82.3% of patients, and 79% of the patients saw a decrease in the use of coagulation factor. Finally, 41.2% had improved knee motility with  $p > 0.001$ . The scintigraphic control showed homogenous distribution of the material with no articular escape. In conclusion, we have seen significant improvement after synovectomy in hemophilic knees with 153-samarium and an increased biological effect with larger used activity. The clinical results of this procedure show excellent potential to decrease bleeding in the joint and to reduce the use of factor, but we observed no gain of movement in the most serious arthropathies.

### **Session B3**

### **Monitoring, Anesthesia, Post-Operative Pain Management, and Nerve Blocks in Hemophilia Surgery**

#### **B3.1: Central Venous Access and Monitoring in Hemophilic Orthopedic Major Surgery**

F RAFFAN

*Anesthesia and Intensive Care, Fundacion Santa Fe de Bogota, Colombia*

The patient with hemophilia who requires an operation represents a challenge for the multidisciplinary team that must handle this special class of patients, especially if this is major orthopedic surgery, where they are exposed to serious bleeding. Although hemophilia is a disease widely recognized in any one of its forms, publications that direct their handling from the anesthetic point of view are rare. From the general point of view, major orthopedic surgery requires invasive monitoring to optimize the perioperative management, which is accentuated in the hemophilic patient. In this way, central venous catheterization (CVC) entails an increased risk, which is why the im-

plementation of safety strategies becomes necessary. We, in the FSFB, have implemented an algorithm for the safe placing of central venous catheters in hospitals in our country (Colombia), where ultrasound exists. Based on evidence, we suggest the use of ultrasound guided CVC in patients with hemophilia to prevent complications, reducing the time of insertion and decreasing morbidity related with the procedure.

#### **B3.2: Pre-operative Evaluation: Risk Assessment**

LP SOLIMENO, OS PERFETTO, C CASADEI AND G PASTA

*Orthopaedics and Traumatology Department and Angelo Bianchi Bonomi Hemophilia Center, IRCCS Maggiore Hospital Foundation, Milan, Italy*

Orthopedic surgery in patients with hemophilia should be planned by the surgeon together with the hemophilia specialist and the physiotherapist in order to establish a long-term program that integrates surgery with medical and rehabilitative interventions. Orthopedic surgery should be performed in centres that are able to provide hematologic expertise, medical support, and laboratory monitoring of coagulation throughout the period of hospitalization. The pre-operative assessment should include medical features (i.e. definition of the severity of the coagulation factor deficiency, information on inhibitor history and current inhibitor status, evaluation of concomitant liver disease, and HIV infection status) and surgical aspects such as X-ray pre-operative planning to evaluate axial deviation and joint deformity. CT scan is recommended to accurately assess bone stock. The authors will discuss all these items in detail.

#### **B3.3: The Use of Thromboelastography in the Hemophilic Patient**

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In the present century there has been an increasing demand on transfusion services as complex surgery associated with significant blood loss and coagulopathy has become more common. Orthopedic, trauma, transplants, obstetric, and cardiovascular interventions have an important impact on the use of blood components. Therefore, the use of methods that help evaluate the bleeding patient on-site based care with a simple and suitable device is recommended. Kang et al pioneered the use of thromboelastography (TEG) for liver transplantation and cardiovascular surgery, and found that the total number of transfusions was reduced when patients were managed with TEG, compared to a control group without this monitoring. This improvement was attributed to improved coagulation, as a result of more intensive monitoring. **TECHNICAL ASPECTS:** A thromboelastogram is a computerized device that measures the visco-elastic properties of blood, from a dynamic and global perspective, since it documents the integration of platelets within the coagulation cascade. This test is performed in vitro, having used a bucket or wins, where they deposit 0,36cc of blood, which

is previously graduated according to the temperature of the patient and during a period of 10 seconds. The variations obtained by the characteristics of the clot and its evolutionary stage are registered by an electromechanical transducer, which turns the rotation of the pin into an electrical signal, documenting the different stages of the coagulation, that is to say, the formation of fibrin, the retraction of the clot, the platelet aggregation, and the lyses of the clot. The patient with hemophilia is prone to have serious bleeding during major surgery, as well in the post-operative period. Hence, the use of TEG could be useful in the management of these patients. It has the potential to provide accurate diagnosis of the coagulation status and it enables rationalization of the use of clotting factors, platelets, and antifibrinolytics, decreasing the need for empirical therapy.

#### **B3.4: Post-Operative Pain Management for Musculoskeletal Surgery : Conceptual Framework and Practical Approaches**

G BLAMEY

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This presentation will address issues surrounding the difference between post-operative pain and the acute and chronic pain experienced by people with hemophilia as a result of musculoskeletal bleeding episodes and joint arthropathy. Management techniques that may be employed by the individual with hemophilia or the physiotherapist will be the focus, as well as how those techniques may best be implemented in order to provide the most effective relief from pain. The positive attributes of pain will be explored as well as the importance of educating people with hemophilia to recognize that the experience of pain post-operatively is not equivalent to active bleeding.

#### **B3.5: Peripheral, Post-Operative Nerve Block**

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Traditionally, pain has been a major concern for both patients and caregivers, but with the variety of peripheral nerve blocks available, pain can be managed much more effectively. Peripheral nerve blocks are used in selected procedures as part of a pre-emptive and multimodal or balanced analgesia technique to provide safe intra-operative and post-operative analgesia with minimal side effects. With advancement in the understanding of coagulation mechanisms and the improvement of factor VIII and factor IX replacement therapy, regional anesthesia may be used in a patient with hemophilia provided that FVIII and FIX activity levels are maintained above 30% throughout the peri-operative period. Repeated bleeding into a joint can cause chronic synovial inflammation and progressive cartilage degeneration, known as hemophilic arthropathy, and which often requires a challenge in the peripherals block technique.

Under close supervision of the anesthetic and hematology teams, similar safety can be achieved while using central and peripheral nerve blocks (PNBs) for elective operations. However, in the literature there are only a few case reports to support wider use of these techniques in hemophilia patients. PNBs are likely to have a lower risk of hematoma formation and neurological deficit. Use of a short bevelled needle, peripheral nerve stimulator, ultrasound technique improve the success rate and further reduce the risks. Communication amongst all the members of the multidisciplinary team is vital for better outcomes in this group of patients.

### **Session B4**

#### **Free Papers IV: Surgical & Invasive Procedures**

##### **B4.1: Arthrodesis of the Ankle and Subtalar Joint in Patients with Hemophilia**

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There have only been a few studies in the literature that reported on the outcome of ankle arthrodesis in patients with hemophilia. Furthermore the number of patients is usually low and the operative technique is not uniform. The aim of this study is to evaluate the outcome of surgery in hemophilic arthropathy of the ankle and subtalar joint using internal fixation. From 1983-2006, 20 fusions were performed in 13 patients with advanced hemophilic arthropathy of the ankle and subtalar joints. There were 11 ankle fusions, one isolated subtalar fusion, and eight combined ankle and subtalar fusions. Three of the latter had a subtalar fusion at a second operation. The mean age at operation was 38.7 years and the mean follow-up was 9.4 years. Pre-operatively the mean modified Mazur score was 47.7. In the majority of cases the ankle fusion was stabilized by two crossing screws, while for the subtalar fusion either staples were used, or the tibiotalar screws were extended to the os calcis. Arthrodesis of the ankle was successful in all but one patient, who was revised and progressed to fusion. The mean post-operative modified Mazur score was 94.9. There was also one painless failed fusion of the subtalar joint, which has never been revised. There was no recurrent bleeding and no deep infection. Arthrodesis using internal fixation with cross screw or staple fixation proved to be an effective and reliable method for fusion of the ankle and subtalar joint in patients with hemophilia.

##### **B4.2: Ankle Prosthesis in Bleeding Patients: Our Experience with 24 Patients with AES Prosthesis, 1 to 7 Years Follow-up**

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Hemophilic arthropathy of the ankle is well tolerated for a long time, often bilateral with consequential effects on the other joints, homo and contralateral. With ankle prosthesis, we tried to improve the quality of life of bleeding patients. Clinical follow-up of 34 ankles: some HIV or hepatitis C, others both; 24 cases AES ankle prosthesis from 1 to 7y. (average 32 m.); 8 cases bilateral; 8 cases unilateral; 10 cases awaiting surgery now. **Methods:** We work with three specialities, together, in close collaboration: The pre-operative check-up needs X-rays in weight-bearing, **Protocol:** The factor VIII or other is indicated daily by the CRTH on the result of daily blood analysis. **Surgery:** total anesthesia, AES ankle prosthesis: + complementary surgery. **Patient stay:** One week in Orthopedics; 6 weeks in rehabilitation; then to physiotherapist. **Complications:** One false embolism of the lung; one respiratory distress; 1 total instability; revision by arthrodesis. **Results:** The clinical check-up (score of AFCP similar to AOFAS score on 100 Pts.). Not only on the functional capacity but also on the recovery of the autonomy, stand up, way of walking, no crutches, up and downstairs, driving, sports, etc. **Conclusion:** In the beginning we started carefully on the indication of ankle prosthesis; it was a new, indeed, experimental surgery. Now we know that ankle prosthesis is a new functional surgery that brings to the bleeding patient as other prosthesis a marvellous advance in haemophilia and in orthopedic complication. The essential play of the three specialities together explains our good results and the few of complications. Now, our concept of ankle care in haemophilia is agreed by some centre in France and they send us patients to care.

#### B4.3: Upper Ankle Joint Prostheses in Hemophilia Patients

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Secondary arthrosis of the upper ankle joint (talocalcaneal joint) is one of the most frequent forms of hemophilic arthropathy. It is a secondary form of arthrosis caused not only by chronic synovitis and cartilage injury resulting from chronic recurrent intra-articular bleeds, but also due to the misalignment of the joint and abnormal joint stress. The consequences are manifest even in young patients and finally lead to subtalar joint ankylosis with the biomechanical disorder of foot drop (talipes equinus). In such clinical situations, implantation of a subtalar joint endoprosthesis is a viable alternative to arthrodesis. **The patient:** A male patient, 52 years of age, suffering from severe hemophilia A (residual factor VIII activity <1 %) with no inhibitor formation. **Course of events:** The patient has a history of several years of painful ankylosis of the right ankle joint and minor talipes equinus and suffers from symptomatic talonavicular ar-

throsis. Open joint cleansing considerably improved mobility in the upper ankle joint and alleviated the pain in the talonavicular joint. However, the recovered mobility of the arthrotic upper ankle joint also activated the patient's arthrosis, associated with severe pain. As there was no contraindication to upper ankle joint replacement, a cement-free prosthesis was implanted. Three months after surgery, the patient was mobile, with good foot rolling properties without orthopedic aids and without pain in his upper joint ankle. **Conclusion:** In terms of biomechanics, an upper ankle joint endoprosthesis is a superior alternative to arthrodesis in hemophilia patients. In order to minimize the complication rate, the treatment of hemophilia patients should be restricted to specially equipped interdisciplinary treatment centres with adequately trained and experienced surgeons as well as hemostaseologists.

#### B4.4: Total Knee Replacement in a Patient with a Below-Knee Amputation with Hemophilia

RSA HOUGHTON-CLEMMY, TKSINGHRAO, AND NJ GODDARD

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Total knee replacement is an established treatment for hemophilic arthropathy. We report the case of a 79-year-old male with hemophilia B and a below-knee amputation who underwent successful total knee arthroplasty in his amputated leg. The patient underwent below-knee amputation following a tibial fracture 41 years previously, developing arthritis secondary to frequent hemarthroses whilst remaining active with the use of a prosthetic leg. We had previously performed successful ipsilateral total hip replacement and contralateral total knee replacement. Arthroplasty has been documented twice for osteoarthritis in below-knee amputees with good functional outcome and has been advocated as an alternative option to arthrodesis or above-knee amputation. Its use has not been reported in hemophilia. Assessment was undertaken using a multidisciplinary approach. Tibial length was adequate for prosthesis insertion and extramedullary alignment. A cemented cruciate-retaining prosthesis (AGC) was used without synovectomy. The patient received continuous factor IX infusion until the fourth post-operative day. Prophylactic tinzaparin was administered until the patient mobilized. Post-operative recovery was uneventful and inpatient rehabilitation was achieved using a pneumatic post-amputation mobility (PPAM) aid, cryocuff, and full range of motion exercises. The patient enjoys independent pain-free mobilization and greater exercise tolerance with an excellent functional outcome.

#### B4.5: Follow-up of Primary Hinge Total Knee Arthroplasty in Patients with Hemophilia

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Total knee arthroplasty (TKA) is the treatment of choice for end-stage hemophilic knee arthropathy. We reviewed the results of all the primary TKAs in a group of hemophiliacs performed by two orthopedic surgeons with the collaboration of our hemophilia centre. Between January 1996 and July 2008, 33 primary TKAs were performed in 26 severe and moderate hemophilic patients, one with an inhibitor against clotting factor. The mean age at the TKA was 40 years (18 to 66). All except one had active hepatitis C infection; nine patients were HIV positive and at the time of TKA their CD4 counts were > 200. The prosthesis used was a hinge total knee replacement for anatomical specificities. The clotting factor replacement was standardized and no chemical antithrombotic prophylaxis was used, except for one patient who had ischemic heart disease. Manipulation under anesthetic was performed if patients had not achieved 70° flexion by the second month of active rehabilitation. Few hemorrhagic, no thrombotic, and no early infectious complications were observed. The main improvements were pain relief and more functional gait without flexion contracture. The mean follow-up was 7.2 years (0.5 to 13). We observed five aseptic loosening between 3 and 11 years (mean 7.3y) and two late infections in two HIV negative patients, 1 and 3 years after the surgery. **Conclusion:** The hinge total knee arthroplasty is suited for hemophilic knee arthropathy with good clinical results. The global survival of this prosthesis is good and has to be validated with more patients.

#### **B4.6: Incidence of Total Knee Replacement Infection in Hemophilic Patients with Negative HIV and HCV Positive Serology**

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**Objective:** This prospective study was to analyse the incidence of this infection in hemophilic patients with negative HIV serology. **Methods:** The study involved 4 total knee replacements in 10 hemophilia A patients with negative HIV serology, performed in a centre with just one surgeon. Twelve were primary surgeries, with two revisions in just one patient. Only one patient had positive HAV serology (10%), none had HBV, and 100% had positive HCV serology. The mean age of the patients was 41 (24 to 63) and the mean follow-up was four years (ranging from 12 to one year). **Results:** There was no early or late infection in this group. The Knee Society score was excellent or good in 90% of the patients. The clinical information more outstanding is the higher mobility than a 90° in all cases, by the years the previous articular rigidity had better quality of life. **Conclusions:** Total knee replacement in hemophilic patients is a complex procedure with a predictable evolution and the potential to improve quality of life. In patients with negative HIV serology, we found no sign of early infection and no sign of late infection four years after treatment

#### **B4.7: The Use of Plasma-Derived Biomaterial in the Treatment of a Hemophilic Bone Cyst: A Case Report** M EL EKIABY<sup>1</sup> AND AR EL DESOUKY<sup>2</sup>

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**Clinical Picture:** A 12-year-old hemophilic patient was referred to our centre with swelling below the knee. An X-ray of the knee joint showed bone swelling with decreased bone density below the head of the tibia. The diagnosis was made as a hemophilic bone cyst. The cyst was almost round with a diameter of about 3-4 cm. **Management:** We have developed in our blood centre a procedure to prepare a plasma-derived biomaterial called platelet glue. The compound is formulated by mixing equal amounts of fibrinogen and platelet concentrate. When thrombin is added to the mixture, fibrinogen is transformed into a fibrin clot and platelets are activated and release their granular contents, among which are several growth factors. Surgery was planned under prophylaxis with FVIII concentrate. The outer layer of the tibia over the cyst was opened and the cyst was evacuated and then filled with a paste of the platelet glue mixed with artificial bone graft (hydroxyapatite and tricalcium phosphate pellets 5 mm diameter). The lower limb was cast between the knee and the foot for four weeks. Serial X-rays were performed immediately after the surgery, as well as one month and two months after the surgery. **Results:** The swelling below the knee disappeared and the cyst disappeared almost completely, with new bone formation in two months, as evidenced by the follow-up X-rays. **Conclusion:** Early diagnosis of hemophilic bone cysts, early intervention, and the use of platelet glue may help in improving the management of this serious complication.

#### **B4.8: Hip Surgery in Hemophilic Patients Under Low-dose Thromboprophylaxis**

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**Background:** It's well known that the risk of venous thromboembolism in patients undergoing total hip replacement is very high. Before thromboprophylaxis was used routinely, deep vein thrombosis (DVT) occurred in 40-60% of these patients. Although changes in surgical and anesthetic techniques and early mobilization may have reduced this risk, routine thromboprophylaxis remains extremely important and is the standard of care. **Aim:** We report a case of a patient with mild hemophilia A where total hip prosthesis was successfully implanted. **Methods:** Case report. **Results:** A 48-year-old patient with mild hemophilia A suffered a traumatic injury to the right hip (femoral prethorchanteric femoris dex.). He underwent surgery with implantation of total hip prosthesis. Purified freeze-dried human rFVIII (Immunate, Baxter) was used for supportive treatment starting with a dose of 50 IU kg<sup>-1</sup> body weight twice daily. High level of rFVIII was achieved between 120 and 160%. Supportive treatment was administered along with low-dose LMWH (20mg enoxaparin s.c. once a day). This treatment was continued the first seven

post-operative days until the patient was completely immobilized. The operation and the post-operative period passed without any complication. **Conclusion:** This is the first case of a successfully implanted total hip prosthesis in a hemophilic patient in our country. We used low dose thromboprophylaxis and had no complications. There are no data of such treatment in literature. So, one question is propounded: Since we performed hip surgery in a hemophilic patient with supportive treatment that provided conditions approximately the same as in a healthy individual, is antithrombotic prophylaxis really necessary?

#### **B4.9: Percutaneous Treatment of Discogenic Pain in Hemophilic Patients**

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In the general population, discopathy is an extremely common and painful disease. The treatment of discopathy in hemophilic patients can be complicated because of the bleeding risk. We present our experience in treating two slipped discs using a minimally invasive and low-cost procedure. Two hemophilic patients with high bleeding risk in surgical treatment were treated by percutaneous methods, with electromagnetic field using pulsed radiofrequency, obtaining an excellent healthy score in the follow-up. The surgical procedures directed at the intervertebral disk led, in a significant number of cases, to instability, which opened the way to arthrodesis as a definitive treatment option. Interventional radiology has provided new opportunities in the treatment of discogenic pain. The employment of steroid infiltrations with radiofrequency has revolutionized the prognosis of these patients. Using thin needles we can access the disc without neurological risks, thanks to the excellent anatomical information provided by the CT scanner.

#### **B4.10: Catastrophic E. Coli-Induced Necrotizing Fasciitis of the Lower Extremities in a Hemophilic Patient with HIV**

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A 30-year-old hemophilic man with HIV, hepatitis B, and hepatitis C developed left thigh minor trauma while he was in Malaysia. He noticed redness, swelling, and pain at the site of trauma that progressed quickly to involve the whole left lower limb. He injected himself with factor VIII and returned to Saudi Arabia four days later. The patient presented to the emergency room (ER) with excruciating pain in the lower extremities with necrotizing skin changes, bluish discoloration, and in severe septic shock. Immediate supportive measures were initiated including antibiotics, IV fluids, dopamine, ICU transfer, and intubation. The patient's condition progressed very rapidly as the necrotizing changes in the skin spread to the upper thigh and extended to the perineum and anterior abdominal

wall. He was taken directly to the operating room to undergo left lower limb amputation and disarticulation of the left hip. The patient returned to the ICU in a very advanced stage of shock with multiple organ failure including respiratory failure, acute respiratory distress syndrome, acute renal failure, lactic acidosis, liver failure, and DIC. The patient went into cardiopulmonary arrest and died 27 hours after his presentation to ER. The wound culture revealed *E. coli*. This case demonstrates the potential serious outcome of trauma in immune-compromised hemophiliacs and represents the second published case of necrotizing fasciitis in a hemophilic patient with HIV infection.

**Wednesday, April 29, 2009**

#### **Session C1**

#### **Superior Extremity Involvement in the Acute, the Chronic, and Sequelae**

##### **C1.1: Physiotherapy for Recurrent Bleeds of the Elbow: Tips, Tricks, and Milestones**

P DE KLEIJN

*Department of Rehabilitation and Sports Medicine, University Medical Center, Utrecht, The Netherlands*

Although bleeding episodes occur most frequently in knees, ankles, and elbows, a PWH will react less adequately in case of an intra-articular elbow bleeding. This is simply caused by the fact that this is not a weight bearing joint, and functional consequences are less severe. This is even worse when the non-dominant side is affected. As the distal translation of the tibia is a known consequence after severe knee bleedings, less is known about the consequent force of the biceps muscle on the radial bone. As the enlargement of the radial head after recurrent bleeding episodes is a common phenomena, we know this part of the elbow joint is often involved. The need for a temporary sling is not a common action to undertake in HTCs after PWH present with bleeding of the elbow. Guidelines, which include referral to a physical therapist, do not yet exist. Stabilizing exercises of the elbow joint and less frequent use of the sling, with special attention to the supination of the forearm and biceps participation in all actions, is a logical physiotherapeutic choice. The end result is full functional recovery, including leisure and sports activities. Fortunately major elbow bleedings seldom occur in PWH visiting our centre in the Netherlands. Two cases of failure in adolescents (male-hemophilia A and in female—von Willebrand disease), with extended limitations of an elbow, will be used to emphasize the value of using guidelines and functional exercise after every acute bleeding episode in every HTC worldwide.

### **C1.2: Diagnosis and Treatment of Synovitis of the Elbow**

M HEIM, R TIKTINSKI, Y AMIT, AND U MARTINOWITZ  
*National Hemophilia Center, Sheba Medical Center, Tel Hashomer Hospital, Israel*

The elbow is one of the most frequently involved articulations in patients with hemophilia. Within the elbow joint complex there are three separate articulations. The entire complex is surrounded by a common capsule and hence hemarthroses affect all facets of the joints. Pain is a common feature of a bleed within the joint and the elbow is held in a position of flexion. Repeated hemarthroses results in synovial hypertrophy. The persistence of the hypertrophic synovium results in osseous changes in young children whose growth plates are still open and progressive cartilaginous damage. The joint loses the ability to full extension very quickly and forced extension may cause additional damage. A bone scan is useful to quantify the extent of the synovial overgrowth. The hypertrophic synovium needs to be controlled and this can be achieved by cessation of the bleeding and protracted physiotherapy. In persons where the synovium continues to bleed and hypertrophy, invasive procedures will be necessary and this may be instituted by the introduction of a sclerosing chemical or a radioactive substance into the joint. Arthroscopic synovectomy or a formal debridement of the joint may be necessary. In persons with a hypertrophic radial head, this part of the radius may require resection for its change in size may provide a mechanical obstruction to a full range of motion.

### **C1.3: Role of Radial Head Resection in the Recuperation of Prono-Supination of the Forearm**

L DA ROCHA LOURES PACHECO  
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The elbow is the second most frequently affected joint in hemophilia. Chronic hemophilic synovitis of the elbow usually leads to enlargement of the radial head and severe arthropathy. A derangement of the proximal radioulnar joint secondary to hypertrophy and marginal irregularities of the radial head is the major source of pain, recurrent bleeding, and restricted forearm rotation commonly seen in patients with hemophilic arthropathy of the elbow. Limitations in forearm rotation often result in significant disability. Loss of supination of the dominant extremity interferes with eating, handling money, and personal hygiene. Excision of the radial head has been successfully used to improve the symptoms associated with advanced hemophilic arthropathy of the elbow. Excision of the radial head is accomplished by a transverse osteotomy of the radial neck, performed just below the level of the ulnar facet. An effort should be made to preserve at least part of the annular ligament in order to improve the stability of the remaining proximal radius. The excision of the radial head has been shown to provide sustained pain relief, reduction in the frequency of the bleeding, and improvement in forearm pronation and supination, though, minimal or no improvements in flexo-extension

range should be expected after the excision of the radial head. But it is a simple, safe, and effective procedure that should be considered in hemophilic patients with advanced hemophilic arthropathy of the elbow associated with disabling limitations in forearm rotation.

### **C1.4: What Can Surgery Do for the Established Flexion Contractures of the Elbow?**

M SILVA  
*Los Angeles Orthopaedic Medical Center, CA, USA*

The elbow is the second most frequently affected joint in hemophilia. While an overall progressive loss of elbow motion has been observed in patients with hemophilic arthropathy, significant fixed flexion contractures of the elbow have been described in severe hemophilic patients as early as during their third decade of life. The development of flexion contractures of the elbow in patients with hemophilia is the result of the recurrent bleeding inside the joint, with subsequent changes to the joint capsule and the articular surfaces. The presence of arthrofibrosis (a phenomenon commonly seen in hemophilia patients in whom the hyperplastic synovial membrane undergoes metaplasia to dense fibrous tissue), the presence of osteophytes over the olecranon fossa and coronoid, and the presence of humeral trochlear groove deepening, with ulno-humeral impingement, are usually responsible for the presence of fixed flexion contractures in these patients. The surgical options to correct fixed flexion contractures in hemophilic patients are limited, and the reported results are not encouraging. The excision of the radial head, with concomitant synovectomy, has demonstrated significant increases in forearm rotation without improvements in the extension range. Even when an extensive anterior capsulotomy has been performed in addition to the radial head excision (with resection of osteophytes from the coronoid and olecranon), the improvements in extension range have been limited. An insight into the limited success of open surgical releases for the treatment of fixed flexion deformities in patients with hemophilia can be gained from the analysis of such procedures in non-hemophilic patients with post-traumatic flexion contractures of the elbow. Although there is increasing evidence to suggest that good results can be expected in most patients, specific subgroups of patients have demonstrated poor results. Those with narrowing of the joint space and anatomic changes of the articular surfaces, and those in who the contracture has been present for a long period of time. Both of these factors are present in patients with hemophilia. Interposition arthroplasty has been used in patients without hemophilia, but there is very limited experience in patients with hemophilia. Improvements in elbow flexion contracture have been demonstrated with the use of total elbow arthroplasty in hemophilic patients. However the risks of elbow arthroplasty in hemophilic patients, and especially the risk of infection, should be weighted carefully against the potential benefits in range of motion. The surgical treatment of fixed flexion contractures in hemophilic patients is challenging. A careful analysis of the patient characteristics, including the age of the patient, the severity of the hemo-



philia, the length of presence of the contracture, and the status of the joint, among others, is necessary to determine the surgical options.

### **C1.5: Acute Compartment Syndrome of The Forearm** R BERNAL-LAGUNAS

*Instituto Mexicano Del Seguro Social, Federacion De Hemofilia De La Republica Mexico*

Compartment syndrome (CS) is a clinical condition caused by increased pressure in a closed fascial compartment. It causes circulatory compromise and ischemia in the tissues, therefore its diagnosis and treatment are urgent. The presence of this condition in hemophilic patients becomes more serious if the patient has factor VIII inhibitors. In hemophilic patients, CS usually originates after a minor trauma or spontaneous muscle bleeding. Patients require special attention when signs and symptoms appear because their onset is faster when the deficient factor is not administered. The diagnosis of CS is merely clinical. Assessment of the compartment is necessary in small children and babies, polytraumatized, and obtunded patients for the diagnosis to be corroborated. Upon evidence of existent CS, action such as a cushioned splint, localized ice, optimal level (40-60%) administration of the deficient factor, and frequent monitoring the presence and evolution of signs and symptoms, should be taken. In these patients, long-term observation (24 to 72 hrs) is recommended, hoping the clotting cascade to be normalized for the patient to have a satisfactory response. If not, it will require surgical resolution through a fasciotomy, having corrected the serum levels of the deficient factor (70-100%). The patient with high levels of inhibitors will require special treatment on a case-by-case basis. The reported complications in hemophilic patients are Volkmann contracture, residual muscle contracture, and ossific myositis. The literature contains very few articles on this topic.

### **C1.7: Conservative Management of the Acute Shoulder Bleed**

P NARAYAN

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The shoulder is the most mobile joint in our body but nonetheless is structurally insecure. The ball-shaped humeral head glides and rotates on the shallow glenoid cup of the scapula. A limited amount of passive stability is provided by the glenoid labrum and ligaments on the anterior and superior surfaces of the capsule of the shoulder joint. This means that the primary source of stability in the shoulder is provided by balanced muscle function. In the PWH, the most common joints affected in the upper limbs are the elbow and the shoulder. There are more commonly muscle bleeds around the shoulder, which are difficult to diagnose clinically and have a high rate of recurrence in varying intensities. This presentation will outline a protocol for the physiotherapeutic management of the acute shoulder bleed. The discussion will include case studies. It will highlight how physical therapy treatment with orthoses can be tempered to suit the repetitive

bleeding shoulder. There will be a focus on how bleeding shoulders can affect quality of life. The role of lifestyle alterations in managing shoulder bleeds, particularly in countries where factors are scarce, will be highlighted. The role of networking with family and workmates of PWH to improve compliance to treatment is emphasized in this talk.

### **C1.8: Shoulder Arthroscopy in Hemophilic Patients**

OS PERFETTO, G PASTA, C CASADEI AND LP SOLIMENO

*Orthopaedics and Traumatology Department and Angelo Bianchi Bonomi Hemophilia Center, IRCCS Maggiore Hospital Foundation, Milan, Italy*

The shoulder has never been considered a target joint in hemophilic patients, even if many adult hemophiliacs suffer from shoulder symptoms. Repeated articular bleeding leads to a hypertrophic and hypervascularized synovia. Simultaneously, intra-articular bleeding leads to direct damage of articular cartilage mediated by proteolytic enzymes, cytokines, and oxygen metabolites. Synovial and articular degenerative processes influence each other, leading to end-stage hemophilic arthropathy. The clinical picture of hemarthrosis is characterized by pain, swelling, and limited abduction. Rotator cuff tears are a common component of hemophilic arthropathy of the shoulder with consequent positive impingement signs, even if swelling and disabling pain are the surgical indication to shoulder arthroscopy. The authors reviewed their shoulder arthroscopic series in severe and mild hemophilic patients.

### **C1.9: Management of Grade IV Arthropathy of the Shoulder**

G PASTA, E CRISTINI, OS PERFETTO AND LP SOLIMENO

*Orthopaedics and Traumatology Department and Angelo Bianchi Bonomi Hemophilia Center, IRCCS Maggiore Hospital Foundation, Milan*

Spontaneous bleedings occurring into joints and muscles represent the more common manifestations of hemophilia A and B. The joints most often affected are knees, elbows, and ankles, however, many adult hemophiliacs suffer from shoulder symptoms. Recurrent bleeding into joints causes synovial proliferation and the progressive destruction of both cartilage and bone. The final result of these changes is referred to as chronic hemophilic arthropathy which is characterized by pain, stiffness, and deformity. Moreover, rotator cuff tears are a common component of hemophilic arthropathy of the shoulder with consequent positive impingement signs. Hemophilic arthropathy can be prevented by giving regular prophylaxis and implementing physiotherapy programs. The first treatment option in hemophiliacs with recurrent shoulder hemarthroses and/or chronic synovitis is synovectomy, shoulder hemiarthroplasty or arthrodesis being limited to the patient with advanced stage of arthropathy and chronic pain. The authors reviewed the role of orthopedic surgery in the treatment of advanced hemophilic arthropathy of the shoulder.

### **C1.10: Total Shoulder Arthroplasty in Patients with Hemophilia**

G PASTA, OS PERFETTO, E CRISTINI AND LP SOLIMENO  
*Orthopaedics and Traumatology Department and Angelo Bianchi Bonomi Hemophilia Center, IRCCS Maggiore Hospital Foundation, Milan, Italy*

Although knees, elbows and ankles are the most often affected joints in hemophiliacs, a number of patients suffer from shoulder symptoms as the result of chronic hemophilic arthropathy. Recurrent bleeding into joints causes synovial proliferation and the progressive destruction of both cartilage and bone, commonly associated with rotator cuff tears. The final result of these changes is characterized by pain, stiffness, deformity and positive impingement signs. Hemophilic patients with such clinical and radiological features remain a difficult challenge in shoulder surgery and the ideal procedure has not yet been found. The severity of soft-tissue and bone destruction determines the choice of the surgical procedure, being the hemiarthroplasty preferred to avoid critical glenoid fixation. The authors reviewed the role of replacement surgery in the treatment of advanced hemophilic arthropathy of the shoulder.

### **C1.11: Rehabilitation of Total Shoulder Arthroplasty**

A FORSYTH  
*Penn Hemophilia and Thrombosis Program, Philadelphia, PA, USA*

Total shoulder arthroplasty is an option in the treatment of advanced hemophilic arthropathy and physiotherapy is an essential component of ensuring an optimal outcome. There are variable rehabilitation approaches, which tend to begin with an early focus on regaining range of motion and then advancing to strengthening and functional exercises. Treatment and exercise progression must also take into consideration concomitant conditions, such as possible rotator cuff repair or other surgical findings, bleeding episodes, and other hemophilic target joints. A review of the literature revealed several studies that highlighted rehabilitation of total shoulder arthroplasty. However, none of these articles were specific to hemophilia. There may be special challenges during this surgery due to characteristics of hemophilic arthropathy, so it can be expected that the rehabilitation program will need to be customized for each individual. Typically, there are several phases of rehabilitation described, which progress along an established timeline. There are goals, precautions, specific exercises, and criteria for advancement in each phase. For example, Wilcox et al published a rehab program which will be reviewed: Phase 1: Immediate Post-Surgical (POD #1 – 4 weeks post-op); Phase 2: Early Strengthening (begin at 4 weeks); Phase 3: Moderate Strengthening (begin at 6 weeks); Phase 4: Advanced Strengthening (begin at 12 weeks). During this session, we will review rehabilitation protocols for total shoulder arthroplasty and their application in hemophilia care.

## **Session C2 Formidable Tasks and Future Outlook**

### **C2.1: Pharmacological Protection of Cartilage from Blood-induced Damage: State of the Art**

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In the last decades more insight into the pathogenetic mechanism of blood-induced joint damage has been gained. This insight yields possible treatment targets, useful to reduce this co-morbidity of hemophilia. In short, when blood is present in the joint cavity it will be taken up by the synovial tissue, leading to chronic synovial inflammation. This leads to damage to the synovial tissue and to the production of pro-inflammatory cytokines, which have a destructive effect on the cartilage. Independent of synovial tissue, exposure of cartilage to blood results in adverse changes in chondrocyte activity. Erythrocytes are taken up by macrophages, which upon this stimulation produce IL-1 $\beta$ . IL-1 $\beta$  activates the chondrocyte and thereby increases the production of hydrogen peroxide by these cells. The hydrogen peroxide reacts with iron from the erythrocytes, forming hydroxyl radicals, that induce apoptosis of the chondrocyte and thereby the cartilage matrix turnover gets disturbed. The release of cartilage matrix components in turn can enhance the synovial inflammation. Thus joint bleeding leads to initially independent adverse changes in both the synovial tissue and the articular cartilage, but these subsequently influence each other: the synovial inflammatory changes enhancing cartilage damage and vice versa. Consequently, effective treatment strategies will have to affect both pathways. In search of possible pharmaceutical options, one can think of chelators that reduce the iron load, scavengers that protect against free radicals, several anti-inflammatory components, and possibly other tissue (cartilage) protective agents. All these possibilities have to be explored.

### **C2.2: Revising the Infected TKA**

J WIEDEL  
*Denver, CO, USA*

The rate of infection following total knee arthroplasty in the person with hemophilia has always been reported as higher than in the general population. This rate has been reported at 7.2 percent. With the occurrence of HIV infection, the incidence increased to reported rates of 11.7 to 18.7 percent and even as high as 26 percent in HIV positive patients with CD4 counts below 200 cells/mm<sup>3</sup>. In addition to HIV infection, other reasons for an increased incidence of late infection after TKA include hepatitis C and the risk of hematogenous infections. Frequent IV infusions and remote infections are a source of hematogenous infection. Making the diagnosis is critical to successfully treating an infection. Joint aspirations should be done and if positive, immediate intravenous antibiotics and surgical debridement should be performed. The prosthesis may be retained if it is stable, the symptoms lasted

for less than 72 hours and the organism is sensitive. The presence of chronic infection and resistant organisms will require a two-stage revision. Options other than reimplantation could include resection arthroplasty and arthrodesis. I will report on the Colorado experience of treating nine septic TKA in six patients. An incidence of 20%. The onset was seen from 1 year to 18 years after the TKA. Four patients (7 TKA) were HIV positive. One had arthrodesis and two resection arthroplasties were performed. Six TKA were successfully salvaged with aggressive surgical debridement, IV antibiotics and second stage reimplantation. The main risk for a PWH living with a TKA is late infection. Early diagnosis and aggressive medical and surgical treatment can successfully salvage this complication.

### **C2.3: Arthroplasty in Patients with High-Titer Inhibitor and Prosthetic Infection: Are the Rates Acceptable?**

N GODDARD

*Royal Free Hospital, London, UK*

The availability of rFVIIa for hemostatic cover has enabled surgery that was previously impossible. Globally, experience of performing orthopedic surgery in hemophilia patients with inhibitors is increasing. The surgery being performed has become more enterprising and is certainly not for the faint-hearted. Thrombotic complications are rare in inhibitor patients and surgeons frequently ignore the concept of thromboprophylaxis. However, bleeding complications and subsequent infection remain a concern. As there may be a bias in the literature towards describing positive outcomes, it is likely that failures of treatment are not well-reported and hence pertinent information on how to prevent failures is lacking. More knowledge relating to the incidence and type of bleeding complications liable to be encountered, together with further information about appropriate rescue treatment, would be valuable. The European Register of Knee Arthroplasty (Eureka), was established to collate retrospective and prospective data, and patients are to be followed for up to five years after their surgery. In general the results obtained to date have been good, but there remain serious concerns about the incidence of post-operative infections, which are probably related to potential problems with hemostasis. The increased risk, coupled with the vastly increased expense of hemostatic cover has led some to question the wisdom of performing surgery in these patients. Clearly there are some fundamental questions of health economics, but in addition we should provide guidance as to which patients are, or are not, suitable for joint replacement.

### **C2.4: Is the Bleeding Tendency in Hemophilia B Less Severe than in Hemophilia A?**

PLF GIANGRANDE, R ALIKHAN, J O'NIONS,

A NAVARATNARAJAH, D KEELING, G HALL

*Oxford Haemophilia and Thrombosis Centre, UK*

The distinction between hemophilia A and B was first made by various groups in the period 1947-1952. Al-

though early clinical reports suggested that the bleeding tendency in hemophilia B was less severe, the conventional view held in recent years has been that both conditions are associated with the same bleeding tendency. However, the results of several recent studies challenge this view. A review of Swedish patients (Schulman et al, 2008) for the period 1990-1999 documented that the hemophilia severity score (HSS) was higher in severe hemophilia A than in severe hemophilia B (median 0.50 vs. 0.24;  $p=0.031$ ). Data from Canada (Pai KM et al, 2005) reported that patients with hemophilia A bled more often and appeared to use more factor concentrate than those with hemophilia B. A review of Scottish patients showed that admission rates for patients with hemophilia A were two- to three-fold higher than for hemophilia B at all levels of severity (Lowe GDO, 2008). We reviewed data relating to our own patients in Oxford involving 85 new diagnoses of hemophilia among children in the period 1989-2008 inclusive. These included 69 with hemophilia A (53 severe; 16 moderate) and 16 with hemophilia B (10 severe; 6 moderate). Preliminary analysis showed that the children with hemophilia B presented with their first symptomatic bleed at an age of 43 months compared to an earlier age of 31 months for those children with hemophilia A. 44 of 53 (83%) of the children with hemophilia A require treatment on a prophylactic basis compared to just 5/10 (50%) of those with hemophilia B. A multinational survey would help to provide a definitive answer to this important question.

### **C2.5: Revision of Infected Total Joint Arthroplasty in the HIV Positive Patient**

LP SOLIMENO, OS PERFETTO, C CASADEI AND G PASTA

*Orthopaedics and Traumatology Department and Angelo Bianchi Bonomi Hemophilia Center, IRCCS Maggiore*

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**Introduction:** Knee arthroplasty is the treatment of choice in advanced hemophilic arthropathy, even if a high complication rate has to be considered. The revision of a total knee replacement (TKR) is a challenging surgery considering lack of bone stock, ligamentous instability, stiffness, and tissues necrosis. **Methods:** We reviewed our experience since 1993 of 116 TKR performed on 98 PWH. Clinical data on 10 revision surgeries for deep infections performed at a single centre in 10 patients with hemophilia A or B (4 HIV positive) were reviewed. The Hospital for Special Surgery knee-rating score (HSS), data on knee flexion contracture, and range of motion were collected before and after surgery and after a short-term follow-up. **Results:** The median duration of follow-up after revision surgery was 27 months (range: 6-132). The two-stage exchange technique was successfully used in four cases of deep infections. For the persistence of infection, of the remaining six cases: three needed resection arthroplasty and three arthrodesis. **Conclusions:** Our results show that knee revision arthroplasty is often complicated with infections. The higher risk of post-surgical infection in hemophiliacs could be correlated to the prolonged post-operative bleeding and the presence of chronic infections.

## **C2.7: The Future of Hematological Management of Hemophilia: What is Brewing?**

M EL EKIABY

*Shabrawishi HTC, Cairo, Egypt*

Hemophilia is a global health problem. Tremendous efforts to control the disease as well as improvement of the quality of life of patients with hemophilia have been done. Today problems of patients of hemophilia are variable according to their country development index. While patients in the developed world are put on elegant protocols of prophylactic safe factor concentrate therapy and enjoy the highest level of specialized medical facilities, their colleagues in the developed world are still striving for proper diagnosis and availability of safe treatment products. Yet, patients in the developed world are not without problems. Inhibitor development and its etiology is one of the problems that is under intensive investigations where multicenter prospective studies are under design. New recombinant factor concentrates, that are claimed to be less immunogenic, are about to be launched. Long-acting factor concentrates are under development, which may improve prophylactic therapy programs. New inhibitor bypassing medicines are in phase two clinical studies. Genetic therapy, though still not close to success, is ongoing. The developing world is also showing a lot of activities to improve hemophilia care. In collaboration with WFH many country programs are active to diagnose new cases of hemophilia. The concept of comprehensive hemophilia care centres, and the integration of hemophilia care into national health system, is gradually improving care delivered to hemophiliacs in this part of the world. A new method for pathogen inactivation of plasma and cryoprecipitate using a medical device will enable blood banks in the developing world to use their local blood resources in a safer way for treatment of patients with hemophilia and help to maintain the supply and availability. The improvement of hemophilia care around the world is in a continuous active process, which insures that the hope for a permanent cure will be achieved one day.

## **C2.8: Unintended Consequences of Modern Treatment.**

CK KASPER

*Orthopaedic Hospital, Los Angeles, CA, USA*

The introduction of concentrate and self-infusion by the early 70s improved control of bleeding and liberated patients and their families. Hemophilia centres saw their patients less often and sometimes lost the ability to track hemorrhages and concentrate consumption. It became increasingly difficult to promote such services as physiotherapy and counseling. Payment for hemophilia care has improved over recent decades; however, where it improves too much, substandard providers start to compete for patients, and where it improves too little, replacement of dedicated caregivers becomes difficult. Hemophilia patients now are expected to have a near-normal life span and to reproduce at a near-normal rate, but that leads to an increased prevalence of mutant genes in the general population.

## **SCCOT Plenary Session**

## **Everything You Wanted to Know about Hemophilia and Were Afraid to Ask**

### **Hemophilia: Past, Present, and Future**

PM MANNUCCI

*Angelo Bianchi Bonomi Hemophilia and Thrombosis Center, Department of Medicine and Medical Specialties, University of Milan and IRCCS Maggiore Hospital, Mangiagalli and Regina Elena Foundation, Milan, Italy*

In the last few decades, the management of patients with hemophilia has witnessed dramatic improvements, through the larger availability of safe plasma-derived and recombinant products for replacement therapy. Another important step forward is the larger implementation of primary prophylaxis in children. Currently the main problem in patients with hemophilia is the onset of antibodies inactivating the infused clotting factor (inhibitors), even though immune tolerance regimens capable of eradicating inhibitors and the availability of products that bypass the intrinsic coagulation defects has dramatically improved the management of these patients. Cure of hemophilia through gene transfer is being attempted, but it is relatively far from being implemented on a large scale. It is likely that further improvements in replacement therapy will occur soon, through the availability of new weapons such as factors VIII and IX with longer half-lives, more potent bypassing agents, and factors extracted from the milk of transgenic animals.

### **Clinical Manifestations and Management of the Acute Articular Bleed**

LP SOLIMENO, OS PERFETTO, C CASADEI AND G PASTA

*Orthopaedics and Traumatology Department and Angelo Bianchi Bonomi Hemophilia Center, IRCCS Maggiore Hospital Foundation, Milan, Italy*

Acute articular bleed is the most common and disabling clinical manifestation of hemophilia, and the complications of repeated bleeding bring the patient to the attention of the orthopedic surgeon. Hemarthrosis is commonly preceded by the patient's perception of an aura or tingling sensation in the joint. The joint is generally in slight flexion and any movement is painful and restricted. The most important initial step in the management of acute hemarthrosis is rest, ice, and factor replacement as soon as possible, preferably when the patient recognizes the aura. Arthrocentesis consists of joint aspiration in order to drain bleeding and is indicated in selected cases.

### **Diagnosis and Management of Chronic Synovitis**

M HEIM, R TIKTINSKI, Y AMIT, U MARTINOWITZ

*National Hemophilia Center, Sheba Medical Center, Tel Hashomer Hospital, Israel*

Chronic synovitis is a clinical sign of failed conservative management of the patient with hemophilia. The hypertrophic synovium represents the destructive element that will

eventually result in malfunction of the articulation. Diagnosis is based upon clinical examination and imaging modalities. The clinical picture may be confusing, for the joint may be hot and swollen or look normal but extremely painful, with a pain-relieving effect after factor infusion. Chronic contracted joints may still contain 'pockets' of synovium that may bleed without external clinical signs. Joint cartilage is damaged by excessive proteolytic enzymes released from the synovium and with the passage of time, joint destruction will prevail. The treatment depends upon the anatomical and functional status of the joint. Joints that still have a functional range of motion will undergo procedures for their preservation while those that are excessively destroyed will need to be changed. Hypertrophic synovia can be destroyed by chemical or radioactive substances or can be removed from the joint surgically.

### Muscular Bleeds and Their Consequences

K MULDER

*Department of Physiotherapy, Children's Hospital, Winnipeg, Canada*

Intramuscular bleeding is second to hemarthroses as a clinical presentation in hemophilia. Muscle bleeds tend to be under-recognized and under-rehabilitated. Uncontrolled bleeds into deeper compartments can give rise to neurovascular compression with subsequent tissue necrosis. Calcified hematomas and formation of hemophilic pseudotumors are serious long-term complications of repeated muscle bleeds. Acute muscle hematomas can result in painful inhibition of muscle function and/or muscle spasm that limit joint function. Once the bleeding is stopped, careful assessment and rehabilitation are necessary to restore muscle length, to regain strength, and to minimize formation of contractures, which can also adversely affect adjacent joints.

### Articular Manifestations of Hemophilia

M SILVA

*Los Angeles Orthopaedic Medical Center, CA, USA*

Intra-articular hemorrhage is the most common musculoskeletal manifestation of hemophilia. Blood is not a normal constituent of the synovial fluid. The presence of blood inside the joint can lead to the progressive degeneration of the articular cartilage through metabolic, chemical, and mechanical pathways. Without proper treatment with clotting factor replacement, joint surface erosions secondary to hemarthrosis and chronic synovitis are often seen in early childhood, progressing to advanced arthropathy by late adolescence. The most commonly affected joints in hemophilia are the knee, the elbow, and the ankle. In early stages of arthropathy, most joints demonstrate similar pathologic changes, including osteopenia, narrowing of the joint space, subchondral irregularities and cysts, and osteophyte formation. As the degeneration progresses, specific changes are observed for each one of the joints. In the knee, angular deformities are commonly seen in advanced stages. While the

epiphyses are oversized, the diaphyses are usually narrower than normal. The tibia subluxes laterally and posteriorly on the femur, rotating externally, leading the patella to dislocate laterally. Absence of the tibial normal posterior tilt is commonly seen, and is usually the result of a knee flexion contracture developed early in childhood. Marked deepening of the trochlear groove is often observed, and can result in significant challenges when reconstruction is considered. Chronic synovitis of the elbow often leads to the enlargement of the radial head, which can impinge against the proximal ulnar facet. Because of its size and shape, the enlarged and irregular radial head can act as a mechanical block to forearm rotation, generating synovial impingement, hemorrhage, and pain. It is not uncommon to observe humeral trochlear groove deepening, resulting in ulno-humeral impingement and a reduced flexion-extension arc. In the ankle, the tibio-talar joint is often the first involved. Abutting anterior exostoses on the tibia and talus develop, and are often associated with an equinus deformity. A lateral tilt of the distal tibia, with resultant valgus malalignment, is often the result of growth asymmetry on the distal tibial physis. In about half of the patients with symptomatic arthropathy of the ankle, involvement of the subtalar joint can be demonstrated. The earliest subtalar erosions are seen at the posterior margin of the posterior facet. Avoiding joint bleeds is the key in maintaining healthy joints in patients with hemophilia.

### Hemophilia and Knee Function: What is the Difference Between Hemophilic and Healthy Children?

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**Material and Methods:** In a prospective multicentre study we examined 443 children with hemophilia (ages 3-18) treated in over 20 German hemophilia care centres since 2003. Data evaluation was done on 276 children with hemophilia A. We did a physical examination and a motion analysis of the knee joints on a treadmill and during squats. For further evaluation, we designed a motion analysis score for joint angle, angular velocity, and angular acceleration, and judged the motion curves by their regularity, rhythm, and sinusoid curve function. Another criteria of the score is axis deviation in two planes during gait and squat and the quality of the roll and glide mechanism. The same criteria were looked at in a group of 200 healthy age-matched children. For the first time, we can compare hemophilic and healthy children on an objective level. **Results:** In the whole study group, no one reached the optimal score (360 points). We found a high correlation of score with age in both groups. The

correlation was higher in the hemophilic group. Looking just at the roll and glide mechanism, there was no correlation with age at all. The inner knee motion seemed to be a more pathofunctional criteria. This was true for both groups. The gait score correlated with age, with a quotient of 0.71 for the hemophilic group and 0.54 in the healthy group. The difference was biggest in the younger children. At the age of three, the average score in the hemophilic group was 45 points out of 120, and in the healthy group 70 of 120. The difference diminished slowly during puberty. **Discussion:** What happens until the age of three in children with hemophilia? Are the parents sheltering too much and thus inhibiting motoric evolution? Are early bleeds or subclinical bleeds the cause of retardation in coordinative development?

### **Diagnosis and Treatment of Cysts and Pseudotumors**

F. FERNÁNDEZ PALAZZI

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It is important to introduce this nosologic entity, because it could be mistaken with a malignant tumor. The hemorrhage in adjacent bone originates located rarefaction areas and lead to a cystic hemophilic pseudo tumor. The etiopathogenic it is not well known when it occurs in hands and feet, it has a quick growth in young patients with epiphysics with fertile growth. An early adequate treatment of the pseudotumor permits more simple surgeries with a master recovery and return to the social and family activities. The initial conservative treatment with the administration of Factor VIII has in those tumors with uncontrollable growth to be accompanied by a surgical treatment. The pseudo tumor will be evaluated for existence of one or more cavities. When small the evacuation of the clot is required, by means of blood suction or a small incision, then the cavity is curettaged, and then fill cavity with fibrin glue (1 cc of fibrin glue for each 4 cc of blood obtained) which is produced in the Orthopaedic Unit National Haemophilia Center Municipal Blood Bank because of the lack of resources for its importation, of industrial preparations. If the tumor is bigger the cavity is filled through a laparoscope with bone grafts mostly commercial and fibrin seal. This percutaneous treatment is a non aggressive method, that requires less AHF than when a surgery is required. A real big pseudotumor invading aggressively the surrounding tissues must be treated by an open surgery performed by skill surgeons and in an specialized center.

### **No Lab Test Echoes Clinical Response**

CK KASPER

*Orthopaedic Hospital, Los Angeles, CA, USA*

We have puzzled over differences in clinical outcomes in patients who supposedly have the same deficient factor level. Factor measurement in one assay system differ notably from that in another system: which assay is correct? The issue is vital when children are selected at an early age for prophylaxis. The nature of the mutation un-

derlying a given factor level may convey the opportunity for improving the factor level with certain drugs, or may make a patient more vulnerable to inhibitors. The coagulation milieu consists not only of the deficient factor, but also of many other pro-coagulant and anti-coagulant factors, affecting the ability to clot. The strength of the musculature, inborn or deliberately developed, may affect the tendency to bleed. Co-existing non-hematologic disorders may provoke joint inflammation. Risk-takers, such as motorcycle riders, may accumulate damage, additive to that of "spontaneous" hemorrhages.

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# Novo Nordisk Lunch Symposium

Modern Management for an Optimal Outcome of  
Elective Orthopaedic Surgery in Haemophilia Patients with Inhibitors

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## WFH 11<sup>th</sup> Musculoskeletal Congress

Tuesday, April 28<sup>th</sup>, 2009 · 12:00–13:30 h · Hilton Cartagena, Colombia · Room Bolivar B/E

12:00–12:15 h	Lunch
12:15–12:20 h	Welcome & Introduction Adolfo Linás, Bogotá, Colombia
12:20–12:40 h	Optimising Elective Orthopaedic Surgery (EOS) in Haemophilia: The Multidisciplinary Approach Arlette Ruiz-Sáez, Caracas, Venezuela
12:40–13:00 h	A Consensus Protocol for the Use of rFVIIa in Pre-/Peri- and Post-Operative Management of EOS in Inhibitor Patients Paul Giangrande, Oxford, UK
13:00–13:20 h	Challenges in Post-Operative Care following EOS in Inhibitor Patients (including a Case Study) Adolfo Linás, Bogotá, Colombia
13:20–13:30 h	Questions & Concluding Remarks Adolfo Linás, Bogotá, Colombia

Lunch will be provided prior to the symposium.

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**Wyeth**