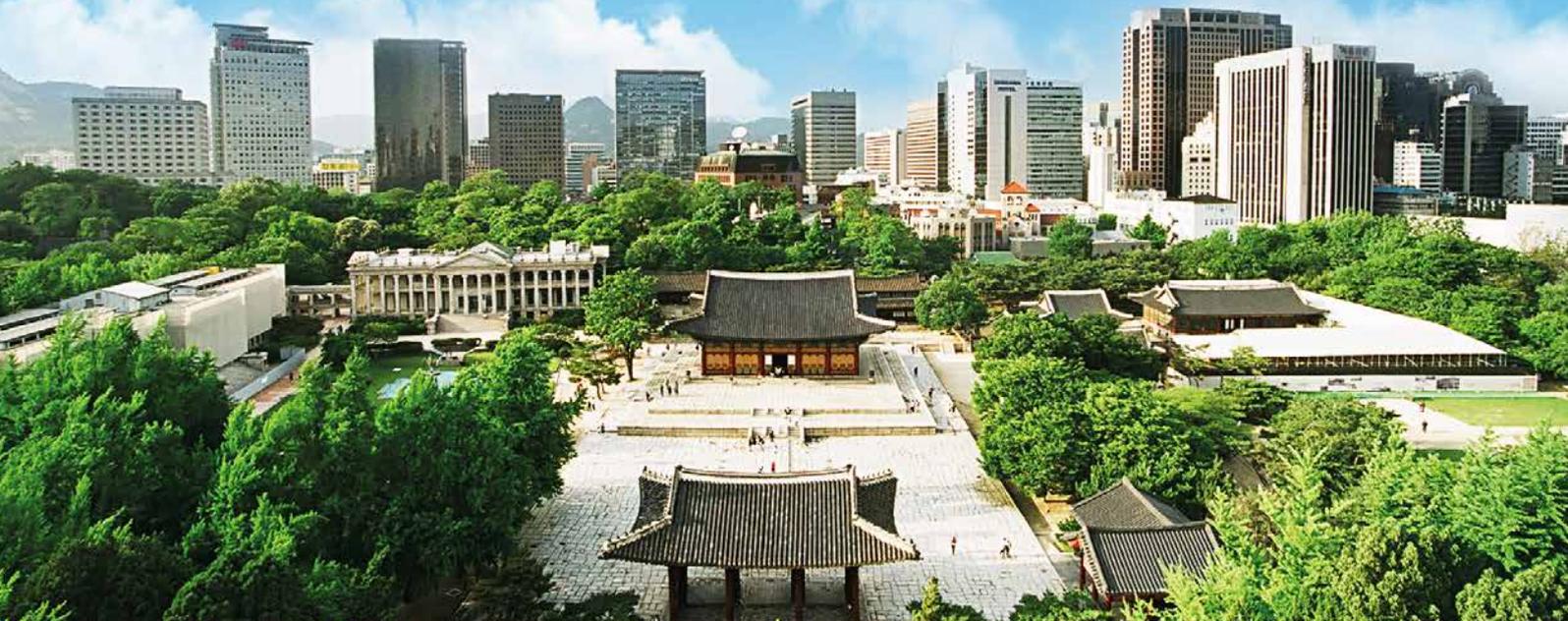


SEOUL



WFH 15TH INTERNATIONAL MUSCULOSKELETAL CONGRESS

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**FINAL
PROGRAM
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WORLD FEDERATION OF HEMOPHILIA
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Abstracts

Driving ahead with one eye on the rear view mirror

What's the future of Physiotherapy in Haemophilia care?

Helen Dixon

NZRP Wellington Hospital, New Zealand

The future of Physiotherapy in Haemophilia care is exciting! The possibilities are endless. We will discuss the advances in product development and how they will help to shape the future- less frequent infusions or possible increases in trough levels. The introduction of treatments for inhibitors has the potential for a very beneficial impact. What's coming and already here from the world of MSK physiotherapy, Chronic pain management, Rheumatology and Orthopaedics? Our colleges in these fields will help us to develop more comprehensive and patient focused treatment interventions.

The use of manual therapy and joint mobilization is used frequently in MSK physiotherapy. Is its use appropriate following acute bleed and during rehabilitation of the arthritic joint? All physical activity requires strength and technical skill from throwing an Olympic Javelin to getting up from the toilet. Appropriate strength, balance, endurance and skill training will help to prevent injury and bleeding episodes. Adherence to exercise regimes is always a challenging part of our practice- how can modern technology help with this? We all understand the value of bracing, strapping and foot orthoses during acute bleeding episodes we will examine its potential use for rehabilitation and injury /bleed prophylaxis. With improved methods of cortical mapping and imaging techniques the understanding of neuroplasticity in chronic pain is an exciting development. Graded motor imagery has developed due to this work and has a definite role within Haemophilia. There is now widespread use and acceptance of acupuncture by physiotherapists to treat chronic pain, could this help PWH.

History of the MSK group – Physiotherapy

Pamela Narayan

Wellington Hospital, New Zealand

It may not be wrong to say that Hemophilia ceases to be a haematological condition after diagnosis and becomes more of a musculoskeletal challenge to the treating health care team. Physiotherapy has been an integral component of treatment of Hemophilia almost since the disorder was first 'discovered' in history. Physiotherapy is an inexpensive but effective way to treat the musculoskeletal sequelae of frequent joint and muscle bleeds especially in regions of the world where there is suboptimal availability of clotting factor concentrates. Physiotherapy

plays an important role in the treatment of Hemophilia in the primary, secondary and tertiary aspects of the disease. It can help reduce the incidence of bleeding in joints and muscle (proactive rehabilitation) as well as help reduce morbidity as a consequence of bleeding. This presentation will make a humble attempt to revisit the history of the physiotherapy in terms of concept evolution in the musculoskeletal care of Hemophilia; how they have changed with time and the availability of blood products for treatment of hemophilia as well as our knowledge of wound healing, the pathophysiology of joint bleeds and the effect of blood on cartilage.

Physical Examination is the traditional method of musculoskeletal evaluation. With the evolution of treatment products, there was perceived a need for more objective, reproducible methods of evaluation. Thus, began a run of MSK outcome measures starting with a simple scoring system in 1985 introduced by the WFH. A very significant observation made in recent studies is that even children on prophylaxis who have no overt joint bleeds show signs of early joint damage on MRI imaging. This is bringing into focus the need for outcome tools which can pick up early joint disease without the use of expensive imaging techniques. Work in this area has been done to a large extent in recent years by the musculoskeletal professionals involved in the IPSPG group. The palette of treatment modalities within the scope of physiotherapy have increased from encouraging simple physical activities to the use of exergaming and other computer simulated programs to improve the MSK status of PwH. However in conclusion, the core principle of physical therapy management of bleeding disorders remains unchanged over history - that encouraging physical activity and exercise among members of the bleeding disorders family has a positive impact on the quality of life.

Skill Acquisition – at what cost?

Typical benefits of exercise: physical/ psychosocial

Greig Blamey

Health Sciences Centre, Winnipeg, MB, Canada

This talk will review the common benefits, both physical and psycho-social, that are normally associated with regular exercise for all people regardless of bleeding disorder status. Particular benefits that may be leveraged for those who have established musculoskeletal changes due to bleeding as well as the preventative effects of exercise to protect against joint and muscle damage will be explored. This talk will serve to frame the session as a whole, which will explore whether or not these exercise based adaptations and impacts are universally positive or whether they may be negatively overshadowed when taken to the extreme.

Phased approach to cross training for peak fitness in hemophilia

Eleonora Forneris

Children Hospital 'Regina Margherita', Turin, Italy

In the last decade the management of patients with haemophilia has changed considerably: physical activity and physiotherapy, associated with an appropriate personalized therapeutic treatment, can greatly improve the clinical condition of the musculoskeletal system, allowing a high standard of quality of life. It is known as physical activity induces an improvement on the psycho-physical state. Training is "the sum of all the stresses applied in a specific period of time which lead to functional and morphological changes of the organism" (Hollmann, 1963). The training methodology must take into account the complications related to the disease. Therefore, it is essential to proceed with an accurate evaluation, which will enable the professional expert to define the clinical and functional status of the subject, as well as the performance level (starting condition). Consequently, it is possible to plan the workout, according to the methods known in the literature. The fundamental criteria to be taken always into account are to adapt and customize the objectives and training loads, according to the potential of each individual. In general, the human body responds to variations of stress imposed, so we have to consider constantly the clinical signs and how muscles or joints are reacting to stress. This adaptation enables one to react to variation expected or unforeseen muscle loads and stresses more efficiently, which decrease risk of injury. The training load is the measure of the work that the athlete must perform in order to induce the necessary adaptations to increase performance. For there to be an adaptation the load must follow precise principles. It also depends on various factors, such as intensity, duration, density, volume and frequency of the stimulus, goals, methods, content and training tools. Periodization helps to reach a higher level of performance. The number, type, length of phases can vary but programs typically include a base, a preparation, and a peak period with each phase building on the previous one. The coordination of a multidisciplinary team (orthopedic surgeon, hematologists, a physiotherapist and a specialist in physical education and sport) is crucial, in order to constantly monitor throughout the period of training.

Pause for concern – The impact of physical exhaustion

Axel Seuser

Center for prevention, rehabilitation and orthopedics, Bonn, Germany

We have a lower limit where we start to benefit from an input and an upper limit above that overloading starts. The musculoskeletal system follows this basic law. Three major areas of concern can be described: 1. Structure; 2. Function; 3. Mind. For the musculoskeletal part the most important structures are cartilage, muscles, tendons and bones (synovia). In literature we find very different scales of limits for these structures; they have been measured and looked for in vitro and in vivo experiences. Function has totally different limits and is even more individual than our structural components. We know a lot about that from high performance sports, where the limits are pushed to its utmost possibilities to increase performance. Just looking at epidemiology we find out, that nearly no high performance

athlete leaves his athletic career unharmed. The overloading of function will lead - caused by basic biomechanical laws - to the overloading of structures. We already proved this true for our PWH (Bleeding – Synovitis – Hemarthropathy). It all starts with diminishing of movement/contraction quality. This is the start of overloading for muscles, tendons and ligaments and the last in the chain might be the cartilage. Emotions and motivation can be overloaded as well. The result is a burn out with all the known consequences like depression, loss of compliance and motivation. So our main task as physicians is to make sure, that our PWH, that are already born with a deficit and with a diminished resistance to loading do not harm themselves but do mainly profit of physical impact, that has to be done without any exhaustion. The task is clear but to get there is the most difficult part. It needs a highly individual approach to figure out the save limits within each PWH can perform physically unharmed at his personal best.

Transformational change from the patient perspective

Andrew Selvaggi

Personal trainer and hemophilia advocate, Melbourne, Australia

Andrew Selvaggi is a 29-year-old from Melbourne Australia living with severe haemophilia A with inhibitors. Andrew was diagnosed with haemophilia at 15 months old and developed inhibitors very soon after diagnosis. Between the ages of 2–10 years, Andrew lived with no treatment for his bleeds and because of this was wheelchair bound through his childhood and teenage years. As a consequence of having multiple target joint bleeds throughout his youth, Andrew developed long-term joint damage and arthropathy. By the age of 20 years, Andrew was in a wheelchair almost all of the time and was very overweight, with poor psychosocial well-being. Something had to change and one day he decided to get on an exercise bike. One year later and 30 kg lighter, instead of bleeding 1–2 times per week it was now 1–2 times every few months, and the wheelchair was no longer needed. Such a change in lifestyle was not without its challenges; heavy exercise for a person with haemophilia can be a shock to the body, but he has managed this with the support of a multidisciplinary healthcare team comprising doctors, nurses, physiotherapists, dieticians, personal trainers and rehabilitation experts. Andrew's lifestyle has remained fitness orientated, and this passion for health led him to become a personal trainer and help others to get fit and healthy. 9 years after undergoing a transformational change, Andrew now exercises 5-6 times per week, and hasn't had a bleed in over a year. With already pre-existing joint damage and both arthrodesis and arthroplasty procedures done, what is the true cost of High intensity training with someone who has Severe Haemophilia with Inhibitors?

High Impact – Structural concerns of physical training after arthroplasty

Adolfo Llinas

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Achieving outstanding athletic performance in persons with hemophilia who in addition have undergone joint arthroplasty implies considerations that are outside the conventional treatment

spectrum. Direct evidence from this high level of achievement is scarce in persons with hemophilia; however, one may extrapolate or speculate from similar clinical circumstances in other patient populations. For example, Multanen et al, in 2014 found that high impact training in previously sedentary postmenopausal women increased bone mineral density by approximately 0.6% (95% CI, -0.2% to 1.4%) during the first year, in contrast with a sedentary control group the mean loss in mineral density was -1.2% (95% CI, -2.1% to -0.4%). The change in baseline, body mass, and adjusted body mass change in BMC between the groups was significant ($p = 0.005$), whereas no changes occurred in the biochemical composition of the cartilage, as investigated by MRI. Balance, muscle force, and cardiorespiratory fitness improved significantly more (3% to 11%). From the skeletal perspective, high impact training under controlled conditions seems to improve the quality of bone without harming cartilage. However, from the perspective of an artificial joint, one can anticipate several challenges. Periprosthetic fractures may result from the high torque applied to the bone during sporting activities in the transition from the rigid bone-implant composite to the more flexible diaphysis. Furthermore, the characteristic steep slope that is associated with the training routines of high performance athletes hypothetically increases the chances of fatigue fractures in this location. Aseptic loosening is the most frequent cause of revision in total joint arthroplasty. An extensive body of literature has associated a lower implant survivorship in young populations, being youth a proxy for activity level. Based on this affirmation, the activity level associated with over-average athletic activity may carry a risk of increase frequency of implant loosening. A surrogate variable associated with activity and loosening is wear, especially polyethylene debris. In a group of 70 patients with hip replacement involved in high-impact sports, Ollivier et al in 2012 found the wear rate in the high-impact group was more than twice that of the rate in the low activity group (0.14 mm/yr vs 0.06 mm/yr). Twenty percent of subjects in the high-impact group underwent revision for mechanical failure (loosening) and 6.5% in the low activity group required revision THA.

Kinetic chain / upper limb

Global effects on lower extremity biomechanics as joint health declines

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Quantifying the extent of joint damage in patients with hemophilia (PWH) is critical to prevent disease progression and compare the efficacy of treatment strategies. In PWH the long-term consequences of repeated haemarthrosis include cartilage damage and irreversible arthropathy, resulting in severe impairments in locomotion. Quantifying the extent of joint damage is therefore important in order to prevent disease progression and compare the efficacy of treatment strategies. This presentation endeavors to improve our knowledge of the biomechanical consequences of multiple arthropathies on gait pattern in adult patients with

haemophilia using gait analysis. The more economical mode of walking is an intermediate strategy in centre of mass (CoM) displacement between extreme flatness and bouncy walking, that is, a CoM sinusoidal pathway of 3-4 cm amplitude. This intermediate strategy in CoM displacement associated with relatively straight-legged walking is achieved by peculiar movements in the lower limb joints that enable our legs to behave neither as stiff struts nor as compliant ones. These peculiar limb joint movements are called "gait determinants" because they were considered paramount in human bipedalism, as they enable a smooth progression of the body through small fluctuations of the CoM displacement to conserve energy. In PWH, with the loss of some of the major determinants as a result of multiple joint arthropathies, the strategy of vertical CoM displacement reduction is compromised leading indirectly to increase in metabolic expenditure, that is, increase of metabolic cost. This theory is confirmed as metabolic cost was dramatically increased in PWH and highly correlated to a loss in joint range of motion at ankles, knees, and hips level. In PWH with isolated ankle arthropathy, the increase in metabolic cost is proportional to ankle dysfunction, that is, the less the ankle power is generated, the more the metabolic energy is consumed and more efficiency of walking is impaired. The disruption to the normal walking process by an orthopedic disorder in PWH thus appears to generate mechanical and metabolic changes that follow a continuum linked to the progressive loss of mobility into the joints.

Clinical assessment – Examining the HJHS

Sharon Funk

PT, DPT, Physiotherapist, University of Colorado, USA

The 8-item (plus Gait) Hemophilia Joint Health Score (HJHS) was developed to identify key markers of joint structure and function in elbows, knees and ankles in persons with hemophilia, and to use this scoring system to quantify joint status. Joint properties include Swelling, Duration of Swelling, Crepitus, Muscle Atrophy, Flexion and Extension Loss, Objective Joint Pain and Muscle Strength in muscles adjacent to the joints most frequently affected with hemophilia joint bleeding. Assigning these joint properties a numeric value allows for quantitative analyses and the ability to track changes in the joint over time. The HJHS provides a systematic way of assessing the elbow, knee and ankle joints in persons with hemophilia and provides a valid and reliable method for scoring joint health status. The HJHS is an impairment measure, identifying primarily structural changes in joints affected by bleeding. Although some of the items on the scale are linked to joint function, the HJHS is not a functional outcome measure and it cannot tell us how a person is able to function within home and community. As the HJHS only includes the 6 index joints, it does not address shoulder or wrist function in the upper extremity. Impairment at one or both of these adjacent joints can impact elbow joint function and vice versa. Structural elbow joint changes leading to flexion contracture can increase abnormal anterior moment forces in the shoulder joint leading to shoulder pain and bleeding. The presence of synovitis in the elbow joint can impair normal ulnar nerve function affecting muscles both proximal and distal to the joint. Comprehensive evaluation of musculoskeletal status of upper extremity joints and the adjacent muscles in persons with hemophilia should include, but not be mutually exclusive to, the HJHS. Full orthopedic and functional assessments of all upper extremity joints should be

included and appropriate interventions implemented to provide optimal joint health and good outcomes. The HJHS is an adjunct tool for informing physiotherapy intervention with people with bleeding disorders. It should not be the focus, but rather another tool in the therapist's toolbox.

Surgical considerations for the lower extremity with multi-joint arthropathy

Gianluigi Pasta, MD

Centro Emofilia Angelo Bianchi Bonomi Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy

Persons with hemophilia (PWH) born before the 1970s have often developed hemophilic arthropathy in more than one joint and have to deal with pain, limited mobility, disability and adverse consequences on their quality of life. In such patients, orthopaedic surgery (joint replacement surgery or arthrodesis) of more than one joint of the lower limb might be indicated. In the general population, multiple-site elective surgery has been associated with an increased risk of perioperative and postoperative complications. In selected patients, improved perioperative management has recently reduced the number of adverse events, so this procedure have been re-evaluated and published studies have not reported any increased risk of complications. In PWH, multiple-site elective surgery could be indicated in order to reduce the time of recovery and return to professional activities as quickly as possible. Moreover, multiple-site elective surgery has the advantage of limiting the consumption of coagulation factors, resulting in a significant reduction of costs. In this setting, a tailored rehabilitation program is needed with special emphasis on the usefulness of hydrotherapy. Staged procedures should be considered when multiple-joint clinical improvement is possible after a single surgery. In conclusion, a thorough pre-operative examination and planning is mandatory in order to achieve good surgical results. Our recommendation is to perform multiple-site elective surgery in selected patients and in experienced multidisciplinary centres.

Surgical considerations for the upper extremity with multi-joint arthropathy

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Successful upper limb function depends upon a complex kinetic chain with the shoulder/elbow/forearm and wrist working in harmony with the objective of placing the hand in a functional position in space. Any break in this linkage will obviously have significant, and sometimes disproportionately adverse functional effects. Fortunately, in patients with haemophilia, the shoulder and wrist are least likely to be affected but loss of movement at the elbow, can have significant result in significant problems with regard to overall hand function. In the early stages this may just pose some minor inconveniences when dressing, discarding a tie and choosing shirts with none or few buttons for example. Later on there may be problems washing, shaving eating, or driving and ultimately can prevent the patient's ability to treat themselves by self-administration of factor replacement. The aim of this session is to highlight some of the less appreciated problems, to then recognise the early manifestations, and to try and implement potential solutions so as to maintain the patients quality of life, dignity and independence for as long as possible.

Manual therapy and therapeutic exercise for the elbow and shoulder in haemophilia; assessment, techniques and contraindications

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The elbow remains the second most affected joint in haemophilia. Clinically shoulder arthropathy is also relatively common, although it remains under assessed and under studied within the literature. What is even less clear is the effect of long term elbow arthropathy on motor control and joint position of the scapulohumeral and glenohumeral joints, and to what extent rotator cuff pathology is caused by or is a side effect of, impaired elbow anatomy and function. Manual therapy as an adjunct to physiotherapy interventions, that also include therapeutic exercise, is commonly cited as an effective treatment modality for relief of joint pain and joint stiffness, as well as improving ROM and function. A few 'schools of thought' exist about how manual techniques are applied (Maitland, Mulligan, Kaltenborn, Osteopathic) and as a result perceptions may vary about which is best. This in turn can lead to misunderstanding and thereby mis-application, of techniques which could in fact be harmful for PWH. This presentation will: highlight the assessment and reasoning process, by which manual therapy and exercise could play a role in the management of the elbow and shoulder in haemophilia; describe how this aids the clinical decision making process to choose manual therapy as an intervention; discuss the current understanding of mechanisms of action of manual therapy and why it may provide positive effect in some patients; examine the various techniques available; stimulate discussion around potential contraindications to some techniques as well as how techniques can be modified to work better with PWH; address rehabilitation concepts of the upper limb kinetic chain and how manual therapy can work alongside these (such and proprioceptive neuromuscular facilitation [PNF], multi-directional activity and muscle training with static and through range resistance).

Spotlight on South Korean issues

Management of musculoskeletal issues in Korea

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Until 30 years ago, only a few of university hospitals focused on haemophilia treatment in Korea, but full-fledged treatment and prevention of haemophilia began with the establishment of the Korea haemophilia Foundation (KHF) in 1991. Since then 13 haemophilia treatment centers were operated nationwide additionally, and a systematic service system for haemophilia patients have been built. By the end of 2016, the number of registered haemophilia patients was 2540, haemophilia A 1840, B 442, vWD 127, and others 131 respectively. The musculoskeletal functional disability is the one of the most deteriorating problem to devastate quality of life of haemophilia patients. From 1994 to 2016, 1436 cases (1239 patients) were carried out a broad variety

of orthopedic procedures; total hip arthroplasty 131 cases (102 patients), total knee arthroplasty 205 cases (185 patients), total ankle arthroplasty 10 cases (9 patients), total elbow arthroplasty 9 cases (9 patients), total shoulder arthroplasty 7 cases (7 patients), arthroscopic surgery 592 cases (556 patients), synoviorrhesis 222 cases (200 patients), Rifampin injection 69 cases (57 patients), and other procedures 216 cases (196 patients). Orthopedic surgeries have been performed mostly in three haemophilia surgery centers, and small number of procedures was performed in other hospitals. Most of post-operative rehabilitation are being carried out in three rehabilitation centers in the KHF. The KHF runs a dormitory in Seoul, most of patients who underwent any orthopedic procedure stayed a week or months in the dormitory for rehabilitation. This is a very efficient system for better functional recovery after surgery and patient convenience. The KHF runs also not only out-patient clinics and rehabilitation centers, but push ahead registry work and follow-up study of patients who carried out orthopedic procedures. This is one of the main reasons why all musculoskeletal issues of haemophilia patients are managed systemically and taken an excellent follow up measures in Korea. The Authors will introduce our patient managing system and trend of musculoskeletal procedures in Korea.

Mid-term outcomes and complications of total knee arthroplasty in hemophilic arthropathy: a review of consecutive 131 knees between 2006 and 2015 in a single institute

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Purpose The purpose of this study was to evaluate the mid-term outcomes and complications of total knee arthroplasty (TKA) in hemophilic arthropathy. **Methods** The study retrospectively reviewed 131 consecutive primary TKAs (102 patients) in single institute. The mean patient age was 41.0 years and mean follow-up time was 6.9 years. The clinical results were evaluated using the Knee Society knee and function score and the Western Ontario and McMaster Universities (WOMAC) score. The radiographic results were evaluated using the mechanical axis and position of the components. The complications or any additional surgeries were categorized according to the classification system of the Knee Society TKA complications. **Results** The average knee score increased from 34.4 to 80.9. The average function score increased from 37.0 to 70.3. The average WOMAC score improved from 66.0 to 24.2. The average flexion contracture significantly decreased from 17.3° to 4.7°, but the average pre and postoperative maximum flexion did not differ (80.9° vs. 85.6°). The average mechanical axis was varus 5.2° preoperatively and valgus 0.3° postoperatively. The coronal position of femoral and tibial components and sagittal position of those components were within 3° in 83.2%, 89.3%, 63.4%, and 73.3%. The complication occurred in 17 knees (13.0%). There were 7 bleeding and hemarthrosis. One medial collateral ligament injury of grade 2 required change of postoperative rehabilitation protocol. Two stiffness of grade 3 required unplanned admission and manipulation under anesthesia. Three deep periprosthetic joint infections were treated with 2 stage revision TKA. There were 4 periprosthetic fractures, and these were treated with

reduction and fixation for 3 distal femur and conservative treatment for one patella. **Conclusions** The mid-term results of TKA in end-stage hemophilic arthropathy were satisfactory with obtaining pain relief, improving function, and decreasing flexion contracture. Bleeding and PJI continues to be a major concern for TKA in patients with hemophilic arthropathy, and risk of periprosthetic fracture has to be taken into account for patient education and appropriate prevention. The TKA can be performed safely if performed under the strict supervision of an experienced hematologists and the meticulous attention to the technical demands by the experienced TKA surgeons. Level of evidence IV

Anterior osteophytes resection of the ankle joint to increase range of the ankle motion in haemophilic patients

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Introduction and Objectives: Haemophilic arthropathy is an important cause of morbidity in patients with severe haemophilia. The ankle joint is the second most common morbid site, and joint pain and stiffness are the most frequent findings. The stiffness of the ankle joint is the most functional disabling sequelae of gait cycle. The main causes of the joint stiffness are osteophytes bone block and the surrounding soft tissue contracture. Osteophytes bone block that occur along the anterior lip of the distal tibia and the neck of talus is a consistent radiographic finding in the ankle joint of haemophilia. The purpose of the present study was to investigate the role of anterior osteophytes bone block to cause limitation of the ankle motion in haemophilic patients. **Material and Methods:** Thirty-seven cases (Thirty-five patients) were enrolled. Two patients had bilateral procedures. Mean age was 30.7 years. Male was thirty-four and female was one. Haemophilia A was 28 patients, B was six patients, and one was factor VII deficiency. A mean follow-up was 41.2 months (22 to 95). No patient was lost to follow-up. Osteophytes resection was carried out through an anterior ankle approach. Some of patients were combined with joint debridement, synovectomy, and articular cartilage shaving. **Results:** The mean AOFAS (American Orthopedic Foot and Ankle Society) score had improved from 62.5 points preoperatively to 82.6 points at the final follow-up ($P < 0.01$). The mean VAS decreased from 8.2 pre-operatively to 1.6 at the final follow-up ($P < 0.01$). The mean dorsiflexion increased significantly from 4.1° pre-operatively to 10.2° at final follow-up ($P < 0.01$). Mean plantar flexion improved from 35.3° pre-operatively to 39.4° at final follow-up ($P < 0.01$). A total of 30 patients (89.2%) were very satisfied or satisfied with the outcome. **Conclusion:** Anterior osteophytes formation was very common, and impingement between the anterior lip of the distal tibia and the head-neck junction of the talus by osteophytes was one of the main causes to block ankle dorsiflexion. Anterior osteophytes resection of the ankle joint was very effective and valuable surgical procedure to improve function of the ankle joint in haemophilic patients.

Intermediate-Term Outcomes of the Total Ankle Replacement (TAR) In Patients with Haemophilic Arthritis of the Ankle Joint

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Introduction and Objectives: The standard treatment for advanced haemophilic arthritis of the ankle joint is ankle fusion. But TAR is still controversial as a treatment option. The purpose of the present study was to evaluate the intermediate-term outcome of TAR in patients with hemophilic arthritis of the ankle joint. **Material and Methods:** Eight haemophilic ankles (eight patients) treated with TAR were followed and retrospectively reviewed. Haemophilic A was seven, B was one. All patients were male. Mean age was 37.5 years, and mean follow-up was 6.5 years. The implant was unconstrained three-compartment ankle implant. **Results:** The outcome was assessed with clinical and radiological evaluations. The AOFAS ankle-hindfoot score increased from 29 preoperatively to 89 postoperatively in average. All patients were satisfied with results. VAS scale decreased from 8.1 preoperatively to 0.4 postoperatively. There were significant improvements in all outcome categories between the preoperative and postoperative evaluations. There was no loosening or osteolysis in radiological analysis. **Conclusion:** This study suggests that the intermediate-term outcome of TAR in haemophilic patient is comparable to that of other total joint replacements. In haemophilic arthropathy, TAR is a valuable and efficacious alternative to ankle fusion in an aspect of pain free mobile ankle arthrodesis.

Metal-on-Metal Hip Resurfacing Arthroplasty (HRA) in Haemophilic Coxitis

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¹Department of Orthopaedic Surgery, School of Medicine, KyungHee University Hospital at Gangdong and ²Department of Orthopaedic Surgery, School of Medicine, KyungHee Medical Center, Seoul, Korea **Introduction and Objectives:** HRA has several advantages; minimal bone resection, wider range of motion, minimal risk of dislocation, no thigh pain, and earlier rehabilitation. But attempt of HRA in haemophilic coxitis is very few. The purpose of the present study was to document the functional outcome following HRA in patient with haemophilic coxitis and to compare between HRA and Total Hip Arthroplasty (THA). **Material and Methods:** From Jan. 2006 to July 2014, Hip Resurfacing Arthroplasty in twelve haemophilic hip joints (eleven patients; 10 males and one female) with mean age of 37.9 years (range: 26-55) were performed. The mean duration of follow-up was 7.2 years. Eight hips were haemophilia A, two hips were haemophilia B, two hips were haemophilia factor VII. **Results:** The Harris hip score improved from 49.4 points to 98.2 points. All patients can make cross leg position and squatting position. In comparison analysis between HRA group and THA group in haemophilic coxitis during the same study period, the Harris hip score and range of hip motion at last follow up of HRA group were much better than that of THA group. Patient

satisfaction was also higher in patient who underwent HRA than that of THA. There were no cases of dislocation, neck fracture, infection, and thigh pain. Cup loosening, osteolysis, and implant position were not detected in radiological analysis. Blood Cobalt and Chrome concentrations were within normal limit. Adverse response to metal debris such as pseudotumor, ALVAL, or metal hypersensitivity were not significant issues in our study. **Conclusion:** HRA provides an excellent alternative to conventional THA in haemophilic patients.

Long term results of cementless total hip arthroplasty in patients with haemophilic arthropathy – minimum 11 years follow up results

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Introduction: Haemophilic arthropathy of the hip joint is less common than that of knee or ankle joints. Total hip arthroplasty (THA) in haemophilic arthropathy has been a valuable tool for relieving pain and improving functional status for patients. There is a paucity of long-term analysis, and most studies are results of cemented THA. The aim of this study was to analyze the clinical and radiographic results of cementless THA for treatment of severe haemophilic hip arthropathy. **Materials and methods:** Between Jan. 1995 and Dec. 2005, 50 cases of cementless THA were performed in 44 patients with haemophilic hip arthropathy. All cases were evaluated in a retrospective study with minimum follow-up of 11 years. Harris hip score, range of motion, pain score (VAS), complications and patients satisfaction was analyzed clinically. For the radiographic assessment, wear, osteolysis, implant loosening, pseudotumors, heterotopic ossification and postoperative infection were analyzed. **Results:** Harris hip score, range of motion, VAS score improved significantly. Patient satisfaction was very high. One case of pseudotumor, three cases of revision two cases of periarticular heterotopic ossification occurred. There was no case of periprosthetic infection. Postoperative coagulation factor requirement reduced remarkably. **Conclusion:** Even the relative high complication rate compared to patients with other causes, patient satisfaction was excellent. Cementless THA in patients with haemophilic hip arthropathy can bring a reliable pain relief and functional improvement in long follow-up analysis. However, an increased risk of infection and bleeding associated remains a cause for concern.

Synovitis

Synovitis in Hemophilia – Pathology/ Aetiology Pathology/aetiology

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Objective: The treatment of patients with hemophilia (PwH) has undergone a significant development through the last decades. Especially the introduction of a prophylaxis in young patients reduced the incidence of severe bleedings. Nevertheless, there still seems to be a lack of evidence of when and how to treat a synovitis in those patients. Since a synovitis after a bleeding

event is the start point of joint degeneration, it is crucially important to start an adequate treatment at the right moment. The intend of our work was, to develop a guideline based on the evidence in literature to support physicians, medical staff and patients in the treatment of synovitis in PwH. **Methods:** All member societies of the The Association of the Scientific Medical Societies in Germany (AWMF) were invited to take part in the work of our guideline group. The group consisted of experts in the fields of hemophilia and hematology, nuclear medicine, pediatrics, physiotherapy, orthopedics, radiology, and sports medicine. A total of 5.561 papers and abstracts were scanned and finally 1.166 was reviewed in detail by the working group. The content was evaluated and put in context to the working thesis. Results: We developed the underlying work for a guideline that may help physicians, medical staff and patients to treat synovitis according to the available evidence in literature. The result is subdivided in diagnostics, treatment and secondary prophylaxis of synovitis. Regarding the etiology of synovitis in hemophilia there is evidence that the first bleeding event results in synovitis. Besides a direct influence on the metabolism of chondrocytes, the inflammatory reaction induces hyperplasia of synovium, a direct apoptosis of chondrocytes and neoangiogenesis. This results in an increase of the inflammatory reaction, a degeneration of the joint and an increase of bleeding events in the affected joint. Nevertheless, the underlying work has been achieved in animal models. **Conclusion:** A combination of biomechanical und immunological reactions following initial synovitis leads to hemophilic arthropathy. The aim of treatment should be to avoid the first bleeding, detect any bleeding as early as possible, reduce the pain and the inflammatory reaction, and maintain the joint function.

Mild synovitis – Early clinical detection and treatment

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Repeated haemarthroses result in haemophilic arthropathy. The pathogenesis of haemophilic arthropathy is not truly understood, but the effects of the iron deposited into the synovial fluid include hypertrophy and hypervascularity of the synovium and progressive osteochondral changes. The clinical assessment of synovitis is subjective. Clinical reviews aim to detect the early stages of joint damage and prevent functional limitations. The HJHS and HEAD-US offer repeatable means of identifying clinical and radiological early joint changes. Ultrasound assessment of children's joints with moderate or severe haemophilia on prophylaxis has identified early joint abnormalities. These findings have mainly been within joints that have clinically identified low HJHS scores. Ultrasound has however identified joint abnormalities in the absence of clinical findings observed on HJHS. In one study none of the joints without reported bleeding showed abnormalities at physical examination and/or ultrasound. Bleed history is essential to recognition of a target joint and as such early identification, monitoring and prevention of chronic synovitis. Intra-articular steroid injections (IAI) are used widely in clinical practice, but to date there are no randomised or large non-randomised studies to inform an evidence-base in relation to the use of IAI

in children or adults with haemophilia. Symptomatic relief has however been reported in chronic synovitis.

The goals of physiotherapy remain the same for all stages of haemophilia management and these are to reduce swelling and pain, improve and or maintain range of movement and muscle strength, re-establish proprioception and to protect and prevent the joint from additional bleeds. These aims are integral to maintaining and where possible improving the physical function of the individual. To date there are no randomized controlled trials on which rehabilitation programme should be initiated for managing mild synovitis as the specifics of rehabilitation are patient specific.

Chemical vs. radiosynoviorthesis

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Synoviorthesis (SO) is the intraarticular injection of materials to destroy or inactivate hypertrophic synovia from chronic hemophilic synovitis. This minimally invasive group of therapies is used for over 50 years. Particularly in inhibitor patients, surgical options are reserved for failure to SO. Satisfactory experiences from chemical synoviorthesis (CS) with osmic acid, rifocine and oxitetracycline were shown. Advantages of CS are the low cost and easiness of handling. Osmic acid is an exception, being highly toxic for workers and operator. Rifocine is not globally available, while oxitetracycline is restricted to veterinary use in some countries. Both demonstrated proteolytic and an fibrinolytic action producing fibrosis and sclerosis of the synovium, consequently reducing bleeds. Disadvantages of CS are poorer results in knees, compared to radiosynoviorthesis (RS) and pain, associated to the weekly injections. RS is an intra-articular application of a beta-emitting radioisotope, irradiating the synovia and reducing joint bleeds. Usually one RS is enough for achieving good results. Today the most used isotopes for hemophilia are Y-90, Re-186, P-32 and Sm-153. Disadvantages of RS over CS are higher costs (yet much lower when compared to surgery) and radiation safety concerns. Costs can be minimized with proper strategies, concentrating groups of 5 to 10 patients/session. RS is considered safe in terms of exposure to ionizing radiation, according to several studies. Thus, RS did not contribute to the development of neoplastic diseases, being cancer development post-RS is half of the control group. It is key to choose the best RP and its dose. It is necessary to evaluate the synovial thickness and select a RP with an appropriate range of beta radiation in soft tissues. Y-90 (mean penetration 3.6 mm) is more suitable for knees, while Re-186 (mean penetration 1.1 mm) is useful for elbows and ankles. Several groups, working with anatomic and mathematical models, are attempting to develop protocols for individualization of doses of radiopharmaceuticals for RS. Cooperation between hematologists, orthopedic surgeons and nuclear medicine (NM) physicians is key for choosing the best therapy for each case. Awareness for early treatment of synovitis, a devastating disease, should include NM international agencies, hemophilia societies and governments.

Synovitis: Arthroscopic vs. Open

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Surgical synovectomy, whether open or arthroscopic, requires extensive resources from an experienced team, a dedicated hemophilia treatment center, and a reliable supply of clotting factor. Surgical synovectomy is seldom necessary today and is only considered when other less invasive and equally effective procedures fail. Historically, synovectomy for chronic hemophilic synovitis was performed through an open arthrotomy. With extensive surgical approaches, most of the synovium can be removed from a joint. The success rate of open synovectomy in controlling recurrent bleeding was over 80%. Most of these procedures were performed in patients who already had extensive joint surface destruction, therefore, a natural progression to end-stage disease was eventually observed. Many patients had difficulty regaining range of motion following open synovectomy. Additionally, the procedure required considerable amounts of clotting factor replacement and prolonged hospitalization. For these reasons, open synovectomies have been largely abandoned and replaced by arthroscopic synovectomies. Arthroscopic synovectomy is recognized today as an effective method of synovial deactivation. While its use as a first option has some advantages, there is consensus that it is best used as a second level of defense after radiosynoviorthesis or chemical synovectomy. The procedure requires surgical expertise and meticulous execution, and allows access to the vast majority of the joint with minimal external incisions. Patients require hospitalization, surgical amounts of clotting factor replacement, and dedicated physiotherapy. The technique allows removal of osteophytes, treatment of chondral lesions and remodeling of meniscal tears, which are characteristic of grade III and IV arthropathy.

Medical management of haemophilic synovitis

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Primary prophylaxis clearly prevents development of haemophilic arthritis and synovitis. At the other end of the spectrum, surgery and chemical or radionuclide synovectomy all have a place in the management of established synovitis. One study has suggested a strong link between predisposition to haemophilic synovitis and HLA B27 (Ghosh K, Lancet, 2003) with the implication that some patients might benefit from early and intensive prophylaxis. Confirmation of the diagnosis of synovitis is important and can be achieved by ultrasound or MRI. Once identified, synovitis should be treated as early and aggressively as possible. The goal is to break the 'vicious cycle' of hemarthrosis-synovitis-hemarthrosis. Almost inevitably, affected joints become 'target joints' (defined as being the site of three or more haemarthroses within a six month period). Initial conservative medical management includes intensive prophylaxis treatment for 3-6 months, aiming for a higher than usual trough level. Perhaps the novel longer-acting concentrates may prove to have an important role in this context. Physiotherapy is important to maintain muscle power and range of movement as well as to prevent the development of contractures. The use of COX-2 inhibitors such as etoricoxib

may be of great benefit, particularly when several joints are affected. The lowest dose should be used and the duration of treatment restricted in order to minimise the potential for cardiovascular complications. Looking to the future, it may prove useful to explore the value of monoclonal antibodies which antagonise the effect of inflammatory cytokines, and which are of great benefit in conditions rheumatoid arthritis. Aspiration of a persistent effusion and injection of steroid can ameliorate symptoms for up to a year after treatment. Angiography may identify a "blush" or microaneurysms in associated with frequent bleeding. Selective microcatheterization and embolization with microcoils can stop the bleeding and control associated synovitis.

Lessons Learned...by Experienced Treaters

The elephant in the room ... the things we don't always ask, but should

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Even with significant improvements in haematological management, for many, haemophilia remains a musculoskeletal disease. In countries with limited access to treatment, joint disease and potential infirmity remain high. In countries with good access PWH are living into older age with significant physical impairments, as well as the ongoing issue of ankle damage in populations on good primary prophylaxis. The patient that we see in our clinic is a demonstration of 'their life lived with haemophilia' – the physical, emotional and psychological consequences of it. So do we as clinicians actually consider them in this context? MSK interventions have a tendency to focus on more physical aspects of a person's being; ROM, strength and pain. A person centred care approach is a good model for PWH and identifying their potential unmet needs. The domains of self-care, self-management, self-esteem and self-efficacy permit consideration of the life with haemophilia – mobility, toileting, eating and dressing, physical and emotional intimacy, drug use, body image to name but a few. How many of us would comfortably instigate a conversation how a PWH and significant elbow arthropathy goes to the toilet? How might you address an assessment of this activity and start to plan potential helpful interventions? What about how they feed themselves and brush their teeth? And how does their joint arthropathy impact on their close physical relationships, being intimate with a partner? Is this just the realm of nursing or psychological therapies, or do we have a responsibility to see the whole person when planning MSK interventions that matter to them? Illicit drug use for pain relief, social participation or body enhancement may be a bigger issue than we realise –but are we assessing its cause and effect? Do PWH actually want to do exercise? Can we aid them to be contented and safe with their joint disease? Joint restriction and pain may only be the tip of the iceberg, what lies beneath may actually be more meaningful to the patient. This presentation aims to highlight potential issues and stimulate thought on how we can best address them in everyday practice.

What's hot and what's not

Knee synovitis, treatment with sinovioangioliolisis

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Haemophilic synovitis is a disabling complication of hemarthrosis that leads to destruction of the articular cartilage and the emergence of hemophilic arthropathy and produces disability, chronic pain, and depression, social and physical limitation. Selective embolization of the knee can prevent joint bleeds and joint damage. Sinovioangioliolisis is a selective procedure that deprives selective vascularization of the synovial hyperplasia of the joint, without affect the residual superficial cartilage. **Material and Method:** 34 knee embolization in 33 patients were performed. Mean age 20 years old. Thirty one type A, two type A with inhibitors and 2 haemophilia type B. Evaluation of the number/year of bleeding episodes and consumption of factor VIII before and after embolization was performed. **Results:** Thirty patients (91%) had satisfactory results. Three patients (9%) needed another embolization, two with favourable outcome, and one need arthroscopic synovectomy for poor outcome. The Average bleeding episodes were 1.21 per month (range 0-3) before the procedure and was reduced to 0 (range 0-1.67, p: 0.001) at three months, 0.17 (range 0-1.67, p: 0.001) at 6 months and 0.33 (range 0-1.67; p: 0.024) at twelve months. Four patients were free of bleeding for more than six months. Also after the procedure there was a significant reduction in the use of factor VIII in the first year. No significant adverse effects were reported. **Conclusions:** Selective embolization of arteries of the knee, is a safe procedure that prevents repetitive episodes of bleeding, which translates into a better joint function, with decreased cartilage damage and less consume of factor VIII.

Free Papers – Surgical

Total hip arthroplasty via direct anterior approach in patients with hemophilia: less bleeding, faster rehabilitation

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Objectives: The aim of this study is to evaluate the clinical and radiologic outcomes of total hip replacement in hemophilic patients with implementation of direct anterior approach. **Materials and Methods:** From January 2010 to December 2014 nine patients with advanced hemophilic arthropathy of the hip underwent total hip replacement (THA) through direct anterior (DA) approach. **Results:** Eight hemophilia A and one hemophilia B cases underwent THR. All of them were male and mean age of the patients were 36±7. The mean operation time was 65±20 min. The average blood loss was estimated 450±100 ml. We did not use drain in this group of patients. We have experienced no postoperative hematoma formation; Compared to the pre-operative hemoglobin level, post-operative hemoglobin

decreased by 1.8 ± 0.7. Only one patient with inhibitor who underwent simultaneous bilateral THA required single unit blood transfusion. The mean Harris Hip Score improved from 46 (ranged, 38-53) to 76 (ranged, 72-85) at the final follow-up that was compatible with good to excellent results in 65% of hips. We identified one acetabular cup loosening 20 months after index surgery that was revised with a larger cementless cup and bone grafting. All other acetabular cups and femoral stem remained well fixed at the last follow-up. **Conclusion:** Cementless THA using DA approach in hemophilic patients with severe hip arthropathy takes along satisfactory short-term outcomes. Patient optimization with adequate factor replacement and a multidisciplinary approach are essential parts of our success.

Intraoperative features of arthroplasty procedures in hemophilic arthropathy

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Introduction and objectives: The arthroplasty procedures are never routine in hemophilic arthropathy. Aim of this study was to evaluate intraoperative difficulties and complications of arthroplasties in hemophiliacs. **Material and methods:** In this retrospective study, 64 procedures of 32 patients with hip and/or knee involvements operated between 2002 and 2017 were evaluated. Of these, 41 primary total knee arthroplasties (TKA) (14 bilateral, six of which were simultaneous), three knee revisions (because of infection in one case, aseptic reasons in two) and seven total hip arthroplasties, four of which were accompanied with a simultaneous TKA, were performed. Preoperative features such as range of motion, knee society and function scores were recorded. Intraoperative features including type of prosthesis, necessity for extensile approaches, bone grafting and complications were noted. **Results:** The mean age of the patients at the time of operation was 38.70±12.48 years. Mean follow-up was 3.7±3.91 years (range 1 month – 14 years). One patient was deceased in the first postoperative year. Preoperative knee society and function scores were 35±16.7 and 51.70±17.42. Mean range of motion was 59.35°±32.51°; 33 knees (76.7%) were stiff and had flexion contracture. Lateral retinacular release was necessary in 16 knees, TTO in five, and rectus snip in three knees. Lateral approach was used in one patient. Bone grafting was performed in 11 knees. In six primary knee cases, stemmed prostheses were implanted. In a bilateral primary case, hinged prostheses were used. Residual flexion contracture, partial avulsion of patellar tendon, femur fracture (a THA patient; treated conservatively) and oblique patella fracture (one each) were the intraoperative complications. In hemophiliacs, the intraoperative complication rate of TKA was found to be 9.76% in primary cases; this rate was 14.29% in THAs. **Conclusion:** Stiff knees are generally encountered in hemophilic arthropathies. In that case, standard approaches for TKAs do not supply sufficient exposures, thus extensile approaches are necessitated more than usual. Instead of primary prosthetic components, most of the time revision prostheses with stems are implanted. Intraoperative complication rates are higher when compared to standard arthroplasties which was reported as 3.8% and 2.3% in TKAs and THAs, respectively in other studies.

Total knee replacement for the patients with severe haemophilic arthropathy in Taiwan: A nationwide population-based retrospective study

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Introduction and Objectives: Total knee replacement (TKR) surgery is a treatment option for advanced hemophilic arthropathy. Due to its rarity and complexity, most previous reports demonstrated the results of single-site studies with limited cases. This population-based study aimed to investigate the major epidemiological characteristics, mean consumption of coagulation factors, length of hospital stay, complications and failure rate of primary TKR surgery for severe hemophilia patients in Taiwan. **Materials and Methods:** A cohort of 996 hemophilia patients registered between 1995 and 2011 were found and a total of 103 primary TKR surgeries were performed on 75 patients (7.5%). Unilateral TKR was performed on 47 patients and 28 patients of them received bilateral TKRs including 12 simultaneous, and 16 staged cases. The mean age at surgery was 32.3 years (range, 17.3-55.7), and the mean follow-up duration was 77.9 months (range, 2.3-176.8). **Results:** Failure was found in eight patients (8.5%) at a mean 32.8 months (range, 2.3-95) after operation. Four patients revealed aseptic loosening whereas infection occurred in 6 cases. The 10-year prosthesis survivorship was 88.6%. For the patients receiving unilateral TKR, the mean length of hospital stay was 15 days (range, 7-32). The mean cost of factor supplement was US\$43,543 in association with a mean consumption of 4-unit packed RBC transfusion (range, 0-38). The total admission cost was US\$48,326 (range, US\$4,165-US\$262,619). **Conclusion:** In comparison with the staged bilateral TKRs, the simultaneous procedures significantly reduce the mean total admission cost from US\$101,923 to US\$61,587 ($P=0.023$). Therefore, bilateral simultaneous TKR surgery is more preferable than the staged procedures in terms of cost-effectiveness.

Drain after total joint arthroplasty in patients with hemophilia: a necessary practice?

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Introduction: There is still controversy around using drain in total joint arthroplasty (TJA) especially in patients with hemophilia. Use of drain is proclaimed to reduce the rate of hematoma, wound problem, limitation in range of motion and infection. As the drain may reduce the tamponade effect in the joint, blood loss is the main potential disadvantage of it. This latter, becomes more important in patients with bleeding diathesis. On the other hand, postoperative hematoma will increase patients' pain and influence the rehabilitation course. We conceived this study to see if no drains protocol has any effect on the outcome of TJA in patients with hemophilia. **Materials and Methods:** Between 2001 and 2008, we routinely use closed suction drain following TJA in our hemophilia patients. During that time period, we did 42 TKA in 39 patients and 4 THA in 4 patients with a mean age

of 35.5 years. From 2010 to 2013, we did 38 TKAs in 27 patients and 10 THAs in 9 patients with a mean age of 35.7 years at the time of surgery. Eight patients underwent simultaneous bilateral TKA and 1 patient received simultaneous bilateral THA. The estimated blood loss was calculated based on highest hemoglobin drop in the post-operative period using the formula proposed by Gross. The rate of complications was observed in the two groups. Patient reported level of postoperative pain (depicted by visual analogue scale) was recorded for all patients. The time (days) to regain a 90 degree of range of motion was also registered. **Results:** There was no statistical difference between two groups in terms of blood loss, postoperative pain, fever, time to regain the range of motion and infection. Two patients in drain group and one patient in no drain group were re-operated because of periprosthetic joint infection. No patients needed blood transfusion in each group. The final functional and knee scores were the same in both groups. **Conclusion:** Our study suggests that "no drain" protocol after TJA in hemophilia patients is as safe as drain application protocol. On the other hand, it decreases the cost of surgery and potentially the amount of blood loss. Further prospective randomized study is necessary to determine the exact role of drain in TJA in hemophilia.

Is it recommended to release the ipsilateral hip for severe flexed knee in patients with haemophilia?

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Introduction: The flexed knee remains a serious problem to solve, especially because it occurs in young patients who do not have adequate access to factor replacement therapy. The literature showed that hamstring release is indicated when the knee flexion contracture is from 30° to 45° or more, associated with repeated Hemarthrosis, and failure of conservative treatment after 6 months. Daffunchio et al have demonstrated the effectiveness of treatment with toxin botulinum type A in patients with knee flexion contracture with less than 45°. When the flexion contracture is more than 60°, hamstring release is not enough and we consider that the release have to make in the ipsilateral hip. The aim of this study was to evaluate the results of sartorius, rectus femoris of quadriceps and fascia latae tendon release in the hip, for the treatment of knee flexion contracture in patients with severe hemophilia. **Methods:** Five patients with nine knees were treated with tensor the fascia latae, sartorius and rectus femoris release. Four patients have bilateral flexed knees. All patients have severe hemophilia A, one has inhibitor. The follow-up was 3 years. The average age at the time of surgery was 12.5 years old (range 9-14). Clinical assessment of the patients was performed at least twice per year and consisted to evaluate knee flexion, knee flexion contracture and the possibility of walking, with or without assistance. **Results:** The average preoperative knee flexion contracture was improved from 87.9° preoperative to 30.7° postoperative ($p \leq 0.001$). The average preoperative knee flexion was 121.4° and the postoperative was 78.6° ($p = 0.001$). Four patients cannot walk before the surgery and use wheelchair, one can walk with crutches but without weight on his affected

lower limb. After surgery all patients were able to walk; only one with crutches. **Conclusion:** Patients with severe degrees of knee flexion contracture have disability. This type of surgery allows patients to walk again, with a good range of motion in the affected knee.

Measures of elective surgery outcomes in haemophilia A patients on turoctocog alfa prophylaxis

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Introduction and objective: Repeated haemarthroses in patients with haemophilia A can result in progressive arthropathy. Orthopaedic surgery aims to relieve pain and restore function and stability. The guardian™ clinical trial programme enables investigation of whether orthopaedic surgery can reduce bleeding frequency. As part of the guardian™2 trial, annualised bleeding rate (ABR) and spontaneous ABR (AsBR) were measured before (from study start to first surgery) and after (from first surgery to study end, excluding additional surgeries) orthopaedic surgery in patients receiving prophylaxis with turoctocog alfa, a B-domain-truncated recombinant factor VIII (Novo Nordisk) and compared with a control, non-surgery haemophilia A population. **Methods:** Nine major orthopaedic procedures were performed in 9 patients (>50 FVIII exposure days; no history of inhibitors; mean age 38.8 years; FVIII level ≤1%). Poisson estimates of mean population ABR were used to account for the number of turoctocog alfa exposure days prior to surgery. A 'paired cases' analysis was performed to determine whether reduction in ABR was due to surgery. For each case, an age-matched (± 1 year) non-surgery control patient was selected. The day of surgery was used to split the study period for the pair-control into 'before' and 'after'. Paired-case analyses are based on AsBR data only. **Results:** Following surgery, mean ABR reduced by 1.98 bleeds/person/year. Mean AsBR reduced by 31.3% in surgery patients and by 26.7% in control patients (Table 1) **Conclusions:** Both ABR and AsBR reduced following orthopaedic surgery. Evaluation of the paired cases analysis showed similar relative reductions in AsBR in control patients. However, the limited cohort size may have prevented a small but significant reduction in bleeding being detected. A randomised trial assessing differences in joint ABR between surgery and non-surgery patients could further investigate whether ABR is a relevant outcome predictor of elective orthopaedic surgery.

Free Papers – Non-Surgical

A PEGylated, vWF-Independent Long-Acting Factor VIII (LAFATE)

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Introduction: Because FVIII circulates in a tight non-covalent

complex with vWF, engineering FVIII alone may not allow improving its half-life beyond that of vWF. Thus by fusing von Willebrand factor (vWF) to its FVIII-binding domain, we are making a very Long-Acting FVIII, termed LAFATE. **Methods:** We sought to make a FVIII having a vWF fragment truncated to harbor only the FVIII-binding region. We produced this molecule in HEK293F cells, purified and further stabilized it by PEGylation. To confirm in vitro properties of this chimera, we measured its affinity of binding to vWF and to the low density lipoprotein receptor-related protein 1 (LRP1) using ELISA. We also measured thrombin generating profile of the molecule using a calibrated automated thrombogram triggered with tissue factor (TF) over time. The pharmacokinetic behavior of the molecule was evaluated in FVIII knockout hemophilia A mice, and the pharmacological property was monitored in the tail-clip bleeding model of the mice. The blood loss was quantified by use of ELISA method for hemoglobin. **Results:** The corresponding result revealed that the LAFATE possesses 3.3-fold more extended half-life than Advate, which has been hardly achieved by engineering FVIII alone. LAFATE corrected the impaired thrombin generation of the FVIII deficient plasma in a concentration-dependent manner, and showed an acute efficacy comparable with Advate in stopping bleeding in tail-clip bleeding model. **Conclusion:** LAFATE has a great potential to overcome limitation imposed by vWF to the pharmacokinetic behavior of FVIII in vivo and also to create a novel FVIII chimera administrable on a weekly basis.

Optimising functional measurement selection to individualise treatment and improve outcomes in patients with haemophilia

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Introduction: Existing musculoskeletal (MSK) assessment tools for joint disease in haemophilia are non-standardised, complex and lack sensitivity and accuracy when used individually, particularly for the early detection of joint dysfunction. There is also limited guidance on the most appropriate tools to evaluate MSK status. **Objectives:** To review current tools for MSK assessment in individuals with haemophilia and suggest recommendations on their appropriate use. **Methods:** An expert panel evaluated current MSK assessment tools in haemophilia through published literature and personal expertise. Assessment tools were grouped according to their purpose and methodology, and their usefulness, validity, sensitivity and specificity was evaluated and rated by the group to summarise their clinical utility. A table with recommendations for the use of MSK assessment tools in different patient groups and different clinical situations was then prepared. **Results:** The functional tools assessed received the highest scores, indicating their utility across several assessment criteria. However, no single tool provided a comprehensive assessment of MSK structure, function or impact of MSK disease in people with haemophilia that would be sufficient to guide therapy. Consequently, multiple tools must be used to provide a complete assessment; our patient

evaluation table provides guidance on the use of recommended examinations for specific patient groups based on age, disease severity and joint deterioration. Essential assessments that should be carried out regularly include clinical examination of the joint, disease-specific structure and function scores and activity/participation scores including quality of life assessment. Joint imaging, used in conjunction with structure and function scores, is also recommended in the prevention, diagnosis and follow-up of haemophilic arthropathy. **Relevance and applicability:** An integrated approach using combinations of various MSK assessment tools will allow early detection and treatment of joint dysfunction and may improve long-term outcomes. An integrated approach could be used for regular long-term follow-up in all patients independent of age and disease stage, and could be especially useful in children to detect and treat early dysfunction prior to the establishment of overt arthropathy.

Survey of physiotherapy practice and MSK facilities available for pwh in India

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Hemophilia Federation (India)

Background: India has high absolute numbers of People with Haemophilia (PwH) with Musculoskeletal (MSK) sequelae; little evidence to support role of physiotherapy on quality of life of PwH. **Objective:** To survey physiotherapy treatment protocols, practices, facilities available for MSK care for PwH in India. **Methods:** Cross-sectional study with mixed methods. We conducted web-based survey for infrastructure and protocols for physiotherapy and interview of physiotherapist about knowledge and practices. We directly observed few physiotherapists and ran in-depth interviews in four facilities. **Results:** 40 out of 76 hospitals from 13 states responded to web-survey. 30 of 40 were tertiary hospitals, 5 Orthopaedic/Physiotherapy clinics and 5 clinics in HFI chapters. There were median 2.5 (IQR 1-8) physiotherapists and median 2 (IQR 1-2) were trained for managing Haemophilia. 20 hospitals had a Haemophilia care team. 90% had interferential currents, ultrasound therapy, muscle stimulator and TENS. 75% had SWD and IR therapy. 17 of 40 hospitals had protocols for management; assessment mostly by HJHS and FISH. 17 hospitals provided CFC, 18 had orthopaedics, 13 had physiotherapy protocols for PwH. 30 hospitals supplied orthotics. 47 physiotherapists from 40 hospitals responded to survey. Most were 22-42 years. 50% were treating Haemophilia patients for < 3 years. 66% received training for Haemophilia management. 64% started physiotherapy as soon as Haemophilia was diagnosed. 21% started only after giving CFC. Bleeding in knee, elbow joints were considered indication of severe disease. 75% knew PwH suffer muscle bleeds. Common muscle bleeds mentioned were iliopsoas, quadriceps and calf muscles. 32% knew of some contraindications for exercise in Haemophilia. 64% of physiotherapists knew HJHS, 19.2% of Gilbert score and 12.8% Pettersson score. Few mentioned FISH. 80% used US therapy, muscle stimulator, TENS and Interferential currents. Physiotherapists were cautious while exercising PwH. Few physiotherapists prescribed accessories like splints/footwear. 58% recommended joint assessment quarterly and 23% annually. Wide variations exist in availability of CFC, departmental coordination and home advice. **Conclusion:** There is wide variation in settings

and no physiotherapy standards exist for PwH in India. Need to allocate resources to improve physiotherapists' capacities in treating PwH.

Measuring limitations in activities and participation in persons with haemophilia: a systematic review of commonly used measurement tools

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Introduction: Monitoring clinical outcome in persons with

haemophilia (PWH) is essential in order to provide optimal

treatment for individual patients and compare treatment groups.

The use of measurement tools on all domains of the International

Classification of Functioning (ICF) model is recommended.

However, there is no consensus on a preferred set of measurement

tools. **Aim:** The aim of this study was to give a comprehensive

overview of the measurement properties of a selection of

commonly used measurement tools developed to measure

limitations in activities and participation. **Methods:** Electronic

databases were searched for articles that reported on reliability,

validity or responsiveness of predetermined measurement tools

in PWH. Commonly used measurement tools were identified by

a survey and selected during a consensus meeting. Included tools

were: the Haemophilia Activity List (HAL), the Pediatric HAL

(PedHAL), the Functional Independent Score for Haemophilia

(FISH), the Impact on Participation and Autonomy questionnaire

(IPA), the International Physical Activity Questionnaire (IPAQ),

the Canadian Occupation Performance Measure (COPM), the Six

Minute Walk Test (6MWT), the Timed Up and Go test (TUG)

and accelerometry. Methodological quality of the studies was

assessed with the COSMIN checklist. Best evidence synthesis

was used to summarize evidence on the measurement properties.

Results: The search resulted in 3453 unique hits; forty-two articles

were included. Eight studies explicitly aimed at investigating

measurement properties in PWH. For most measurement tools

only hypothesis testing (convergent and discriminant validity) was

investigated sufficiently. The HAL, PedHAL and FISH were studied

somewhat more extensively but with low COSMIN scores when

considering other measurement properties. Measurement error,

cross-cultural validity and responsiveness were rarely reported.

Conclusion: the PedHAL and HAL are the recommended self-

reported measurement tools to assess limitations in activities. The

FISH is the recommended performance based tool in populations with clinical significant arthropathy. Further research needs to focus on cross-cultural validity, measurement error and responsiveness of the self-reported tools and performance based tools that are suitable for patients with minimal arthropathic complaints.

The Effects of Physical & Mental Health Rehabilitation Program (PMHRP) for Hemophilic arthritis patients

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Introduction and Objectives: Most of the rehabilitation program for patients with hemophilic arthritis are focused on only the improvement of physical activities. However, the actual hemophilic arthritis patients are accompanied by mental problems as well as physical disabilities, so a rehabilitation program to improve physical and mental problems simultaneously is needed. PMHRP was developed to solve these problems through increasing the interpersonal relationship, developing each potential, self-development and understanding others. PMHRP was analyzed by this study groups to verify the clinical effectiveness. **Material and Methods:** The subjects for this study were a total of 75 patients with hemophilic arthritis who underwent lower extremity joint surgeries at the hemophilia surgery center in Kyung Hee university at Gangdong. The study was attended PMHRP 5 times(4hours/time) for 4 weeks. The measurement tools of this study were Numerical Rating Scale for 100mm Pain VAS, 100mm Fatigue VAS, self-efficacy, self-esteem, SCL-90-R (Symptom Checklist-90-Revision). The data was analyzed with t/ANOVA test using SPSS/Win18.0. **Results:** After PMHRP application, self-efficacy score increased significantly in the study group ($p<.001$) and self-esteem score also increased ($p<.001$). On the contrary, 100mm Pain VAS & 100mm Fatigue VAS scores decreased, and SCL-90-R scores increased significantly ($p<.001$) after the 4 weeks in the group. **Conclusion:** In conclusion, PMHRP showed much more satisfactory results than the simple physical therapy to treat the physical and mental disabilities including psychosocial stresses in patients with hemophilic arthritis by increasing the self-esteem and quality of life by themselves. These results suggest that PMHRP is highly recommended as a distinguished method of rehabilitation for patients who underwent any orthopedic procedure with hemophilic arthritis.

Early post-surgical derivation after orthopaedic surgery from a high complexity centre to a specialized haemophilia centre

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Introduction: Orthopaedic surgery in PWH is complex and must be performed by a specialized team at a highly specialized centre. This work shows a model of care in orthopaedic surgery performed in a high complexity centre and three to six hours

after surgery patients were derived to a specialized Haemophilia Centre. The aim of this work is to show the effectiveness of this care programme. **Patients and methods:** Inclusion Criteria: Patients under 80 years old requiring orthopaedic surgery of the upper or lower limbs. Exclusion criteria: Severe lung disease, Diabetes poorly controlled; severe arterial hypertension; symptomatic anaemia; unstable coronary syndromes; heart failure. 419 orthopaedic surgeries were performed in 273 PWH, between June 1981 and December 2016 in a single centre. 269 were men and 4 women, mean age was 32 years old (4-78). 251 were haemophilia type A, 16 type B, 5 vW and 1 type B carrier. 229 were severe, 25 moderate, and 13 mild. Fourteen patients had inhibitors. Surgeries were classified: low, medium and high complexity. After surgery patients stayed at the post-surgical recovery room and then are derived at the Haemophilia Foundation for total recovery, until they are discharged. The transfer will be carried out in an ambulance with an assistant physician. Parameters to be transferred: Vital signs stable for 1 hour: with blood pressure and heart rate plus 20% of pre-anaesthetic levels and oxygen saturation greater than 90%. Patient must be oriented in time and space. Pain control: in surgeries that will have high painful impact, guided regional block will be performed under ultrasound guidance, if not, opioid analgesics were administrated; Absence of nausea, vomiting and haemostasis control. **Results:** 68 low complexity, 215 moderate complexity and 136 high complexity orthopaedic surgeries were performed with this model programme. Only one patient without inhibitor, undergoing a highly complex surgery (femur fracture) required to be hospitalized at the intensive care unit. None of the patients had to be readmitted after the derivation to the haemophilia centre. **Conclusion:** This is an unique care programme that allows to perform highly complex orthopaedic surgery in PWH with excellent results even in inhibitors patients.

Posters Surgical

PS-01

Anthropometric study of the hemophilic knee joints undergoing total knee arthroplasty

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Introduction and Objectives: Total knee arthroplasty (TKA) is the standard treatment in severe stages of hemophilic arthropathy (HA). HA usually starts in childhood when open physes are vulnerable to growth alteration and this may alter the anatomy of the knee joint. So especial considerations may be required when operating on a hemophilic knee for which little data is available about anthropometric characteristics. We studied the anatomical dimensions of hemophilic knees compared to usual osteoarthritic ones. **Materials and Methods:** In a prospective case-control study, anthropometric characteristics of 24 patients with hemophilic arthropathy and 28 patients with osteoarthritic knee were recorded. To eliminate the sex bias, only male osteoarthritic patients were enrolled. Several pre- and intra-operative indices were measured for each patient. The data was introduced to IBM

SPSS software and were analyzed using appropriate methods. **Results:** Fifty-two patients were enrolled (24 with HA). Patients with HA had significantly wider medial-lateral diameter of distal femur (87.5 ± 5.2 mm) compared to OA patients (76.2 ± 4.2 mm, $p < 0.05$) while the anteroposterior size did not differ significantly between the two groups. The patients with HA had significantly higher medio-lateral to antero-posterior size aspect ratio compared to OA group ($p < 0.05$). The patellae were also larger in hemophilia group. **Conclusion:** The anatomic characteristics of hemophilic knee are different from osteoarthritic knee. This disparity may have an impact in intraoperative decision-making and selection of the type of the prosthesis to avoid mismatches in size and gap balancing.

PS-02

Clinical outcome of 73 total joint arthroplasties in patients with congenital bleeding disorders

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Introduction: Persons with hemophilia and allied congenital bleeding disorders suffer from severe joint damage. Total knee/hip/ankle replacement (TKR/THR/TAR) performed under adequate replacement therapy offers a chance of pain relief and significant improvement of physical function. **Material and methods:** We report on 73 consecutive joint replacements (35 TKR, 37 THR and 1 TAR) performed in 54 patients with congenital bleeding disorders at our center between 1993 and 2016. **Results:** Median age at the time of surgery was 49 (range 25-72) yrs. Fifty, 3, 15 and 5 procedures were performed in 39 hemophilia A (HA), 3 hemophilia B (HB), 7 FVII deficiency (FVIIdef) and 5 patients with v. Willebrand disease (vWD), respectively. Postoperatively intensive factor replacement was administered for 10-16 days employing continuous infusion and bolus injections in 11 and 62 procedures, respectively. Enhanced prophylaxis for postoperative physiotherapy was employed during 3-5 weeks. Only nine patients received thromboprophylaxis (5 HA, 3 FVIIdef and 1 vWD type 1). Early complications comprised 5 large hematomas of the thigh after TKR despite sufficient hemostasis and 2 local inflammations, one patient with vWD type 1 developed thrombosis. Median follow up is 8 (0.5-23) yrs. TKR performed in 35 patients (30 HA, 2 HB, 2 FVIIdef and 1 vWD) resulted in a disappearance of pain in all. The ROM evaluation 3 mths after TKR showed an improvement with reduction in the deficit of extension (from $22.5 \pm 6.4^\circ$ to $3.7 \pm 5.3^\circ$), however, only a slight increase in the flexion (from $79.6 \pm 35.8^\circ$ to $90.0 \pm 21.4^\circ$) was observed. In the long term follow up the mean ROM was of $89.0 \pm 14.6^\circ$. All 37 THR patients (19 HA, 1 HB, 13 FVIIdef and 4 vWD) reported disappearance of pain and the ROM was improved in 29/37 (85%) patients. Late complications comprised 2 aseptic loosening requiring revision 7 and 6 yrs after the first THR in 1 HA and 1 FVIIdef patient, respectively. All knee prostheses are still in situ, one patient developed severe posttraumatic inflammatory hemarthrosis.

Conclusion: Joint replacement is safe and effective procedure in hemophilia and related bleeding disorders if performed by highly experienced surgeon under meticulous hemostatic management by hemophilia comprehensive care center.

PS-03

Haemophilic pseudotumor of the limbs: 25 years' experience of mini invasive surgery

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Introduction: The treatment of Haemophilic Pseudotumour (HP) is resection of Pseudotumour and pseudo-capsule. The HP is nothing more than a hematoma, which grows wildly damaging surrounding tissues, the pseudo-capsule is fibrous tissue surrounding the periphery of the hematoma. We consider that the capsule was not necessary drying it because it was just a defensive per fibrous tissue hematoma. The purpose of this paper is to show the experience of one centre and one surgeon treating pseudotumours of limbs with mini invasive technique for 25 years in patients with haemophilia with and without inhibitors. **Materials and Methods:** Thirty three patients with 39 HP were treated conservatively for 6 weeks. Nine (with ten HP) were inhibitors patients. After six weeks treatment, an X-ray or new MRI of each Pseudotumour was performed to evaluate the response to treatment (reduce to less than half of their original size). Surgery were performed on the patients did not response to conservative treatment. The mini invasive technique consisted in evacuate each HP cavity, and filled it with Hidroxipatyte coralline (granules) in bone pseudotumours and with spongostane® in soft tissue pseudotumours. **Results:** Two (6%) patients responded to the conservative treatment and cured. One was an inhibitor patient. They continued with conservative treatment for an additional 6 weeks and cured. These HP were the most recently formed (3 months). Surgery was performed on the other thirty one (94%) patients with thirty seven pseudotumours. The mini invasive technique of the HP has been effective in 35 of 37 HP (94.6%). Only two cases failed (5.7%) treated with this technique. In both cases the wall of the pseudotumours were not collapsible after the evacuation of the cavity, they need a second surgery with resection of Pseudotumour and pseudo-capsule. **Conclusion:** The incidence of this disease decreased with patient access to treatment. In this series, 6% heal with conservative treatment, even in the presence of inhibitors. We believe, after 25 years' experience, that the mini-invasive technique is the easiest way to treat this complication and that the pseudo-capsule should be excised when the walls are not collapsible.

PS-04

Heterotopic ossification of iliopsoas muscle in hemophilia patient: a case report from Colombia

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To describe a clinical case of a patient with hemophilia who develop and Heterotopic ossification of iliopsoas muscle. A

32-years-old patient with severe hemophilia A (factor VIII: <1%), receiving "Secondary Prophylaxis" treatment (20 IU Kg⁻¹ three times per week in his local treatment center, with recombinant factor VIII - Xyntha®) was referred to our Center. He presented with pain in the upper third of his right thigh and with incapability to bend the hip and he has had recurrent minor spontaneous bleeding episodes in the hip in the last ten years, despite prophylaxis. The physical exam showed a right hip with a range of motions of 70° flexion; 20° extension; 10° abduction; 25° internal rotation; 30° external rotation. Besides, there were a palpable mass, hard, adhere to surface plans, not painful in the upper third of his right thigh. The 3D-computed tomography scenography showed a major lytic defect in the right iliopsoas muscle depending of the Minor trochanter (8 x 4 cm). A Heterotopic ossification of iliopsoas muscle diagnosis was made and patient underwent surgical correction. No complication occurred, except for hypoesthesia in the territory of the femoral nerve. He underwent early physical rehabilitation and after 6 weeks he presented a slightly improvement in the range of motion of the hip. Three months after surgery and final follow-up, patient showed a right hip with a range of motions of 100° flexion; 30° extension; 20° abduction; 35° internal rotation and 30° external rotation, but femoral nerve involvement persisted. The inhibitors titers were negative (0.3 B.U./mL). We advised to adjust prophylaxis treatment at a dose of 30 IU Kg⁻¹ three times per week with the same recombinant factor VIII and to continue physical rehabilitation including nerve rehabilitation. There are two major aspects we want to emphasize: 1) Early referral to a major hemophilia treatment center could avoid bleeding complication and improve quality of life in hemophilia patients; 2) Risky orthopedic surgery for hemophilia patients it is feasible in South-America as has been show by Argentina, Chile and Brazil.

PS-05

Joint distraction in a boy with severe haemophilic ankle arthropathy

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Ankle is the most affected joint in children with severe haemophilia, as a result of frequent and even asymptomatic haemorrhages, causing arthritic damage. Joint distraction has already been applied in adults with haemophilic ankle arthropathy in order to preserve the joint and postpone arthrodesis. We present the case of a 9-year-old boy with severe haemophilia A, who had suffered many haemarthroses in his right ankle that had not been appropriately treated due to inadequate compliance and poor venous access. He experienced ankle arthropathy, presented with chronic pain, synovitis and gait disturbance and crippling. His condition was steadily deteriorating although he was receiving prophylaxis every day through a port-a-cath. X-rays and MRI revealed subchondral cysts and narrowing of the joint space. Ankle joint distraction, using Ilizarov external fixator, was applied for ten weeks, in order to allow space in the affected joint to restore. Clinical effectiveness was evaluated using physical examination. X-ray and MRI evaluation were

performed at baseline, 6-month and 1-year follow-up. During distraction, the patient received prophylaxis with tapering doses (25 – 20 U/kg) of FVIII concentrates every day. He experienced no bleeding. After removal of the Ilizarov fixator, the patient continued prophylaxis with the same dose, every other day. Pin tract infection occurred in week 3 and was treated effectively with oral antibiotics. Physical examination of the patient's right ankle, using the Haemophilia Joint Health Score 2.1, was considered as progressively improved, from 18/20 at baseline, to 13/20 at the 6-month and to 7/20 at 12-month follow-up. No pain through active range of motion was detected at 12-month follow-up. The Global Gait Scoring showed also a great improvement, from 4/4 at baseline, to 3/4 at the 6-month, and to 1/4 at the 12-month follow-up. Plain X-rays showed satisfactory restoration of the ankle joint space and improvement of subchondral sclerosis. MRI revealed a decrease in volume of subchondral cysts and bone oedema and improvement of the joint space width at the 12-month follow-up. Conclusion: Joint distraction by Ilizarov external fixator, for the management of severe haemophilic ankle arthropathy, seems effective and promising in children, who have a rapidly growing skeleton.

PS-06

Orthopaedic surgeries for haemophilia patients with inhibitors using pd-FVIIa/FX

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[Introduction and Objective] A new bypassing agent, Byclot[®] (Kaketsuken, Kagoshima, Japan) reached the market in Japan in November 2014. This agent is a complex concentrate of plasma-derived activated factor VII (FVIIa) and factor X, (FX; pd-FVIIa/FX). The aim of this study was to evaluate the effects of pd-FVIIa/FX during surgery for inhibitor positive patients with haemophilia. [Materials and Methods] Four orthopaedic surgeries (2: total knee arthroplasty (TKA), 1: debridement after infection of TKA, 1: ankle arthroscopic synovectomy) in three high responded inhibitor positive patients with haemophilia were performed. One of them was already reported as the first case describing the successful use of pd-FVIIa/FX as a bypassing agent during major orthopaedic surgery (Takedani H, et al. Haemophilia, 2016). According to the simple pharmacokinetic study results, a sequential administration schedule for pd-FVIIa/FX and recombinant activated factor VII (rFVIIa) was planned during the perioperative period to achieve haemostasis. Pd-FVIIa/FX was administered 1 hour before the skin incision and every 36 hours thereafter. A single dose of pd-FVIIa/FX was 120 µg kg⁻¹, and the medication was administered three times. rFVIIa was administered 1 hour after pd-FVIIa/FX administration during the skin incision; it was the administered every 3 or 4 hours. Each regimen was arranged depending on each operative procedure. Haemostatic effect was evaluated using several coagulation assays, including thromboelastograms (TEM), thrombin generation assays (TGA), activated prothrombin times (APTT), prothrombin times (PT) and plasma coagulation factor activities. The safety of this bypassing agent was evaluated by D-dimer, fibrinogen degradation products (FDP) and platelet counts, as well. [Results] There were no unexpected bleeding episodes or thromboembolic

events, and no additional treatments were needed.

[Conclusions] Pd-FVIIa/FX has the potential to become a good partner for parallel therapy during the perioperative periods of major orthopaedic surgeries. Further clinical studies will be required to establish the optimal administration regimen.

PS-07

Surgical treatment of pseudotumors in patients with hemophilia

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One of the most often manifestations of hemophilia are muscle hematoma, less often we meet with subperiosteum and intraosseous localizations. When the treatment is deferred or not full such hematoma can be transformed into pseudotumors. The most severe complication of pseudotumor is its infection and spontaneous dissection with destruction of underlying tissues. That can be accompanied by massive uncontrolled bleeding especially in hemophiliac patients with an inhibitor. Surgical extirpation of pseudotumors usually is technically difficult and can be complicated with bleeding. Nowadays we meet pseudotumors rarely due to current opportunities of hemostatic therapy. However, a full scale hemostatic therapy and possibilities of early pseudotumor surgical treatment aren't available everywhere, which results in serious complications. Therefore, we believe that an experience of treatment of massive pseudotumors would be of interest. Within the last 3 years more than 10 patients have been treated in our center for massive pseudotumors with destruction of subject's bones and muscles. Generally all patients with massive pseudotumors were from developing countries where there is no haemostatic therapy in due volume. In all cases surgical intervention included an extirpation of the pseudotumor. The capsule and the destroyed fragments of the bones and the muscles were eliminated. The wounds were plugged with antiseptic swabs and a vacuum drainage system was installed. The hemostasis during the operation and in the post-surgical period was supported with FVIII at patients with hemophilia and rFVIIa at patients with inhibitor hemophilia. Only in one case at the patient with a huge pseudotumor of all hip with destruction of all muscles hip bone wasn't succeeded to keep an extremity. In all other cases we managed to avoid amputation of the extremities and preserve their functionality in the patients with hemophilia.

PS-08

The efficacy of arthroscopic debridement of ankle in patients with bleeding disorders

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Introduction and objectives: Arthroscopic debridement is performed in the treatment of early stage hemophilic arthropathy.

The aim of this study was to assess the patient reported outcomes of arthroscopic debridement in the management of hemophilic arthropathies of the ankle. **Material and Methods:** 17 patients with bleeding disorders as well as 17 with ankle problems other than hemophilic arthropathy who had undergone arthroscopic debridement between 2008 and 2016 were included in this study. Preoperative AOFAS (American Orthopedic Foot and Ankle Society Score), FFI (Foot Function Index) with its subscales were obtained. The patient reported outcome was measured utilizing FFI in the latest follow-up. Degree of hemophilic arthropathy was assessed with Pettersson score on the preoperative x-rays. Statistical analyses were performed utilizing SPSS v18. **Results:** Comparison of both objective and subjective scores (preoperative & postoperative) between the two groups did not reveal statistically significant difference (table 1); however, patients in both groups experienced improvements by means of FFI and its subscales which was statistically significant for all but one (disability subscale in study group, $p=0.05$) (activity subscale in study group: $p=0.006$, other p values <0.0001). Despite the comparison of the improvement in FFI between two groups was not statistically significant, the subjective gain was greater in the control group. The median value for the Pettersson score was 6 (min 2, max 9) which was not correlated with the scores. **Conclusion:** With the perspective of the patients, the pain and functional restrictions associated with hemophilic arthropathy of the ankle joint could be improved following an arthroscopic debridement. The findings of this study prove that by means of subjective measures this procedure is less efficient than it is in the nonhemophilic population despite this is not statistically significant. The gains appreciated by the patient are irrespective of the radiological stage of the disease.

PS-09

Total hip arthroplasty in a patient with severe haemophilia A plus high-response inhibitors with FEIBA allergy.

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Introduction: Hemophilia patients may experience severe destruction of their joints as a result of advanced hemophilia arthropathy, with severe pain and functional deficit; in the last 40 years, these patients with severe hemophilia have benefited with elective orthopedic surgery, all thanks to the multidisciplinary treatment and the administration of the replacement therapy. However, 10 to 30% with severe hemophilia A and 5.2% with severe hemophilia B; may develop inhibitors against factor VIII or factor IX, this being the most common and serious complication of replacement therapy in patients with hemophilia A or B. As such, the use of recombinant factors or bypass agents in patients with high response inhibitors make orthopedic surgery can be performed safely and with high expectation of success. **Objective:** report a case of total right hip arthroplasty in a patient with advanced hemophilia arthropathy, severe type A with high response inhibitors, allergic to FEIBA. **Material and**

Methods: We report the case of a 26-year-old male with severe hemophilia A developing high-dose inhibitors, positive HCV, which presents FEIBA allergy. Treated with rFVIIa. in a V stage on the Arnold-Hilgartner radiological scale and with irreversible joint damage of the Petterson radiological scoring scale. **Results:** Prosthetic replacement surgery, which is performed without trans-surgical complications, placing a total cemented hybrid prosthesis, being treated with rFVIIa exclusively. **Conclusion:** Arthroplasty can alleviate pain and improve the function of symptomatic hemophiliac patients with advanced arthropathy, thus increasing their quality of life, being the knee and hip the most common and with better results; With the advent of Bypass agents and recombinants of type rVIIa orthopedic and non-orthopedic surgery in haemophilia patients with inhibitors can be performed with high expectations of success and safety.

PS-10

Total knee replacement in patients with hemophilia in a pronounced axial strain and instability

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Objective: Improved results of Primary Total Knee Replacement (TKR) in patients with Hemophilia A and B by applying structural implants of different types and sizes.

Materials and methods: Results of treatment of 37 patients with III-IV stages of hemophilic arthropathy, who were operated on for severe combined instability and various types of deformations: varus – 17, valgus – 22, rekurvation – 2, totally 41 TKR were performed. In 39 cases (95,1%) deviation of the limb axis was > 10°. We used: X-ray, topogram lower extremities, CT. Function was evaluated before at 1–7 years postoperatively using KSS scales, Oxford and VAS. Statistical analysis – STATISTICA 6.0; p=0,05. **Results:** We used the following endoprosthesis models: unconstrained – 34 (82,9%), semiconstrained – 3 (7,3%) and fullyconstrained – 4 (9,8%). As a result, the complex treatment of patients with the use of developed by us technology of preoperative planning, the sequence of certain surgical techniques, combined with the selection of an appropriate model of the endoprosthesis in the period of 1 year after the operation on the scale of assessment: KSS obtained excellent results in 35 (85,4%) cases, good in 5 (12,2%), satisfactory – 1 (2,4%); by Oxford excellent results were obtained in 33 (80,5%) cases, good in 7 (17,1%), satisfactory – 1 (2,4%). After 2–7 years postoperatively there was no statistically significant differences in the results of the data in the period of 1 year after the operation. **Conclusions:** 1. The main components of that hinder restoration of the right axis of the limb during TKR under: 1) varus deformity: medial and posterior osteophytes, the deep portion of the tibial collateral ligament, posterior capsule inner joint card; 2) valgus deformity: lateral and posterior osteophytes, joint capsule posteroexternal department; 3) rekurvation requires implanting the femoral component of a more distal to the posterior edge, increase «sloped», implantation thickened tibial insert. 2. Selecting the implant model (bicondilar unconstrained, semiconstrained, fullyconstrained) should be based on: the axial relationship of

the lower limb, the state of joints and ligaments, muscle balance assessment of the femur and tibia, a bone.

PS-11

Use of platelet rich in fibrin (prf) in patients with haemophilia

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Introduction: Platelet-rich plasma (PRP) is a platelet-rich fraction of plasma and clinically available as a source of growth factors to facilitate tissue repair and regeneration. Choucroum et al have described a technique for producing PRF without exogenous additives. This material placed in the surgical wound of the patients would fulfil 3 functions: biodegradable membrane that would guide the regeneration of tissues; biodegradable scaffolds for tissue engineering; reservoir of growth factors in PRP gel form, which would act as tissue regenerator and bone inductor. Use it as a haemostatic plug in the surgery area. The aim of this study is to observe the feasibility of using PRF in PWH. **Materials and Methods:** For PRF preparation, venous blood is obtained without any anticoagulant and directly centrifuged. The blood is placed in 10ml glass test tubes, and immediately centrifuged at 3000 rpm for 10 minutes. The biological steps of this centrifugation involve that the circulating blood thrombin turns fibrinogen into fibrin: this fibrin then places at the centre of the test tube. The PRF obtained will be used in two ways: a part of it is used as a filling material, with a gelatinous consistency and an amorphous shape; the remaining part is shaped in order to form a resistant fibrin membrane which can be transfer. Two haemophilia type A severe patients were treated. The average age was 42 years old (41-43). One had an elbow pseudotumour without inhibitor and the other a varus arthrosis of the knee with inhibitor that was submitted to a valgus osteotomy. In both, post- surgical pseudotumor and osteotomy beds were filled with PRF. **Results:** We have not observed any difficulty in the preparation and use of PRF, since the biologists have made their learning curve for PRF preparation. Both patients did not present hematoma in the wound and evolved favourably of their pathology wich originated the surgery. **Conclusions:** We consider that PRF should be used in haemophilia for its properties and because is it simple and low cost treatment.

PS-12

Haemophilic Psoas pseudotumor mini-invasive treatment

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Introduction: The hemophilic pseudotumor of the psoas muscle is infrequent. Usually it occurs in severe forms of hemophilia. Its growth can compress the kidney, affecting its function. The treatment is usually surgery, with the risks involved in hemophilic patients. In this study we present a mini-invasive treatment for hemophilic psoas pseudotumor. **Methods:** The study included 2 patients with hemophilia A and presumptive diagnosis of psoas pseudotumor. The average age was 35 years (28-43). The average

pseudotumor time was 18 months (4-32). Both had vertebral compression and kidney displacement without involvement of its function. One of them had a fistula in the ipsilateral lumbar region. The mini-invasive treatment consists in the addressing of the pseudotumor in the Grynfeldt's triangle lumbar region. The patient is placed in ventral decubitus position and a transverse incision of 3 cm, centered of the medio-lateral wall was made. Once the skin is open, fascia transversalis is accessed, leaving the medial spinae erector exposed. Palpating the transverse apophysis and lateral to it, it is incised and the psoas pseudotumor is accessed. After removing the pseudotumor, it was filled with OH-apatite coralline. In the patient with fistula, it was resected. In any patient drainage was left after surgery. Neither, intensive or intermediate care was required. Patients were treated with replacement doses of factor VIII or IX for 7 days. Both patients received prophylactic first generation cephalosporin intravenously 30 minutes before surgery and 24 hours after the same. **Results:** Both patients progressing favorably without any complication. The average volume aspired from pseudotumor was 3.375 cm³ (2.900-3.950 cm³). Both patients were cured of their pseudotumor using this minimally invasive technique. The fistulized patient had no postoperative infection. **Conclusion:** Grynfeldt's triangle is an area that facilitates the access to the psoas muscle. The location in ventral decubitus makes the tumor mass moves forward and lateral, which facilitates the approach. Filling with OH-apatite coralline has been effective in pseudotumours. The weakness of this study is the small number of patients included due to pseudotumours are uncommon in hemophilia. The strength of the study are the excellent results applying this technique.

PS-13

Resection of an abdominal pseudocyst in a patient with severe haemophilia A

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Introduction: Iliopsoas muscle bleeds are often seen in severe haemophilia patients between 10 and 20 years. It is a serious bleeding with up to 42 % of recurrence. When not treated adequately with clotting factor followed by rehabilitation complications are frequently seen: on the short term anaemia and compression of large nerves resulting in loss of the Quadriceps muscle function. On the long term: contractures and pseudotumour, which may be progressive involving muscle, pelvis and abdominal organs. **Objective:** We describe a resection of an abdominal pseudo-tumour of the ilio-psoas muscle in a 51 year old severe haemophilia A patient with antibodies against factor VIII (FVIII) in the past who suffered from recurrent iliopsoas muscle bleeds (left and right) between 1975-1982. Since then he is treated with factor VIII on a regular base, the last ten years with daily 500 U. He has no spontaneous bleeds, but has severe arthropathy of both knees for which he does not want to have surgery. At routine check-up in October 2015 he complained of abdominal pain and swelling, which started 6 months earlier. At physical examination a 15 cm elastic circumscribed swelling in the abdomen was found. Ultrasound and later CT and CT-IVP showed a multi-locular cyst in the pelvic region originating from the iliopsoas muscle with dislocation of the right kidney,

diameter 20 cm. Prophylaxis was increased to 1000 U FVIII daily. CT 6 months later showed no regression. **Methods:** In September 2016 abdominal surgery by an orthopaedic and general surgeon was carried out. Pre-surgery an angiogram was made showing no abnormalities and embolisation was not necessary. By a lumbotomy the tumour was resected. As the tumour involved the Vena Cava Inferior, resection was not complete. CT postoperative showed a rest-collection with a diameter of 4 cm. **Result:** Patient recovered well without any complications. FVIII was gradually tapered from 2000 U to 1000 U daily 3 months post-operative. Repeated CT scans will be made to follow-up progression of the rest tumour. **Conclusion:** Uncomplicated but incomplete resection of an abdominal cyst originating from the right iliopsoas muscle.

PS-14

Hemarthrosis among pediatric persons with hemophilia in southern Philippines

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Background: Hemophilia A and B is an inherited disease due to deficiency of clotting factor. Joint bleeding initially leads to independent adverse changes in both the synovial tissue and the articular cartilage. **Objective:** The aim of the research is to determine the prevalence of hemarthrosis among pediatric patients' ages 1-18 years old with Hemophilia A and B in Southern Philippines. **Design and Subjects:** A prospective cross-sectional study design was conducted on 20 pediatric persons with hemophilia registered at Mindanao Hemophilia Center. **Methodology and Statistical Analysis:** The subjects or their parents provided assent and informed consents; very young subjects provided verbal assent, as appropriate. The participants were sequentially approach and were studied at a single study visit. Data analysis was done using the descriptive and analytical statistics. **Results:** Out of 20 hemophilic patients, 17 (85%) patients has hemarthrosis. The mean age of the participants was 8 years old. Clinical evaluation using Health Joint Health Score (HJHS) to assess and quantify hemophilic arthropathy using Pettersson Score revealed that when scores are compared against severity there were no significant differences observed (p value 0.835, 0.715 respectively). However, looking at the distribution of scores across the parameters comprising HJHS the test revealed that only 3 parameters and for Pettersson score only 2 parameters did not differ significantly and all other parameters showed significant result of p value of <0.05. It is found out to be more commonly seen among persons with hemophilia with severe deficiency. Their scores with the above mentioned parameters is inversely proportional to their factor level with a p value of <0.05. **Conclusion:** A significant decrease in functional ability was demonstrated on the basis of the severity of hemophilia. Both HJHS and Pettersson Scoring system is important in measuring the degree of hemophilic arthropathy or hemarthrosis to facilitate early and appropriate intervention. **Keywords:** Hemarthrosis, Hemophilia, Hemophilia Joint Health Score, Pettersson Score

Posters Non-surgical

PN-01

Syrian experience of chemical synoviorthesis in chronic hemophilic synovitis

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Introduction and objectives: In a country where antihemophilic factors are lacking, practitioners have to choose the most cost effective treatment of recurrent haemarthroses. Chronic synovitis being the main cause of recurrent haemarthroses synoviorthesis seems to be the best solution. In the absence of radioactive components, we performed chemical synoviorthesis using rifampicin. **Patients, material and method:** Thirty-six patients were treated. They received 600 mg of rifampicin diluted in 10 mL of saline solution for knees (32 cases) and 5 ml for elbow (2 cases) or ankle (3 cases). Knees required frequently repeated procedures (1 to 6). **Results:** results evaluated by lack or reduction of recurrent haemarthrosis were considered as excellent in 13 patients, good in 11 patients, fair in 5 patients and poor in 7 patients. The main complication was moderate to mild pain, and local inflammation. **Conclusion:** In developing countries, Rifampicin induced synoviorthesis might be the best treatment for recurrent haemarthrosis related to chronic synovitis.

PN-02

Sports in patients with haemophilia: toward a new classification

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Sports and physical activities have long been contraindicated for haemophiliacs, or they were restricted to a few activities such as swimming or walking. Recent progress in therapy has meant that haemophiliacs are now encouraged to participate in sports, although various guidelines have recommended that they avoid activities where risks outweigh benefits. In order to evaluate the type of sports undertaken by haemophiliacs and the benefits or risks associated with each sport, we conducted a questionnaire-based study. The questionnaire was sent to 124 patients. Among the 80 patients who responded, 71 (90%) currently participated in or had previously participated in a physical activity. Among them, 56 (70%) still played a sport. Nearly all sports were cited in the answers, the most frequently played by these 56 haemophiliacs being swimming (59%), cycling (44%), walking (43%), table tennis (33%), soccer (33%) and tennis (31%). Fifty-six patients reported a benefit in doing a sport: somatic (79%), psychological (80%) or social (62%). Thirty haemophiliacs (42%) reported at least one accident or incident linked to sport. From these data and the literature, we propose a new functional classification based on the type of movement and gestures involved in the sport. This classification classifies sports in eight categories: aquatic sports, sports involving walking or running, static sports, sports involving a gliding movement, sports with a ball, team sports, throwing or jumping, and combat sports. Recommendations adapted

for haemophiliacs can be drawn from this classification, which now requires validation in larger series.

PN-03

A cohort of type 3 von Willebrand Disorder individuals and their joint health status

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Introduction and Objectives: Type 3 Von Willebrand Disorder (VWD 3) is a rare bleeding disorder characterised by markedly reduced or absent Von Willebrand Factor (VWF) protein accompanied by a parallel reduction in VWF function and factor VIII (FVIII) levels. Type 3 VWD defined as VWF:Ag < 0.1-0.5 iu/dl is inherited as autosomal recessive or codominant manner and the index cases have homozygous or compound heterozygosity for two VWF null alleles. VWF gene is on chromosome 12 and the estimated prevalence of VWD type 3 is of 1-5/million. Mucosal bleeding predominates the clinical symptoms but reports on joint bleeding in this cohort have been described ranging from 32-57% of the patients. In this retrospective cohort study of type 3 VWD individuals correlation of their joint health status with the level of VWF Ag and FVIII levels and the use of replacement therapies in either demand or prophylaxis manner will be examined. **Materials and Methods:** VWD individuals who fit the criteria of diagnosis of type 3 VWD have had their baseline VWF, FVIII levels, multimers, HJHS joint scores and joint imaging performed. Results: 9 patients had VWF:Ag levels <0.1-0.5iu/dl which is currently defined for VWD type 3 patients. FVIII levels ranged from 1-8 iu/dl and VWF multimers were virtually absent except for one patient. Joint scores ranged from 0 to 50. **Conclusion:** Correlation between 1) baseline FVIII levels and spontaneous joint bleeds and 2) prevention of joint bleeds with regular prophylaxis or on demand replacement was difficult to be ascertained.

PN-04

Pseudotumor and successful management in hemophilia patient with inhibitors: Case experience from a hemophilia center in Barranquilla-Colombia

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Introduction: Pseudotumors are a major complication in patients with hemophilia and can have a long life devastating effects, especially in the presence of inhibitors. The frequency and severity of hemophilic pseudotumours have decreased with the use of factor VIII replacement therapy. However, they may still occur in developing countries where facilities for diagnosis and treatment of hemophilia are not available. Only a few cases have been reported in the literature. **Objectives:** The purpose of our work is to showcase the experience in our Hemophilia Center in Barranquilla, Colombia and to highlight conservative management of hemophilia pseudotumor with long-term replacement therapy. **Methods:** we report a 50 years old patient with severe haemophilia A plus low response inhibitor diagnosis with inguinal and pelvic pain, deformity in hip flexion, limp and palpable mass that was diagnosed with magnetic resonance as

a pseudotumor of psoasiliac muscle with no involvement of the iliac crest. This patient underwent conservative medical treatment with rest, long term replacement of factor VIII therapy and physiotherapy with a remarkable clinical improvement; it was decided not to perform surgical treatment at the moment. Patient has developed satisfactorily and is still in periodical control by the orthopedic service with imaging (US and MRI) observing the localized mass without increase in size or bone involvement of the iliac crest. Discussion: The behavior of the pseudotumour is different in children than in adults. In adults, they are usually observed at proximal bones, and do not usually respond to replacement treatment. The associated cost/benefit of interventional treatment versus replacement therapy, and the risk of recurrence should be taken into consideration when treating a hemophilic pseudotumor with the presence of inhibitors. **Conclusion:** The management of hemophilic pseudotumour aims at preserving function and includes conservative methods (immobilization, therapy), whenever this is feasible.

PN-05

Ankle arthropathy in pwh

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Introduction: This pathology reduces joint movement, chronic pain, which usually needs pain killers, restricts normal activity such school or work, causes depressive episodes and decrease of quality of life. The physiopathology has not been fully elucidated. The aim of this paper is to propose the pathophysiology of ankle arthropathy in PWH. **Materials and Methods:** 3654 ankle joints were evaluated in 28 years. 1923 were right and 1731 were left. All patients were evaluated clinically and radiologically presented different degrees of arthropathy. The evaluation of the X-rays and the local ankle anatomy allowed us to propose the pathophysiology of the ankle arthropathy. **Results:** The ankle joint anatomy has an important role in the natural history of the disease. In the posterior face, it is a strong capsule and the reinforcing ligaments (PTE, IT, PT). On the sides are the medial and lateral malleolus so, the best place to distend this is the anterior capsule. When the ankle haemartrosis occurred, the anterior capsule is distended and produced a mass inhibition which affects the tibial muscle and equine tendency. The capsular were seen like expansions in the anterior capsule from the ankle by the tibia and the astragalus. The smallest resistant zone is previous to the capsule that could be distended by haemartrosis and/or synovitis, and ends with the calcification of the insertion. X-ray images could show the osteophytes, with a poor or null compromise of the joint surface. These osteophytes will limit progressively joint movement, generating an impingement and carrying the feet to the equine. The synovial, which is in anterior expansion, was involved generating repeated bleedings episodes. Intra-articular bleeding generates another 2 types of osteophytes. The marginal osteophyte can be observed in the medial region by its overgrowing and can make entrapment of the tibial tendons or the tibial nerve. The central osteophyte is the definitely arthropathy that ends in chronic pain, reduce ROM and affects quality of life of the patient. **Conclusion:** Anatomy of the ankle and joint bleeds with the development of synovitis and

osteophytes explain the pathophysiology of the ankle joint.

PN-06

Training for orthopaedic surgeons in Latin America

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Orthopaedic surgery in haemophilia needs a highly specialized surgeon training. The problem is that there are not many surgeries and the learning curve is very long. To facilitate the learning of orthopaedic surgery in haemophilia, our Centre in 2006 designed a 4-days intensive course. The aim of this work is to show the result of this teaching model. **Materials and Methods:** Eight courses were held. 49 participants from thirteen Latin American countries attended the course. 35 were orthopaedists and 14 haematologists. The activity began on Monday at noon and the first activity was to share an entire afternoon with the rehabilitation team and the patients in the physiotherapist department. On Tuesday morning theoretical activity had been performed and in the afternoon all patients that should be operated on the subsequent days were examined and the cases discussed. Also until 2010 a chemical synovitis and a radioactive synoviorthesis were performed that same afternoon. Since 2011 a chemical synoviorthesis and a synoviolangiolytic were performed. On Wednesday and Thursday surgeries were performed. On Thursday afternoon all the cases operated were examined and the results discussed. The average number of surgical procedures per course was 6 (4-10). **Results:** Two of the 35 surgeons who attended the course died but they continued in the haemophilia practice after the course. The remaining 33 course attendees were consulted if they continued the haemophilia practice. 26 (74.28 %) continued in the haemophilia practice. **Conclusions:** The personalized and intensive training of the orthopaedic surgeons allows the understanding of the multidisciplinary treatment of this pathology and motivated them to continue being part or to form a multidisciplinary team of treatment.

PN-07

Testicular hematoma in haemophilia: The importance of evaluation through Magnetic Resonance (MR).

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Introduction: Testicular haematoma in patients with haemophilia, diagnosed doubt leads to removal of the testicle unjustifiably. The purpose of this work is to show 2 cases of testicular haematoma after microtrauma, diagnosed and follow by Magnetic Resonance (MR), that allows the detection of the hemosiderin in the hematoma and confirms the diagnosis. **Material and Method:** Two patients were included. Both were haemophilia type A severe without inhibitors and asked for pain and enlargement of testicles. First patient, 24 years old, right

testicle, history of micro trauma during sexual intercourse. Second patient of 43 years old, left testicle, history of micro trauma during horse riding. In both patients the diagnostic was performed by testicular ultrasound (US) (Philips® HD 15) with a 5- to 12-MHz linear transducer. In both patients a well-circumscribed mass in the testicle surrounded by a hypochoic halo measuring 1.5 to 2.0 mm and the contents was debris with multiple small septa. Both patients were studied with blood test (β -chain chorionic gonadotropin and α -fetoprotein) with negative results. MRI (General Electric® HDXT 1.5 Tesla) were performed in both patients. In the MRI demonstrate an intratesticular mass, which low signal intensity on T2-weighted images, had high signal intensity hypointense on the T1-weighted image, and shows a low-signal-intensity with gradient-echo T2, loss of signal intensity caused by the presence of hemosiderin. MR study confirmed the diagnosis of testicular hematoma and were treated with factor VIII replacement therapy. **Results:** Patients respond to factor VIII replacement therapy, reducing pain and progressively testicle size. After a positive US, an organized intratesticular hematoma was strongly suspected. Ultrasound and MRI was performed to dismiss out other possibilities, allowing continued monitoring the haematoma until resolution. The follow-up images showed decrease in size of the lesion in the MRI. Years ago at the Foundation of Haemophilia, 3 patients were treated of the same pathology and were diagnosed by US. Biopsy were not definitive and removal of the testicle injured were performed. Pathology results later informed a haematoma. **Conclusion:** This work shows that with correct diagnostic and suitable studies images like MRI, haematoma of the testicle can be resolved with conservative treatment.

PN-08

Risk of osteoporotic fracture following haemophilia: A nationwide population-based cohort study

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Introduction and Objectives: Low bone mineral density occurs more commonly in patients with haemophilia (PWH) than the general population. However, the fracture risk of haemophilia-related osteoporosis has not been well established. We explored the relationship between haemophilia and the subsequent development of osteoporotic fracture. **Materials and Methods:** We selected patients who were diagnosed with haemophilia, according to the data in the Taiwan National Health Insurance Research Database. A comparison cohort was formed of patients without haemophilia who were matched according to age and sex. The incidence rate and the hazard ratios (HRs) of subsequent new-onset osteoporotic fracture were calculated for both cohorts. **Results:** The haemophilia cohort consisted of 75 patients, and the comparison cohort comprised 300 matched control patients without haemophilia. The risks of osteoporotic fracture (HR = 5.41, 95% confidence interval [CI] = 2.42–12.1, P < .001) was higher in the haemophilia cohort than in the comparison cohort. After adjustments for age,

sex, comorbidities, urbanizations, and socioeconomic status, patients with osteoporosis were 4.53 times more likely to develop multiple osteoporotic fractures (95% confidence interval, 1.93–10.62, p = .001) as compared to matched patients. In addition, the incidence of newly diagnosed osteoporotic fracture remained significantly increased in all of the stratified follow-up durations (1-5, ≥ 5 y). **Conclusions:** Haemophilia may increase the risk of subsequent osteoporotic fracture. The risk ratios are highest for PWH diagnosed more than 5 years. Clinicians should pay particular attention to osteoporotic fracture in PWH.

PN-09

Course of target joints and bleeding rates in pediatric hemophilia A patients on twice weekly prophylaxis with extended half-life, PEGylated, full-length recombinant factor VIII

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Introduction and Objectives: In patients with severe hemophilia A, prophylaxis with factor VIII (FVIII) may prevent joint bleeding and subsequent development of target joints and arthropathy. Bleeding rates and target joint status were determined in pediatric patients on prophylaxis with BAX 855, an extended half-life, full-length, recombinant FVIII built on ADVATE. **Materials and Methods:** Patients <12 years with severe hemophilia A and no history of inhibitors to prior FVIII exposure received twice weekly infusions of 50 ± 10 IU/kg of BAX 855 over 6 months. A target joint was defined as a single joint with ≥ 3 spontaneous bleeding episodes in any consecutive 6-month period. **Results:** Sixty-six pediatric patients received a mean BAX 855 dose of 51.1 IU/kg at a frequency of 1.8 infusions/week. During a mean of 48.5 prophylactic exposure days/patient, 5 (7.6%) patients had ≥ 1 target joint bleeds. The point estimate for the mean (95% CI) annualized bleeding rate in 52 patients without target joints was 2.9 (2.0 - 4.2; median: 2.0) and was similar in 14 patients with target joints at screening at 3.5 (1.9 - 6.6; median: 2.1). During 6 months of prophylaxis with BAX 855, target joints decreased from 23 in 14/66 (21.2%) patients to 8 in 6 (9.1%) patients (Table 1). Ten patients lost ≥ 1 target joints. In 8 of these patients, all target joints resolved. No new target joints developed.

TABLE 1: Target Joints at Screening and After 6 Months of Prophylaxis with BAX 855 in Children <12 Years with Severe Hemophilia A (N = 66)

# Target Joints	# Subjects with Target Joints	Target Joints			
		Ankle	Knee	Elbow	All
Screening					
1	6	2	2	2	6
2	7	5	4	5	14
3	1	2	1	0	3
All	14	9	7	7	23
Completion					
1	4	2	0	2	4
2	2	1	2	1	4
3	0	0	0	0	0
All	6	3	2	3	8

Conclusion: Results demonstrate that twice weekly prophylaxis with BAX 855 is effective in preventing target joint bleeding and may resolve target joints in pediatric patients with severe hemophilia A.

PN-10

Frequency analysis and automaticity of postural sway during quiet stance in haemophilia and non-haemophilia subjects

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Introduction: In adults with haemophilia and arthropathy (HA) the sensory strategies of postural control are still poor understood. The contributions of these systems to postural control have been assessed by spectral analysis during the sway balance, dividing low, medium and high frequency, corresponding to visual/vestibular, cerebellum and somatosensory strategies respectively. The automaticity of postural control could be assessed through the sample entropy (SampEn), which determines the complexity or regularity of signal in a time series of data. A high SampEn express more irregularity of signals and could be interpreted in more automaticity and less attention during quiet stance. **Aim:** To determine the sensorial postural control strategies through frequency analysis of postural sway during bipedal quiet stance with deprivation of visual stimulus in haemophilia (HG) and non-haemophilia group (NHG). Secondly, to determine the automatic control of postural balance through sample entropy analysis (SampEn). **Methods:** A tri-axial accelerometer attached at L3 level was used to determine the displacement and acceleration of the center of mass (DCoM) during closed eyes (CE) conditions. Sensorial strategies were studied by spectral analysis of the DCoM signal, dividing in high, medium and low frequency. The SampEn of DCoM also was analysed. **Results:** Fifteen patients with hemophilic arthropathy and fifteen healthy subjects were included. Only the mediolateral direction showed differences between groups ($p < 0.05$). The HG showed minor energy in high and medium frequency values ($p < 0.05$), and mayor in the low frequency band. The NHG showed the highest SampEn in comparison to HG in anteroposterior and mediolateral axis ($p < 0.05$). The positive relationship was found between automaticity of postural control and high frequency in NHG and HG in both axis ($R^2 > 0.40$ and $R^2 > 0.70$, respectively). **Conclusion:** The results confirm that the patients with HA have a less automaticity of postural control with less contribution of the somatosensory system, compensated with more contribution of vestibular inputs compared to NHG. This approach could be useful to assess the sensory strategies of postural control during quiet stance and determine the motor control progress of the postural exercise program in people with haemophilia.

PN-11

Platelet rich plasma for chronic synovitis treatment in patients with haemophilia

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Introduction: Repeated bleedings episodes into the joints cause chronic synovitis in PWH. The aim of this work is to evaluate the effectiveness of the application of platelet rich plasma (PRP) into the affected joint with chronic synovitis after 3 months, six months and one year follow up. **Patients and methods:** Forty eight patients with 74 chronic synovitis joints were treated. Forty six patients were haemophilia type A and two type B (43 severe, 4 moderate, 1 mild). Two patients had inhibitors. Mean age was 26 years old (8-48). Forty were located in the knee (54%), 20 in the elbow (27%) fourteen in the ankle (19%). Patients were divided to analyze data according to follow-up times: 1 year (18 knees, 8 elbows and 2 ankles); 6 months (17 knees, 8 elbows, 8 ankles); 3 months (5 knees, 4 elbows, 4 ankles). Joint perimeter, visual analogue scale (VAS), number of bleedings (BE) and the Haemophilia Joint Health Score (HJHS) were evaluated prior to and again 3 months, 6 months and 1 year after treatment. PRP was obtained by intravenous blood centrifugation for 8 minutes at 1600 rpm and were injected into the joint cavity of each patient. **Results:** All patients reported pain relief ($p < 0.001$), decrease in joint bleeding episodes ($p < 0.001$) 3 months, 6 months, 12 months after treatment. No statistically significant differences were shown for BE after 3 to the year. Joint perimeter measures also improved 3 months, 6 months and a year after treatment for knee and ankle joints and after 3 months and 6 months for the elbow joint ($p < 0.001$). The difference in HJHS before and 3 months, 6 months and 1 year after treatment was statistical significant ($p < 0.001$). This significance is maintained even up to 6 months. **Conclusion:** All patients reported benefit of the PRP therapy. The improvement was more important in the first 3 months, and continues until one year up follow up. We believe that PRP injection is a simple, safe and inexpensive treatment for joint chronic synovitis, improving the quality of life of PWH.

PN-12

Usefulness of a simple self-administered joint condition assessment sheet in the management of children with hemophilia.

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Introduction and Objectives Hemophilic arthropathy is the serious complication of hemophilia deteriorating quality of life. Recently, a simple self-administered joint condition assessment sheet for the management of hemophilic arthropathy was developed and validated for adults. The objective of the work is

to verify the sheet for children. **Materials and Methods** From 2012 to 2016, 33 patients (25 children with hemophilia A and 8 children with hemophilia B, 5 inhibitor positive children with haemophilia) were enrolled in this research. The average age of them was 10.5(2-20) years old. The patients were asked to answer the self-administered sheet regarding to the condition of bilateral elbow, knee and ankle joints. The sheet comprises four items about bleeding, swelling, pain and physical impairment. At the same time, the joints are evaluated with Hemophilia Joint Health Score (HJHS) 2.1, DePalma and Arnold-Hilgartner (A-H) radiological classification. Each items of the sheet were correlated with those evaluations using Spearman's rank correlation. **Results** Three of four items were correlated with HJHS 2.1 in elbow and knee joints. Two of four items correlated with it in ankle joint. Especially, the item for pain was strongly correlated with HJHS 2.1 in knee and ankle joint (correlation coefficient: 0.77 and 0.77, respectively). It was also moderately correlated in elbow joint (0.56). The item for swelling was moderately correlated in every joints (0.56, 0.49 and 0.40 in elbow, knee and ankle joint, respectively). In elbow and ankle joints, the item for bleeding moderately correlated with DePalma classification (0.65 and 0.54, respectively) and A-H classification (0.65 and 0.55, respectively). **Conclusions** Using this concise self-administered joint condition assessment sheet would be helpful in the management of children with hemophilia.

PN-13

Specialist in action

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Introduction: Our country is an extensive territory with a highly specialized haemophilia treatment centre in Buenos Aires. Many years ago patients had to leave their homes and their tasks (work or studies) to be evaluated by the orthopaedic surgeon specialist in haemophilia. In 2006 a care orthopaedic program was established. The orthopaedist travelled to different centres of the country to evaluate the patients together with the local medical team. In 2007 it was decided to incorporate a physiotherapist into the programme that was called "specialists in action". The purpose of this paper is to show the experience of this program between 2007 and 2016. **Materials and Methods:** Sixteen haemophilia care centres in our country and one in Uruguay were included in this programme. Thirty-one visits to centers in Argentina and one in Uruguay were performed between January 2007 and December 2016. The average distance from the centres to the specialized care centre in Bs As was 873.78 km (58 km-1492 km). The total number of patients seen in Argentina was 831. The average number of patients seen per year was 85.8 (64-168). The number of patients seen in Uruguay was 27. To calculate the savings in the health system, we calculate the cost of the patient's bus tickets from the place of origin to the care centre, round trip and the costs of transporting a doctor and a physiotherapist to the same specialized care centre and the

corresponding meals. **Results:** The advantage of this programme is that involves the multidisciplinary team which treats the patient implicating them into the evaluation and treatment decisions. The patient's family can also be involved since they are not limited by the costs of moving to a distant centre of more complexity. Total savings of the program for the health system was U\$307, 222. The average saving per centre was U\$ 16,170 (1,163-65,917) depending on the distance of the centre and the number of patients attended. **Conclusions:** We developed a program that facilitates the multidisciplinary team work between different treatment centres that also generates marked saving costs for the health system.

PN-14

Physical activity in haemophilic patients: one year evaluation of hemorrhagic risk and factor levels (myPKFit)

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Introduction and Objective: The increase in life expectancies in haemophiliacs reinforces the need to promote a physically active lifestyle. According to expert recommendations, adults should perform 10,000 steps/day. To reduce the risk of bleeding in severe patients, it is necessary to adjust the amount of physical activity with prophylaxis and trough levels. **The objective of this study is** to determine the amount of physical activity by Fitbit Charge HR and joint bleeds in patients with haemophilia in prophylactic treatment controlled by myPKFiT. **Materials and Methods:** A group of 7 severe haemophilic patients type A (Age: 37.00±8.23 [28.00-49.00] years old; Weight: 84.77±29.36 [67.20-150.20] kg; Height 1.75±0.11 [1.54-1.85] m; IMC: 27.56±8.28 [20.69-44.85] kg/m²) from the Haemostasis and Thrombosis Unit of the Hospital La Fe, Valencia, Spain were included. The Fitbit Charge HR activity wristband was used to monitoring the amount of daily physical activity performed during 12 months. To adjust the prophylaxis treatment, a standalone web-based software that uses a published population pharmacokinetic (PK) model together with a Bayesian algorithm (myPKFiT, Baxalta) was used. myPKFiT was developed to predict PK parameters in patients with hemophilia A with only two blood samples. The PK values and joint bleeds were compared with the values of the previous 12 months. **Results:** Table 1 depicts the results obtained in the 12 months follow-up and in the 12 previous months. In general, in the follow-up year, patients suffer less bleedings with high trough levels and similar rFVIII consumption and amount of physical activity. **Conclusions:** myPKFit allows customize prophylaxis in severe patients with haemophilia according to the amount of physical activity monitored with an activity wristband, reducing the risk of joints bleedings. However, further studies are needed to include a larger sample size.

TABLE 1: Arthropathy, joint bleeds, pharmacokinetic parameters and activity data

Patient	1	2	3	4	5	6	7
Gilbert score	36	19	23	17	23	14	5
Previous 12 months							
Joint Bleeds	2	1	1	0	1	2	1
rFVIII consumption / day (IU)	797.8	633.9	473.2	721.1	522.8	1107.0	521.2
half-life (hours)	14.7	13.8	13.9	21.0	17.4	11.1	12.7
Trough level (IU) 48 / 72 hours	5.5 / 1.4	3.0 / N.A.	N.A. / 1.6	N.A. / 5.3	N.A. / 2.5	1.5 / 0.85	1.7 / N.A.
Steps /day (mean ± sd)	12323 ± 3634	14398 ± 1693	9151 ± 3426	9748 ± 2861	13763 ± 6244	6936 ± 2376	10436 ± 2703
12 months follow-up							
Joint Bleeds	1	0	0	0	2	0	0
rFVIII consumption / day (IU)	906.6	642.9	489.0	714.3	500.0	1191.0	474.6
half-life (hours)	15.9	13.6	14.5	21.8	16.5	11.1	12.6
Trough level (IU) 48 / 72 hours	5.5 / 2.3	3.0 / N.A.	N.A. / 1.6	N.A. / 6.1	N.A. / 3.4	1.5 / 0.85	2.3 / N.A.
Steps /day (mean ± sd)	11178 ± 3508	13628 ± 3013	9606 ± 3295	10087 ± 3734	13885 ± 4951	6761 ± 3621	10358 ± 3277

N.A.: Not available.

PN-15

Acute musculoskeletal injuries in the hemophilic patient: clinical and ecographic protocol

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Introduction: The musculoskeletal injuries are a characteristic problem with serious consequences in the haemophilic patients. Diagnosis, treatment and evolutive control of this injuries, impacts on hemophilic arthropathy. The early diagnosis of acute hemarthrosis is basic and, in our center, is based on clinical and ultrasound criteria. In our protocol for acute episode, we evaluated the following physical symptoms: pain, swelling, range of movement (ROM) and functional impotence. The ultrasound evaluation is quickly and allows to objectify the presence of alterations, even though the clinical symptoms have disappeared. **Objectives:** To present our clinical and ultrasound protocol. To describe the evidence of lesions by ultrasound and the non-operator dependent characteristic of this protocol. **Materials And Methods:** Four parameters were explored: 1) Pain 0-3 points (no pain, mild, moderate and severe); 2) swelling 0-3 points (no, present, remarkable, "tension"); 3) active ROM without pain (flexion / extension); 4) The effort / load: 0 points (possible and even a previous situation), 1 point (impossible or worse previous situation). The ultrasound protocol consists of obtaining a longitudinal cut in concrete references of the elbow, knee and ankle joints. The clinical score= 0, with ROM equal to previous situation, should be contrasted with ultrasound image without evidence of injury or comparatively equal to previous image in the absence of acute episode. **Results:** 37 hemophilic

patients with 61 lesions (38 knees, 15 elbows and 8 ankles) were examined by two independent observers. Clinical examination did not show significant differences using the described score. On ultrasound, the diameter of the lesion, as well as the skin-bone distance, did not provide differences. **Discussion And Conclusions:** The evaluation of pain, swelling and ROM are parameters sufficiently validated by scores recommended by the WFH, we agree to support its importance for clinical diagnosis. The benefit of ultrasound, for diagnostic confirmation of acute process, is also cited by most authors. In our opinion, the use in consultation of a simple protocol would improve clinical care and could influence the prevention of haemophilic arthropathy.

PN-16

A case note review of diagnostic ultrasound—identifying pathology masquerading as a bleeding event

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Introduction – Technological advances and the reduced costs of diagnostic ultrasound is allowing this tool to be exploited within a variety of clinical environments. In haemophilia care diagnostic ultrasound is used to improve disease surveillance through the use of scoring systems such as the HEAD-US scale (Haemophilia Early Arthropathy Detection with Ultrasound) and to assess for haematomas and joint haemarthroses. **Aim** – To report on the value of diagnostic ultrasound in identifying the presence of pathology that may masquerade as a bleeding event. **Methods** – The case notes of four patients were interrogated. The patients were known to have undergone ultrasound examination for a suspected bleed and subsequently found to be suffering from non-haemophilia related pathology. **Results** - All four patients had a diagnosis of severe haemophilia A. The patients all presented with symptoms regularly associated with a bleeding event. In each case clinical ultrasound excluded a bleeding episode. Following ultrasound each patient was urgently referred on for further diagnostic tests within dedicated imaging departments. One patient went on to receive a diagnosis of gout, one patient was diagnosed with superficial thrombophlebitis, the third patient was diagnosed with a large ganglion and the final patient was diagnosed with spindle cell sarcoma. **Conclusions** - When patients present to haemophilia centres' with bleed-like symptoms it is understandable that this is often the initial default diagnosis. The use of clinical ultrasound can help inform this initial diagnosis, direct or re-direct a patient's care and improve the efficiency and effectiveness of clinical pathways.

PN-17

Comparing objectively measured physical activity and sedentary behaviour between adults with haemophilia and healthy controls

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Introduction: Haemophilia arthropathy leads to pain, loss of range of motion and muscle atrophy, resulting in limitations in activities and participation. Earlier research in children with haemophilia who benefit from prophylactic treatment showed that physical activity (PA) levels are similar to activity levels of healthy peers. Limited research was published regarding the PA behaviour of adults with haemophilia. **Aim:** The aim of the current study was to determine differences in physical activity and sedentary behaviour between persons with haemophilia (PWH) and healthy controls. **Methods:** Data of adult PWH were extracted from medical health records at the van Creveldkliniek, the Netherlands. Data of PA behaviour of healthy male adults were collected prospectively. Physical activity and sedentary behaviour were measured with a three-axial accelerometer (activ8) which can distinguish between; lying down or non-wear, sitting, standing, walking, running and cycling. Time spent on these activities was compared between persons with severe haemophilia, mild/moderate haemophilia and controls. Comparisons between groups with normal distributed data were made with ANOVA and Tukey correction for multiple testing and for not normal distributed data with the Kruskal-Wallis test. **Results:** In this interim analysis 105 PWH (73 severe and 32 mild/moderate) and 95 healthy controls were included. Mean (SD) or median [IQR] time per day spent in each activity was calculated. Differences between groups were found for all activities except biking. Compared to controls, persons with severe haemophilia spend more hours sitting (9.0 ± 2.0 vs 8.2 ± 1.8 ; $p < 0.05$) and standing (3.0 ± 1.4 vs 2.4 ± 1.2 ; $p < 0.05$), less hours lying/nonwear (9.7 ± 1.5 vs 10.6 ± 2.2 ; $p < 0.05$) and walking (1.9 ± 0.8 vs 2.4 ± 1.0 ; $p < 0.05$) and less minutes running ($0.5 [0.2-1.1]$ vs. $3.0 [0.6-7.8]$). Persons with mild/moderate haemophilia spend less hours lying/nonwear (9.5 ± 1.9 vs 10.6 ± 2.2 ; $p < 0.05$) than controls and more minutes running ($1.9 [0.9-3.4]$ vs. $0.5 [0.2-1.1]$) than persons with severe haemophilia. **Conclusion:** Persons with severe haemophilia showed more stationary (sitting and standing) behaviour and less physical active behaviour (walking and running) than healthy controls. This may be a risk for further decline in physical condition and limitations in activities.

PN-18

Current UK use of intra-articular steroid injection (IAI) and radiosynovectomy (RS) in adults and children with haemophilia

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Introduction: Repeated haemarthrosis leads to inflammation of the synovium, which if recurring can lead to chronic synovitis, repeated haemarthrosis and haemophilic arthropathy. There are procedures for controlling synovitis and these include IAI and

RS. These procedures aim to decrease recurrent haemarthrosis and slow down the bone and cartilage damage. IAI with corticosteroids are known to have potent anti-inflammatory properties and provide localised pain relief from synovitis. RS involves joint injection with a radioactive material into the inflamed synovium. The resultant effect causing synovial fibrosis and reduced haemarthrosis. The purpose of this study was to survey the current UK use of IAI and RS in adults and children.

Methods: Questionnaires were sent to 23 UK Comprehensive Care Haemophilia Centres to explore the current use of IAI and RS in adults and children with haemophilia in the preceding year 2015. This study was exempt from ethical review as no patient identifying details were used. **Results:** The response rate was 70%: 16/23 centres completed the questionnaires (3/16 centres returned 2 questionnaires for both adult and paediatric services (total 19 questionnaires). All centres referred patients for IAI; 85 adults (7 inhibitors) and 11 children (9 inhibitors) were injected. The most commonly injected joint in adults and children was the ankle. The majority of centres did not immobilise the joint following injection, though advocated reduced activity for between 24 hours – 2 weeks. 11/16 (69%) centres had referred patients for RS; 29 adults (4 inhibitors) and 11 children (4 inhibitors) were injected. All centres immobilised the joint following RS; duration of immobilisation ranged from 24 hours - 5 days. All centres offered rehabilitation following RS, with the majority suggesting this started 2 weeks post-RS. **Clinical Implications:** The survey objective was to identify the current use of IAI and RS in the UK based cohort of people with haemophilia, with a view to guiding future studies. There is variation in the management of patients in the UK that requires standardisation before comparison of outcomes can be completed.

PN-19

Effect of physiotherapeutic methods on joint problems and functional activity on patients with haemophilia

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Background: Musculoskeletal problems like joint pain, deformity or arthritis and changes in muscle strength are more common in patients with haemophilia (PWH) due to recurrent hemarthrosis or muscle bleed. Availability of factor concentrates is a major concern and only 1% of PWH in India are on regular prophylactic therapy. More than 75% of PWH in India are affected with musculoskeletal problems. With limited availability of factors, intermittent prophylaxis is best possible option. There limited data on the effect on joint scores and functional activity before and after physiotherapy with intermittent prophylaxis. **Methods:** Twenty PWH with diagnosed musculoskeletal problems who came to clinic were included in this study after due consent. Baseline measurements of haemophilic joint health score (HJHS) and functional independency score of haemophilia (FISH) were taken. The subjects underwent physiotherapeutic protocols for maximum of 10 days. HJHS and FISH were done after the intervention and the comparisons of pre and post intervention scores were analysed using paired t test. **Result:** Twenty patients

with median age of 18 years took part in the study. None of the patients were on regular prophylaxis regimen. All patients underwent 10 days of physiotherapy under the intermittent prophylaxis plan. The mean pre and post HJHS were 38.35 ± 3.11 and 18.8 ± 2.51 respectively showing a significant reduction in the HJHS ($p=0.00$). The pre and post intervention FISH scores were 17.05 ± 1.70 and 22.95 ± 1.58 respectively which also showed a significant improvement ($p=0.00$) in functional activity. **Conclusion:** This study highlights the effectiveness of physiotherapy under intermittent prophylaxis on joint score and functional activity. Although regular prophylaxis is the best option to reduce musculoskeletal complications, intermittent prophylaxis could be an option to reduce morbidity in resource constrained settings. Larger sample size and longer follow up will be required to watch for repeated bleeds and further joint damage.

PN-20

Evaluation of a Canadian video-based educational tool for assessment of acute joint bleeds in children with hemophilia

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Introduction and Objectives: In a pediatric hemophilia care setting, parents are often the ones to first recognize and evaluate a possible joint bleed. Lack of parental/caregiver knowledge of assessment and recognition of joint bleeds can result in delayed treatment and prolonged treatment times and resolution, making parent/caregiver education an essential component of care. Current practice is to teach and review joint assessment with families at review visits or when their child is seen for acute bleeding episodes. Printed materials are often provided. Previous studies have shown that educational videos can be effective modes of information delivery and can improve patient/caregiver recall of information. Videos can be viewed by parents at convenient times or real-time during acute episodes. Our objective is to evaluate the efficacy of an educational video in meeting the learning needs of parents of children with hemophilia in the assessment of an acute joint bleed. **Materials and Methods:** Participants included parents of children with hemophilia age 4-12 with proficient English. Parents were asked to view an educational video demonstrating an assessment of the ankle joint for an acute bleed. The video included clinic contact information and personal stories on experiences with hemophilia and joint bleed assessment. Parents were asked to complete a self-administered questionnaire rating the importance, relevance, clarity and usefulness of the videos. **Results:** Parents/caregivers viewed the video positively and voiced that it helped them with review of their joint assessment skills. They also valued the perspectives that parents in the video shared. **Conclusions:** Parents/caregivers may have knowledge gaps in joint bleed recognition and assessment. Videos demonstrating joint assessment skills can be a useful tool for parents/caregivers of children with hemophilia. While it does not replace education provided during face to face visits, it can provide reminders that are easily accessible between clinic visits.

PN-21

Hemophiliac iliopsoas bleeding: a 20-year experience

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Introduction: Iliopsoas bleeding regarding hemophilia is problematic to manage in developing countries. Over 20 years, the treatment concept of hemophilia has been altered from on-demand treatment to prophylaxis. In Thailand, the additional use of factor concentrate combined with cryoprecipitate/cryosupernatant plasma has increased since 2003. After initiating nationwide hemophilia treatment supported by Thai government in 2006, more hemophilia patients have received adequate factor replacement to treat emergent bleeding. **Materials and Methods:** We conducted a 20-year retrospective chart review to evaluate the management and outcome of iliopsoas bleeding at the International Hemophilia Training Center: Bangkok. **Results:** A total of 39 episodes of iliopsoas bleeding confirmed by ultrasonography among 15 hemophilia patients (12 hemophilia A, 3 hemophilia B) were analyzed. Seven patients had inhibitor (6 hemophilia A, 1 hemophilia B). The median inhibitor among 6 patients with high titer was 17.9 BU while one patient had low titer of 4.2 BU. Eight of 10 bleeding episodes among patients without inhibitor were treated with adequate factor replacement defined by peak factor activity $\geq 80\%$ on the first 2 days of bleeding and maintained trough factor activity $\geq 30\%$ at least 3-5 days. All 6 episodes in one patient with low titer were adequately treated. In contrast, only 7 of 23 episodes among 6 patients with high titer were sufficiently treated with bypassing agents at least 3-5 days due to limited resources. Recurrent/repeated iliopsoas bleeding was significantly found in patients with inhibitor ($5/7=71.4\%$) compared with those without inhibitor ($1/8=12.5\%$) with a p-value of 0.02. The complications involved anemia ($n=34$), femoral nerve paresthesia ($n=15$), infected hematoma ($n=11$), hemorrhagic shock ($n=5$) and pseudotumor ($n=2$). Follow-up ultrasonography was determined in only 14 episodes (36%) revealing 3 complete resolutions of hematoma, 7 stable hematomas and 4 progressions of hematoma. Two patients with inhibitor succumbed to uncontrolled bleeding. Three patients still had abnormal gait. The remaining 10 patients could walk normally. **Conclusion:** Iliopsoas bleeding requires comprehensive evaluation and adequate factor replacement until complete resolution of hematoma by ultrasonography. Patients with inhibitor are at risk of recurrent/repeated bleeding and experience difficulty to manage in a resource-limited country.

PN-22

How are we assessing therapeutic relationships in the care of patients with hemophilia?

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Background: Hemophilia is a genetic condition characterized by recurrent bleeding into joints and muscles, often resulting in impairment of musculoskeletal (MSK) structures and function. Prevention of this process is a priority in hemophilia treatment, and patients and physiotherapists work together to deter MSK issues. This purposeful partnership of patient and physiotherapist is described as the “therapeutic relationship.” Researchers have used various measures of therapeutic relationship when studying associations between these relationships and the outcomes of physiotherapy interventions. In the physiotherapy care of patients with hemophilia, it is similarly unclear whether a standard method of assessing therapeutic relationships has been established. **Objective:** The purpose of this project was to characterize the nature and extent of the research evidence concerning assessment of physiotherapy therapeutic relationship in hemophilia. **Methods:** We conducted a systematized literature search in consultation with a health research librarian from the University of Alberta. MEDLINE, EMBASE, CINAHL, and PsycINFO were searched using a combination of subject headings and keywords for the concepts *physiotherapist*, *therapeutic relationship*, and *hemophilia*. We included studies that used a measurement instrument to evaluate the physiotherapist-patient relationship or a related construct (e.g. communication, treatment satisfaction) in hemophilia. **Results:** Our search identified 37 records. After conference abstracts and duplicates were removed, 27 records were screened for relevance. Three articles met inclusion criteria, and one additional article was found through citation searching. No studies assessed the therapeutic relationship between physiotherapists and patients with hemophilia. A related construct, patient satisfaction with treatment, was assessed in all four articles. However, a validated measurement instrument was not used in these studies. **Conclusions:** We were unable to find a measure of therapeutic relationship used in physiotherapy care of patients with hemophilia. Future research should involve validation testing of existing measures or potentially the development of a new measure in the hemophilia population. A validated measurement instrument will be necessary before researchers can begin examining associations between therapeutic relationship and outcomes of hemophilia treatment.

PN-23

In persons with severe haemophilia bone fractures occur at significantly younger age than in those with mild disease

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Introduction and objectives: Osteoporosis and fractures are uncommon in male population under 50 years of age. In persons with haemophilia reduced bone mineral density (BMD) associated with an increased risk of fracture has been reported. Chronic arthropathy, gait instability and muscle weakness may additionally predispose these subjects to higher risk of falls and fractures, yet real-life data on incidence of fractures in individuals with haemophilia are lacking. In this study we investigated incidence rate of fractures in a cohort of subjects with haemophilia treated in our haemophilia center. **Patients**

and methods: In this retrospective cross-sectional study we included 148 male subjects with haemophilia (57 with severe, 27 moderate and 64 mild haemophilia) mean age 41.2±16 years. All previous fractures were recorded over the last two decades (January 1996 - January 2016). Data was gathered from a total of 2960 patient-years of observation and incidence rate was calculated. **Results:** During the observation period for whole cohort of subjects with haemophilia 36 fractures have been recorded, giving total incidence of 12.2 fractures per 1000 patient-years. Although the incidence of fractures in patients with severe haemophilia (12.2 per 1000 pts years) was higher than in the group of subjects with moderate (9.6 fractures per 1000 pts years) and mild haemophilia (10.9 fractures per patients years), the difference between these groups was not statistically significant ($p < 0.05$). Interestingly, patients with severe haemophilia were significantly younger at the time of fracture (mean age 25.75±14.5 years) than patients with mild haemophilia (mean age 41±14.4 years) ($p = 0.0081$), while difference was not significant when compared to subjects with moderate hemophilia (mean age 32.6±14 years). **Conclusion:** Although the incidence rate of fracture for whole cohort of subjects with haemophilia is similar to data from literature for general population (12.2 versus 9.6 per 1000 patients years) patients with severe disease fracture at significantly younger age compared to those with mild disease, suggesting the influence of factors related to disease severity on the risk and time of fracture. Mechanisms underlying skeletal changes and prediction of fracture risk in patients with haemophilia need further investigation.

PN-24

Monitoring joint health with HJHS in patients with hemophilia with low bleeding rates: predictors of deterioration

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Objective Joint bleeds in persons with hemophilia may result in hemophilic arthropathy. Monitoring joint health is essential for identifying early signs of deterioration and allow timely treatment adjustment. The aim of the present study was to describe changes in joint health over a 5-10 years follow-up and identify factors associated with joint health deterioration in patients with hemophilia. **Methods** A post-hoc analysis was performed on data from previous cohort studies in patients with moderate/severe hemophilia, ≥ 16 years at T0. Joint health of ankles, knees and elbows was measured with the Hemophilia Joint Health Score (HJHS) from 2006-2008 (T0) to 2011-2016 (T1). Analyses were performed on patient level (Δ HJHS-total) and joint level (Δ HJHS-joint). Deterioration was defined as Δ HJHS-total ≥ 4 and/or Δ HJHS-joint ≥ 2 . **Results** Sixty-two patients (median age 25, 73% severe hemophilia, 48% of joints ≥ 1 joint bleed T0-T1) were included. After median 8 years,

HJHS-total deteriorated in 37% and HJHS-joint in 17%. Ankle joints (31%) showed more deterioration than the elbows (19%) and knees (3%). Deterioration of HJHS-total was associated with severe hemophilia only. Deterioration of HJHS-joint was associated with a higher number of joint bleeds between T0-T1, lower HJHS at baseline, presence of synovitis and more limitations in activities. **Conclusion** In 37% of patients with moderate/severe hemophilia and low joint bleeding rates joint health deteriorated over 5-10 years. Ankle and elbow joints showed most deterioration. Most non-impaired joints with ≤ 1 joint bleed and no synovitis during follow-up stayed non-impaired during five to ten years follow-up. Factors found in the current study help to identify which joints need less frequent monitoring in PWH with access to prophylaxes from an early age.

PN-25

Multidisciplinary collaboration to improve musculoskeletal care in people with hemophilia in thailand: a pilot project

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Background: Standardized musculoskeletal (MSK) assessment is a crucial part of comprehensive hemophilia care in order to identify early joint changes and prevent arthropathy. This is particularly important when prophylaxis is implemented. To accomplish this goal, multidisciplinary collaboration and enhanced MSK training between hematologists, physiatrists, physiotherapists, nurses and radiologists is important. In 2016, funding was obtained for the translation of outcome measurement tools, training and education, and the development of an MSK registry in Thailand. **Materials and methods:** The Hemophilia joint Health Score (HJHS) 2.1 and Functional Independence Score in Hemophilia (FISH) were translated into the Thai language for use by Thai medical personnel assessing PWH. An intensive three-day MSK training session took place in Bangkok in September, 2016 for multidisciplinary teams from two hemophilia treatment centers (HTCs); the experienced international hemophilia training center (IHTC), Ramathibodi Hospital, Bangkok and a new regional comprehensive HTC from Chiang Mai University Hospital, Chiang Mai. The training which focused on hands-on teaching of the HJHS and FISH was co-led by experienced physiotherapists from The Hospital for Sick Children, Toronto Canada and the Mountain States Regional Hemophilia Center, University of Colorado, Denver, U.S.A. **Results:** A total of fourteen HTC multidisciplinary team members, , trainers, invited speakers and six PWH from five families were involved in the training session. After a series of educational sessions with hands-on training and assessments, the participants demonstrated increased assessment skills and indicated increased understanding of the evaluation tools on the post training evaluation forms. As a result, the centers in

Ramathibodi and Chiang Mai have started to perform both the HJHS and FISH on their PWH. **Conclusion:** Through the collaboration between the experienced and the newly-established teams, the program of comprehensive MSK assessment and care could be systematically established in HTCs using a Train-the-trainer approach, with Ramathibodi and Chiang Mai as the trainers. This project will be the model for improving national hemophilia assessment and care in HTCs around Thailand with the potential for collaborative clinical research.

PN-26

Patient reported use of pain medication among patients with hemophilia at the Helsinki University Hospital Hemophilia Comprehensive Care Center

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Objectives: Pain, mainly caused by bleedings and chronic arthropathy, is a common symptom among patients with hemophilia (PWH). In patients with chronic arthropathy there is often a discrepancy between the levels of pain, functional joint status and imaging results. The aim of this study is to evaluate the level of pain by the use of pain medication related to joint function and to identify questions to be addressed in the future. **Methods:** We interviewed 28 consecutive patients regarding the use of pain medication during hemophilia clinic visits. Hemophilia Joint Health Score (HJHS 2.1) was performed when appropriate. **Results:** The mean age of patients was 48 (range 27-72). HJHS was available for 19 patients with a mean of 35.9 (range 6-66). Of sixteen patients using pain medication on demand, 15 had HJHS (mean of 34.0, range 12-66). For nine patients using pain medication regularly, four had HJHS (mean 50.5, range 43-58). Two patients reported no analgesic usage; one of them had a HJH score of 6. COX-2-inhibitors (etoricoxib or celecoxib) were used by 53.6% of our patients, and paracetamol (acetaminophen) by 25%. Coxib or paracetamol was used as first line treatment by 75% of all patients. Mean HJHS for these patients was 39.8 (range 6-66). Three patients reported regular, continuous use of coxib. Six patients used mild opioid, mainly codeine combined with paracetamol, one patient used tramadol. HJHS was available for five patients, mean 34, range 13-65. Strong opioids were used by six patients; five had severe hemophilia and four also an inhibitor. HJHS score was available only for two of these patients; 57 and 51 respectively. Only three patients used targeted medications for chronic or neuropathic pain (duloxetine, venlafaxine, gabapentin and pregabalin). **Conclusions and relevance for hemophilia care:** In our treatment center higher HJHS seems to correlate with increased regularity of pain medication usage and risk for opioid usage among PWH, although there were significant exceptions. Patients' attitudes and fears of side-effects, addiction and tolerance also limits use. More research is needed to understand the motivations; resilience, perseverance, and perceptions among PWH concerning usage of pain medication.

PN-27

Radiosynoviorthesis with ⁹⁰Y in patients with hemophilic arthropathy: a single institution experience

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Introduction: Arthropathy is the main cause of morbidity in the patient with hemophilia (PWH). Continuous articulation bleeding causes damage by chronic inflammation, joint structure deterioration and deforming contractures resulting in chronic pain and disability. Thus for target joint, an alternative treatment is radiosynoviorthesis (RS), which consists of the application of a radioactive isotope into the joint cavity, causing necrosis and fibrosis of the synovium. **Objectives:** To describe the results of radiosynoviorthesis in a prospective cohort of PWH. **Material and Methods:** A 140 patient cohort, under informed consent, 33 RS were performed (23 knees, 6 elbows and 4 ankles) in 23 PCH. The indication was to have a target joint. Every patient was in secondary prophylaxis (SP) 24 months prior to inclusion. To prevent new hemarthrosis they were admitted to the hospital 12 hours prior to the procedure, and Factor VIII (FVIII) was applied at a dose of 50 IU / kg for 22 PWH without inhibitors and to for one patient with a high-response inhibitor, 100 IU / kg FEIBA was prescribed. The same prescription was repeated half hour prior and 12h after RS. The dose of Yttrium 90, for intra-articular application, was 10mCi. During the first 24 hours they remained bed ridden, with ice applied locally to affected joint, ketorolac, celecoxib and two 8 mg doses of iv dexamethasone were given for pain and swelling. Ceftriaxone was given as an only dose antibiotic. Factor VIII at 25 U/kg/day continued for one week, and the patient with inhibitor continued with FVII 90 ug / kg / day, for the same time. Patients were encouraged to undertake aquatic rehabilitation 7 days after RS, and then continue for at least 6 months as well as SP and monthly Zoledronic Acid for a year. **Results:** Average age was 18.8 years (9-45). After RS, the hemorrhage rate decreased. 80%, 19 joints without hemorrhage, two patients had dermatitis in the area of infiltration. Ingestion of analgesics decreased by 60%. **Conclusions:** RS with ⁹⁰Y decreased the incidence of hemarthrosis and pain.

PN-28

Signposting the need for joint imaging in Haemophilia Clinic: using the Haemophilia Joint Health Score as a guide.

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Identification of arthropathy development at the earliest opportunity enables the clinician to initiate management aimed at limiting disease progression. The Haemophilia Joint Health Score (HJHS) may be useful in indicating this where specific domains within the score might be particularly useful. Ultrasound scanning has been proposed as a high utility alternative to MRI imaging in the surveillance of joints in haemophilia. A haemophilia-specific score, the HEAD-US, has been developed and is in use clinically. This cross-sectional study of 38 people was carried out

to evaluate the correlation between the HJHS and HEAD-US tool in order to identify any indicators for early arthropathy as indicated by the "Synovitis" domain of the HEAD-US tool. A consultant radiologist conducted the scans and an experienced clinical specialist physiotherapist completed the HJHS. The "Synovitis" domain was compared to the HJHS domains for each joint. Correlational analyses were carried out and as the underlying prevalence of arthropathy at each joint is not known, likelihood ratios were calculated. Results indicate that for each joint different domains are more highly correlated and that there were specific domains that showed least or no correlation repeatedly. Six out of ten measured domains were not significant for ankle synovitis with the best correlation being "Swelling" with a moderate result ($\rho = .363, p = .002$). Whereas "Atrophy" had a strong correlation at the knee ($\rho = .757, p = .0005$) and elbow ($\rho = .619, p = .0005$). Likelihood ratios of 4.16 and 7.08 are present at the knee and elbow indicating a moderately increased likelihood of disease in the presence of atrophy. A likelihood ratio of 5 at the ankle also confers a moderately increased likelihood of disease in the presence of swelling. Clinicians should consider referral for US scan at the earliest signs of muscle atrophy at the knee and elbow, and swelling at the ankle as measured by the HJHS. Further investigation would be beneficial to identify the best pathways to early intervention for arthropathy prevention and amelioration.

PN-29

Spontaneous lumbar facet joint hemarthrosis in hemophilia a patient presented with lower back pain and sciatica

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Introduction and Objectives: Lumbar radiculopathy most commonly is caused by herniated intervertebral disc. We would like to report a rare case of spontaneous hemarthrosis of the lumbar facet joint in hemophilia A patient manifesting as severe lower back pain and sciatica. **Patient and Methods:** A 61-year-old hemophilia A male suffered acute onset of severe lower back pain and left-side sciatica without history of trauma. Magnetic resonance imaging revealed hematoma around the left L5/S1 facet joint with left S1 root compression. There was no herniated intervertebral disc. **Results:** After factor replacement therapy and adequate pain management, the lower back pain improved but left -side sciatica and numbness persisted. The patient received physical therapy including hot pack, short wave diathermy, Interferential current therapy, therapeutic exercise and core muscle strengthening subsequently. The patient's symptoms were relieved 6 months later after conservative treatment. **Conclusions:** We reported the first case of spontaneous lumbar facet joint bleeding in hemophilia patient. The etiology and pathomechanism of the hemorrhage at the lumbar facet joint are unclear, but that could be an association with coagulopathy of hemophilia and degenerative change of the facet joint.

PN-30

Stability limits evaluation on hemophilic patients using Kinect v2

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Introduction: Hemophilic patients are prone to suffer loss of balance control capabilities due to the joints deterioration, and are therefore advised to perform specific training. Currently, new devices related to video games are being used in physical therapy programs and appear as a promising tool for balance and rehabilitation. **Objective:** Develop dedicated software for Kinect v2 for stability limits evaluation and training on hemophilic patients with different degrees of arthropathy. **Methods:** The study has been carried out using a Kinect v2, a Windows PC and dedicated software designed in Matlab/C++. The center of mass calculation is implicitly done in the code using a table of anthropometric measurements and taking into account the 3D joint positions calculated by Kinect v2. The Kinect v2 is placed facing the test subject at a distance of 2.5 m. The test subject is asked to sway his body from the base position to reach eight target positions sequentially in different directions and a score is awarded considering the percentage of time spent in the target spots. Three levels of difficulty, with increasing sway requirement have been implemented. The hemophilic population consists of 8 individuals (height=1.77±0.06m, weight=88.95±29.79kg, age=34.62±10.42 years old, Pettersson=5.00±6.24 / 5.43±5.29 [knees / ankles]). **Results:** Results of this pilot study are summarized in TABLE 1. As expected, the score is lower as the level of difficulty increases. However, only significant differences are obtained between levels 1 and 3. This fact can be explained by the rapid motor learning process during exercise execution. **Conclusion:** Kinect v2 can be used in the evaluation and training of control postural in hemophilic patients at a fraction of the cost of other custom-made balance platforms. Additionally, this setup can be installed in the patients' home. However, further studies are needed to validate this software.

	Patient								mean
	1	2	3	4	5	6	7	8	
Level 1	76.4 ± 6.8	80.0 ± 6.3	77.4 ± 10.8	62.8 ± 17.8	68.8 ± 14.7	52.3 ± 15.3	48.0 ± 9.1	54.5 ± 15.7	65.0 ± 4.4
Level 2	77.6 ± 7.1	79.4 ± 3.7	79.0 ± 12.1	57.4 ± 23.0	52.9 ± 36.5	13.1 ± 13.3	48.1 ± 25.9	35.0 ± 27.1	55.3 ± 8.3
Level 3	72.8 ± 8.4	77.5 ± 5.7	69.9 ± 17.5	43.1 ± 35.2	53.3 ± 30.6	8.9 ± 14.1	37.5 ± 24.5	39.6 ± 28.3	50.3 ± 8.1*

TABLE 1: Mean score (%) ± standard deviation across all balance positions per individual per difficulty level. *p<0.05 between levels 1 and 3 according to the ANOVA test.

PN-31

Synovitis in hemophilia – The German guideline

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Introduction and Objectives: The treatment of patients with hemophilia (PwH) has undergone a significant development through the last decades. Especially the introduction of a prophylaxis in young patients reduced the incidence of severe bleedings. Nevertheless, there still seems to be a lack of evidence of when and how to treat a synovitis in those patients. Since a synovitis after a bleeding event is the start point of joint degeneration, it is crucially important to start an adequate treatment at the right moment. The intend of our work was, to develop a guideline based on the evidence in literature to support physicians, medical staff and patients in the treatment of synovitis in PwH. **Material and Methods:** All member societies of the AWMF were invited to take part in the work of our guideline group. The group consisted of experts in the fields of hemophilia and hematology, nuclear medicine, pediatrics, physiotherapy, orthopedics, radiology, and sports medicine. Based on the regulations of the AWMF, the present and past literature on synovitis and hemophilia was acquired, systematically reviewed and thereafter scored with the Oxford rating system. A total of xxx papers and abstracts was scanned and finally xx were reviewed in detail by the working group. The content was evaluated and put in context to the working thesis. **Results:** We developed the underlying work for a guideline that may help physicians, medical staff and patients to treat a synovitis according to the available evidence in literature. The result is subdivided in diagnostics, treatment and secondary prophylaxis of synovitis. Besides a comprehensive paper we aim for short versions for physicians and patients that offers a quick guide in the daily routine work. **Conclusions:** The guideline may be a valuable tool for physicians, medical staff and patients for the treatment of early and late onset synovitis. In the future, treatment of the PwH can be evidence based on what has been published in literature. Nevertheless, the review of the literature has shown that many established treatment options can't be substantiated with sufficient evidence in literature and are only recommended by experts' opinion. This knowledge may be the basis for further research.

PN-32

The impact of a group educational intervention to teach parents of children with hemophilia and pediatric hemophilia patients about joint assessment

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Introduction and Objectives: Hemarthroses are a concern in patients with hemophilia and, if not adequately managed, can lead to long term joint damage, pain and disability. Joint assessment includes looking for swelling, pain and changes in range of motion at the first sign of hemarthrosis and

throughout the course of resolution. Lack of parental and/or patient knowledge of joint assessment may delay treatment and prolong treatment time and resolution. We hypothesize that with increased knowledge and better assessment skills, parents/patients will be able to recognize and initiate management of joint bleeds earlier, and effectively communicate their findings to healthcare professionals. Our objectives for this study were to determine baseline joint assessment skills and increase subject competence in performing a joint assessment on themselves or on their child. **Materials and Methods:** We recruited parents of children with hemophilia age 1-13 and children with hemophilia age 14-18. Subjects were asked to perform an assessment of the knee and ankle joints, on their child or on themselves, and evaluated using an assessment checklist. They attended a 30 minute education session during which they were taught basic joint anatomy and physiology as it relates to hemophilia and joint bleeding, and how to conduct an assessment of the knee and ankle. Subjects were re-evaluated on their assessment skills immediately after the education session and 3 months later. **Results:** 36 subjects completed the study. At baseline, median checklist score was 12 out of a possible 24 items. Median scores immediately after and 3 months after the intervention were 14 and 16, respectively. Data analysis showed a statistically significant improvement between pre and post intervention scores. Checklist items subjects scored well on included checking for swelling, presence of pain (at rest and with movement) and range of motion. Items that were commonly missed included pain location, pain with palpation, comparison of range of motion between the affected and unaffected side, and bruising. **Conclusion:** Our study shows that a group educational intervention was effective in improving the practical joint assessment skills of parents of children with hemophilia and adolescents with hemophilia.

PN-33

To study the dosage & effects of continuous passive mobilization

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Background: Sub-acute stage of post knee joint bleed, approach is to gradually increase weight bearing, loading on the joints, increase range of motion (ROM) of knee to facilitate faster functional activities. Benefits of CPM, structural & functional changes in the connective tissue, one must judiciously monitor CPM to adjust parameters in accordance with the patient's outcome measurements. CPM prevents intra-articular, peri-articular adhesions by maintaining proper constituents within ground substance. CPM also increases range, dynamic mechanical loading on joints. Biomechanics of body and angle 70° for stair case activities are required (Mow & Hayes). To prevent the wrong biomechanics, achieve ROM in knee joints timely, appropriate mobilization is required. CPM motion is passive so muscle fatigue does not interfere with movement. CPM machine is able to apply controlled motion continuously. (Robert Salter). There are no dose response studies to guide

in treatment parameters of CPM use in studying magnitude of motion, velocity & force of CPM so FREQUENCY & DURATION parameters of CPM were studied in patients with knee target joint. Age- 21 to 45. **Method:** 25 patients in each group. ROM documented with pre and post CPM usage before study and periodically once weekly. Also swellings, pain, quality of life scale were measured. Group A- **No factor** + exercises active assisted+ CPM for 20 minutes **Time** in supine position, 5 times a week for 4 weeks. Group B- Same like Group A +CPM for 45 minutes. Group C – **Factor** + exercises active assisted+ CPM for 20 minutes in supine position, 5 times a week. Group D- Same like Group C +CPM for 45 minutes. **Results:** Group A, improved ROM median 30°, HJHS 7. Group B, improved ROM 55°, HJHS improved 9. Group C, improved ROM 50°, HJHS 10 Group D, improved ROM 70°, HJHS 10. **Conclusion:** A & B, Group B ROM improved faster rate, muscle efficiency. C and D better ROM faster, increased muscle efficiency. Functional and Quality of life score improved in all groups with lesser bleeding episodes.

PN-34

Ultrasonographic findings in hemophilic joint treated with radiosynovectomy

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Introduction and Objectives: Radiosynovectomy is an effective non-surgical option in the treatment of recurrent hemarthrosis and chronic synovitis in patients with hemophilia. Clinical effects of radiosynovectomy, including decreased bleeding frequency, reduced factor consumption, and pain relief, have been widely reported. However, only a few studies have explored the influence of radiosynovectomy on articular structure based on imaging. Currently, ultrasonography has become a useful tool in evaluating hemophilic joints owing to its diagnostic capability in detecting synovial hypertrophy, joint effusion, and osteochondral changes. At present, no study investigated the ultrasonographic changes in hemophilic joint treated with radiosynovectomy. The aim of our study is to use ultrasonography to evaluate the influence of radiosynovectomy on articular structure of hemophilic joints. **Materials and Methods:** Radiosynovectomy was performed in five joints (2 ankles, 2 elbows, 1 knee joint) of three patients with hemophilic synovitis by using ⁹⁰Y citrate colloid in the knee and ¹⁸⁶Re sulfide colloid in the elbow and ankle joints. The effect of radiosynovectomy was measured by clinical response and ultrasound examination. The joint range of motion and pain scale (visual analogue scale) was used to evaluate clinical response. The ultrasonographic evaluation included synovial thickness, changes in cartilage, and joint effusion. Difference percentage in synovial thickness was used to compare the changes after radiosynovectomy. **Results:** The mean patient age was 20 years. The mean follow up time was 1 year and 10 months. Clinical responses included pain relief, and increased joint range of motion was observed in all of the participants after radiosynovectomy. The thickness of

synovium increased within one month after radiosynovectomy. Afterward, the thickness of synovium began to decrease at approximately one month after the treatment. The effect of synovium shrinkage persisted up to three years in one of the three participants. No changes in cartilage surface were found after radiosynovectomy, and no joint effusion was observed during follow up. **Conclusions:** Ultrasonographic evaluation can provide more information about the influence of radiosynovectomy on articular structure. Further investigation of large series and long-term follow-up is needed for better understanding of the effect of radiosynovectomy.

PN-35

Utilising the weight-bearing lunge test as a screening tool for early ankle arthropathy in haemophilia

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Introduction: The bleeding disorder community's goal of preventing joint bleeds in haemophilia has yet to be realised. While joint bleeds continue, children are at risk of developing chronic arthropathy. At present the ankle joint is most vulnerable which is challenging on many levels due to difficulties in symptom management, identifying those most at risk and putting into place strategies for prevention. A recently developed logistic regression model explained 81.1% of the variance in ankle joint disease group ($p < .0005$). This model included Weight-Bearing Lunge Test (WBLT) for dorsiflexion motion.

Objective: It has been suggested that a left-to-right difference of greater than 1.5cm on the WBLT is indicative of the presence of pathology. A multi-centre cohort study attempting to identify factors that could predict presence of haemophilic arthropathy at the ankle included this test. **Methods:** A case-control study assessed three groups (each N=30): PWH with ankle disease, PWH with healthy joints and normal volunteers. Within subjects data analysis was conducted to assess the clinical utility of the WBLT. Results were dichotomised into values greater or equal to 1.5cm difference or less than 1.5cm difference. A Chi Square test of Independence was carried out to assess if there were differences between the groups with a Cramer's V to indicate the strength of association. Spearman's correlations were carried out to explore any relationships between factors that might be potential explanatory anthropometric variables. The value of the test to clinicians was evaluated by assessing likelihood ratios.

Results: Participant group and side-to-side differences were significant, $p = .0005$ with a moderately strong association of .426, $p = .0005$. There were significantly more participants in the HmAk group with a dorsiflexion asymmetry. No relationships existed between the WBLT and anthropometric data. A positive likelihood ratio of 10.13 indicates that in the presence of a

positive test, the person has a ten-fold increased likelihood of having ankle arthropathy. **Conclusion:** The WBLT is a useful test with high clinical utility. Its use is highly recommended in PWH in order to aid clinicians in their screening and monitoring of the musculoskeletal health of those with haemophilia.

PN-36

The World Bleeding Disorders Registry: A Tool to Increase Capacity of Data Collection Around the World

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Background/Aims: The World Bleeding Disorders Registry (WBDR) (ClinicalTrials.gov NCT02776826) was designed to collect patient data through participating Hemophilia Treatment Centres (HTCs), with the goal of increasing the quantity and quality of data available on patients with hemophilia. The experience and capacity of HTCs to collect quality patient data varies greatly around the world. A Universal Case Report Form (UCRF) was developed to provide a standardized tool to increase the capacity of data collection, and is the basis for the CRF to be used in the WBDR. The feasibility of conducting a global patient registry using the minimal data set of the U-CRF was assessed in a pilot study.

Methods: The pilot was an observational, global registry of patients diagnosed with hemophilia A or B, replicating the methodology of the planned WBDR. A web-based data entry system (McMaster University, Canada) was developed, using the minimal data set of the U-CRF. HTCs from regions around the world were invited to participate in the pilot study of the WBDR. Along with patient and HTC interest in participating in the registry and ability to obtain ethics approval, the ability to collect and enter quality data in the web-based data entry system was one of the primary outcomes of the pilot study.

Results: A total of 26 HTCs representing 25 countries, and 356 patients participated in the pilot study of the WBDR. Using the minimal data set of the U-CRF, all 26 HTCs collected standardized demographic and clinical data on at least 10 patients each. The number of joint bleeds among all patients was 261 (73%), muscle bleeds was 134 (38%), CNS bleeds was 10 (3%) and other bleeds was 125 (35%).

Conclusions: Preliminary data on bleeding events demonstrates the potential to collect clinical data on hemophilia patients on a global level. Pending approval of the WBDR, HTCs with diverse geographic and economic backgrounds will be invited to participate in mid-2017. Plans are underway to include musculoskeletal outcomes in the full scale WBDR.