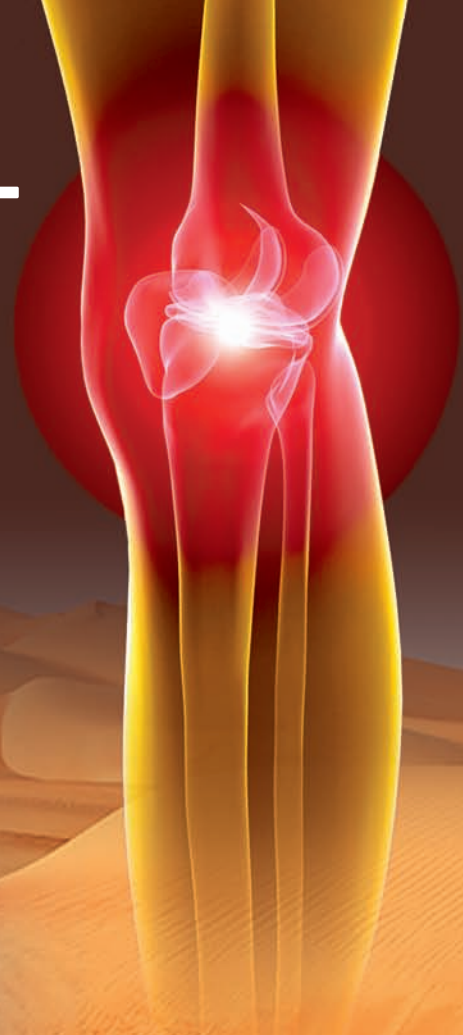


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Abstracts

Session A1 - PWH: The Way of Life

A1.1

Primary Prophylaxis in Young People with Hemophilia

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Decades of clinical experience support the concept that primary prophylactic clotting factor replacement for severe hemophilia A and B can decrease the incidence of joint bleeding and prevent joint damage, when compared to replacement in response to bleeding or secondary prophylaxis begun to treat established joint damage. Although different definitions of primary prophylaxis have been used, the onset of prophylaxis prior to the onset of joint damage is critical. Even in countries with abundant replacement clotting factor, institution of primary prophylaxis does not necessarily imply immediate institution of "full prophylaxis" schedules. Instead, approaches tailored to minimize morbidity and optimize adherence and empowerment of families may be pursued. An analysis of 10,262 patients in a U.S. database has recently confirmed that there is additional benefit that prophylaxis is associated with a significant risk reduction for intracranial hemorrhage. Epidemiologic data suggesting that early institution of primary prophylaxis in the absence of intensive treatment or bleeding may decrease the risk of FVIII inhibitors is more controversial. Randomized clinical trial data now also demonstrates superior clinical outcomes using primary prophylaxis compared to a very aggressive on-demand replacement schedule. (Manco-Johnson et al, N Engl J Med 2007). An unanticipated finding in that trial, however, was that the number of clinically observed hemarthroses correlated weakly with joint outcomes as determined by MRI. The finding suggests the possibility that, even without clinically evident joint bleeding episodes, repeated osteochondral microhemorrhages may contribute to deterioration of joints in the absence of prophylactic replacement. Additionally, animal models suggest that adequate initial hemostasis in response to bleeding does not ensure normal wound healing, which instead may require maintenance of some hemostatic potential during the course of wound healing. New insights into the scope of mechanisms by which clotting factor protects the joint may be required to optimize primary prophylactic strategies.

Key words: primary prophylaxis, hemophilic arthropathy

A1.2

Living with Hemophilia: The Physiotherapist's Role in Adolescence to Young Adulthood

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Adolescence into young adulthood is a time when many changes occur at varying rates and intensity including physical, emotional, social, and psychological alterations. Body size and structure can change dramatically, resulting in self-image concerns. The adolescent strives to gain independence, often testing boundaries, and peer acceptance is important. For boys with hemophilia, this transition period can present many challenges, often occurring simultaneously and potentially becoming overwhelming. Joint health may be a low priority for the youth at this time.

The hemophilia program physiotherapist can play a significant role in educating and supporting adolescents/young adults during this transition. During childhood, parents make decisions regarding bleed detection/management, return to activities, potential consequences, and activity selection. During adolescence, the emphasis changes from passive to active involvement. Gradually building upon the youth's knowledge base to develop a good understanding of the effects of blood in the joint, the potential development of chronic synovitis and joint arthropathy will help them find a balance between optimal bleed recovery and what they want to do. Stressing that factor replacement alone is not sufficient to treat joint and muscle bleeds is essential. Keying in on the adolescent/young adult's priorities in exploring the risks/benefits of activities and potential physical jobs/careers and recommending possible modifications will assist the physiotherapist to make a positive influence in the young man's choices.

Group education sessions that generate topics of interest/concern to youths and young adults that can be addressed by the physiotherapist may be of particular benefit to shy or less confident individuals. The venues can also be varied, more casual, and less boring or intimidating.

Starting the transition process from passive to active participation earlier in adolescence may result in instilling good habits at a younger age. Truly tailoring the approach to match the youth's priorities will ensure his commitment.





Abstracts (cont'd)

A1.3

Surgical Treatment in Patients on Primary Prophylaxis: What is the Incidence?

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Prophylaxis is recommended as first choice therapy for patients with severe hemophilia in countries where factor concentrates are largely available. Primary prophylaxis refers to preventive treatment started in children before the onset of joint damage. Although the benefits of primary prophylaxis are well documented, only the Scandinavian and northern European countries have adopted, since long time, prophylaxis as the standard treatment in all children with severe hemophilia. Usually, the joint status is considered the primary endpoint of prophylaxis studies; however data on the impact of primary prophylaxis in reducing the need for orthopedic surgery are still scarce. Moreover, the wide availability of safe and effective replacement therapy has favoured an increasing number of elective orthopedic surgeries (i.e. joint replacement, arthroscopic procedures). These changes in surgical practice occurred through the last decades in a different way across the world, having rendered difficult the comparison between different patients' cohorts. Furthermore, the use of primary prophylaxis has allowed the participation to high-risk activities of young hemophilic patients. Consequently, an increasing need for several surgical procedures such as sports traumatology may be expected. The assessment of long-term outcomes of primary prophylaxis is warranted and, in this light, the claim for orthopedic surgery represents a relevant outcome, not only by the clinical standpoint but also in terms of pharmacoeconomics.

A1.4

Hemophilia and Ageing: Medical Aspects

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The hemophilia population, particularly in developed countries, is ageing. For example, 2% of patients in the U.S.A. with FVIII and FIX deficiency are over 65 years of age with a further 15% over 45-years-old. There are problems specific to the hemophilia population of chronic arthropathy and long-term effects of transfusion transmitted infection which is compounded by the wider community ails of cancer and vascular disease, concurrent with the ageing process. Risk factors for vascular disease are identical in patients with hemophilia as for the general population, independent of the severity of hemophilia. Hemophilia centres may provide, though not necessarily be resourced for, medical care beyond the usual array of

bleeding disorder specific conditions. Diagnostic, screening, and preventive strategies for age-related conditions need to be identified and delivered. Clinical data to inform treatment are scant, and multicentre observational and treatment studies required. Treatment centres should soon be adding geriatricians to the multidisciplinary team.

A1.5

Hemophilia and Ageing: Addressing Musculoskeletal Problems

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In older men with hemophilia, musculoskeletal (MSK) complications of bleeding contribute to increased morbidity that occurs with age. Hemophilic arthropathy, similar to osteoarthritis, causes pain and structural joint deterioration that often impairs functional ability and impacts physical activity. In addition, persons with hemophilia (PWH) also experience the general MSK complications of ageing such as declining balance, incidence of falls, and osteoporosis. These problems may be intensified in PWH, due to concomitant MSK bleeding issues. A regular fitness program that incorporates aerobic exercise, strength-training, and balance and flexibility activities is important to help improve functional mobility and manage hemophilic arthropathy symptoms, balance and falls risk, osteoporosis, and osteoporotic fractures. Research from the non-bleeding disorders population has shown that exercise is an effective tool to address symptoms of osteoarthritis, reducing the risk of falls and helping to maintain bone density. Because of the special challenges associated with hemophilia, which include both the underlying coagulopathy and, in many cases, extensive MSK involvement, patients beginning an exercise regime should be referred to appropriately trained physiotherapists for evaluation, education and instruction, and follow-up. Although some information from the general population can be applied to PWH, more studies specifically addressing these topics in regards to PWH are needed.

Session A2 - Hemophilia and Cartilage

A2.1

Hemophilia and Cartilage: The Blood Effect

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The inflammatory synovitis that results from hemophilic bleeding into joints has been implicated in the subacute and chronic secondary osteochondral destruction that





Abstracts (cont'd)

characterizes hemarthropathy. Nevertheless, intraarticular blood is directly toxic to cartilage, as iron-catalyzed formation of destructive oxygen metabolites results in chondrocyte apoptosis and loss of the cartilage matrix. Gaps in the mechanistic understanding of the progress of bleeding-induced joint damage leave us unable to answer the key question: Why do some PWH progress rapidly to cartilage loss and subchondral deformation in response to bleeding that is relatively well tolerated in other individuals? Much has been learned from models of blood-induced changes in joint tissues in vitro and in hemostatically normal animals in vivo, including the implication of proinflammatory (e.g. TNF α ; IL-1 β) and anti-inflammatory (e.g. IL-10) cytokines in hemarthropathy. Ultimately, some processes that lead to osteochondral degeneration in human hemophilic arthropathy (e.g. abnormal wound remodelling after bleeding) may only be fully elucidated by in vivo preclinical investigations in hemophilic animals, as a path to human clinical investigation.

Key words: cartilage, hemophilic arthropathy

A2.2 Joint Aspiration, Viscosupplementation, Growth Factors, or Nothing?

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The pathogenetic mechanism of hemophilic arthropathy is not fully understood, however general agreement exists on the fact that the repeated extravasation of blood into the joint cavity is the factor responsible for synovial and cartilage changes. The proper treatment strategy to prevent and/or cure cartilage damage in hemophilic arthropathy is still an open issue. Several experimental studies suggest the potential role of joint aspiration in case of hemarthrosis to prevent the blood-borne damage of the cartilage. Nevertheless, the management of hemarthrosis in hemophilic patients is still controversial; often the conditions to perform a safe arthrocentesis are not fulfilled, and very few hemophilia centres are used to practising such a procedure. Since cartilage and bone damages have very limited ability to heal spontaneously, procedures able to generate cartilage and bone tissue are needed. It has been suggested that intra-articular administration of hyaluronan (viscosupplementation) as growth factors may improve articular cartilage defects. This issue has been poorly investigated so far, however preliminary results are promising. The recent research achievements in this field and the potential clinical applications of new technologies in the treatment of chronic hemophilic arthropathy will be reviewed and discussed.

A2.3 The Role of Movement

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Cartilage is an essential structure for the joint. Blood can destroy cartilage cells and start a vicious cycle that leads to inflammation and more deterioration of cartilage. We observe worsening of joints that never experienced symptomatic bleeds. Cartilage is a bradytrophic tissue that needs pressure and release of pressure for nutrition and regeneration. Movement therefore plays a major role in maintaining and preserving cartilage. In 3D motion analysis we can measure the movement of the joints and can predict the loading of the cartilage. Optimum loading is rhythmic, regular and sinusoid, with no acceleration peaks and a good distribution of the pressure on to the cartilage. Loading should always be perpendicular because of the vertical lining of cartilage cells. Acceleration peaks, sagittal loading like gliding in the knee joint or disturbed pressure distribution will hurt the lamina splendens. There is enough biomechanic proof that pressure on the superior layers is much higher than on the bony endplate. False movements as described above occur routinely in younger children. They have the advantage of a higher regenerative power as cartilage is still in growth. They occur as well if the joints are overloaded by vigorous exercise. The mechanically destroyed cartilage will act just like the chemically destroyed cell, which results in faster deterioration of the joint. **Conclusion:** Early moderate movement of all joints and a good musculature that is well trained concerning strength, flexibility, and coordination will provide vertical loading of cartilage a good distribution of pressure.

A2.4 Surgical Treatment: Is It Useful?

M. Silva

No Abstract Submitted.

Session A3 - Hemophilia and Bone

A3.1 Osteolytic Lesions: Differential Diagnosis and Treatment

H. Caviglia

There are two types of cysts in patients with hemophilia, which have different pathophysiology and clinical presentation. The subchondral cyst is always presented in the presence of arthropathy, in contrast, the intraosseous cyst is secondary to intraosseous bleeding. The first signs of arthropathy on the X-ray are narrowing of the joint space and small abnormalities in the subchondral bone.





Abstracts (cont'd)

The subchondral cyst appears later, and is multiple, and irregularly distributed. The X-ray show these cysts later, however in the CT scan or MRI they are observed early. These cysts are connected and exposed in the articulation accompanied by the disintegration of the subchondral bone. (1-2-3-4). The connection of the cyst with the joint causes increased intraarticular pressure during synovitis and leads to progression of the cyst.

The clinical presentation of the subchondral cyst is not specific and is related to the symptoms of synovitis or arthropathy.

Sometimes the cyst progression results in big osteolytic lesions that may lead to a pathological fracture. The intraosseous cyst are caused by intraosseous bleeding, the locations is in general in the bone metaphysis and are not related with the arthropathy. Most of the time, its location is related to the location of the nutritive artery of the affected bone. Clinical presentation is usually with acute pain in the affected bone, but unlike acute osteomyelitis, patients don't usually present fever. Early diagnosis is made by MRI. Radiological images are done later and are characterized by an osteolytic lesion in the metaphysis of the bone without the presence of arthropathy

Surgery is indicated for subchondral cysts when:

- 1- The size of the cysts is greater than 15% of the area of the joint, especially if these are load articulations. In the knee and in the elbow each medial and lateral compartment is considered separate and should therefore be evaluated separately.
- 2- A control X-ray shows that cyst size has progressed; even if its subchondral extension is smaller than 15%.

The purpose of treatment is to avoid the crumbling of the joint and to reconstruct bone stock when it is necessary to carry out definitive arthroplasty. Intraosseous cysts require surgical treatment when they have not responded to replacement therapy during 6 weeks. In both subchondral and intraosseous cysts, the joint must be studied by computed tomography and a magnetic nuclear resonance MRI, which allow appropriate pre-operative planning.

A3.2 Is Bioengineering the New Solution?

F. Forriol

No Abstract Submitted.

A3.3 Osteoporosis: Clinical Implications

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Osteoporosis compromises the quality of life of persons with hemophilia (PWH) associated with arthropathy. Low

level of physical activity, low BMI (<19), liver dysfunction, concomitant hepatitis C, HIV sero-positivity, systemic steroid therapy, and prolonged immobilisation are some of the common causes leading to low peak bone mass (PBM) in the first 20-25 years of life. This leads to reduced bone marrow density (BMD) subsequently resulting in osteopenia and eventually osteoporosis. Various literature shows that the incidence of osteopenia is up to 43% and that of osteoporosis is 25-27% in PWH. However, the sample size is not adequate enough to show a true picture worldwide. A U.K. consensus group published that 30% of hip fractures and 20% of vertebral fractures occur in men in the general population, though there is no published data showing incidence of osteoporotic fractures in PWH. Dual energy X-ray absorptiometry (DEXA) scan done to measure BMD is the hallmark of diagnosis. It measures the density around the hip, wrist, and spine of the patient compared with age and gender matched healthy individuals. It is correlated with laboratory values of normal serum calcium, phosphorus, and alkaline phosphatase values. High alkaline phosphatase values suggest associated osteomalacia, which can be confirmed with values of serum vit.D3. Use of factor prophylaxis since early childhood may preserve normal BMD in severe hemophilia. Vitamin D supplementation to prevent bone loss and fractures for "at risk" population contemplated. Weight bearing physical activity, physiotherapy, and surgery to remobilise diseased joints, and calcium and vit. D supplements can be recommended for anyone at any age. Treatment with anti-resorptive medication like bisphosphonates may be indicated for patients in whom rapid bone loss of bone mass has been confirmed by sequential BMD measurements, or who have suffered fragility fractures. Surgical treatment is a challenge in presence of osteoporosis. Load sharing implants like intramedullary locking nails are the preferred method compared to load shielding implants for fixation of long bone fractures. Locking compression plates have been helpful to a certain extent. Using sharp drills, avoiding tapping after drilling, use of bone grafts/bone cement are some of the techniques for conventional fixation. Cemented replacement arthroplasty for femoral, neck, and humerus fractures are choice in the older population.

A3.4 Osteoporosis and Exercise

A. Sabbour

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The word osteoporosis comes from the Greek osteo and poros: osteo means bone and poros translates to porous (holey). Even healthy bones have holes in them as part of their intricate honeycomb-like design. Bones affected by osteoporosis, however, have bigger holes, making them weaker and more prone to breaking. Osteoporosis is common, occurs throughout the world, and has become a major public health concern. Adequate bone mass





Abstracts (cont'd)

accumulation in early life is important in preventing osteoporosis. Lifetime plots of bone density and age show that late childhood and adolescence are important periods of bone mineral acquisition. Persons with the greatest bone mass at the end of adolescence have the greatest protection against the gradual decline in bone mass that occurs with aging. Physical activity has been considered an important factor for bone density and as a factor facilitating prevention of osteoporosis. Bone density has been reported to be reduced with hemophilia. The main cause for reduced bone mass in the hemophiliac is most probably due to hemophilic arthropathy, typically associated with chronic pain and loss of joint function subsequently leading to inactivity. Evidence shows that exercise may help build and maintain bone density at any age. Studies have seen bone density increase by doing regular resistance exercises, such as lifting weights, two or three times a week. This type of weight-bearing exercise appears to stimulate bone formation and the retention of calcium in the bones that are bearing the load. The force of muscles pulling against bones stimulates this bone-building process. Therefore, any exercise that places force on a bone will strengthen that bone. Weight-bearing exercises are the most effective to build bones. These include activities such as walking, stair climbing, running, hiking, and weight lifting. However, swimming and bicycling are not considered weight-bearing exercises. Key words: osteoporosis, weight-bearing exercises and hemophilia.

Session A4 – Free Papers: Non-Surgical

A4.1

Assessment of Musculoskeletal Function and Mood in Hemophilia A Adolescents: A Cross-Sectional Study

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Introduction: Hemophilia A is characterized by the occurrence of frequent spontaneous intra-articular and intramuscular bleeding. If inadequately treated, it results in progressive damage to joints and muscles, leading to crippling deformities and musculoskeletal dysfunction. These complications result in lifelong chronic pain and disability that may greatly affect the patients' mood. We aimed to evaluate the musculoskeletal function in our hemophilia A patients and its correlation to depressed mood in these patients, and determine the impact of degree of factor VIII deficiency, different replacement therapy regimens, and frequency of hemarthrosis on both musculoskeletal function and mood. **Methods:** A cross-sectional study was carried out on 50 adolescent

hemophilia A patients. Musculoskeletal function was assessed using Functional Independence Score for Hemophilia (FISH), and mood status was assessed using Beck Depression Inventory—Short Form (BDI-SF). **Results:** The mean FISH score was 23.32 ± 4.69 (range 13-28) and the tasks that obtained the lowest score were step climbing, squatting, and walking. Of our 50 patients, 16 (32%) were not depressed, 18 (36%) had mild depression, 11 (22%) had moderate depression, and 5 (10%) had severe depression. There was highly significant negative correlation between mean FISH score and mean BDI-SF score ($P < 0.001$). **Conclusion:** The better the replacement therapy regimen, the better the musculoskeletal function that could be obtained in hemophilia A patients and the better their mood.

Key Words: Hemophilia A, musculoskeletal, FISH, adolescents

A4.2

Measuring Balance in Individuals with Hemophilia Using the Community Balance and Mobility Scale: A Pilot Study

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Introduction: This study aimed to explore the scoring patterns of individuals with mild, moderate, and severe hemophilia A and B using the Community Balance and Mobility Scale (CB&M). This study also aimed to investigate any relationship between Hemophilia Joint Health Score (HJHS) and CB&M score, as well as to investigate any relationship between Body Mass Index (BMI) and CB&M score. **Methods:** This pilot study used a single-centered, cross-sectional study design. The CB&M and HJHS were administered to 15 participants. Correlation analysis, t-test, and a descriptive analysis were performed. A subgroup analysis was conducted on individuals with severe hemophilia A and B. **Results:** No significant differences were found in CB&M scores between individuals with mild and severe hemophilia A and B. A significant correlation was observed between CB&M scores and HJHS ($r = -.805$, $p < 0.001$), age, and total CB&M scores ($r = .765$, $p = 0.045$). There was no significant correlation between BMI and total CB&M scores. Specific item CB&M scoring pattern was generally comparable to normative values in the sample population. **Conclusion:** As the hemophilia population ages, balance and falls are becoming issues of primary importance. This pilot study suggests that individuals with hemophilia show decreased scores on the CB&M compared to the healthy population, regardless of hemophilia severity. It is the first study to investigate the relationship between dynamic functional balance performance and joint health. It is also the first study to





Abstracts (cont'd)

suggest a single validated measure, the CB&M, as a useful clinical tool to measure balance in the hemophilia population.

A4.3

Assessment of Musculoskeletal Status of a Lebanese Hemophilia Patient

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Introduction: It is important that adequate data on outcomes be documented to develop an optimal protocol for management. In hemophilia care, musculoskeletal outcome remains the major hallmark. The purpose of the study was to assess joint status of Lebanese hemophilia patient (PWH). **Methods:** We conducted a cross-sectional study at the Lebanese Hemophilia Association care centre. 65 severe PWH out of the 159 registered were assessed. Patients were subdivided into 5 groups according to actual age. Group A patients were less than 5-years-old (n=8); group B, 5 to 10-years-old (n=9); group C, 10 to 20-years-old (n=12); group D, 20 to 40-years-old (n=25); and group E over 40 years of age (n=11). Musculoskeletal status was measured using the Hemophilia Joint Health Score (HJHS) and the Functional Independence Score for Hemophilia (FISH). **Results:** Mean age was 23 years. The means HJHS and FISH scores were 25.57 and 23 respectively. Mean gait score was 2.34. The most affected joints were knee, followed by elbows and ankles. Locomotion then transfers and self care were affected. Mean HJHS and FISH score according to age group were 2 and 33 in group A, 8.77 and 32 in group B, 19.08 and 30.33 in group C, 35.36 and 25.48 in group D, and 43.09 and 23.09 in group E, respectively. Scores were significantly more impaired in older patients. **Conclusion:** According to our result new strategies of care must be undertaken. More data must be collected then used for lobbying to enhance standards of care with the health authorities.

A4.4

Guideline for the Treatment of Chronic Synovitis in Pediatric Hemophilia: Case Report Involving Synovitis of the Ankle Joint in Two Boys with Hemophilia A

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Objective: Review and reflection on the use of developed guidelines in the treatment of chronic synovitis in boy A & boy S. **Methods:** Joint review by physiotherapist identified acute bleed, with subsequent chronic synovitis, in boy A (right ankle), and clinical evidence of bilateral

ankle chronic synovitis in boy S. Both individuals were treated according to developed guidelines including the use of: daily factor; P.R.I.C.E (compression being very limited); splint immobilisation and reduced activity; a 6-week course of Ibuprofen 10mg/kg with covering Lansoprazole; intra-articular steroid injection; regular rehabilitative physiotherapy; use of combined podiatry clinic in boy S; baseline MRI imaging. **Results:** Both boys appeared to respond favourably to the used guidelines and showed a reduction, boy S, in clinical signs of chronic synovitis, and complete resolution in boy A. **Conclusion:** Chronic synovitis may be addressed successfully with combination therapy and the support of an extended team including pediatric rheumatology. Follow-up joint reviews continue to be conducted to describe the potential longevity of treatment. Ideally repeat MRI's may provide more objective data.

Key words: chronic synovitis, hemophilia A

A4.5

Aerobic Capacity in Egyptian Adults with Hemophilia

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Background and objectives of the study: It is well known that physical activity is very important for hemophilic patients and can contribute to better quality of life. Physical training also reduces the risk of bleeding episodes and prepares the body to manage hemophilia better. In spite of these benefits, participation in physical activity for children with hemophilia is discouraged in Egypt. The purpose of this study was to investigate the feasibility of maximum cardiopulmonary exercise test (CPX) to evaluate aerobic capacity of hemophilic patients, also to determine the maximum oxygen consumption (VO_{2max}) and total work load (TWL) in adult Egyptian patients with hemophilia A. **Study design:** VO_{2max} and TWL obtained during CPX test (breath by breath) from 30 subjects with hemophilia A (16 moderate and 14 severe); their mean age was 17.6 ± 1.92 years; were compared with normal matched for anthropometric data and age. **Result:** All subjects were able to perform at maximal or near maximal level on exercise tests and none





Abstracts (cont'd)

of them reported bleeding or other adverse events. The aerobic capacity indices; the VO_{2max} and TWL were less for hemophilic subjects by 56.5% and 32.5% respectively than normal peers. There was no significant difference between moderate and severe hemophilic subjects. **Conclusion:** It was concluded that the maximum exercise test was safe and useful for hemophilic subjects. Egyptian adults with bleeding disorders have impaired aerobic capacity compared with their peers. This may be attributed to lack of appropriate treatment, patient's overprotection and socioeconomic factors. The severity of the bleeding disorder did not contribute to aerobic capacity deficiency. Key words: aerobic capacity, hemophilia

A4.6 Development of a Chatterbot Specialist in Hemophilia Care

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Introduction: In the past several years, Information Technology has supported medical areas. The purpose of this project is to present a Chatterbot specialist in hemophilia. The prefix 'chatter' means to establish an informal conversation, while the suffix 'bot' comes from the word robot. Thus, this computer program aims to set up a chat with the user about any particular issue. Its development is a result of an integration of Artificial Intelligence and hemophilia care. **Methods:** The development of Chatterbot used the Java programming language to implement its web interface and the Prolog artificial programming language to handle its knowledge base. The Chatterbot uses Portuguese as its default language, but other languages will be supported in the future. Its knowledge base was constructed based on hemophilia courseware, with the help of hematologists and physiotherapists, and can be improved on demand. **Results:** The goal of this computer program is to teach children how to deal with hemophilia issues. Children can interact by asking questions and getting answers through text, images, and videos. Chatterbot is available on the Internet and has been evaluated by specialists in hemophilia care. **Conclusion:** It is challenging for children to deal with hemophilia. They have to know which activities they can perform and how to proceed if something unexpected happens. Chatterbot was developed to support children to understand their restrictions while having fun. It shows how multidisciplinary areas can help children with hemophilia learn about hemophilia by using a computer program.

A4.7

Physiotherapy Care Profile in Brazil: An Evaluation of 18 Hemophilia Treatment Centres

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In 2009 the Brazilian Federation of Hemophilia nominated a group comprising physiotherapists (PT) from all geographic regions of Brazil. The main goals of the Brazilian MSK Group are to discuss MSK manifestations in hemophilia, and develop trainings. In order to obtain a profile of the physiotherapy hemophilia care among different centres, the group sent an e-mail to 85 PTs with a questionnaire covering demand for treatment, availability of auxiliary resources, multidisciplinary interaction, and patient adherence to the programs. We obtained complete answers from 18 PTs (21.1 %), 44.4% of these dedicated exclusively to hemophilia care, and most attending patients referred by hematologists (83.3%). PTs who replied use at least 2 auxiliary equipments for electrotherapy. All maintain exercises programs as well as cryotherapy (94.4%) and some kind of hydrotherapy (66.6%). The preferred modalities of treatment are exercises programs (100%), therapeutic ultrasound (88.8%), and cryotherapy (83.3%). Patients have 3 appointments per week (77.7%), averaging 66 minutes per session, with factor infusion prior to physiotherapy (50.0%). Public transportation is used by 72.2% of the patients attending physiotherapy. When intensive program is needed, 77.7% can count on housing. Except for one centre, all have a multidisciplinary team. One of the greatest obstacles to adhesion to treatment is transportation. Only 3 centres (16.6%) regularly provide exercise programs for home and only 2 (11.1%) provide written electronic guidance to other PTs. We concluded that more efforts should be done in order to guarantee physiotherapy care focusing on the patient needs and education, since the infrastructure seems adequate.





Abstracts (cont'd)

A4.8

Hemophilic Arthropathy: Can Non-Hematological Factors Affect the Burden of Disease?

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There is an increasing awareness that non-hematological factors may influence the presentation of hemophilia and therefore the potential to develop hemophilic arthropathy. Clinical observation seems to suggest a group of factors that may have the ability to influence the likelihood of a person to bleed more readily than others. These include hypermobility, proprioceptive acuity, and biomechanical factors. Prophylaxis remains the standard of care, however recently evidence has emerged that would appear to indicate that some boys cope very well on an enhanced on-demand regimen. Although there are some potential hematological explanations for the differences between people who do and don't bleed with severe hemophilia, such as maintenance of trough levels, there is also the potential that other factors are at work. All presentations of Ehlers-Danlos Syndrome include tissue hypermobility and easy bruising in their symptom spectrum. Proprioception and muscle weakness are also features. When considered alongside hemophilia it presents a paradigm that may explain why some persons with hemophilia persist in bleeding in the presence of adequate prophylaxis. Biomechanics of the foot and ankle are also regularly discussed within the hemophilia literature where a presumption exists that non-ideal biomechanics and bleeds have a cause and effect relationship, however the author cannot identify robust evidence to wholly support this and would at least partly challenge this view.

This presentation will discuss the potential influence of these factors and an upcoming program of work that will begin investigating their influence.

Key words: Hemophilia, Hypermobility Etiological Factor, Proprioception

A4.9

The Mild Hemophilia Experience: Attitudes, Behaviours, Communication with the Health Care Team and Determining when Treatment is Needed

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The Canadian Physiotherapists in Hemophilia Care (CPHC) conducted qualitative research with young adults with mild hemophilia. **Objectives:** To discuss how these men make decisions about their injuries and their ability to recognize an injury needing medical attention, to explore the attitudes towards physical activity, to identify effective communication strategies. **Method:** 18 semi-structured interviews using grounded theory were performed, in person or phone, with men aged 18-30-years-old with mild hemophilia from across Canada. **Results:** Many themes emerged from these interviews including the importance of previous experience with injuries, the reluctance to access care, and participation in vigorous physical activity. **Conclusion:** The interviews demonstrated that communication between these young men and the healthcare team was not optimal. Gaps in knowledge also emerged regarding bleed identification and management. Young men with mild hemophilia feel that they are "normal", so partake in aggressive sports. **Contribution to the practice:** This qualitative research helped develop a tool to assist young men with mild hemophilia assess injuries and to determine when factor replacement is necessary. Recommendations regarding sports and physical activity with this population should be re-examined. It also led to the development of a set of questions suggested for use by the healthcare team. It is hoped that enhanced communication will lead to more effective interactions between the clinic team and these young men.

Key words: Mild Hemophilia, Self-management

A4.10

Proprioceptive Training in Hemophilic Children

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Efficient movement function and the maintenance of balance during dynamic tasks are more complex than only force production; it requires a primary sensory mechanism for motor control which is proprioception. The objective of this work is to study alterations in proprioceptive performance to subsequently evaluate the appropriate therapeutics. **The purpose of the study:** To determine whether exercise intervention improves balance in children with hemophilia following participation in a balance and proprioceptive training program using the Biodex Stability System. Thirty hemophilic boys aged between 7 and 14 years-old with hemophilia (type A & B) participated in this study. They were classified randomly into two groups of equal number (control and study). The control group received factor replacement as prophylactic medical management. The study group received the proprioceptive training program and factor replacement. Proprioception parameters were assessed using the Biodex Stability System in both groups with eye opened and with eye closed, before and after three months of the





Abstracts (cont'd)

application of the treatment program. The results of this study revealed statistically highly significant improvement in nearly all of the measuring variables of the study group ($P < 0.01$) when comparing its pre- and post-treatment results, and when comparing the post-treatment results of the control group. From the obtained results of this study, it can be concluded that proprioceptive training is a beneficial modality that may be used to improve standing postural control with minimal stress to the joint in hemophilic children.

Key words: hemophilia, hemarthrosis, proprioception

A4.11

Clinical and Research Uses of the CDC Normal Range of Motion (ROM) Public Use Dataset

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Objective: To describe the potential clinical and research uses of a public use dataset of normal ROM measurements taken on 674 healthy normal male and female subjects aged 2 – 69 years. **Methods:** ROM life-span charts for each joint by gender can be made using the 95% confidence intervals and population percentiles of the mean values for ROM obtained from healthy subjects in four age groups. One possible research use of the normal ROM dataset would be to combine these data with bilateral measures of ROM taken on the shoulders, elbows, hips, knees and ankles at the most recent clinic visit of 4,655 patients with severe hemophilia participating in a public health surveillance system in the U.S. to generate summary measures of the proportionate reduction from normal ROM. **Results:** In the clinical setting a normal ROM life-span chart could be used to plot a patient's ROM. This would provide a visual tool to help a patient understand how his joints compare to normal joints for his age group. In a research setting the average overall summary measure of proportionate normal ROM can be plotted for hemophilia patients over the life span. The plots can be used to compare joint outcomes among hemophilia severity groups. **Relevance to hemophilia care:** Plotting ROM measurements obtained during clinic visits on a life-span chart can help monitor disease progression. The public use dataset can be used by researchers in comparison studies and to study patterns of ROM changes in patient populations with chronic joint disease.

A4.12

Postural Adjustment after an Unexpected Perturbation in Children with Hemophilia

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Introduction: The ability of individuals to recover from a sudden loss of balance is essential for preventing falls and subsequent injuries. Balance recovery depends on the selection of a proper postural response to maintain the centre of pressure (COP) in the limits of the base of support. Patients with hemophilia often bleed inside the joints and muscles, which could lead to poor sensorial information about the position of the body segment. The purpose of the study was to evaluate postural adjustment after a sudden perturbation in children with hemophilia compared to a control group. **Methods:** 11 children with hemophilia (10.3 ± 1.3 years-old) and 11 able-bodied children (9.7 ± 1.1 years-old) comprised the hemophilia group and control group. A controlled external posterior perturbation (EPP) was applied by means of horizontal traction to the subject's dorsum, using a fixed pulley system. The EPP was unexpectedly removed, producing postural adjustment. COP displacement in the anterior-posterior direction was obtained by means of a force plate. Postural responses were compared between groups using unpaired student t test in eight intervals of 1 second (t1 to t8), beginning at the moment of EEP removal. **Results:** COP displacement was higher in the hemophilia group in t2 ($p=0.02$), t6 ($p=0.02$), and t7 ($p=0.04$) intervals. These results indicate that children with hemophilia have a poorer ability to control their posture under a sudden loss of balance. **Conclusion:** Children with hemophilia use greater centre of pressure displacement to control balance after an unexpected perturbation when compared to a control group.

A4.13

Early-Onset Administration of IL-4 and IL-10 Prevents Blood-Induced Cartilage Damage in Vitro

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Introduction: It was reported that interleukin (IL)-10 limits blood-induced cartilage damage in vitro. In addition, IL-4 has cartilage protective properties. Hence our aim was to study whether IL-4 in addition to IL-10 can prevent blood-induced cartilage damage when administered during (prevention) and after the onset (treatment) of blood-exposure. **Methods:** Human articular cartilage explants were cultured for 4 days in presence or absence of 50% v/v homologous blood (mimicking a joint bleed). IL-4, IL-10, or a combination of these two cytokines was added during blood exposure, and 24 or 48 hours after start of blood-exposure. After 4 days the medium was refreshed and cartilage was cultured for an additional 12 days to determine cartilage matrix turnover. **Results:** Cartilage exposed to blood showed a decrease of





Abstracts (cont'd)

proteoglycan synthesis rate, an increase of -release, and a decrease of -content (all $p < 0.05$). This blood-induced damage to the cartilage matrix was limited both by IL-10 and IL-4 alone. Proteoglycan synthesis rate improved by 125% and 512% respectively, release decreased by 34% and 50%, and content increased by 23% and 22% (all $p < 0.05$). Moreover, the combination of IL-4 and IL-10 was clearly more protective than IL-10 alone. Addition of IL-4 and IL-10 after the onset of blood-exposure did not prevent cartilage damage anymore unless blood was first removed. **Conclusion:** The combination of IL-4 and IL-10 is most protective when added within 24 hours after start of blood-exposure. Therefore treatment should be started as soon as possible and duration of blood-exposure should be limited to prevent hemophilic arthropathy.

Session B1 - Synovitis

B1.1 Pathogenesis

L. Valentino

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Synovitis and arthropathy are frequent and serious complications of repeated joint bleeding in patients with hemophilia, resulting in pain, deformity, disability, and reduced quality of life. While the pathogenesis of hemophilic arthropathy has not been fully elucidated, it appears to have similarities with the degenerative joint damage that occurs in osteoarthritis and the inflammatory processes associated with rheumatoid arthritis. This presentation will review the potential actions of various blood constituents on joint components that culminate in the development of hemophilic synovitis and arthropathy.

B1.2 Conservative Management of Hemophilic Synovitis

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Introduction: Radiosynovectomy (RS) is an efficient procedure for the conservative management of chronic hemophilic synovitis. However, quantification of its efficacy has not been fully investigated in the literature. We have analyzed ten articular parameters to try and measure the efficacy of RS. **Materials and Methods:** In a group of 78 persons with hemophilia we performed 156 RSs in 104 joints, under the diagnosis of hemophilic synovitis. The mean age of patients was 18-years-old (range: 7-51). Fifty-eight patients required RS in a single joint while 20 patients required RS in more than one joint. Of the 104 joints treated 33 were elbows, 24 ankles, and 47 knees. We used Yttrium-90 for the knees at a dose of

185 MBq, Rhenium-186 for the elbows (56-74 MBq) and the ankles (74 MBq). The procedure required 1 to 3 injections of the appropriate radioactive material (RS-1, RS-2, RS-3) with 3 to 6-month intervals. Of the 104 joints, 68 required RS-1, 20 RS-1 plus RS-2, and 16 RS-1, RS-2, and RS-3. Thus, the total number of RSs was 156. In eight cases (7.6%) surgical treatment (arthroscopic synovectomy) was eventually required after the failure of RS. We studied ten articular parameters before RS and 6 months after RS to assess its efficacy: number of hemarthroses, articular pain (VAS), range of motion -ROM- (in flexion and in extension), muscular power (in flexion and in extension), degree of clinical synovitis, size of the synovium (mm) measured by image (ultrasonography and/or MRI), WFH clinical score and radiological score. **Results:** RS significantly improved all the parameters except for the WFH's radiological score. Moreover, such parameters improved in an independent way for each of the RSs (RS-1, RS-2, RS-3). On average, the number of hemarthroses and the degree of pain improved by 70%. Synovial size improved by 35%. The WFH's clinical score improved by 19%. Muscular power improved by 12%, while ROM improved by 3%. We also observed that the efficacy of RS was not related to patient's age, type and grade of hemophilia, previous hematological treatment (on demand or prophylaxis), presence or absence of inhibitor, synovial size, type of joint (elbow, knee, ankle), previous physical activity, presence or absence of joint degeneration (arthropathy), radioactive material used, and presence or absence of complications after RS. We also found that after RS-1 the knee had 3.4 and 3.2 times more risk of not improving pain than elbows and ankles, respectively. Regarding ROM after RS-1, severe hemophilia has 2.1 times more risk of lack of improvement of ROM than moderate and mild hemophilia. Moreover, the ankle has 6 times more risk of not improving ROM than elbows and knees. **Conclusions:** We have confirmed that RS is an efficient procedure for the conservative management of chronic hemophilic synovitis, although it may require 1 to 3 injections (RS-1, RS-2, RS-3) with 3 to 6-month intervals. On the other hand, studying ten articular parameters before and 6 months after RS, we have been able to measure the efficacy of RS and also define some prognostic factors.

B1.3 Role of the Physiotherapist in Prevention and Treatment of Synovitis

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Hemophilic synovitis (HS) - an inflammation of the synovial membrane - is directly and indirectly related to the effects





Abstracts (cont'd)

and consequences of hemarthrosis. Research over the past 50 years has progressively offered insight and better options for the management of joint bleeds and chronic HS. Nevertheless, the long-term consequences of joint bleeding and chronic synovitis in people with hemophilia still remain a challenge for members of the comprehensive care team, both in the developed and developing world. Prophylaxis to help prevent or reduce joint bleeds is presently considered to be more effective than on-demand treatment following a joint bleed. Similarly, a preventative approach with respect to physiotherapy may also be more effective in decreasing joint bleed frequency and directly-related complications such as HS and joint degeneration. Specialists such as hematologists, orthopedists, rheumatologists, researchers, as well as other members of the comprehensive care team, such as physiotherapists, nurses, and psychologists each have an important role in the management of HS. This presentation will look at some of the common approaches presently used in physiotherapy treatment of HS, attempt to put them in perspective, as well as offer prevention rationales based on an amalgamation of the current evidence.

B1.4

Surgical Options, Including Angiographic Embolization

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Surgical synovectomy, whether open or arthroscopic, requires extensive resources from an experienced team, a dedicated hemophilia treatment centre, and a large supply of clotting factor. Surgical synovectomy is seldom necessary today and is only considered when other less invasive and equally effective procedures fail. The population with synovitis refractory to non surgical treatment constitutes a challenging subgroup to treat. Identifying the reasons that lead to failure of previous, less invasive approaches is key to achieve success. The explanation for the failure is most probably multi-factorial, and may include undetected inhibitors, early bleeder phenotypes, association with other coagulopathies, intra-articular and peri-articular mechanical problems. Examples of the latter are torn meniscus, intra-articular loose bodies, and quadricipital palsy secondary to previous injury of the femoral nerve during a psoas-iliacus muscular hemorrhage.

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, however, 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 general consensus that it is best used as a second level of defence after radiosynoviorthesis. 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 remodelling of meniscal tears, which are characteristic of grade III and IV arthropathy.

Angiographic embolization of peri-articular arteries is a promising percutaneous procedure. Published case series suggest this intervention modifies the natural course of chronic synovitis in hemophilia. This accomplishment is materialized in a decrease in bleeding frequency and in volume of the synovial tissue. In the absence of comparative data it is early to conclude on the role this therapeutic modality may play in the armamentarium for the treatment of chronic synovitis. Given the intra-arterial nature of the procedure, generating data on safety is paramount before recommending for widespread use.

Session B2 – Inhibitor Patients: The Truth

B2.1

Open Issues: Prophylaxis and ITI

E. Santagostino

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Thanks to the progress achieved with by-passing therapies, elective orthopedic surgery has become feasible in hemophiliacs with inhibitors; however only centres already experienced can tackle this challenge. A strict collaboration amongst surgeons, hematologists and physiotherapists is essential for a successful long-term outcome. The choice of the therapeutic strategy is based on the current inhibitor titre and the information on historical peak titres. Bypassing agents, activated





Abstracts (cont'd)

prothrombin complex concentrate (aPCC), and recombinant activated factor VII (rFVIIa), represent the first choice in patients with high-responding inhibitors. No bypassing product can guarantee sustained hemostasis and no laboratory assay is available to predict the efficacy of treatment. Caution is recommended administering bypassing agents in elderly patients or in those with thrombotic risk factors. High-dose factor replacement may neutralize low-titre inhibitors, allowing achievement and maintenance of the desired factor levels to cover surgery. A preliminary pharmacokinetic study is useful in these cases to establish the optimal dose regimen. Factor replacement therapy represents the first-choice approach for patients with low-responding inhibitors and it could also be considered in high-responders who have a temporarily low inhibitor titre. Laboratory monitoring is crucial for dose adjustment and factor recovery should be measured at least daily until discharge. The cornerstone of post-operative management is to avoid bleeding complications and if bleeding occurs it must be treated without delay. Early mobilization should be individually tailored and started with caution; a single concentrate infusion may be given before the physiotherapy session in order to reduce the bleeding risk.

B2.2

Before Surgery: What the Surgeon Wants to Know

P.L. Solimeno

Ortho-Trauma Unit, IRCCS Cà Granda Foundation, Policlinico Hospital, Milan, Italy

The management of recurrent hemarthroses, chronic synovitis, and chronic arthropathy is more challenging in patients who developed an inhibitor against FVIII/FIX, the most common and most serious complication of replacement therapy in patients with hemophilia A or B. In such patients, although the frequency and severity of hemarthroses is similar to non-inhibitor patients, we registered a severe degree of arthropathy related to minor treatment response. The introduction of bypassing agents as rFVIIa has improved the management of recurrent bleeding episodes and synovitis in hemophilic patients with inhibitors, and today it is possible to perform surgeries in inhibitor patients, even if we have a higher complication rate in comparison with non-inhibitor patients. The author describes the pre-op management from a hematological and orthopedic point of view to avoid risk of complications.

B2.3

Before Surgery: A Hematologist's View

P. Giangrande

Oxford Haemophilia & Thrombosis Centre, U.K.

The life expectancy of people with hemophilia in affluent countries is now very similar to that of the general population. This good news is counterbalanced by the fact

that patients with inhibitors often have a significant burden of disability with inevitable adverse impact of their quality of life. Elective orthopedic surgery can certainly be undertaken with good results nowadays, although this requires careful planning and should only be undertaken in centres with the requisite experience. Full evaluation of hemostasis, including platelet function tests, should be carried out prior to surgery. Patients with liver disease or HIV infection may also require treatment with fresh frozen plasma and/or platelet concentrates. Fibrin sealants can help to reduce blood loss during surgery and tranexamic acid should be used in conjunction with NovoSeven. Laboratory monitoring of peri-operative therapy is generally not needed but a number of groups are seeking to apply thromboelastography and thrombin generation testing. Both FEIBA and recombinant activated factor VIIIa (NovoSeven) have been widely used to cover elective surgery, with good success. The cost of both blood products is very high but may be recovered in subsequent years as a consequence of reduced bleeding frequency. More conservative options may need to be considered as alternatives to surgery. These include treatment with COX-2 anti-inflammatory agents such as Rofecoxib, radionuclide or chemical synovectomy, and embolization.

B2.4

Inhibitor Patients: The Rehabilitation Program, Is It Different?

A. Forsyth

Penn Hemophilia and Thrombosis Program, Thorofare, NJ, U.S.A.

Even with advances in preventing and treating bleeding episodes, arthropathy continues as a common comorbidity in persons with hemophilia (PWH). Arthropathy may result in the need for elective orthopedic surgery (EOS) to address pain and functional limitations. In PWH, EOS is common in countries where it is readily available, and with bypassing agents and accruing experience in hemophilia treatment centres (HTCs), EOS is now being carried out more in PWH with inhibitors. The physiotherapist is an integral member of the comprehensive, multidisciplinary team. For optimal functional outcome, physiotherapy is critical leading up to EOS in PWH with inhibitors, and during post-operative recovery. Pre-operative goals include preparation of the PWH for the rehabilitation program and assuring realistic expectations of the specific EOS. While addressing the muscles and joints adjacent to the operative site, as well as the affected joint, the experienced physiotherapist may be able to judge the PWH's level of participation. This may in turn help predict adherence to prescribed treatments after surgery, a factor that will have a substantial impact on recovery. During post-operative rehab, the physiotherapist caring for the PWH with inhibitors must negotiate a fine balance between intervention and minimizing the risk of bleeding into the operative joint or at other sites. Bleeding





Abstracts (cont'd)

into the operative joint will not only hinder recovery but may also set the stage for infection, which may lead to further complications, including subsequent need for surgery. The rehab program in a PWH with inhibitors has unique challenges above and beyond rehab of PWH without inhibitors, which will be highlighted.

B2.5 **Before Surgery: Are the Results of Surgery Different?**

N. Goddard

Royal Free Hospital, London, U.K.

The availability of by-passing agents (rFVIIa or FEIBA) for hemostatic cover in patients with inhibitors has permitted major surgery that was previously considered to be impossible. Globally, experience of performing orthopedic surgery in patients with inhibitors is increasing. The surgery being performed has become more enterprising and is certainly not for the faint-hearted. Thrombotic complications are rare in inhibitor patients and surgeons frequently ignore the concept of thromboprophylaxis. However, bleeding complications, problems with wound healing and resulting increased infection rates remain a concern. Despite our best endeavours it has not been possible to establish a worldwide register of patients undergoing such surgery and we are still somewhat in the dark as to the ultimate outcomes. There are still only a limited number of publications in this field, often single case reports, small series or pooled data from multiple centres with no standardised treatment regimes. Current published guidelines are seemingly based on a "best guess" scenario rather than true "best practice" and a reliable evidence base. It is probable that there is a bias in the literature towards reporting positive outcomes and consequently failures of treatment are under reported. It is vital to obtain this information lest we repeat the errors of others since only with appropriate data can we make suitable recommendations for peri-operative management. As far as I can tell the published results for major surgery (TKR or THR) in patients with inhibitors have been good. However there is sufficient anecdotal evidence to suggest that there are still serious concerns about the incidence of post-operative infection and premature failures. The increased risk, coupled with the vastly increased expense of hemostatic cover has led some to question the wisdom of performing surgery in these patients. Clearly there are some fundamental questions of health economics, but in addition we should provide guidance as to which patients are, or are not suitable for surgical intervention and to provide robust recommendations for peri-operative hemostatic treatment regimes.

Session B3 - Musculoskeletal Issues in Patients with VWD and RBD

B3.1 **The Hematologist**

D. Quon

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The Hematologist's Perspective on Musculoskeletal Issues in Patients with VWD and RBD.

Von Willebrand disease (VWD) is the most common inherited bleeding disorder, occurring in approximately 1% of the general population. It is caused by a qualitative or quantitative deficiency in von Willebrand factor (VWF) which results in an increased risk of bleeding. In contrast, rare bleeding disorders (RBD) affect less than 5% of all patients with bleeding disorders (hemophilia and VWD combined constitutes the remaining 95%). The clinical manifestations of VWD may vary substantially from patient to patient with the majority of the patients experiencing mucocutaneous hemorrhage. Despite the prevalence of VWD, there is a paucity of published information regarding musculoskeletal or orthopedic complications in these patients, and even less is known in the cases of RBD. The majority of the information that is available is extrapolated from hemophilic arthropathy as the disease shares many similarities with other coagulopathies. Many studies have "lumped" taken cases of VWD and RBD into the cohorts of hemophilic patients. The current status of these issues will be discussed.

B3.2 **The Orthopedic Surgeon**

O.S. Perfetto

Sant'Ambrogio Clinic, Milan, Italy

Von Willebrand disease (VWD) is the most common hereditary bleeding disorder affecting both males and females, with an estimated prevalence of 1.3% in the general population. Clinical manifestations of the diseases primarily consist of excessive and prolonged mucocutaneous and post-operative bleeding that result from quantitative or qualitative defects in the adhesive glycoprotein von Willebrand factor (VWF). Rare bleeding disorders (RBDs) represent 3 to 5% of all the inherited coagulation deficiencies and include the inherited deficiencies of fibrinogen factor (F) II, FV, FV + FVIII, FVII, FX, FXI, and FXIII, and are usually transmitted as autosomal recessive traits, meaning that women represent about half of the affected patients. The authors described their surgical experience (surgical indications, peri-operative management and complications) in the last years in these patients: shoulder replacement, knee replacement, hip replacement, and shoulder and knee arthroscopy.





Abstracts (cont'd)

B3.3

The Physiotherapist

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This presentation will focus on the musculoskeletal issues related to von Willebrand disease (VWD) and rare bleeding disorders (RBD), and how they compare to those most commonly experienced with hemophilia. Key differences between the coagulopathies themselves as well as the types of patients that they affect present the physiotherapist with challenges not usually encountered when treating hemophilia. Some of the issues faced are due to the pathology of the disorders themselves, while others relate to the medical treatment that is provided. The presentation will address how the impact of gender, longer factor half lives, tissues targeted by the various bleeding disorders, and whether their impact poses a direct or an indirect threat to the MSK system. This presentation will assist the physiotherapist to determine the types of issues that will need to be addressed with VWD and RBD, and therefore in selecting the treatment techniques that are most appropriate.

Session B4 – Free Papers: Surgical

B4.1

Flexion Deformity in Knee: Femoral Extension Osteotomy in Pediatric Patients with Hemophilia

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Objective: Display experience with the use of femoral osteotomy in inverted "V" for the correction of knee flexion deformity. Is a very stable metaphyseal osteotomy that provides a wide contact surface and easy fixation with cross Steinman wires. **Material and Methods:** A total of 7 patients with knee flexion contractures were treated between 2006 and 2009, they had severe hemophilia A. Average age was 11 years (8 to 15 years). No response to previous physiotherapy treatment. Showed flexion contracture of the knee between 40° and 70° (3 cases: 40°, 2 cases: 50°, 1 case 60°). All had pain in active-passive mobility and gait. The average number of hemarthrosis was 13.4 times per year (1.11 times per month). The 7 cases

(4 right and 3 left) were treated surgically with inverted "V" distal femoral osteotomy, the internal fixation was with Steinman wire and external immobilization with long cast. **Results:** Bone healing was achieved at an average of 9.1 weeks (8-10 weeks). The follow-up was 2.8 years (maximum 4 and minimum 1). Postoperatively, patients were sent to physiotherapy for muscle strengthening and gait training. Extension was achieved in 100% of cases. The average active motion was 60°. No patient had pain in the mobility and gait. The average number of hemarthrosis was 3.1 times per year. **Conclusions:** The inverted "V" osteotomy is very stable, easy to perform and has low risk, requires minimal internal fixation that is removed at the same time as the external immobilization and is useful in countries with few resources because requirements are minimal.

Key Words: contracture, femoral osteotomy

B4.2

Cysts in Patients with Hemophilia: Bone Restoration by Means of Hydroxyapatite Coralline

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Introduction: Subchondral bone cysts are frequent in hemophilic patients and progress to articular damage.

Objective: The aim of this study is to show the restoration of the bone architecture filling with hydroxyapatite coralline in hemophilic patients with subchondral cysts.

Materials and Methods: 31 male patients with 39 lesions were treated. 29 patients suffer from hemophilia A and 2 from hemophilia B. 2 patients had inhibitors and 1 was a low responder. The mean age was 23 years (6-47). The mean follow-up was 7 (1-16). The most frequent location of the cysts was tibia (19), talus (7), ulna (5), humerus (3), femur (2), and radius (1). Surgery was indicated when the size of the subchondral cyst is greater than 15% of the area of the joint, particularly if it includes load articulations or when the control x-ray shows the progression of the cyst size, even if its subchondral extension is less than 15%. Every cyst was evaluated with X-ray, CT scan and MRI before surgery. All patients were treated in the same way, guided with image intensifier, cyst aspiration, and filled with hydroxyapatite coralline. **Results:** 38 lesions progressed to healing. Restoration took 10 months. In 1 patient the filling of the cyst was inadequate and required further surgery to complete the filling, which finally evolved to healing. **Conclusions:** Filling subchondral cysts with hydroxyapatite coralline prevents the progression of joint damage and facilitates reconstruction with a prosthetic implant in the future.





Abstracts (cont'd)

B4.3

Correlation Between Musculoskeletal Function and Radiological Joint Scores in Hemophilia A Adolescents

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Key Words: Musculoskeletal; Hemophilia

Introduction: The Functional Independence Score in Haemophilia (FISH) is a performance-based instrument used to assess musculoskeletal function in patients with hemophilia. We aimed to evaluate the functional independence of hemophilia A adolescents and its correlation to radiological joint scores. **Methods:** A cross sectional study was carried out on 50 adolescent haemophilia A patients. Musculoskeletal function was assessed using the FISH score and individual joints were assessed radiologically using the Pettersson score and MRI scale. Data were checked, entered, and analyzed using SPSS version 11. **Results:** The mean age of our patients was 16 ± 1.1 with a mean FISH of 23.32 ± 4.69 (range 13-28), and a mean Pettersson score of 2.32 ± 3.09 (range 0-13) for the knees, 1.86 ± 2.67 (range 0-11) for ankles, and 1.42 ± 2.17 (range 0-10) for elbows. The mean MRI score for the knees was 3.92 ± 2.74 (range 0-10) while that for ankles was 3.16 ± 2.64 (range 0-10), and for elbows was 2.34 ± 2.63 (range 0-10). There was highly significant correlation between both radiological joint scores and FISH and between degree of factor VIII deficiency and each of FISH, Pettersson score and MRI score. MRI was superior to conventional radiography in detection of subchondral cyst formation and erosions at joint margins. **Conclusion:** Given the highly significant correlation with both radiological joint scores, FISH appears to be a reliable tool for the assessment of functional independence in adolescents with hemophilia A. MRI is more sensitive than conventional radiography in detection of early joint abnormalities.

B4.4

Micronucleus Evaluation for Potential Chromosomal Breakages in Hemophiliac Patients Who Have Undergone Radioisotope Synovectomy

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Introduction: Radioisotope Synovectomy (RS) is defined as the intra-articular injection of radioisotopic agents in the target joint of patients with hemophilia. The aim of this study was to investigate the potential genotoxic effects of

Y⁹⁰ and Re¹⁸⁶ in the young population with hemophilia. Micronucleus (MN) assay on peripheral blood lymphocytes has been reported as one of the best established in vivo cytogenetic assays in the field of genetic toxicology. **Methods:** 19 patients exposed to RS were enrolled in the study. Mean age was 20.6 ± 10.4 years-old (range: 6-52), and 17 patients had hemophilia A. 18 patients with hemophilia who were not exposed to RS procedure were selected as a control group (mean age: 22 ± 10.6 years-old, range: 5-45 /13 hemophilia A). MN assay was performed using cytochalasin-B and 1000 binucleated cells were scored under light microscope in the Medical Genetics laboratory. **Results:** There was no significant difference between patients (18.2 ± 10.3 per 1000 cells) and controls (16.6 ± 7.8) in respect of MN values. However, both values obtained in RS exposed patients and control group were much elevated than values reported from healthy controls ($4.4-9.8/1000$ cells). The mean values of patients below 20 years old (15.4 ± 8.2) were much lower those above 20 years old (22.2 ± 12.0), however there was no difference. MN frequencies between Re186 (22.5 ± 10.0) and Y90 (19.3 ± 11.3) groups were also analysed, and no significant difference was observed. **Conclusion:** In conclusion, radioisotope synovectomy seems to be a safe procedure, however further studies including larger series of patients are needed to better understanding the effects on health status.

B4.5

Nerve Entrapment and Neurodiagnostics, Our Experience in Hemophilia

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Introduction: Often the presence of hematoma in patients with hemophilia can occasionally cause entrapment neuropathies of various nerves according to their location. Although the neurological symptoms of the nerve entrapment is very typical, it is necessary to confirm the same non-invasive methods that allow us to quantify the damage, from which the most appropriate treatment can be designed. **Methods:** Neurophysiological techniques were used, motor and sensory nerve conduction in nerve stimulation and recording through surface electrode bar or rings. **Results:** Clinical suspicion of entrapment neuropathy was found in 9 patients with positive studies, of which all but one were resolved with physiotherapy and medication methods, only one required surgical intervention for resolution. The nerves affected were mainly found in the median nerve in the ulnar nerve second, then the radial nerve, and finally a single case of femoral cutaneous lateral nerve. **Conclusion:** In this paper we show our experience in the detection, confirmation, and treatment of nerve entrapment in hemophilia patients with the use of nerve





Abstracts (cont'd)

conduction techniques for noninvasive neurophysiological treatment.

B4.6

Inter-Observer Reliability of Three Types of Radiographic Scores for Adult Hemophilia

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Introduction: The purpose of this study is to evaluate the inter-observer reliability of radiological assessment systems for hemophilic arthropathy. **Methods:** Three senior orthopedic surgeons with expertise in hemophilia independently evaluated a total of 527 joint radiographs of adult hemophilia patients, without any knowledge of the clinical data. **Results:** This study was the largest study to evaluate the reliability of radiological assessment systems. As for the results, the Arnold-Hilgartner staging system showed moderate reliability (kappa value: $\kappa=0.44$, $p=0.000$), and the De Palma grading system and the Pettersson scoring system showed fair reliability ($\kappa=0.40$, $p=0.000$) and slight reliability ($\kappa=0.12$, $p=0.000$), respectively. As for the reliability of the eight findings in the Pettersson scoring system, three findings showed substantial or moderate reliability. These were "narrowing of joint space" ($\kappa=0.70$, $p=0.000$), "irregular subchondral surface" ($\kappa=0.58$, $p=0.000$), and "erosion of joint margins" ($\kappa=0.56$, $p=0.000$). Other findings showed fair or less reliability. The traditional radiological assessment systems showed poor inter-observer reliability. Both progressive scales showed higher reliability than the additive scale, and the three findings in the Pettersson scoring system showed good reliability. **Conclusion:** Our results suggest that the progressive scale, including the three reliable radiological findings, might be a more reliable radiological assessment system.

B4.7

Pharmacovigilance in Patients Who Underwent Radiosynovectomy with Colloidal Chromic Phosphate P-32

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Introduction: Radioisotopes as localized radiotherapy for resistant synovitis have been widely implemented for the

treatment of inflammatory arthropathies. Their efficacy has been addressed by several trials, but safety concerns due to the theoretical risk of stochastic effects and reported cases of malignancy have precluded their use in many countries. **Methods:** We approach the issue of safety through 15 years of patient follow-up after P-32 radiosynovectomy performed by our multidisciplinary hemophilia group in collaboration with nuclear medicine specialists. **Results:** Since 1995, 287 individual joint radiosynovectomies were performed in 148 patients aged 4 to 71-years-old (mean 21.9). The main underlying condition leading to arthropathy was hemophilia (71.6%), followed by rheumatoid arthritis (9.5%), degenerative arthritis (6.1%), PVNS (6.1%), and seronegative arthritis (4.1%). Joints treated were knees (53.3%), elbows (27.5%), ankles (8.3%), hips (5.9%), and shoulders (3.1%). Doses of P-32 in colloid form per joint range from 0.25 to 1 mCi, and the number of joints treated per patient from 1 to 10 (mean 1.97). Average follow-up for all joints is 8.5 years (range 16 months to 15.8 years). There have been no documented cases of malignancy in patients exposed to the radioisotope. During follow-up, one patient died from a hemorrhagic complication of hemophilia. **Conclusions:** With this series we contribute cumulative data to the universal pool dedicated to establishing the long term safety of radiosynovectomy. Verifying the long term safety of radiosynoviorthesis is relevant given that when paired with its known efficacy for controlling synovial hypertrophy, this procedure is a highly risk-effective intervention. Maintaining strict clinical observation on this cohort will provide additional safety information as the population ages.

Key words: hemophilic arthropathy, radiosynovectomy

B4.8

CT Guided Percutaneous Decompression of Iliopsoas Hematoma in Hemophiliac Patient: Case Report

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Iliopsoas hematoma is a complication in hemophilia. It occurs in 0.3-13% depending on the cited source. It is potentially life threatening with a high risk of compartment syndrome (CS) and femoral neuropathy. The current treatment is conservative, with high doses of coagulation factor and bed rest. Percutaneous decompression is used on different pathologies, but on iliopsoas hematomas there are only 3 successful cases published on patients with anticoagulant therapy. We report a case of a 14-year-old male patient, with moderate hemophilia A and severe left groin pain, hip in flexion, positive psoas sign and hypoesthesia on femoral nerve





Abstracts (cont'd)

territory. He received factor VIII replacement and analgesics. CT reported acute left iliopsoas hematoma. We decided to perform a CT guided percutaneous decompression under strictly sterile conditions, with a No. 18 lumbar puncture needle. Intracompartmental pressure was measured with a manual manometer. 30 mmHg was the first measurement, 10 ml of blood was drained and posterior pressure was 23 mmHg with immediate pain relief and improvement on the leg's sensibility. This procedure is a valuable alternative in severe iliopsoas hematoma treatment with CS. It has to be performed by trained physicians in a hemophilia treatment centre.

Key words: Iliopsoas hematoma, percutaneous decompression

B4.9

Long-Term Outcome of Total Elbow Endoprosthesis in Hemophilia Arthropathy May Be Superior to Radial Head Prosthesis – Some Case Reports

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Introduction: The elbow represents the second most involved joint in hemophilic arthropathy. In contrast to hip and knee, total elbow replacement is not yet an established therapy (less functional impairment, no long-term results, early loosening of the prosthesis, design of the implants). Long-term outcomes are still awaited.

Methods: Since 2001, 6 patients (35-64 years-old) with advanced arthropathy of the elbow received a total elbow prosthesis, and 1 patient (27-years-old) a radial head prosthesis. We evaluate every patient at least once per year including functional outcome in daily life, range of motion (ROM), bleeding pattern, and pain assessment. **Results:** Functional outcome in everyday life is highly satisfactory in all patients. ROM depends mainly on pre-operative stiffness of the joint capsule. The median increase of motion was 20°, full extension in most cases is not achieved. 5/7 patients had complete relief of pain. Remaining pain was observed in one patient with total elbow replacement, suffering from dysesthesia of the hand improving under gabapentin therapy. No complete pain relief and increasing pain in the meantime was seen in the one patient with radial head prosthesis in 2007. There seems to be a continuous trauma by the radial head prosthesis eroding the opposite joint partner, leading to repeated micro bleedings. **Conclusion:** Total and partial elbow arthroplasty reduces significantly or even resolves pain, improves the range of motion and the capacity of the elbow joint. Radial head prosthesis is only a minor surgical procedure but may have disadvantages in hemophilic arthropathy, causing micro bleedings in the opposite bone.

B4.10

Arthroscopic Synovectomy with Joint Distraction Using a Patella Tendon Bearing Brace for Severe Hemophilic Ankle Arthropathy

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Introduction: Although arthrodesis is a gold standard for severe ankle arthropathy, we would like to avoid it, especially for young patients. We devised treatment with joint distraction using a patella tendon bearing (PTB) brace after doing synovectomy. The purpose of this study was to evaluate the clinical results of our procedures. **Methods:** Nine patients (all boys, 5-18-years-old, mean 10-years-old) with progressed ankle arthropathy were treated. Careful arthroscopic synovectomy was performed and defective cartilage areas were treated with bone marrow stimulation technique. The PTB brace was applied for 1 year post-operatively. Clinical results and radiographic findings using weight-bearing views were evaluated. Follow-up durations were from 24 to 64 months, with an average of 48 months. **Results:** Pain and disturbance of ADL were dramatically improved. An average AOFAS score was improved from 59 points to 88 points. Episodes of intra-articular bleeding were significantly decreased after the treatment. Erosive changes were repaired and narrowing of joint space was recovered to nearly normal. The Arnold-Hilgartner stages also were improved. The average Pettersson score was improved from 7.7 to 4.7. Autism in one patient was cured post-operatively and he plays drums in his band now. **Conclusion:** Arthroscopic synovectomy is an established therapeutic option for hemophilic ankle arthropathy. However, it was well known that synovectomy could not improve existing joint degeneration. Our devised treatment could improve radiographic stages for progressed ankle arthropathy. It should be done before indicating arthrodesis for young patients.

B4.11

Feasibility of Continuous Peripheral Nerve Block for Post-Operative Analgesia in Patients with Severe Hemophilia

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Background: Hemophilic patients with hemarthropathy are chronic pain patients. Pain treatment is essential in orthopedic surgery. Procedures are often associated with relevant post-operative pain. **Aim:** Moreover in





Abstracts (cont'd)

hemophiliac patients, the implementation of surgery requires early mobilization for rehabilitation in order to optimize functional recovery (a further cause of major pain). Continuous peripheral nerve block (CPNB) has become the standard approach; this study assessed their realization (safety) and effects in these patients. **Material & Methods:** At our hospital, under adequate factor replacement, CPNB (Contiplex B.Braun®) can be performed before (35% of cases) or after the patient was awake from general anesthesia (65%), using ultrasound or electrical nerve stimulation guidance. The interruption of the sensitive transmission can be obtained with low doses of anaesthetic, in the absence of motor block with the use of elastomeric pump (EasypumpR8ml/h) with a solution of bupivacaine 0.125%. 27 patients were included. They underwent a five-day programme of early joint mobilization - factor activity level was maintained above 30%, and they were evaluated for placement of catheter as the primary outcome; secondary outcomes included: VAS visual analogue scale, adverse effects and their satisfaction of pain control and compliance with physiotherapy. **Results:** The success rate of catheter placement is higher using US (100%) compared to electrical nerve stimulation (92%). VAS diminished to 2.5 in all patients except three. Adverse effects=1; satisfaction and good compliance of the patient with mobilization (all but two). **Conclusion:** CPNBs may be safely performed in hemophiliac patients and provides excellent post-operative analgesia. The anesthesiologist should be in charge of adopting most adequate approaches in order to contribute to improve treatment quality.

Keywords: pain, CPNB, safety, rehabilitation

B4.12

Efficacy of Zoledronic Acid to Normalize the Lumbar Bone Mineral Density in Children with Hemophilia

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Pediatric patients with hemophilia often have low bone mass for their age. Bisphosphonates are used for the treatment of adult bone diseases associated with excessive resorption, but there has been increasing and successful application in pediatric bone diseases. Trials using Zoledronic acid have never been performed in this population. **Objective:** To determine the safety and efficacy of Zoledronic acid to improve bone mass in children with hemophilia. **Methods:** In an open-label phase II trial, patients with hemophilia and osteopenia (defined as lumbar spine bone mineral density (BMD) Z-score less than 2 standard deviations (SD) below the mean for sex and age at baseline), were treated with Zoledronic

acid every four months for a six-month period. The primary outcome was change in lumbar spine BMD (LSBMD) Z-score at 6 months determined using dual X-ray absorptiometry, secondary outcomes included adverse events. **Results:** Six patients with on-demand treatment and a median age of 12-years-old were included. At baseline the median value of LSBMD Z-score was -2.6 SD compared with a -0.95 Z-score value at 6 months ($p = 0.027$), this significant difference persisted until 12 months. The treatment was well tolerated; adverse reactions consisted of flu-like symptoms and pruritus, which resolved without any further action. **Conclusions:** Our data suggest that Zoledronic acid seems to be a safe and effective option to get the LSBMD back to normal after 6 months in all children with hemophilia, however, this was an open and uncontrolled study, and double-blind, placebo-controlled studies are needed.

Keywords: Children, Osteoporosis

B4.13

Perioperative Clotting Factor Replacement and Infection in Total Knee Arthroplasty

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Hemophilia is an X-linked inherited bleeding disorder which can cause severe arthropathy. The knee is the most commonly affected joint, and is associated with the most significant disability and impaired quality of life. Over the past few decades, total knee arthroplasty has become the most common surgical procedure performed in adult hemophiliacs. The outcomes of total knee replacements (TKR) in the published literature is, however, highly variable, with infection rates of 1-16% reported in studies. One contributing factor may be the differing clotting factor replacement protocols used. Current World Federation of Hemophilia guidelines for perioperative plasma factor VIII levels recommend a pre-operative level of 80-100%, tapering down to 30-50% at 14 days. Recent animal studies have suggested that high initial levels of the deficient plasma factor are not sufficient to normalise wound healing, and that normal wound healing requires adequate hemostatic function for an extended period of time. The purpose of this study was to compare the differing clotting factor replacement protocols used in the published series of TKR for hemophilic arthropathy, in order to determine if a relationship exists with infection rates. A meta-analysis of the published literature shows a statistically significant relationship between infection and clotting factor replacement. This would seem to support the use of 100% factor replacement perioperatively and the maintenance of high levels for 2 weeks post-operatively. A cost analysis of this protocol shows that whilst there may be an increased "up-front" cost implication, the ultimately reduced complication rate makes the high dose regime more cost-effective in the





Abstracts (cont'd)

long term. We believe that our findings support a change in the current international guidelines.

Key words: Knee arthroplasty, peri-operative management

Session C1 - Ankle Arthropathy

C1.1

Conservative Management: Physiotherapy

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Ankle arthropathy is one of the most common complaints in patients with hemophilia, even in young and adolescent patients. There are many treatment options, but the conservative way is the first option. It is very important to learn and understand what the typical evolution of the arthropathy of this joint is like and to identify the boys and adolescents with high risk of developing this condition. Once it is settled, it is essential to recognize the typical patterns that include loss of flexibility and range of motion, decrease of the strength of certain muscle groups, and impoverishment of proprioception and neuro-muscle control. In addition, it is important to emphasize that the majority of these patients have not only the ankle but many joints affected, so their walking patterns are involved. In our experience treating many cases of patients with this condition, we have obtained very good results adequately applying modalities, strengthening and flexibility exercises, proprioception drills, and walking re-education. This presentation hopes to transmit the way in which this condition can be treated choosing the conservative option, and describe in detail the aforementioned points.

C1.2

Orthoses: When and Why?

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Despite the constant improvement in hemophilia management, arthropathy remains the major cause of significant clinical problems in patients with hemophilia since joints are the most frequent location where bleeding occurs. In literature, it is often stated that the ankle joint is less frequently affected by arthropathy than other joints, yet it is well known that the ankle joint demonstrates early onset of bleeding in severe hemophilia, so in this population it is quite frequent. Treatment of ankle arthropathy should always begin with simple steps including footwear changes, activity modifications, and use of orthoses. Orthotic management can be very useful for ankle arthropathy when is used in its early stages. There are many types of orthoses. In mild arthropathy a brace can be applied to help hold the ankle joint in position, or in moderate cases they can be custom-made and are called ankle-foot-orthoses or AFOs. Application of

orthoses has two main goals. Firstly, to reduce the forces through the ankle allowing weight to bypass the inside of the joint. Secondly, to physically restrict the movements of the ankle reducing the forces passing through the joint in this way also prevents the impingement of altered synovia. As long as symptoms are limited to weight bearing activity and not present when resting or during sleep, an orthotic is useful. Once the symptoms are not alleviated by wearing orthotics then more invasive treatments such as surgical options may be considered.

Key words: Hemophilia, Ankle Arthropathy, Orthoses.

C1.3

Pre- and Post-Operative Physiotherapy Management of Hemophilic Ankle Arthropathy

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The ankle is a common site for spontaneous hemarthrosis in patients with hemophilia. Bleeding episodes principally affect the tibiotalar and/or the subtalar joints, and may lead to severe degenerative changes. Several mechanical and functional deficits have been identified that may contribute to associated pain and functional loss. Associated muscle atrophy, decreased tibio-talar and subtalar range of motion, muscle flexibility, and proprioception because of the pre-existing target joint are generally evident. One of the hidden symptoms is decreased range of motion of the midtarsal and tarsometatarsal joints. This deficit could predispose the patient to increased pain, stiffness, and disrupted proprioceptive input to the sensorimotor system. Therefore, improving accessory and physiological motion at the entirety of the ankle and foot joints is a clinical consideration. Localized joint mobilizations could be an effective intervention. The role and effectiveness for conservative treatment, such as medication, patient education, shoe modification, and physical agents will be discussed in this presentation. Surgical procedures can include synovectomy, synoviorthesis, debridement, or arthrodesis. Synovectomies are advocated to help stop uncontrolled, repetitive cycles of joint bleed and/or chronic synovitis. As the involved synovial membrane is destroyed, joint bleeding is often resolved or significantly decreased. The key is in taking advantage of the subsequent, relatively safe, bleed-free period to propose a tailored physiotherapy program. After ankle arthrodesis, mobilizations to unfused joints may begin after two weeks post-op. Once the fusion has completely healed, low level balance and proprioceptive exercises can be started. An insert in the shoe or an orthopedic rocker sole will facilitate the foot to move through a step.





Abstracts (cont'd)

C1.4

Ankle Fusion: A Solution for Developing Countries Only?

TBC

No Abstract Submitted.

C1.5

New Developments in Total Ankle Replacement

J.F. Asencio

No Abstract Submitted.

Poster Session 1: Surgical

P1.01

Knee Surgical Synovectomy in Young Hemophiliacs: A Report of 8 Cases

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In Algeria in 2010, 524 hemophiliacs from a total of 1465 were under 19 years of age.

The aim of this study is to assess the results of synovectomy in the treatment of hemophilic arthropathies and demonstrate the interest of a multidisciplinary collaboration (hematologist, orthopedic surgeon, and physiotherapist). **Method:** Between 2008 and 2010, 24 hemophilic patients (type A and B) had a total of 29 surgical procedures, of which 14 orthopedic surgical procedures were carried out on 11 children. Eight synovectomies were performed in the Orthopedics Unit in 7 severe hemophilic children, of which 2 were carried out on the same patient. Patients were between 7 and 16-years-old. Viral contamination (HIV, HCV) was absent in all patients as were inhibitors. Indications of the synovectomy: frequent hemarthrosis, chronic hemarthrosis, pain and deterioration of function, stage III and IV arthropathy (Arnold-Hilgartner scale), the mobility of the knee was 102°. Substitution by antihemophilic factor was maintained during a couple of weeks postoperatively, then twice a week to ensure functional rehabilitation. During hospitalization the patients used on average 50000 IU of factor VIII and 22500 IU of factor IX, and between 48000 IU and 96000 IU of each factor to cover functional rehabilitation. No hemorrhagic complications, infections, or appearance of inhibitors occurred after the surgery. The results were evaluated with

13 months follow-up period. **Results:** Synovectomy yielded good and satisfactory results. In more than half of cases, no bleeding episodes occurred; its effect on the range of motion was moderate with a usual loss of mobility between 8 and 14°, recovered in some patients after rehabilitation. **Conclusion:** Synovectomy in moderate to severe chronic arthropathy produces good results in terms of bleeding recurrence and overall function when performed in a specialized centre with a multi-disciplinary approach.

P1.02

Primary Prophylaxis Therapy in Hemophilia with Low Dose and Low Intensity: Results from a Centre in Turkey

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Primary prophylaxis with coagulation factor concentrates has become the standard of care for children with hemophilia. However, while reducing the risk of bleeding and related morbidity, the optimal use of prophylaxis is not well known. We tried to tailor the dose and schedule of prophylaxis and reduce costs without decreasing efficacy. The aim of this study was to evaluate the effectiveness of low dose primary prophylaxis therapy in children with severe hemophilia A or B. We documented the patients from June 2005 to June 2010. 15 patients (10 hemophilia A patients and 5 hemophilia B) who had been on prophylaxis therapy for at least 6 months were gathered. These hemophilia A and B patients received 20-30u/kg of factors VIII and IX, one or two times per week, respectively. All except 1 hemophilia A patient were treated with recombinant factors (Recombinant, Kogenate FS), only one patient began prophylaxis with plasma-derived factor concentrates and then shifted to recombinant factor. All hemophilia B patients were treated with plasma-derived factors. Twelve were diagnosed during the first year of life, usually when they were 7-8 months old (3.5-11 month) except three who were diagnosed when they were 4-5-years-old. The orthopedic evaluation was performed, 86.6% of patients who had used factor regularly did not present any clinical or radiographic changes in their joints; only one patient who was diagnosed when he was 4-years-old and put on prophylaxis had radioactive synovectomy. The preliminary results of low dose primary prophylaxis confirm its effectiveness in preventing hemophilic arthropathy and bleeding.





Abstracts (cont'd)

P1.03

The Results of Staged Bilateral Total Knee Arthroplasty in Congenital Factor XIII Deficiency

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Introduction: Although hemarthrosis is not a rare manifestation of congenital factor XIII deficiency, destructive changes in the joints are relatively uncommon, in contrast with arthropathy changes seen in patients with classic hemophilia. Factor replacement has significantly helped to reduce the complication rates of total knee arthroplasty (TKA) in classic hemophilia; making TKA an effective method to achieve pain relief and gain better function in this group of patients. According to our knowledge, there is not any published data regarding the results of TKA in patients with congenital factor XIII deficiency. **Methods:** Bilateral staged total knee arthroplasty with a six-month interval was performed for management of disabling arthropathy in a 56-year-old female with congenital factor XIII deficiency. Stemmed condylar constrained knee prosthesis was used because of varus and bony deficiency. According to the recommendations of our hematologist consultant, plasma-derived factor XIII concentrate and Transamin were prescribed pre- and post-operatively. The pre- and post-operative range of movement was assessed and complications after surgery were noted. Functional results were assessed using the Hospital for Special Surgery (HSS) knee scoring system, both pre- and post-operatively.

Results: There was neither abnormal bleeding nor infection. The range of movement was increased. The knee score (HSS) was improved considerably (from poor to good); hence, the patient's quality of life has been improved. **Conclusion:** Like classic hemophilia, total knee arthroplasty may be a safe and effective procedure for the management of severe arthropathy in congenital factor XIII deficiency after infusion of plasma-derived factor XIII concentrate and Transamin.

Keywords: sequence of joint damage, strategy of compensation

P1.04

Musculoskeletal Study of Algerian Severe Hemophiliacs: Primary Finding, and 27 Cases of Elective Orthopedic Surgery

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Aim: This study gives the musculoskeletal situation of an Algerian sample population of hemophiliacs. It gives sequence analyses of joint events, and provides a first draft theory of sequences, vertical or passing from limb to other of joint damage. **Method:** Since January 2008, we received 95 severe hemophiliac patients who have musculoskeletal problems. We perform a systematical review of their medical history; take photos and videos of physical and radiological anomalies. We insist on the sequence of joint manifestation using interview and medical documents. We statistically analyse all the data.

Results: 70% are under 30-years-old. In 55.4% there are 2 hemophiliacs per family, and more than 3 in 35.3%. Inhibitor factor status: 5% positive, 58% negative, and 37% unknown. Viral serology: 10% positive and 37% unknown. One joint's lesion: 32%, more than 3 joint's lesion: 52.5%. The sequences of joint lesion: 72.7% are passing from limb to the other; the diagram (A) gives all found sequences. Discussion: There is segmental joint adaptation in response to progressive joint function deterioration (M. Fearn 2010), the result of our study shows that the sequence of joint lesions is an adaptation of this deterioration. Our study shows that the sequence of joint damage is secondary to different compensation strategies, and joint's damage timing: vertical or cross from one limb to another, is guided by the first affected joint.

Conclusion: The sequence of joint damage in hemophilia is the consequence of different strategies of compensation and allows to define a joint at risk.

Keywords: sequence of joint damage, strategy of compensation

P1.05

Hemophilic Chronic Synovitis: Hemarthrosis Therapy Using Endovascular Embolization in Knee and Elbow

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Introduction: Chronic synovitis produce loss of quality of life (QOL). Objective: To show the effect of endovascular embolization therapy in patients with synovitis. **Material and Methods:** 23 patients, 2 with inhibitors, ranging from 12-56 years-old, and presenting at least 3 hemarthrosis events in 6 months, and synovitis. We performed 26 procedures. Angiography of genicular arteries in the knee (18) and ulnar and radial arteries in the elbow (6) was followed by selective gelfoam or PVA particle embolization in blush or hypervascular lakes using standard endovascular technique. MRI was performed before and at 2 and 6 months control. Follow-up between 6 and 42 months, evaluating number of hemarthrosis events, amount of coagulation, clotting factors used, and QOL.





Abstracts (cont'd)

Results: All patients have improved QOL and made improvements in physiotherapy. In the knee group the average number of days without clinical events was 112 ± 76.8 . There was a reduction in the number of events and the severity of them, and consumption of coagulation substitutive factors decreased ($p < 0.01$) at 6 months in the whole study and ($p < 0.01$) at 18 months in patients having more than 3 bleeding events in 3 months. One patient had to be re-embolized at 6 months. Technical success was 96.15% without major complications or adverse effects. In the elbow group 1 patient had bad results which coincided with an insufficient technical embolization.

Conclusion: In hemophilic patients with chronic knee/elbow synovitis, endovascular embolization seems to offer a safe therapeutic option to treat hemarthrosis and reduce synovitis.

P1.06 Botulinum Toxin-A Intramuscular Injections in a Boy with Severe Hemophilia and Cerebral Palsy

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Introduction: We report a case of a boy with severe hemophilia A and cerebral palsy (CP) with spastic diplegia who received intramuscular injections of Botulinum toxin-A (BtA) to reduce his lower-limb hypertonicity. BtA injection was indicated for dynamic contractures which interfered with walking and wearing splints. No cases of children with hemophilia receiving BtA for CP were found in the literature. **Methods:** Prior to BtA injection, the modified Tardieu Scale was used to assess spasticity and contracture, and the Gross Motor Function Measure – GMFM-66 was used to measure change in gross motor function. Before the BtA injection and the day following the procedure he received factor VIII cover at 60IU per kg, tranexamic acid 250 mg tds for 7 days. BtA was injected bilaterally into hip adductors, hamstrings, and gastrocnemii. Dosage of BtA injection was calculated from international consensus guidelines. Orthotics was amended to accommodate the effects of BtA. **Results:** Eight weeks later, parents reported improvements in activities of daily function. There was absence of dynamic clonus in hips and knees and marked improvement at the ankle joint. Before BtA injection, he scored 83% and 26% in the GMFM-66 crawling/kneeling and standing dimensions respectively. On reassessment, scores were 95.2% and 33% respectively, which were considered clinically significant. **Conclusion:** Assessments demonstrated improvements in ambulation, tolerance of orthotics, functional abilities, and range of movement. There were no bleeding side effects. BtA is documented in the literature for the management of hypertonicity in children with CP and should be considered in those children with hemophilia and CP.

P1.07

Synoviorthesis Induced by Rifampicin in Hemophilic Arthropathy; Report of 24 Treated Joints

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Introduction: Intra-articular injection of chemical or radioactive substances in order to produce fibrosis of the hypertrophied synovium has proved effective in the treatment of chronic hemophilic synovitis. The aim of this study is to investigate clinical outcomes of synoviorthesis induced by rifampicin in 21 patients (which included 24 joints) with hemophilic arthropathy. **Methods:** This quasi-experimental study was conducted between December 2006 and July 2007. We treated 21 patients with hemophilic arthropathy by synoviorthesis with rifampicin (500mg) once a week. The procedures were performed on 14 knees, 5 elbows, 4 ankles, and 1 shoulder (24 joints and 113 injections). **Results:** Radiological staging using the Arnold-Hilgartner scale and Fernandez-Palazzi classification were performed. Mean age was 16.7 years (range 7–49 years). According to the Fernandez-Palazzi classification, synoviorthesis was considered satisfactory (excellent or good) in 21 (87.5%) joints and unsatisfactory (fair or poor) in 3 (12.5%). A mean reduction of 6.3 bleeding episode per month was obtained ($p < 0.0001$). The mean pain score was reduced from 2.62 (range: 2–3, SD: 0.49) before treatment to 0.79 (range 0–2 SD: 0.65) after treatment. The mean World Federation of Hemophilia (WFH) score was 5.45 (range 2–8) before synoviorthesis and 3.5 (range 1–7) after treatment. **Conclusions:** This method effectively reduced hemarthrosis and pain, and also improved range of motion in patients with hemophilic arthropathy. Chemical synoviorthesis with rifampicin appears to be efficient, inexpensive and simple, and also especially practical in developing countries where radioactive agents are not easily available.

P1.08

Our Follow-up Study After Synovectomy of Hemophilic Knee Joints

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Synovectomy of the hemophilic joints controls bleeding; however, significant loss of motion frequently results. Over the last 16 years we performed 56 operations in our orthopedic centre, on hemophilia patients with hemophilic arthropathy. Among these, 25 were





Abstracts (cont'd)

arthroscopic knee synovectomies, combined with joint debridement. Two other knee synovectomies were made by open surgical procedure. Pre-operatively, all of the patients had experienced an average of three bleeding episodes into the affected joint per month, and had been unresponsive to at least three months of substitution management. We are reporting on 10 patients who underwent arthroscopic knee synovectomy and had decreased bleeding episodes. The average flexion contracture before operation was 11.5 degrees, post-operatively flexion contracture reduced to 9.5 degrees. The average loss of range of knee motion was 19 degrees. The average age of the patients at the time of the knee synovectomy was 37-years-old and the average length of follow-up was 5.65 years (range, 1-16 years). It was concluded from our study that bleeding frequency and the pain associated with persistent synovitis in the hemophilic knee can be effectively decreased with knee synovectomy. All of our patients required less factor after synovectomies to control bleeding, although almost all of our patients had loss of motion of the knee post-operatively.

P1.09 Hemophilic Arthropathy: Diagnosis and Treatment of Subchondral Knee Cyst

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Introduction: The purpose of the study is to review the result of the surgical treatment of the subchondral knee cyst with impacted morselized bone graft in a patient with severe hemophilia. These cysts cause pain, loss of bone stock, and result in restricted range of movement and deterioration of quality of life. **Methods:** We evaluated the results of the surgical treatment of a subchondral knee cyst in a 54-year-old patient with severe hemophilia A. The patient had a history of recurrent bleeding, mainly in the knee. The cyst was diagnosed by conventional radiographs and MRI. The indication for surgical treatment was based on the size of the cyst, which was larger than 50% of the size of the tibial plateau.

This procedure consisted of aspirating the contents of the cyst and packing it with impacted morselized bone graft. In addition to surgery, the patient underwent both pre- and post-operative physical therapy. **Results:** After three months, the patient had no joint pain and had improved his activity level. The range of movement increased by 40° (from 70°-110°). Signs of allograft resorption were not detected. **Conclusion:** The treatment of subchondral knee cysts with morselized bone graft provides biological tissue and increases the bony reservation, improving the

patient's quality of life and delaying the symptoms of hemophilic arthropathy.

P1.10 The Role of Colloid 32P Synoviorthesis in Treatment of Hemophilic Arthropathy

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Introduction: Radioactive synoviorthesis by injection of safe radioisotopes into the joints affected by chronic arthritis is accounted as a novel method to treat hemophilic arthropathy. The main goal of this therapy would be to decrease frequency of hemarthrosis and consumption of coagulation factors. In this study we assessed the effect of radioactive synoviorthesis on the frequency of hemarthrosis, factor consumption and other related parameters. **Methods:** In an interventional study in Imam Khomeini Hospital in Tehran, Iran, after meeting the inclusion criteria and taking written consent, colloid 32p radiosynovectomy was performed for 56 joints with hemophilic arthropathy. After local anesthesia of injection site, one mCi of 32P for large joints (knee) and 0.5 mCi for small joints (ankle and elbow) were injected, respectively. Half of these doses were considered for children (age <12 years). **Results:** The mean age was 16.78-years-old (range: 2.5-36; SD: 7.46), and 98.2% of cases were male. Injected were knee 80.35%, ankle 12.5%, and elbow 7%. The mean follow-up was 43.63 months (range: 3-102), the result was 62% decrease in frequency of hemarthrosis (p=0.0001) and 84% decrease in factor consumption (p=0.0001). However, the involvement of other (non-injected) joints during follow-up could lower the decrease of mean total factor consumption. **Conclusions:** Radioactive synoviorthesis can be a cost-effective alternative to decrease hemarthrosis and factor consumption in hemophilic arthropathy.

Keywords: Hemophilic arthropathy, synoviorthesis

P1.11 Cubital Tunnel Syndrome in Patients with Hemophilia

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Introduction: The elbow is the second most common joint involved in patients with hemophilia; however, there is little data about the involvement of the ulnar nerve at the elbow in patients with hemophilic arthropathy. The purpose of this study was to address this problem in the





Abstracts (cont'd)

elbow and evaluate the results of anterior subcutaneous transposition of the ulnar nerve in a small group of patients with hemophilia who had been managed in two institutions. **Materials:** Information on 6 patients who were diagnosed with tardy ulnar nerve palsy in two institutions was retrospectively collected. All patients suffered from severe hemophilia A. Anterior subcutaneous transposition of the ulnar nerve had been performed in all except one. **Results:** The mean age of the patients at the time of procedure was 45.8-years-old and the mean duration of follow-up was 60.2 months. No post-operative complication or recurrence was observed. No additional surgery was required in operated patients. Evaluation was performed using subjective and objective measures, and a modified Bishop score. After operation, subjective sensory and motor disturbances were improved or resolved in all of the operated patients, while objective measures improved less well. **Conclusion:** The ulnar nerve can be involved in cubital tunnel syndrome in patients with hemophilia. Anterior subcutaneous transposition of the ulnar nerve is an effective procedure for improving patients' symptoms, with a low risk of complications.

P1.12 Osteoporosis in Hemophilia: Preliminary Results of a Densitometric Study

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In hemophilic patients, the loss of bone mineral density is associated with osteoporosis, hemophilic arthropathy, and a decreased quality of life. The pathogenesis of osteoporosis is multifactorial, and in hemophilic patients the risk factors are increased by musculoskeletal injuries that reduce mobility and by the common presence of viral diseases and drugs that can affect bone metabolism. In our hemophilia treatment centre, densitometry is included in the patient's assessment protocol. The aim of this study is to describe the epidemiology of impaired bone mineralization in a group of patients with hemophilia. We performed axial and hip densitometry to 70 patients controlled in our centre. Regarding disease severity, 36 were severe, 9 moderate, and 25 mild. Mean age was 31-years-old (range 14 - 62). Mean body mass index was 24.4 (40% were overweight). 88% of the patients were infected by hepatitis C virus and 51.4% by the human immunodeficiency virus. The results of bone densitometry showed loss of mineralization in 70% of our patients: 34 had osteopenia (48.6%) and 15 had osteoporosis (21.4%). We conclude pointing out the importance of assessing osteoporosis considering it as a major clinical problem in the hemophilic population.

Keywords: Hemophilia, Osteoporosis

P1.13

Sonography Versus MRI in Hemophilic Arthropathy: A Comparative Study

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Introduction: Hemophilic arthropathy is the most severe complication of hemophilia. Physical examination and the Pettersson score are routinely used to grade the severity of arthropathy and monitor the effects of the treatment. Magnetic resonance imaging (MRI) is currently the gold standard for evaluating joint damage, but it is expensive and younger children need to be sedated to undergo this investigation. In recent years, sonography has become a useful complementary modality in the evaluation of hemophilic arthropathy because of its prompt availability.

Methods: We evaluated knee arthropathy, comparing the physical examination score and the different imaging techniques in three adult patients with severe hemophilia A treated with secondary prophylaxis. **Results:** While the clinical examination and plain radiographs tended to underestimate the extent of joint destruction, with MRI and sonography we obtained comparable information about synovitis, subchondral bone erosions, and chondral damage. **Conclusion:** While MRI is presently considered the gold standard for the evaluation of arthropathy, the need for sedation in children and its high costs preclude its use as a method for monitoring progression of joint disease. Although sonography is an operator-dependent technique and a systematic protocol for sonographic assessment is needed, we think that it may provide a useful, more readily available and less expensive tool than MRI for evaluating musculoskeletal involvement on a regular basis. In addition, the association of power-Doppler studies could help to detect and quantify synovial vascularity improving the quality of assessment of chronic synovitis.

P1.14

rFVIIa for Orthopedic Surgeries in FVII Deficient Patients

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Background: Inherited factor VII (FVII) deficiency has an estimated incidence of 1/500,000 in the general population and an autosomal recessive pattern of inheritance. The hemorrhagic manifestations in affected





Abstracts (cont'd)

individuals are variable and correlate poorly with plasma FVII activity levels. In severely affected cases, however, significant bleeding problems including spontaneous hemarthroses resulting in advanced arthropathy have been observed. We report here good results of orthopedic surgeries performed in five FVII deficient patients who received recombinant activated factor VII (rFVIIa) in the peri- and post-operative period. **Patients and Methods:** The study comprised 1 male and 4 females, aged 20-78-years-old, with FVII baseline plasma levels <10IU/dl. Two patients required total hip replacement and three required various arthroscopic procedures. All surgeries were performed under hemostatic cover of rFVIIa. **Results:** Doses of rFVIIa ranged from 18 to 37µg/kg bw and were given every 8 hours on the day of surgery (first dose given 10-15 minutes before the procedure), followed by 13-30µg/kg bw given every 12-24 hours for 9 to 14 days. Total dose of rFVIIa per procedure ranged from 16 to 38 mg. Lab monitoring consisted of daily determination of prothrombin time and FVII plasma activity. In all patients peri- and post-operative clinical course was uneventful and there was no need to use other preparations containing FVII. Blood loss ranged from 30 to 700 ml per procedure and none of the patients required blood transfusions. Pharmacologic thromboprophylaxis was used in two patients undergoing hip surgery. **Conclusion:** rFVIIa is an effective, safe, and cost-effective haemostatic agent in hypoproconvertinaemia patients undergoing orthopedic surgery.

P1.15 Radiosynoviorthesis in the Treatment of Hemophilic Joints - Hungarian Experience

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Aim: To present our promising results with radiosynoviorthesis in the treatment of hemophilic joints. **Materials and Method:** Hemophilic hip, knee, and ankle joints have been treated with radiosynoviorthesis at our department since 2003. 110 MBq Rhenium-186 sulfide, in colloidal suspension, is used for hip and ankle joints, while Yttrium-90, in colloidal suspension is implanted into knee joints. Isotope implantations are performed in a very sterile operating room. Local anesthesia with 1% Lidocaine is used for the procedure. There were no cases of injury caused by leakage of the isotope. The patients are admitted to the hospital for three days and bed rest and immobilization of the joints with a brace is prescribed for the same time period, to reduce extra-articular migration of the radiopharmaceutical. Hip joints are not braced. **Results:** We observed an overall reduction in joint bleeding frequency, from 18.7 to 2.4 per year post-

radiosynovectomy. Similar results were obtained in patients with advanced osteoarthritis and in inhibitor cases. There was a significant reduction in pain in all joints. Average range of motion was maintained or increased from 5 to 15 degrees of motion 1 year post-radiosynovectomy in most joints. Radiosynoviorthesis was successful in 80% of the patients after 1 year of isotope implantation. **Conclusion:** Radiosynoviorthesis represents an important alternative for the treatment of chronic hemophilic synovitis, markedly reducing joint bleeding frequency and pain, irrespective of the stage of arthritis and inhibitor status.

P1.16 The Results of ¹⁸⁶Re Radiosynovectomy with Individual Physiotherapeutic Protocol in the Treatment of Pediatric Hemophiliacs with Inhibitor

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Introduction: To evaluate the influence of ¹⁸⁶Re radiosynovectomy with individual physiotherapeutic protocol on reduction of hemarthroses in children with hemophilia and factor inhibitor. **Methods:** We performed 8 ¹⁸⁶Re radiosynovectomies on 4 knees and 4 elbows in 5 patients aged 9 to 15-years-old (median – 11.6) with severe hemophilia A (4) or B (1) with inhibitors. The activity of ¹⁸⁶Re colloid ranged from 60-180 MBq. We used rFVIIa (NovoSeven) before the injection, after 2 hours, after 3 hours, and then after 3 more hours. Then aPCC (FEIBA) was administered. Post-therapeutic scanning was performed 1 hour after the procedure and on the 2nd and 3rd days. The number of hemarthroses and the clinical state were assessed in 3 month intervals. Ultrasound evaluation was performed at each visit. Physiotherapy was adjusted to patient's age and the state of the joint, and aimed to regain a full range of motion, muscle strength, and proprioception. Efforts were made to reduce residual rotational contracture and regain proper walking pattern. Progression-free survival was assessed using the Kaplan-Meier method. **Results:** Significant reduction in hemarthroses was noted (before: 1.5/months; after: 0.4/months). Post-therapeutic scanning revealed even intra-articular distribution of isotope and no leakage. Physiotherapeutic protocol allowed for recovery and improvement of joint function. **Conclusion:** The study suggests that the use of ¹⁸⁶Re radiosynovectomy with tailored physiotherapy reduces the number of hemarthroses in young hemophiliacs with inhibitors.





Abstracts (cont'd)

P1.17

Factor VIII Replacement in the Peri-operative Period in Joint Arthroplasty in Hemophilia A – A Series of 7 Cases

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Introduction: Orthopedic surgeries in patients with congenital bleeding disorders require thorough a logistic plan, an experienced team of specialists, and adequate supplies of clotting factor concentrates. Access to elective joint replacement surgeries in patients with hemophilia in Poland is far below the demand. Since June 2010 the Cracow Rehabilitation Center, working in strict cooperation with the Department of Hematology in Cracow, is the second orthopedic hospital in Poland performing these procedures. We describe factor VIII replacement therapy in the peri-operative period in the first 7 patients with hemophilia A operated on in the Cracow Rehabilitation Center. We point out the issues concerning optimal dosage, monitoring strategy, and factor VIII levels in the peri-operative period. **Methods:** Factor VIII concentrate (Immunate, Baxter) was administered three times a day. Pre-dose factor VIII activity (FVIII:C) was measured daily from Monday till Friday with one-stage clotting assay (BCS XP analyzer, Siemens, Germany). Factor VIII doses were adjusted on a daily basis, based on the clinical status, FVIII and hemoglobin levels. Low-molecular-weight-heparin was used in only one patient due to prolonged immobility. **Results:** Pre-surgery FVIII level was 95-159%. Within 14 days of hospitalization a mean of 1042.6 IU/kg (range 678-1974) of factor VIII was utilized. In 4 of 7 patients mild to moderate bleeding complications were observed that did not affect the outcome of surgery and rehabilitation. **Conclusion:** Routine joint replacement in patients with hemophilia requires coordination between the surgeon, hematologist, laboratory diagnostician, and Blood Bank. Bleeding/thrombotic risk must be assessed individually for each patient.

Key words: joint replacement, hemophilia, factor VIII replacement therapy

P2.01

Physical Therapy of Patients with Hemophilia without a Replacement Therapy: Clinical Cases

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There is a shortage of replacement therapy drugs for hemophilia patients in the Ukraine. Therefore, it is important to use different methods of physical rehabilitation for people with bleeding disorders. In many cases we are using physical therapy for hemophilia patients without a replacement therapy. In this work we present the treatment of a patient with severe hemophilia A, complicated with inhibitors (patient 1), and a patient with von Willebrand disease (third type) (patient 2). Both of them have a 1-year-old combined contracture (flexion and extension) of the knee joint. Both patients were evaluated before rehabilitation and every month during the treatment. Evaluation includes measurements of range of movements in the joints, manual muscle test, and "Up&Go" test. Individual rehabilitation programs include active, active-passive exercises, post isometric relaxation, elements of Maitland's manual therapy, and exercises for muscle strengthening. Both patients received individual rehabilitation programs, which lasted for half a year. After this period patients show significant improvements in knee joint movement: range of movements has increased from -20° – 75° to 0° – 100°, and from -35° – 85° to 0° – 120° for patient 1 and 2, respectively; manual muscle test in knee extension and flexion has increased by 2 points for both patients; "Up&Go" test time has decreased from 22 to 13 seconds and from 18 to 9 seconds for patient 1 and 2, respectively. These results show that our proposed rehabilitation program is very effective for the comprehensive care of patients with hemophilia.

Keywords: hemophilia, physical rehabilitation

P2.02

Acute Effect of Exercise in Persons with Hemophilia

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Hemophilia is a bleeding disorder that concerns experts. Physical activity has shown some changes in hemostasis related to improvements in the coagulation cascade of normal people, such as: increasing FVIII, increasing VWF, decreasing APTT, decreasing PT and increasing fibrinogen.

Goal: This is a preliminary study that aims to analyse the acute effect of exercise on people with hemophilia.

Methods: The sample consisted of 4 severe hemophilia A and 1 moderate hemophilia A patients, physically active for at least six months, aged 15-31 years-old (mean 19.6). The sample had blood samples taken before and immediately after a session of 20 minutes of exercise without the use of clotting factor for 48 hours before. Statistical procedure for analyzing the results was statistical average of the values before and after exercise.

Results: We found decreased APTT from 74.88 to 72.88; a reduction of PT from 17.88 to 13.56; a FVIII increase from





Abstracts (cont'd)

1.75% to 2.59%; and fibrinogen increasing from 299.4% to 304.6% with the heart rate average 142 bpm. **Conclusion:** In the present study we found that there was a change in the parameters observed. However the sample was not sufficient to statistically evaluate the acute effect of exercise on hemostasis. We suggest further studies to evaluate the acute and chronic effect of exercise on hemostasis, the long life of FVIII and other components of the coagulation cascade.

P2.03

Why Physical Activity for Hemophiliacs?

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Sedentary behavior in people with hemophilia is associated with a risk of being overweight, having joint problems, repeated bleedings, and loss of function. The actual recommendations for sedentary people are to accumulate physical activity for at least 30 minutes per day almost every day of the week (5-7days). The ideal for health is to accumulate approximately 28kcal/kg/week. One kcal corresponds to 1 MET (metabolic equivalent). The aim of this study was analyze the chronic effects of physical activities in hemophiliacs participating in regular physical activities (3 times/week) for at least one year. **Methods:** The group had 10 males with hemophilia (A, B) aged between 15 to 34-years-old, stimulated to practice physical activities for one year. The group was analyzed by IPAQ questionnaire, body mass index (BMI), waist-hip ratio, rest blood pressure, and rest heart rate. The statistical analyses were done by averaged and standard error. **Results:** The group were identified: Less active (<600METs), moderated active (600-1499 METs), active (1500-2999METs), and very active (>3000 METs). We found 50% of the group was very active, 30% active, and 20% moderately active. Almost every result of the physical fitness test (BMI, waist-hip ratio, blood pressure, and heart rate) was associated with health conditions and less bleeding episodes. **Conclusion:** The chronic effects of regular physical exercise were associated with maintenance of energy, and we could observe the low level of bleeding during this time. We suggest more studies relating the recommended expenditure of energy in regular physical activity and the frequency of bleeding in joints, muscles, and factor usage throughout the process.

P2.04

Pseudotumors in Patients with Inhibitors

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Introduction: Pseudotumors are a major complication in patients with hemophilia and can be devastating, especially in the presence of inhibitors. Only a few cases have been reported in the literature. **Objectives:** The purpose of our work was to show the experience in treating 7 pseudotumors in patients with inhibitors at one center. **Materials and Methods:** 6 patients with 7 pseudotumors were treated. All the patients have severe hemophillia A. The mean age was 21.4 years (13-60). 1 patient presented soft tissue pseudotumors in the arm. The most frequent location of the pseudotumors was the femur (3), tibia (2), and calcaneus (1). 2 patients had a pathological fracture, one in the femur and the other one in the tibia. 1 patient had a pseudotumor of the femur and calcaneus on the same lower limb at the same time. In this patient, both pseudotumors were embolized before surgery to control bleeding. All patients with true pseudotumors were treated surgically except the one with the soft tissue pseudotumor who was under conservative treatment. The bypass agent used in all patients was rFactor VIIa. **Results:** 5 true pseudotumors recovered with surgical treatment. 1 patient who had a large femoral pseudotumor with a pathological fracture died of sepsis secondary to necrotizing fasciitis. The patient with the soft tissue pseudotumor healed with conservative treatment. **Conclusions:** Our results show that pseudotumors in patients with inhibitors could be treated with surgery and a bypass agent.

P2.05

Physiotherapy Treatment in Patients with Hemophilia and Chronic Ankle Arthropathy: Experimental Study

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Objective: This article tries to evaluate the effectiveness of two programs about physiotherapy in patients with hemophilia A and B, adults and with arthropathy of the ankle (figure 1), without previous surgery and also without the parameters that can interfere in improvement. **Methods:** 9 patients took part in this double-blind study, who randomly received treatment composed of physiotherapy and medication with concentrates of FVIII and FIX, over six weeks, two times per week. They were evaluated before and after the intervention, and after 6 months range of motion, pain perception, quality of life, and stability of the lower limbs was evaluated (figure 2). **Results:** It was noticed the effectiveness of both of the interventions ($p < 0.05$) in eleven and five of the studied variables, of the first and second group respectively. Physiotherapy treatments using thermotherapy, mobilization and passive stretching of the ankle, proprioception and cryotherapy, and manual orthopaedic therapy, were effective in the improvement of arthropathy related to the studied sample. After six months,





Abstracts (cont'd)

physiotherapy interventions were effective, obtaining improvement in pain perception and the reversal of both ankles, left ankle eversion, and mental health. **Conclusions:** It is necessary to mix both of the physiotherapy models, with a higher sample, to determinate effectiveness and to delay surgery as the last therapeutic option.

Keywords: ankle, physiotherapy

P2.06

Postsurgical Mobility of Total Knee Replacement in Patients with Hemophilia

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Introduction: We have analysed 8 international publications and the median range of motion (ROM) post surgery is 22°. **Objective:** To evaluate the range of post-surgical motion in patients with TKA in our treatment center. **Materials and Methods:** From 2001 to 2009, 19 TKAs were performed on 12 patients with severe hemophilia A, 7 bilateral and 5 unilateral. Mean age at arthroplasty was 47.6 years, mean follow-up was 3.5 years (range 1- 9). ROM was evaluated pre-operative, post-operative at 6 months, in one year, and in 2010. All patients received physiotherapy, including postural treatment. **Results:** The mean knee flexion improved from 67.6° pre-operatively to 84.7° at the final follow-up, with an improvement of 17.1°. The mean flexion contracture diminished from 24.5° to 8.4°, with an improvement of 16.1°. The mean total ROM increased from 43.2° pre-operative to 76.3° at the final follow-up, with an improvement of 33.1°. TKAs with total ROM pre-operative below 70° had an increased total post-operative ROM of 35°-55°, whereas those with total ROM of 70°-84° gained 25°-30°. Those with total ROM between 85°-100° remained without changes. One case with a loss of total ROM of 15° had a pre-operative ROM of 110°. ROM obtained in 6 months post-operative was maintained up to the final follow-up. **Conclusion:** TKA with the techniques applied obtains acceptable mobility, allowing patients to improve their functionality.

Key Words: ROM (range of motion) , TKA (total knee replacement)

P2.07

Physical Therapy Management of Chronic Ankle Synovitis: A Literature Review

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Hemophilic arthropathy secondary to recurrent hemarthroses and chronic synovitis is a major orthopedic complication in hemophilia. The ankle is a commonly involved joint, trailing behind the knee and ankle, and is a frequent cause of functional limitation and disability in individuals with hemophilia. Physical therapy is commonly prescribed for conservative management of chronic ankle synovitis. The goal of rehabilitation is to help prevent the progression of chronic synovitis to end-stage arthropathy and to maximize the function and quality of life of patients. This article reviews the musculoskeletal impairments concomitant with chronic ankle synovitis and outlines the current treatment approach, along with the evidence surrounding rehabilitation of this specific condition. Multiple databases including PUBMED, COCHRANE LIBRARY, MEDLINE, and CINHALL were utilized to obtain relevant studies published between 1995 and 2010. Current physical therapy interventions to address impairments associated with chronic ankle synovitis are comprised of manual therapy, cryotherapy, and hydrotherapy, strengthening and stretching exercises, and balance and proprioceptive training. With the exception of manual therapy, these interventions, in conjunction with the use of Factor VIII replacement therapy, have been shown in the existing literature to improve joint function, minimize bleeding episodes, and prevent further deterioration in the knee and elbow. There is a paucity of research investigating the effects of conventional interventions on chronic ankle synovitis in the hemophilic population. More research needs to be conducted in this area to clearly delineate the most effective physical therapy treatments to address impairments and functional limitations and prevent the development of hemophilic ankle arthropathy.

Keywords: chronic ankle synovitis, physical therapy

P2.08

Vertical Ground Reaction Forces during Hemophilic Arthropathy Gait: A Case Study

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Introduction: Hemophilic arthropathy (HA) is the clinical manifestation with the greatest morbidity in hemophilic patients. Furthermore, this degenerative disease in the joint produces disturbances in motor control (e.g. balance, proprioception). The aim of our study was to compare the variables related to vertical ground reaction force (VGRF) depending on the arthropathic severity in the supporting





Abstracts (cont'd)

leg. **Methods:** One patient, suffering from hemophilia A with HA, volunteered. The reaction force signals were acquired with a force plate NedSVE/iBV (Institute of Biomechanics of Valencia, Valencia, Spain) at 100Hz frequency. Non-parametric tests were applied (U de Mann Whitney) for independent samples to determine if there were significant differences between both legs. **Results:** The landing force (N), the take-off force (N), the take-off ratio (N.s-1), and the stance time were greater in the leg with less arthropathic affectation ($p < 0.05$). On the other hand, for the landing ratio (N.s-1) the values were significantly higher in the leg with less articular affectation ($p < 0.05$). **Conclusion:** VGRF produced during walking are affected by the HA, this fact shows the importance of gait pattern re-education in these patients.

P2.09

Kinetic and Kinematic Analysis of the Ankle Joint: A comparative Analysis between Children with Hemophilia and Normal Children

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Hemophilia is an inherited, congenital life-long blood disease. Children with hemophilia are vulnerable to a high percentage of musculoskeletal lesions. These lesions predominantly affect lower limbs, influencing postural control, standing, and walking. The purpose of this study was to evaluate sagittal kinetic and kinematic changes of ankle joint during stance phase of gait. 100 boys with a mean age 10.560 ± 1.110 years participated in this study. The study group examined 50 children with hemophilia and the control group included 50 normal volunteers. The two groups were evaluated using a 3D motion analysis system, the Qualysis motion capture system which has 3 Pro-Reflex 120 infra-red cameras, with Q-trac, Q-capture, and Q-view. The Q-gait software was used to capture and analyze data. Ground reaction was also collected using a force plate unit. Data were compared between the two groups using unpaired student t test. Children with hemophilia were found to be significantly different in all parameters of joint angles, and moments during initial contact, loading response, mid-stance, terminal stance, and pre-swing phase. These results suggested an alteration in range of motion and decrease in all forces around ankle joint which affect dorsi- and plantar-flexor movements. In addition children with hemophilia were found to have joint changes, and alteration of proprioception and pain.

Key word: ankle joint, hemophilia

P2.10

Spontaneous Iliopsoas Hematoma in Patients with von Willebrand Disease and Hemophilia A

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Introduction: Iliopsoas hemorrhage can be developed in individuals with bleeding disorders such as hemophilia A and von Willebrand disease (VWD). This is a review of two cases of severe hemophilia A and type 3 VWD with spontaneous iliopsoas hematoma from south-west Iran.

Case Presentation: **Case1:** A 26-year-old male Iranian with severe hemophilia A had right groin pain. No history of trauma. Pelvic ultrasonography showed a small echo-free lesion on the right iliopsoas muscle. With impression of iliopsoas hemorrhage he was given recombinant factor VIII at a dose of 50 IU/Kg body weight, twice daily. **Case 2:** A 20-year-old male Iranian with Type 3 VWD had onset of right flank pain. Upon arrival, there was limited mobility of the hip without antecedents of trauma. The patient exhibited an anti-pain posture and held his right leg flexed in the hip joint (Figure 2). MRI showed a mass in right iliopsoas muscle (Figure3). An initial clinical diagnosis of an iliopsoas hematoma and partial femoral nerve palsy was made. He was given Humate-P at a dose of 40 IU/Kg body weight twice daily. **Discussion:** Iliopsoas hemorrhage is encountered in a variety of coagulation disorders such as hemophilia A, VWD, rare bleeding disorders, and those using anti-coagulant drugs as Heparin, Warfarin, and Ticlopidine, but most iliopsoas hemorrhages occur in hemophilia patients. The first case was explained by Tallroth in 1939. Iliopsoas hemorrhage is a potentially serious complication because delay in starting treatment may result in permanent femoral nerve palsy. Diagnosis of retroperitoneal hematoma is obtained by ultrasonography, computed tomography scan and MRI. The treatment of iliopsoas muscle hemorrhage remains controversial. Surgical decompression and a medical approach are recommended. In conclusion, these cases with spontaneous iliopsoas hematoma show a good response and outcome with medical intervention.

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Abstracts (cont'd)

P2.11

Successful Conservative Management of Hemophilic Pseudotumour: A Case Report

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Objective: To highlight conservative management of hemophilic pseudotumor with long-term replacement therapy with factor concentrate. **Method:** Retrospective review of patient chart. **Results:** A 14-year-old native American male with moderate hemophilia B (baseline factor (F) IX level 2%) presented to the emergency department with severe left gluteal and thigh pain following injury. X-rays of the left hip were normal. He received 70-80units/kg of FIX concentrates (dosed to the closest vial) daily for 5 days. A computerized tomography (CT) scan obtained for worsening pain showed left pelvic fluid collection and possible pseudoaneurysm. An Ultrasound (US) and Magnetic Resonance Imaging (MRI) revealed a 12.7 cm x 6.2 cm x 7.9 cm hematoma with no evidence of a pseudoaneurysm. Following a central venous access device (port) placement, the patient received daily FIX concentrates for a period of 2 months followed by infusions every other day for 6 months, and then weekly infusions for a period of 1 month. Serial follow-up CT showed interval resolution of the hematoma. At the end of 9 months, US and MRI showed complete resolution of the hematoma. Patient was able to attend school throughout the duration of treatment. **Relevance to hemophilia care:** Early institution of replacement therapy followed by long-term prophylaxis is effective in the management of hemophilic pseudotumors. The cost/benefit of interventional treatment versus replacement therapy, and the risk of recurrence should be taken into consideration when treating a hemophilic pseudotumor.

Key words: Hemophilia B and pseudotumour

P2.12

Joint Hyperextensibility in von Willebrand Disease (VWD): A report of Six Cases

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Objective: To raise awareness of the occurrence of joint hyperextension in VWD, that may mask joint-range-of-motion loss due to bleeding. **Methods:** Retrospective review of charts from a single Hemophilia Treatment Center. **Results:** In 6 female cases of VWD (5 from a single family), all had moderately severe type 1 or type 2 VWD. Joint hyperextension of elbows and knees were seen in all

as compared to age-appropriate normal values. Joint range-of-motion (JROM) of other was within normal limits. None of the affected individuals had joint hypermobility or hyperelasticity of the skin; genotyping for Ehlers-Danlos syndrome was not obtained. All were non-responsive to Desmopressin and required von Willebrand factor (VWF) concentrates for treatment of bleeding episodes. Other members of the family with VWD had normal JROM. There were also members in these families who were not affected by VWD. **Relevance to hemophilia care:** The above cases demonstrate the occurrence of joint hyperextension in family members with VWD. All cases experienced bleeding episodes of varying severity; however none had experienced an overt joint bleed. Unlike hypermobility, which results from a joint abnormality, hyperextension is the stretching of a body part beyond what is normal, although sometimes the terminologies are used interchangeably. To the best of our knowledge the occurrence of joint hyperextensibility in VWD has not been reported. Skeith et al described that baseline joint hypermobility in hemophilia may be a risk factor for chronic arthropathy. This paper highlights the importance of obtaining JROM in patients with VWD.

Key words: Von Willebrand Disease, Joints

P2.13

Management of Ankle Pain in an Adolescent with Severe Hemophilia A: Using Orthotics and Rehabilitation to Improve Outcomes

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Objectives: To highlight the importance of physiotherapy intervention in the management of ankle pain; outcomes measures that are clinically and financially valuable in patient management; the value of patient completed treatment records and the monitoring of them.

Introduction/Report: Patient A (SHA, prophylaxis) at his routine review appointment complained of recurrent left ankle pain, denying any injury or trauma. The pain worsened on weight-bearing activity and he had difficulty hopping and running. Extra FVIII had only mild effect. Physiotherapy assessment identified no swelling, bilateral pes planus and a mild laxity of the talocrural joint. Gait analysis highlighted poor heel strike and foot control with global muscle weakness. The Beighton score was 5/9 and the Haemophilia Joint Health Score (HJHS) was 7. The 'Treatment Direction Test' (TDT) (lessen pronation with strapping) improved pain on assessment. Clinical impression was pain from pes planus and associated muscle weakness. Treatment consisted of orthotics provision, lower limb strengthening, and education.

Outcomes: Treatment record review for 12 months prior





Abstracts (cont'd)

to physiotherapy highlighted 16 excess treatments for ankle pain, in the 10 months following input there were only 4 extra. The cost difference in FVIII use was £11,600. The HJHS reduced to 0, muscle strength significantly improved, and he was pain free. **Discussion and Clinical Relevance:** The cause of ankle pain in this instance was not related to bleeding or hemarthropathy, although arguably if it continued, movement compensations may have led to a bleeding episode. Application of the TDT refined the assessment and reasoning behind orthotic provision. Although the actual biomechanical mechanism by which orthotics have their effect remains inconclusive, they clearly had a positive effect. They should not however be considered a universal treatment, but as a targeted intervention alongside strengthening.

Key words: ankle pain; orthotics

P2.14 Physiotherapy and Synoviorthesis in Hemophilic Patients

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Introduction: The aim of this study is to describe a protocol of physiotherapy for functional recovery after radiosynoviorthesis in severe hemophilic patients.

Methods: Isotopic synoviorthesis in knees were performed with Itrium90, using an average dose of 4-5 mCi. In elbows and ankles we used Renio86 with an average dose of 3 mCi. The dose of replacement factor concentrate used was 25-30 IU/kg of FVIII or FIX before the procedure and after 24 hours, followed by two doses every 48 hours. Patients with inhibitors were treated with bypassing agents. The protocol of physiotherapy after radiosynoviorthesis consisted of: 1) During the first 24 hours: joint immobilization, rest and isometric exercises followed by 10 minutes of cryotherapy. 2) Days 2-7: isometric exercises, active and strength exercises, performed 3 to 5 times a day, followed by cryotherapy. We also allowed a progressive increase in daily activities, using crutches for those patients with lower limbs involved (knees or ankles). 3) During the next 6 months, patients followed individualized recommendations with the aim of reducing swollen joints, avoiding re-bleeding during specific exercises and daily activities, maintaining or improving muscular strength and trophism and range of movement. **Results:** Between 2004 and 2010 we performed 122 radiosynoviorthesis (60 knees, 35 elbows, and 26 ankles) in severe hemophilic patients. There were no re-bleedings due to early physiotherapy after

radiosynoviorthesis in our group of patients. **Conclusion:** An early physiotherapy programme for severe hemophilic patients, with the appropriate replacement coverage, allows rapid and adequate functional recovery after radiosynoviorthesis.

Key Words: Physiotherapy, radiosynoviorthesis

P2.15 Improving Quality of Life in Women with von Willebrand Disease

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Objective of the study: This study was conducted to estimate the effect of aerobic exercises on quality of life in women suffering from bleeding symptoms and menorrhagia due to VWD. **Study design:** 30 women suffering from type 1 von Willebrand disease participated in this study. All patients were divided into two groups; each group consists of 15 patients. Group (A) patients were evaluated by blood test (Ristocetin Cofactor Assay), Congenital Rare Bleeding Disorder Questionnaire, cardiopulmonary exercise tests (Bruce Protocol), and SF-36 QoL Questionnaire, before participation in the study and after three consecutive months. Patients in group B were enrolled in the exercise program using a motor driven treadmill for about three months, and were submitted to the same methods of evaluation as group A. **Results:** The results of this study showed negative findings in VWF, VO2max, VE, limitations of activities, physical functioning problems, general health, bodily pain and bleeding score for patients in group A. The results for patients in group B showed a statistically highly significant increase ($P < 0.01$) in VWF, VO2max, VE, decreased limitations of activities, physical functioning problems, general health and a statistically highly significant decrease ($P < 0.02$) in bodily pain (as a domains for QoL Questionnaire), and highly significant decrease ($P < 0.01$) in bleeding score after the end of three consecutive months of walking treatment program which reflect improvement in the QoL. **Conclusion:** The results of this study concluded that walking exercises are excellent conservative treatment for von Willebrand disease through improving the aerobic capacity which improves the quality of life for such cases.

Key words: VWD, aerobic exercises





Abstracts (cont'd)

P2.16

Improving the Number of Patients with Severe Hemophilia Undergoing a Physiotherapy Annual Review

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Introduction: All patients with severe hemophilia should undergo a formal physiotherapy annual review (AR) (Haemophilia Chartered Physiotherapist Association (U.K.), 2002). Aims were to measure the percentage of patients reviewed in 2009 against the target of 100%, to identify reasons for any shortfall and instigate changes to reduce it. The number of reviews performed in 2010 would be monitored to assess the impact of changes made. Another aim was to record the outcome of the reviews. **Methods:** A retrospective review of the physiotherapy notes of patients with severe hemophilia registered at the NCHCD was used to identify those that attended for AR in 2009. The outcome of the review was also recorded. The NCHCD's electronic patient record was used to establish patient's attendance record at the hemophilia clinics. Descriptive statistics were used to present the findings to the multidisciplinary team for discussion. **Results:** Barriers were identified as lack of time, lack of space, other demands on the service, as well as the poor attendance record of some patients. Changes agreed to address these barriers, such as dedicated physiotherapy space and time being available during hemophilia clinics, resulted in a higher percentage of patients being reviewed in 2010. A need for further intervention was identified in 48% of patients reviewed in 2010. **Conclusion:** With almost half the patients who had physiotherapy AR needing further in-put, maximising efforts to ensure patients are reviewed each year is important.

