

ABSTRACTS

CHALLENGES OF HEMOPHILIA AND MUSCULOSKELETAL ISSUES DURING COVID19

FP-T-01.01 | COVID-19 and its impact on the pediatric bleeding disorders population: Soaring BMIS

Deirdre Tetzlaff; Caroline Malcolmson; Manuel Carcao; Cindy Wakefield; Vanessa Bouskill
The Hospital for Sick Children, Toronto, ON, Canada

Introduction: The COVID-19 pandemic has resulted in children being less active and spending more time at home with easy access to food. We wanted to evaluate the impact of this on weight/BMI in children with severe bleeding disorders on prophylaxis. We retrospectively reviewed the charts of patients with severe bleeding disorders on prophylaxis treated at SickKids (Toronto) from March 01, 2020 - March 01, 2021. We evaluated the following pre- and post-COVID variables: weight (kg), weight percentile, BMI (kg/m²), BMI percentile, HJHS score, and prophylactic dosing (units/kg). We accepted any variables taken within 12 months pre-March 1, 2020 to any time at least 6 months post.

Methods: Eligible patients were 3–18 years of age with a severe bleeding disorder on prophylaxis.

Results: Ninety-eight patients (95 males:3 females) were included in the final analysis. Baseline diagnoses were: 71 hemophilia A; 12 hemophilia B; 9 Type 3 von Willebrand disease; 3 FVII deficiency; 1 FXIII deficiency; 1 FX deficiency; and 1 with afibrinogenemia. Median age at the time of baseline assessment was 11.9 years (IQR 8.13-15.47 years). Median interval time from pre-COVID measurements to latest follow-up was 10.0 months (IQR 7.0-12.0 months) during which patients gained a median of 4.6 kg and increased 1.72 weight centiles (IQR: -2.2325 - +5.7125). Patient BMI increased a median of 1.36 kg/m² and percentile BMI increased by 1.75 centiles. The group that gained the most weight centiles, BMI and BMI centiles were 6–12 years old, closely followed by the 12–18 year-old group. Median HJHS score (6) was the same pre- and post-COVID-19 although data was more limited.

Conclusions: There was a trend to weight gain over the study period. The lack of a worsening of HJHS scores in the short-term could be secondary to children having been less active, however, overtime this may lead to worse long-term joint outcomes. Additionally, there may be increased healthcare costs associated with weight gain for children as prophylaxis is dosed according to patient weight. We will follow these patients to see if these trends continue/reverse with hopefully the end of the pandemic.

P-002 (1018237) | Promotion of physical activity in haemophilia patients: Hemo-olympics online

Ana Chimeno-Hernández¹; Pilar Alberola²; Manuel De diego²; Luis Vaño²

¹Physiotherapy department, University of Valencia, Spain; ²Association of haemophilia in the Valencian Community (ASHECOVA), Spain

Introduction: A low percentage of people with haemophilia (PWH) perform physical activity (PA), despite the benefits of their practice. Thus arises the need to promote the PA in this population. To this end, the aim of this study was to develop an online Hemo-Olympics to encourage the practice of PA in PWH.

Methods: In 2020, two one-week Hemo-Olympics (HO) were held, organized by the Haemophilia Association of Valencia Community (ASHECOVA). A month before the HO, the event was publicised via social media. Participants were sent a video explaining: how this event works (record for 1 minute doing the tailored exercises indicated each day -6 strength and 1 balance exercises- and sent to the organizers), the points to be taken into account in the competition (Movement Control, Repetitions and Animation in the video) and the different categories of the awards. The data were collected through surveys with the Google Forms platform. The survey's questions have been divided into three groups: i) publicity of the event; ii) satisfaction with Olympics' development iii) adherence to the exercise.

Results: A total of 20 severe PWH A were recruited (1stHO:9, mean age (SD):35.2 (8.85) years; 2ndHO:11, age:32.09 (12.74) years). The data collected from both surveys will be presented together below: i) Level of satisfaction with the promotion of the Olympics was 9.89 out of 10. The main motivation for participating was "participating in association events" (88.89%), followed by "testing myself with training" (61.11%) and "by my fellow competitors" (39%). ii) The score of the explanation of the exercises was 9.56 out of 10, and 16 participants qualitatively assessed that they liked the exercises "very much" (88.89%). No serious adverse effects were reported during exercise. iii) Adherence to the exercises was 96.77%. After the Olympics, 88.89% of participants said they wanted to continue exercising, 72.22% of participants noted benefits in their musculoskeletal health and 61.54% dared to do activities they previously thought they could not do.

Conclusions: The online Hemo-Olympics is a fun, accessible and safe way to motivate and encourage exercise in PWH, contributing to increased adherence to physical activity.

P-001 (1036109) | Comprehensive distance treatment focused on physical rehabilitation in patients with hemophilia in Mexico

Francisco González Martínez and Cecilia Gallardo Pérez
Federación de Hemofilia de la República Mexicana, Distrito Federal, Mexico

Introduction: Physical rehabilitation plays a fundamental role in patient's life, it promotes the prevention of arthropathies generated by constant joint bleeding, increasing muscle strength, reducing the risk of deformities and maintain social well-being. Due to actual health contingency, it was not possible for patients to attend their appointments, so they adopted a sedentary lifestyle, as consequence, increase risk of muscular atrophy, lose of joint stability, bleeding and irreversible damage.

Methods: Understanding the need for comprehensive treatment in order to opt for a better quality of life, social worker, psychologists and the physical rehabilitation department, made a survey at Mexican Republic level, interviewing engaged patients to their condition and their treatment to determine in accordance to the results and answers- a remote method to keep monitoring patient's physical activity. For 9 months, a treatment scheme was carried out, beginning with the evaluation of the rehabilitation physician, followed by the execution of the individualized exercise program by the physiotherapist. The follow-up was carried out weekly with video chat and instant messaging applications. The Federation of Hemophilia of the Mexican Republic granted doses of Antihemophilic Factor to support patient's treatment at home and not have bleeding due to physical activity. As a complimentary and self-control, patients did their own perimetry.

Results: During these nine months, we held almost 1000 video-calls between the 52 patients, having as results, notable improvements on the physical and social areas: Independence from wheelchairs or crutches, decreased hemarthrosis, increased muscle strength, increased ranges of motion, greater joint stability and decreased pain. Cost-benefit: the average monthly cost to attend the appointments in person is approximately 1720 MXN, compared to distance appointments, the cost is significantly reduced to 500MXN. Also effective therapy days increase from 2 to 4 times a week.

Conclusions: The commitment was greater while at home, the patient chose the moment and the time to dedicate to their routines; complementing the fact that the economic expense was significantly reduced. According to previous data, the project proves to be a high-efficiency and sustainable alternative for patients who cannot keep their appointments or for those whose home is very far from the HTC.

CLINICAL ASSESSMENT

FP-TH-01.03 | Utilization of point-of-care musculoskeletal ultrasound for management decisions in routine hemophilia care – focus on physical therapy

Nicolas Gallastegui¹; Christopher Hanacek¹; Rebecca Kruse-Jarres²; Bruno Steiner²; Doris Quon³; Cindy Bailey³; Annette von Drygalski⁴; Richard Barnes¹

¹Division of Hematology/Oncology, Department of Medicine, University of California San Diego, San Diego, CA; ²Washington Center for Bleeding Disorders, Seattle, WA; ³The Orthopaedic Hemophilia Treatment Center, Orthopaedic Institute for Children, Los Angeles, CA; ⁴Division of Hematology/Oncology, Department of Medicine, University of California San Diego, CA, USA

Introduction: Musculoskeletal ultrasound (MSKUS) for point-of-care (POC) evaluation of hemophilic arthropathy and acute painful events is thriving. However, the extent to which MSKUS influences medical decisions on routine hemophilia care is unknown.

Methods: A prospective, multi-center study evaluated the routine utilization of POC-MSKUS in adult patients with hemophilic arthropathy from May 2016 to April 2019 at three large hemophilia centers in the USA. Bilateral elbows, knees and ankles were assessed clinically [Hemophilia Joint Health Score (HJHS)] and with POC-MSKUS by Joint Tissue Activity and Damage Exam (JADE) protocol at baseline and ~annually thereafter (= 3 visits). Treatment decisions [(PT) and "medical" [hemostatic agents, interventions, referrals, pain medications] were recorded in relation to POC-MSKUS.

Results: Forty-four patients [median age 37 (IQR 29, 51), mostly with severe Hemophilia A on factor prophylaxis] completed 129 routine visits, yielding 792 joint exams by POC-MSKUS and HJHS [median at baseline 27 (IQR 18, 42)]. Among 157 management decisions, 70% were PT (n = 110), and 30% were "medical". Overall, 50% were informed by POC-MSKUS (Figure 1). New/Adjusted PT plans (n = 110 in 37/44 patients) were proposed during 73/129 (57%) visits. POC-MSKUS influenced 47/110 (43%) PT plans, mostly informing the treatment of specific arthropathic joints (45/47 plans). The median total HJHS of patients with plans informed by POC-MSKUS was higher compared to patients with plans based on physical examination only [median 34, (IQR 27, 51)] vs. [median 22; (IQR 18, 33)]. Of note, 20/47 (43%) POC-MSKUS plans included de-escalation strategies with lesser intensity, mostly for joint instability and subclinical hemarthroses. Generally, PT plans influenced by POC-MSKUS triggered more directed manual therapy/therapeutic exercises, while plans based on physical exam were more focused on global exercises and wellness. Specific

abnormalities identified by POC-MSKUS (osteophytes, impingement, inflammation, and other arthropathic changes) influenced 16/47 plans where MSKUS was used for decision-making.

Conclusions: Routine joint evaluations with POC-MSKUS resulted in few changes regarding medical management decisions but had a profound effect on PT plans. POC-MSKUS informed targeted strategies in arthropathic joints, often de-escalating previously more aggressive treatment plans. Studies are required to determine if POC-MSKUS-driven PT improves outcomes.

P-006 (998458) | Infectious complications in patients with hemophilia

Heghine Khachatryan; Nelli Sargsyan; Lusine Sahakyan
Hemophilia center of Armenia, Yerevan, Armenia

Introduction: Replacement therapy increases factor VIII plasma level, which temporarily corrects its deficiency in the blood plasma and prevents bleeding. Before the use of modern factors, treatment for patients was traditionally with plasma, which has never been virally inactivated. Most people with hemophilia who treated with fresh frozen plasma were infected with the HIV virus, Hepatitis C (HCV), Hepatitis B (HBV) and other blood-borne viruses. These cases of transmission of the viruses have stimulated development of the latest ultra sensitive diagnostic test system over the past 30 years and their introduction into clinical practice. Over the past 20 years, not a single case of transmission of the virus by blood components has been recorded, which confirms the safe use of our blood components. Currently, plasma is used as an alternative treatment option only in countries where there is a deficiency of factors.

Methods: The study was based on the registration data of the Armenian Hemophilia Center. Conducted a retrospective analysis of 271 patients (4.85% younger than 18 years of age) indicated the outpatient cards with HCV infection, HbsAg-in, HBcore-on in education. A prospective serological examination was carried out in the department of the Hematology center using an AWARENES analyzer. The data obtained were processed by statistical methods SPSS, χ^2 and Student.

Results: According to the data obtained, infectious complications were registered only in patients of the I group. Infectious complications were found in 71 (33%) of 212 examined adult patients. Hepatitis C prevailed - 62 cases (87%) among the observed infectious complications; hepatitis B was detected in 8 patients (11%); hepatitis B and C were combined in 1 case (2%). According to the data obtained, only two of II group patients (3.22%) had the development of inhibitors compared with 22 patients of group I (19.8%). Based on the examination results, 91.9% of patients in group I have joint arthropathies, while only 64.5% of patients in group II have joint arthropathies, respectively.

Conclusions: The use of RT excludes infection complications. The immunologic complications, the development of inhibitors, as well as articular complications in mild forms of diseases include a low percentage.

P-005 (1033563) | Hyaluronic- acid and platelet-rich plasma treatment for knee arthropathy

María Landro^{1,2}; Carla Daffunchio²; Guillermo Cambiaggi²; Gustavo Galatro^{1,2}; Horacio Caviglia^{1,2}

¹Hospital General de Agudos Juan A. Fernández; ²Ciudad Autonoma de Buenos Aires, Argentina

Introduction: Repeated joint bleeding leads to chronic synovitis, cartilage damage and bone alterations which result in haemophilic arthropathy and are associated with pain, functional impairment and poor quality of life. There is evidence of the use of hyaluronic acid (HA) and platelet-rich plasma (PRP) for the treatment of knee arthropathy in the literature but they have different mechanisms of action in the disease. We decided to use both components and compare the evolution of treatment. The aim of this study is to compare, the efficacy, safety and duration of a single intra-articular injection of PRP or PRP+HA for pain, bleeding episodes and the Hemophilia Joint Health Score, in the same patient with bilateral hemophilic knee arthropathy.

Methods: Twenty-one men with haemophilia and bilateral knee arthropathy (42 knee joints) were treated with intra-articular injections of PRP or PRP+HA. All of them were haemophilia type A severe. The mean age was 36.6 years (21-72). All patients were evaluated for: Hemophilia Joint Health Score (HJHS), pain (VAS), the number of bleeding episodes (BE) in the last 30 days, before treatment, at three and six months after treatment.

Results: Statistically significant improvement were shown for both knee joints at three and six months after treatment for VAS and BE ($P < .00001$). There were no statistical differences for VAS and BE at three- and six-months follow-up. The HJHS score did not significantly improve for either knee in the 6-month period after injection.

Conclusions: Both treatment (PRP and PRP+HA) are safe and effective in treating haemophilic arthropathy of the knee for up to 6 months follow-up, reducing pain, the number of bleeding episodes, and delaying total knee arthroplasty.

P-003 (1035583) | Diagnosis of hemophilic hemarthrosis: development of a patient-derived symptom assessment tool using musculoskeletal ultrasound (MSKUS) for validation

Srila Gopal¹; Richard Barnes¹; Lena Volland²; David Page³; Annette von Drygalski¹

¹Division of Hematology/Oncology, Department of Medicine, University of California San Diego and ²National Hemophilia Foundation, San Diego, California, USA; ³Canadian Hemophilia Society, St-Michel-de-Bellechasse, Quebec

Introduction: Hemophilia patients experience painful joint episodes which may or may not be associated with hemarthrosis. Point of care (POC) musculoskeletal ultrasound (MSKUS) is used increasingly to rapidly detect hemarthrosis and direct management. Patient-derived symptom questionnaires also have been proposed for diagnosing

hemarthrosis, but are not validated. We sought to validate a questionnaire developed by an experienced patient group, using POC MSKUS to confirm hemarthrosis.

Methods: The questionnaire was developed by Mr. Page and derived from an open-text survey, sent to a group of “experienced” individuals with severe hemophilia A or B. We administered this questionnaire comprising 20 questions (10 each associated with hemarthrosis and arthritis pain) to 32 adult hemophilia patients experiencing acute painful episodes and reporting to the Hemophilia Treatment Center (University of California San Diego). Questions were scrambled and patients and providers were blinded to question assignment. We confirmed the presence (or absence) of hemarthrosis using POC MSKUS [Joint Activity and Damage Exam (JADE)]. We fitted univariate and multivariate generalized estimating equations to identify symptoms associated with hemarthrosis.

Results: We evaluated 79 painful episodes (patient median age 38 years; range 21–74 years); 23 patients had hemophilia A. The majority of episodes occurred in patients with severe hemophilia ($n = 22$, 69%). POC MSKUS detected hemarthrosis in 36 (46%) episodes. The weakest predictor for pain associated with hemarthrosis was ‘no feeling of sponginess with movement’ (odds ratio [OR] .28 [CI .11; .75]); the strongest predictor was ‘like a balloon swelling with water’ (OR 2.84 [CI .75; 10.77]). Both symptoms fit with their original classification of arthritic pain or hemarthrosis pain. We identified 5 questions with the strongest OR for differentiating hemarthrosis pain from arthritic pain and developed a “yes/no” question/answer algorithm for hemarthrosis prediction. Answering these 5 questions in “yes/no” fashion yielded a probability of hemarthrosis ranging from 6% (more likely arthritic pain) to 93% (more likely hemarthrosis pain).

Conclusions: Objective diagnosis of hemarthrosis by MSKUS facilitated a prediction tool by informed selection of the most meaningful patient perceived indicators of arthritic versus hemarthrosis pain. The tool requires further validation and will be particularly helpful in situations where MSKUS is not readily available.

P-004 (1036638) | Haemophilia early arthropathy detection with ultrasound (head-us) for monitoring joint health status in children and adolescents with haemophilia on prophylaxis

Aikaterini Michalopoulou¹; Athina Dettoraki¹; Dimitris Sireggelas²; Dimitra Politi¹; Ioanna Stamati¹; Kaiti Margariti¹; Dimitra Venizelou²; Helen Pergantou¹

¹Haemophilia Centre/Haemostasis and Thrombosis Unit and ²Physiotherapy Department, Aghia Sophia Children's Hospital, Athens, Greece

Introduction: In recent years, imaging and follow-up of joints of haemophilia patients is feasible with Point-of-Care ultrasound (POCUS), implemented by trained operators. The aim of this study is to assess the utility of POCUS using the HEAD-US protocol performed by a physician of Haemophilia Comprehensive Care Center (HCCC), for the evaluation of joint status in children with haemophilia.

Methods: The HEAD-US for the bilateral examination of synovitis, cartilage or bone changes on 6 joints (elbows, knees, ankles) was performed from January to December 2020, by the same trained physician in children and adolescents aged 6–18 years with haemophilia A and B on prophylaxis, followed in a single Greek HCCC. In almost all cases, HJHS 2.1 (inflammation, duration, atrophy, crepitation, flexion deficit, extension deficit, pain, strength, gait) was performed simultaneously by a physiotherapist and relationships between the two scales were analyzed using the chi-square statistic.

Results: A cohort of 53 patients (mean age: 13 ± 3.5 years, 47 haemophilia A, 40 severe haemophilia A) were included and 314 joints (102 elbows, 106 knees and 106 ankles) were examined. Thirty per cent of the HEAD-US assessments yielded a score of zero. In cases of pathologic findings the most frequent lesion was mild synovitis, followed by cartilage derangement and bone involvement. There were 54 concurrent measurements of HJHS and HEAD-US. There was a discordance between the total scores of the two measurements ($p = .028$), which is largely explained by the following: In 33 cases the total HJHS score was zero but this was confirmed by the total HEAD-US measurement in only nine cases. In the other 24 measurement the HEAD-US score ranged from 1 up to 8. Most discrepancies occurred at the ankle measurements and less at the knee measurements, while the elbow measurements had a fairly good agreement.

Conclusions: The HEAD-US in combination with clinical examination (HJHS) can unmask early joint alterations. However, both assessment tools appear to provide complementary data on joint health and contribute to a better understanding of early anatomical changes of joint disease in children with haemophilia.

CLINICAL CASES

P-007 (1036223) | Myositis ossificans in haemophilia: A rare case of post-traumatic heterotopic ossification at the hip

Richard Davis¹; Joanna Farrant¹; Mary Mathias²; Haroon Mann¹; Paul McLaughlin¹

¹Katharine Dormandy Haemophilia Centre, Royal Free Hospital and ²Great Ormond St Hospital, London, United Kingdom

Introduction: Modern treatment permits many young men with haemophilia to participate safely in exercise activities such as soccer. Whilst the use of regular prophylaxis minimises the risk of spontaneous musculoskeletal bleeding in people with haemophilia (PWH), bleeding secondary to trauma or injury remains a hazard. Severe muscle bleeds associated with sporting injury can present difficulties with haematological management and adequate tissue healing.

Methods: ‘John’ is a 25 year old man with severe haemophilia on prophylaxis. He presented to the Emergency Department complaining of right groin pain following a forced hip abduction injury sustained playing football. In the hours following injury, he was still able to walk but groin pain increased and he had significantly limited hip flexion. He was admitted on bed rest for enhanced haematological and pain

management. MRI confirmed a haematoma in the right Quadratus Femoris muscle (deep hip lateral rotator and adductor). Medical management in the subsequent weeks focused on intensive factor replacement, pain management and physiotherapy. Interval MRI six weeks later showed ossification within the haematoma. Clinically whilst walking was pain free, there remained apprehension and limitation of hip flexion and rotation.

Results: He was lost to follow up for four months. He returned for review complaining of increasing pain and functional impairment. A graded rehabilitation plan was implemented and follow up MRI requested. At this point a national lockdown took place associated with the COVID-19 pandemic, delaying his MRI and rehabilitation. A repeat MRI and CT 10 months later confirmed evolving myositis ossificans and mature calcification in the muscle, with bridging bone extending to the greater trochanter. Clinically, symptoms of limited hip flexion and occasional discomfort in weightbearing positions remained but were not worsening. He remains under review by the Orthopaedic team, where surgical excision is being considered.

Conclusions: Myositis ossificans is usually associated with contusion injury to large muscles of the lower limb and is rarely reported in haemophilia. This case highlights the potential complexity of apparently benign sports injuries in young men with haemophilia, and the need to better understand the medical, rehabilitation and psychological management as well as pathophysiology of this condition

IMAGING

P-010 (1014259) | MRI T1/T2 Mapping for accurate non-invasive detection of minor joint bleeding

Flora van Leeuwen¹; Beatrice Lena²; Lize van Vulpen³; Kathelijn Fischer³; Clemens Bos²; Wouter Foppen¹

¹Department of Radiology; ²Image Science Institute; ³Van Creveld Clinic, University Medical Center Utrecht, The Netherlands

Introduction: Severe haemophilia is characterized by repeated joint bleeding. The clinical symptoms of joint bleeds and non-bleeding/arthropathy related symptoms overlap and a non-invasive reference standard for differentiation between simple and haemorrhagic effusion is lacking. Although Magnetic Resonance Imaging (MRI) is considered the reference standard for musculoskeletal imaging, standard MRI protocols are not sensitive to detect low concentrations of intra-articular blood. Since blood and synovial fluid have different T1/T2 relaxation properties, quantitative evaluation by T1/T2 mapping might be used for differentiation. This phantom study investigated T1/T2 mapping for detection of low concentrations of blood in synovial fluid.

Methods: Pooled synovial fluid residuals from patients with inflammatory joint diseases and deoxygenated heparinised blood from a healthy male volunteer were mixed into different synovial fluid-blood ratios. Samples with blood concentrations of 0, 2.5, 5, 10, 25, 50, 75 and 100% were scanned at a constant temperature of 37°C using 1.5, 3 and 7

Tesla MRI scanners. Multislice T1/T2 mapping protocols, with acquisition times under 6 minutes, were used to quantify the samples' T1/T2 relaxation times with standard deviations. The lowest blood concentration of which the relaxation times could significantly be differentiated from synovial fluid was determined at all field strengths.

Results: The samples' T1/T2 relaxation times showed an inverse relation with the blood concentration, making discrimination of synovial fluid and different synovial fluid-blood concentrations possible. The lowest blood concentrations that could accurately be detected using T2 mapping were 50% at 1.5 Tesla, 5% at 3 Tesla and 25% at 7 Tesla. For T1 mapping the detection limits were 10% at 1.5 Tesla, 25% at 3 Tesla and 50% at 7 Tesla.

Conclusions: These in vitro results indicate the use of T1/T2 mapping for differentiation of simple and haemorrhagic joint effusion. At clinically available field strengths, low blood concentrations could be detected with these methods: 10% using T1 mapping at 1.5 Tesla and 5% using T2 mapping at 3 Tesla. The short acquisition times of the multislice protocols promote use in clinical settings. After validation in clinical studies, T1/T2 mapping may be used for accurate non-invasive detection of low intra-articular blood concentrations.

P-008 (1033585) | Comparison of two trained ultrasound specialists in the detection of musculoskeletal joint bleeding

Carla Daffunchio²; Gustavo Galatro²; Guillermo Margulies²; Gustavo Perez Ballester²; María Landro^{1,2}; Horacio Caviglia^{1,2}

¹Hospital General de Agudos Juan A. Fernández and ²Ciudad Autónoma de Buenos Aires, Argentina

Introduction: Detecting subclinical intra-articular bleeds is a big challenge to provide an adequate coagulation clotting factor management and to prevent joint damage. Repeated bleeding episodes may lead to degenerative arthropathy. Ultrasound (US) has been proven capable of detecting and quantifying the most relevant biomarkers of disease activity (joint effusion and synovial hypertrophy) and degenerative damages (osteo-chondral changes). US provides objective and dynamic information about articular status and has risen as an important tool for haemophilic joint evaluation and follow up. The aim of this work is to compare the minimum pathological volumes detected by a musculoskeletal ultrasound specialist vs a general ultrasound specialist in ankle, elbow and knee joints.

Methods: Ten corpses were studied, in two different studies. A total of 60 joints were studied (elbow, knee, ankle). The fluids were injected intra-articularly at 1 cm at a time, under ultrasound control. A liquid similar to synovial fluid (LSSF) was injected on the right side and blood from the blood bank on the left side respectively until achieving the visualization of a pathologic image (HEAD-US grade 1) in the US image. In the ankle the US was placed in the anterior window, between the anterior tibial muscle and the hallux extensors muscle. The knee was located superior to the kneecap and the elbow in the rear window on

the olecranon. Each study was evaluated by a different ultrasound specialist. One of them is a general ultrasound specialist, and the other is a musculoskeletal ultrasound specialist.

Results: That the minimum intra-articular volume of blood to reach HEAD-US grade 1 by ultrasound imaging in elbow joints was 4 ml, in ankle joints was 2 ml and in knee joints was 7 ml, with the general ultrasound specialist. The minimum intra-articular volume of blood to reach HEAD-US grade 1 by ultrasound imaging in elbow joints was 2 ml, in ankle joints was 2 ml and in knee joints was 7 ml, with the musculoskeletal ultrasound specialist.

Conclusions: Results were the same in knee and ankle joints, the elbow joint has an increased difficulty for specialist not used to evaluating joints.

P-009 (1036285) | Intra or extra-articular bleeding?:

Musculoskeletal ultrasound support in children haemorrhagic coagulopathies

A. Patricia Nally; Francisco Nally; Gabriela Sliba; Gisella Dolabella; Fabian Morales; Elsa Barahona; Monica Martinez
Fundacion de la Hemofilia MdQ, Buenos Aires, Argentina

Introduction: Clinical evaluation cannot always show whether the bleeding is intra-articular or extra-articular, Pain and physical limitation are not enough to determine whether the bleeding is within the joint or not. Even less in children. As this differentiation is so important, we highlight the usefulness of Musculoskeletal Ultrasound (MSKUS) to decide following steps.

Methods: MSKUS was performed in 15 patients with bleeding disorders, all suspected of acute bleeding. Patients with hemophilia A, B and factor V deficiency (2-14 years old) were included. Depending on the complexity of the lesion, an average of 3 to 5 studies per patient were performed between initial diagnosis and follow-up until resolution. A total of 328 evaluations were carried out in order to determine if effusions were intra or extra-articular, monitor their evolution and resolution, looking forward to eventual complications, including Haemophilic Pseudo-tumor and Miositis Ossificans. All studies were performed by the same operator, a pediatric radiologist specially trained in MSK ultrasound.

Results: Half of the patients demonstrated intra-articular hemorrhage, with varying degrees of underlying cartilage damage and bone involvement, including chondroepiphyseal degenerative changes. Remaining 50% patients presented accumulations of extra-articular hemorrhage in muscles, tendons and subcutaneous tissue as well as bleeding collections surrounding these structures (example). Special attention should be paid to lesions close to the bone in the follow-up of pseudo-tumor.

Conclusions: It is important to routinely evaluate children with bleeding disorders, with systematic MSKUS, which gives us certainty of location, risk of joint damage and evolution over time. Musculoskeletal bleeding has an extra-articular location in 50 percent of our patients, with no risk of direct joint damage, emphasizing the usefulness of

adjunctive kinetic treatment. The opposite will occur if there is intra-articular hemorrhagic occupation, where the follow-up will be more rigorous and the response to the factor will be assessed, considering the possibility of eventual evacuation.

ORTHOPEDIC ISSUES

FP-TH-01.01 | First experiences of major orthopaedic surgery in subjects with inhibitors with the association of emicizumab and rFVIIa

Christian Carulli¹; Giancarlo Castaman²; Domenico Prisco³; Massimo Innocenti¹

¹Orthopaedic Clinic, University of Florence; ²Center for Bleeding Disorders and Coagulation, Careggi University Hospital; and ³Interdisciplinary Internal Medicine, University of Florence, Toscana, Italy

Introduction: Persons with Haemophilia (PWH) and inhibitors needing surgery have been effectively treated for a long time by the use of activated prothrombin complex concentrates (aPCC) or recombinant activated FVII (rFVIIa). Emicizumab is a brand new molecule introduced for the haematological prophylaxis of PWH and inhibitors: brilliant clinical outcomes have been obtained. Recently, isolated experiences of surgery in such subjects have been performed with this drug, but very few major surgeries have been reported. The present is the first report of a consistent series of PWH and inhibitors undergoing major orthopaedic surgery conducted by a protocol with emicizumab and rFVIIa at a single Haemophilia centre.

Methods: Between 2018 and 2020, three PWH and high titer inhibitors underwent 5 major orthopaedic surgeries: one above-the-knee amputation and a Total Knee Arthroplasty (TKA) in a 56-years old subject; a Total Hip Arthroplasty (THA) in a 59-years old patient; a partial revision knee arthroplasty (rTKA) and an acetabular revision on a failed THA in a 49-years old subject. All previous surgeries were managed by rFVIIa prophylaxis. Visual Analogic Scale (VAS), Haemophilic Joint Health Score (HJHS), and a postoperative radiologic study were adopted for the evaluation. All subjects were managed by a regimen of weekly emicizumab and bolus infusions of rFVIIa for two weeks.

Results: All patients were successfully treated by a single surgeon, without any complications in the perioperative period and at the mean follow-up of 18.1 months (range: 7–28). An effective bleeding control was confirmed during and after surgery. Drains were used only for the above-the-knee amputation for 36 hours. No adverse event was recorded. All patients were regularly discharged and rehabilitated at the same hospital. The mean hospital stay was 13.0 days (range: 12–15). All patients reported satisfaction, pain reduction, and improved joint function at VAS and HJHS.

Conclusions: Major orthopaedic surgery with emicizumab and rFVIIa in PWH and inhibitors has been efficaciously performed with brilliant clinical outcomes and effective bleeding control. This experience witnesses the safety and effectiveness of the combination of emicizumab

and rFVIIa for major orthopaedic surgery, even if a larger number of procedures is required to validate such protocol.

FP-TH-01.02 | Does Digital Templating of total knee arthroplasties reliable in hemophilic arthropathies?

Erdem Er¹; Elcil Kaya Bicer¹; Fahri Sahin²; Kaan Kavakli³; Semih Aydogdu¹

Departments of ¹Orthopaedics and Traumatology; ²Haematology, and ³Paediatric Haematology, Ege University, Faculty of Medicine, Turkey

Introduction: Preoperative digital templating on x rays is a useful method to estimate size of the prosthetic components before a total knee arthroplasty (TKA) procedure. Since haemophilic arthropathy (HA) of the knee joint is associated with severe morphological changes, a question has raised whether preoperative templating is consistent enough as it is in primary osteoarthritis (OA) patients. Aim of this study is to evaluate the reliability of preoperative TKA templating with the actual size of the prosthesis in HA and OA.

Methods: In this retrospective study, 36 knees of 34 HA (mean age: 41.75±10.87) and 36 knees of 36 OA patients (mean age:72.54±9.52) who had TKA between 2011–2020 were included. Templating of femoral and tibial components both on anteroposterior (AP) and lateral x rays was performed utilizing Sectra (Sectra Workstation IDS7 for Windows Version, Sectra AB, Sweden). Actual sizes of the prosthetic components were recorded. Statistical analyses were performed utilizing SPSS v25. Agreement analyses between templating data and actual sizes were performed; intraclass correlation coefficients of average measures were calculated with 95% confidence intervals (CI). Significance level was set at .05.

Results: The agreement level of templating of the femoral component was moderate in both groups in the AP plane whereas it was good for lateral plane in HA group and moderate for OA group. Templating of the tibial component on AP plane had a moderate level of agreement in HA, good level in OA groups. On the contrary, on the lateral plane the level of agreement was good in HA and moderate in OA groups (Table 1).

Conclusions: : The findings of this study showed that reliability of preoperative digital TKA templating was comparable between HA and OA patients with an agreement level of moderate to good. Since templating offers estimation of the prosthesis size with a moderate precision, under or above sizes should also be prepared before the operation.

P-012 (1029723) | Effectiveness of viscosupplementation in the treatment of hemophylic arthropathy: Systematic Review

Samilly Martins¹; Erion de Andrade²; Rodrigo Pagnano¹; Alessandro Zorzi¹; Gustavo Constantino¹

¹University of Campinas - Unicamp, Sao Paulo, Brazil; ²University of Campinas - Unicamp, Cleveland, Ohio

Introduction: In hemophilia, episodes of hemarthrosis lead to specific changes in the synovium and cartilage defined as Hemophilic Arthropathy (HA). The goal of the HA treatment is to control hemarthrosis as fast as possible to avoid secondary synovitis. Among the therapeutic options, viscosupplementation with Hyaluronic Acid (ACh) has been widely used in an attempt to improve the characteristics of synovial fluid and reduce pain. Viscosupplementation has shown an important role in offering patients a useful treatment alternative capable of postponing aggressive interventions, and as an additional option in those refractory to conventional treatment. We aimed to do a systematic review of the data available about the treatment with viscosupplementation in the management of patients with HA.

Methods: A systematic review based on the PRISMA recommendation, without year or language limitations.

Results: The literature search yielded 127 articles on the major medical databases. After applying the eligibility criteria, 10 articles were selected for data extraction and qualitative analysis. A total of 216 patients were included. The average age of the participants was 33.9 years. Six studies used a population of patients with a high-titer inhibitor. When reported, Hemophilia A and severe coagulopathy was highly prevalent. All articles had the description of the intervention, being performed from one to five applications, with weekly to monthly intervals. Clinically, pain and function were assessed using the Visual Analog Pain Scale (VAS) and the WOMAC scores (The Western Ontario and McMaster Universities Osteoarthritis Index), SF-36 (Study 36-item Short-Form Health Survey), Lequesne Algofunctional questionnaire, with evidence of positive clinical effects. No major adverse effects were observed.

Conclusions: Although the level of evidence of the publications available on the topic is still low, viscosupplementation with hyaluronic acid is a safe and effective therapeutic option in hemophilic arthropathy. It can be performed as early initial treatment or when the non-invasive method has failed, with positive results in improving pain, functional capacity, quality of life, and postponing aggressive invasive treatments.

P-011 (1036149) | Complications following total knee arthroplasty (TKA) in the setting of severe haemophilic arthropathy (HA)

George Thivaio¹; Styliani-Despoina Christidi¹; Giannis Vasilopoulos²; Anna Kouramba²; Olga Katsarou²

¹Orthopaedic Department and ²Blood Transfusion Centre and National Reference Centre for Congenital Bleeding Disorders, Laiko General Hospital, Athens, Greece

Introduction: Total Knee Arthroplasty (TKA) is the “gold standard” for severe haemophilic knee arthropathy (HA). However, intraoperative difficulties can compromise the result even for experienced surgeons. The main problems that one must deal with are: 1) poor bone quality, 2) soft tissue fibrosis, 3) increased postoperative bleeding episodes and 4) severe anatomical deformities often requiring custom made implants (angular deformities, narrow medullary canals,

mismatch between anterior/posterior and medial/lateral width and severe condylar bone defects).

Methods: We retrospectively reviewed the charts and operative data of 32 patients who underwent TKA from 2012 to 2017 with a 2-year minimal follow-up. All patients had severe haemophilia (A or B) while 2 of them had positive inhibitor titres. Mean age was 42.9 years (31–62 years); 6 patients had chronic HIV+ under cART. Recombinant clotting factor was infused preoperatively to achieve normal factor levels and postoperatively until hospital discharge. Further prophylaxis was suggested throughout the rehabilitation period. Patients with inhibitors were managed with recombinant factor VII (rFVII) or a PCC.

Results: We recorded postoperative complications in 9 of 32 patients (complication rate 28%) i.e.: 1) early prosthetic joint infection (PJI): treated with debridement, antibiotics, irrigation, and retention of the prostheses (DAIR). 2) Periprosthetic fracture: one lateral condyle fracture treated with fixation with 2 lag screws and one femoral shaft fracture treated with ORIF with plate and strut allografts. 3) Postoperative joint stiffness: treated with arthroscopic adhesions release. 4) Postoperative hemarthrosis: treated with evacuation and infusion of 15ml NS solution containing 1000mg tranexamic acid and 1mg adrenaline. 5) Aseptic implant loosening in one patient with severe valgus deformity requiring a revision TKA.

Conclusions: Total knee arthroplasty in haemophilic patients especially in those with inhibitors and/or immunosuppression is associated with increased complication rates even in experienced centers. Therefore, patients should be aware in detail about the risk-benefit ratio of such a surgical procedure. Of all possible complications, prosthetic joint infection (PJI) is the most serious as it can be life-threatening and difficult to treat.

PAIN MANAGEMENT

P-014 (996204) | Comparison in improvement of symptoms during the treatment of Hemophilic Arthropathy in Nicaragua between Methylprednisolone and Hyaluronic Acid Infiltration

Sergio Luis Gustavo Aguilera Covarrubias¹; Oswaldo José Estrada Vargas¹; Anthony Howay Galarza¹; Christian Gabriel Pasquier Pastrán¹; Armando Raúl Hernández Salgado²; Jonathan Josué González Martínez²; Alfonso alexander Ramirez Chavarria¹; Lilia Ma. Teresa Espinobarros Trujillo²; Jairo Baltazar Campos Valerio¹; Engels Rolando Reyes Sánchez¹

¹Nicaragüense de Hemofilia, Managua, Nicaragua; ²Instituto Mexicano del Seguro Social, Distrito Federal, Mexico

Introduction: The average income of \$1 to 2 USD per day per family limits access to hyaluronic acid treatment costing more than \$100 per injection versus a less expensive \$10 alternative with methylprednisolone. The present study seeks to find the cost-benefit relationship between both treatments in intra-articular infiltration (IAI).

Methods: A clinical evaluation was carried out together with a multidisciplinary team on 8 patients with Hemophilic Arthropathy (HA) can-

didates for IAI, their files were supplemented with Clinical History, X-rays, Gilbert, Petterson and HJHS Joint Health Scales. The clinical evaluation was carried out, Factor was applied at 30 IU per Kg and IAI; 3 received Hyaluronic Acid and 5 methylprednisolone 40mg with 3ml of lidocaine. 2 required prior arthrocentesis. A cane was given to each patient and 7 months later a health survey was applied.

Results: From a sample of 147 patients, 8 between the ages of 22 and 49 years old were selected; 75% do not have formal employment; 50% were asymptomatic at the time of IAI; 1 had moderate type B hemophilia, 7 had type A hemophilia (2 mild, 2 moderate, and 3 severe). The IAIs were: 6 left and 4 right knees, 1 left and 1 right ankle. Those who lived more than 2 hours away from the Treatment Center, needed IAI in both knees and reported the use of opioids for pain management. Obesity represented an increased risk of joint damage.

Conclusions: The use of methylprednisolone is recommended for the management of HA as it provides the same symptomatic improvement reported and at a 90% lower cost. The number of joints affected depends on your access to prompt treatment. All the patients reportedly drink alcohol, representing a greater risk of articular accidents and injuries. 50% have been smoking tobacco for 1 to 6 years, which affects their microvasculature health and joint toxicity. Those who live further away from the capital require opioid analgesic therapy and this implies a greater impact on their joint health due to the lack of access to treatment, the complexity of the terrain and the limited availability of public transport. All patients recommend IAI,

P-016 (1035646) | Moving multidisciplinary pain management online

Alison Gorman¹; Jessica Hedden²

¹Cardiff Comprehensive Care Centre, Wales, United Kingdom; ²Swansea Haemophilia Centre, United Kingdom

Introduction: Between 35–50% of people with haemophilia (PWH) experience chronic pain and mobility issues from haemarthropathy [1]. Pharmacological interventions for pain have been widely reported in PWH however there is a paucity of research on non-pharmacological treatments. Pain is often under-reported by PWH, perhaps because they are so used to living with pain that they only report 'flare-ups' and there is low frequency with which pain and quality of life is investigated by physicians in this field [2]. Swansea Haemophilia Centre implemented a pain management programme using a combination of physiotherapy and psychological tools. Patients were invited to a Hydrotherapy and Mindfulness programme involving weekly hydrotherapy and a monthly psychology intervention with a combination of discussion and teaching around pain and pain management using the Mindfulness for Health approach [3]. An evaluation of this programme found positive patient feedback both in terms of pain, function and coping. In addition, there was a significant social benefit to this approach. With the onset of the Covid pandemic, these F2F sessions had to cease. Four months into the pandemic physiotherapy and psychology recognised not only the need for redesigning the sessions but the importance of

social interactions between this group of patients during the isolation of the lockdown restrictions.

Methods: As a result, online Stretch and Mindfulness sessions were developed. These involved a short seated stretch programme and mindful meditation alongside a social component. On average 4–5 patients attend each session with various inherited bleeding disorders and varying levels of pain and function. Feedback was gathered using semi structured interviews after 9 months of fortnightly sessions. Subjective measures of pain and coping were used to monitor progress alongside qualitative feedback.

Results: The feedback acknowledged that the sessions are not as physically beneficial as hydrotherapy, however the consensus of the participants is that the sessions have brought both physical and emotional benefits.

Conclusions: This study shows that whilst online stretch and mindfulness may not be as beneficial to reported levels of pain and mobility as other F2F interventions, improvements are shown in terms of coping and well-being.

P-017 (1035886) | The impact of social isolation on hemophilia patients with persistent joint pain during the COVID-19 pandemic

Glenda Feldberg^{1,2}; Janaina B S Ricciardi^{1,2}; Andressa Ornelas¹; Alessandro R Zorzi³; Margareth C Ozelo^{1,2}

¹Hemocentro; ²Departments of ²Internal Medicine, School of Medical Sciences and ³Orthopedic and Traumatology, Faculty of Medical Sciences, University of Campinas, UNICAMP, Campinas, Brazil

Introduction: Since the end of 2019, the world has been struck by a new disease (COVID-19) caused by the coronavirus (SARS-CoV-2) that has spread from then on, reaching a worldwide pandemic status. In an attempt to reduce the rates of transmission due to the rapid spread of the virus, measures of social isolation have been included in the protocol and put into action. Thus, the decrease in social interaction produced by social distancing can have a negative impact on the physical and mental health of patients with hemophilia. The objective of the study was to evaluate the effects of the pandemic on the health of hemophilia patients persistent joint pain and their experiences during this period.

Methods: This was a transversal study evaluating the impact of COVID-19 pandemic on patients at the Hemophilia Unit of UNICAMP. The survey in the online format included information regarding the presence of pain, physical activity, work and experiences related to health care during the pandemic.

Results: In total, 33 out of 46 hemophilia patients with persistent joint pain responded the survey. Median age of 34 years (range: 16 to 59 years). 36.4% of the patients had pain in one joint, 30.3% in two and 18.2% in three or more joints. 57.6% reported bleeding in the last 12 months, and 63.6% were less active due to social isolation. Concerning work, 32.3% of patients increased working hours, and 54.8% remained the same. 25% of patients fell into the COVID-19 risk group regard-

ing health conditions that they experienced during the pandemic: 33.3% suffered from work overload, 24.2% eating disorders, 27.3% sleep disorders, 27.3% psychological disorders (stress, anxiety, and panic).

Conclusions: This study showed that patients with debilitating sequels from hemophilia had a negative impact on psychological health and physical activity, with an overload of work, and eating and sleeps disorders during the period of isolation due to COVID-19 pandemic. Thus, strategies of physical and mental approach are highly recommended for this population during this challenging period.

P-015 (1036192) | Haemophilia, pain and exercise: conflict, control and uncertainty. The views, beliefs and experiences of people with haemophilia and healthcare professionals: A qualitative study

Paul McLaughlin¹; Kate Khair²; Pratima Chowdary¹; Michael Hurley³; David Stephensen⁴

¹Katharine Dormandy Haemophilia Centre, Royal Free Hospital, London, United Kingdom; ²Haemnet, United Kingdom; ³St George's University of London and Kingston University, United Kingdom; ⁴Kent Haemophilia & Thrombosis Centre and Royal London Hospital, United Kingdom

Introduction: Chronic pain associated with haemophilic arthropathy restricts daily activities and has a negative effect on the quality of life of people with haemophilia (PWH). It presents a clinical challenge and management options are mostly medications and orthopaedic surgery. Exercise for pain management is used with some success in other arthritises. Little is understood about what PWH and healthcare professionals (HCP's) think of this as a possible strategy. The aim of this study was to explore the views and beliefs of PWH and HCPs on pain experiences, management approaches and using exercise for pain management.

Methods: PWH and haemophilia HCP's volunteered to participate in a focus group or semi-structured interviews. Topic guides were used to guide the groups and interviews. All proceedings were recorded and transcribed verbatim. Reflexive thematic analysis was used to analyse the data.

Results: 14 PWH and 6 HCPs took part in 2 focus groups (n = 11) and interviews (n = 9). All PWH self-reported chronic pain and had severe haemophilia (A or B). The HCPs were two physiotherapists, two haematologists, one specialist nurse and one psychologist. Four themes developed from the data: 'Haemophilia and pain- an evolving life biography', 'My normal is not normal- identity and self-agency', 'Haemophilia management and pain management is discordant' and 'Uncertain about exercise but clear on what matters'. This report presents data on the last two themes. Historical messages and behaviours associated with pain and bleeding continue to undermine the potential of using exercise as an approach for pain. PWH want validation of their life experiences and to be part of a solution that works for them. Living better with pain was identified as important and using exercise was seen as plausible, but with uncertainty about effect and potential risk. Greater

individualised support by knowledgeable clinicians is fundamental to develop and enhance motivation. Partnership and trust are crucial to manage fear and anxieties.

Conclusions: Many PWH have a lifetime of accommodating pain into their daily life, positioning them with unique perspectives on their experiences. These, along with trusted therapeutic relationships are key when trying to incorporate exercise as an option to manage chronic pain.

P-013 (1036818) | Chronic pain and psychosocial aspects in patients with hemophilic arthropathy

Roberto Ucerro-Lozano¹; José Antonio López-Pina²; Rubén Cuesta-Barriuso³

¹Department of Physiotherapy, European University of Madrid;

²Department of Basic Psychology and Methodology, University of Murcia; ³Department of Physiotherapy, University of Murcia, Spain

Introduction: Chronic pain is a feature of hemophilic arthropathy. Identifying the psychosocial variables affected by pain can help in the management of these patients. The objective of the study was to evaluate the relationship between the perception of the intensity of chronic pain with joint damage, kinesiophobia, catastrophism, anxiety and the perception of quality of life in adult patients with hemophilic arthropathy.

Methods: Multicenter cross-sectional descriptive study. Seventy-seven adult hemophilia patients were recruited. The intensity of the usual and maximum pain (Visual Analogue Scale), joint health (Hemophilia Joint Health Score), kinesiophobia (Tampa Scale of Kinesiophobia), catastrophism (Pain catastrophizing scale), anxiety (State-Trait Anxiety inventory) and the perception of quality of life (36-Item Short Form Health Survey) were evaluated.

Results: The mean age of the sample was 44.04 (SD: 10.17) years. The habitual perception of pain in patients with hemophilia was positively correlated ($p < .001$) with catastrophism ($r = .68$), kinesiophobia ($r = .67$), state anxiety ($r = .58$) and trait anxiety ($r = .39$). The patients' maximum pain perception correlated ($p < .001$) with all variables: catastrophism ($r = .49$), kinesiophobia ($r = .52$), state anxiety ($r = .49$) and trait anxiety ($r = .39$). The total physical health score was inversely correlated ($p < .001$) with the usual pain ($r = -.64$) and the maximum pain ($r = -.66$) of the patients. Patients with severe hemophilia showed a greater perception of pain ($p < .001$) than patients with moderate hemophilia, with respect to the usual pain (4.73 vs 3.10) and maximum (8.16 vs 6.80). The patients with inhibitor showed differences ($p < .01$) in the usual intensity of pain (5.67 vs 4.31). There were no differences ($p > .05$) in pain intensity between patients with prophylactic or on-demand treatment.

Conclusions: Psychosocial factors influence the painful experience of patients with hemophilic arthropathy. The intensity of pain affects the quality of life of these patients.

PATIENT REPORTED OUTCOMES

P-019 (1035680) | Relationship of EQ-5D Pain Domain and Bleeds - Insights from the PROBE Study

Mark Skinner¹; Declan Noone²; Randall Curtis³; Neil Frick⁴; Federico Germini⁵; Alexandra Kucher⁶; Michael Nichol⁷; Brian O'Mahony²; David Page⁸; Jeff Stonebraker⁹; Alfonso Iorio¹⁰

¹Institute for Policy Advancement Ltd, Washington, District of Columbia;

²Irish Haemophilia Society, Ireland; ³Factor VIII Computing, Berkeley;

⁴National Hemophilia Foundation, New York City; ⁵Department of Department of Medicine, McMaster University, Hamilton; ⁶Patient Outcomes

Research Group Ltd, Washington; ⁷University of Southern California, Sol

Price School of Public Policy, Los Angeles, California; ⁸Canadian Hemophilia

Society, St-Michel-de-Bellechasse, Quebec; ⁹North Carolina State Uni-

versity, Poole College of Management, Raleigh; ¹⁰Department of Health

Research Methods, Evidence, and Impact, Hamilton

Introduction: Bleeding in people living with haemophilia (PWH) has a significant impact on Health-Related Quality of Life (HRQoL). This impact has been used in terms of disutilities in cost-effectiveness analysis. The aim was to assess the impact on overall HRQoL by severity for a bleed being reported in the last two weeks and assess the level of impact on outcomes.

Methods: Data collected between 2018–2021, was analyzed to determine the relationship of bleeding events being reported within the past two weeks and the PROBE and EQ-5D-5L utility scores. 1168 questionnaires from male PWH A and B \geq Age 18 (mean[SD] age 41.1[14.5]), were collected in 50 countries. Descriptive analysis and generalised linear models were produced using Stata 13.

Results: The level of pain reported using the EQ-5D-5L pain domain increased across all severities when a PWH reported having a bleed in the past two weeks. Overall, the EQ-5D utility scores were .077 ($p = .029$), .058 ($p = .087$) and .092 ($p < .001$) higher for mild, moderate and severe patients, respectively, when a bleed was reported in the last two weeks. Using the PROBE score, they were .074 ($p = .074$), .080 ($p < .001$) and .089 ($p < .001$) higher. Using a generalised linear model accounting for age, severity and current treatment regimen the disutility associated with a bleed in the last 2 weeks was .08 ($p < .001$) for EQ-5D-5L and .086 ($p < .001$) for the PROBE score.

Conclusions: Bleeding has a significant impact on PWH HRQoL. A disutility in HRQoL measured by the EQ-5D and PROBE score is evident even two-weeks after a bleeding event. The temporal limitations of the EQ-5D may mask the length of impact of bleeding events unless the context of recent bleeding events is considered. The resulting utility scores may lead to a misrepresentation of the true impact of pain within the severe haemophilia population.

P-018 (1036845) | Predictors of quality of life in adult patients with hemophilic arthropathy

Roberto Ucero-Lozano¹; Rubén Cuesta-Barriuso²; José Antonio López-Pina³

¹Department of Physiotherapy, European University of Madrid;

²Departments of Physiotherapy and ³Basic Psychology and Methodology, University of Murcia, Spain

Introduction: Hemophilic arthropathy is characterized by chronic pain, degenerative joint damage. Kinesiophobia and catastrophism are characteristics of degenerative joint pathologies. Hemophilic arthropathy affects the quality of life of the patient. The objective of this study was to evaluate the perceived quality of life of adult patients with haemophilic arthropathy and its relationship with pain, joint condition, kinesiophobia and catastrophism.

Methods: Eighty-three adult patients with haemophilia were included in this multicentre, cross-sectional, descriptive study. Perceived quality of life (36-Item Short Form Health Survey), perceived usual and maximum pain (visual analogue scale), joint condition (Haemophilia Joint Health Score), kinesiophobia (Tampa Scale of Kinesiophobia) and catastrophism (Pain Catastrophizing Scale) were assessed.

Results: The mean age of the sample was 43.87 (SD: 10.10) years. Catastrophism was negatively correlated with all domains of perceived quality of life ($p < .05$). Kinesiophobia was negatively correlated with all domains of quality of life ($p < .05$) except for emotional role. The maximum perceived pain was negatively correlated ($p < .05$) with most of the domains of quality of life except emotional role and total perceived mental health. The usual pain perceived by the patients was negatively correlated with all domains except the emotional role and the total perceived mental health. The overall joint condition was negatively correlated ($p < .05$) only with mental health and the total mental health score. In terms of individual joints, elbow joint deterioration presented an inverse correlation with social functioning ($r = -.22$) and mental health ($r = -.27$), while the degree of ankle arthropathy was negatively correlated with the total perceived mental health ($r = -.22$). Age was positively correlated with perceived quality of life. There were differences in quality of life as a function of the severity of haemophilia.

Conclusions: The perceived quality of life of adult patients with haemophilia is worse than that of the Spanish population. Pain, kinesiophobia, catastrophism and haemophilia severity influence the quality of life of these patients.

PHYSICAL THERAPY**P-023 (1027049) | Multimodal exercises program to improve balance in patients with haemophilia: preliminary results**

Ana Chimeno-Hernández¹; Marta Aguilar-Rodríguez¹; Sofía Pérez-Alenda¹; Juan J. Carrasco²; Carlos Cruz-Montecinos³; Santiago Bonanad⁴; Felipe Querol¹

¹Physiotherapy Department; and ²Intelligent Data Analysis Laboratory, University of Valencia, Valencia, Spain., Spain ³Laboratory of Clinical Biomechanics, Department of Physical Therapy, Faculty of Medicine, University of Chile, Santiago, Chile; ⁴Haemostasis and Thrombosis Unit, University and Polytechnic Hospital La Fe, Valencia, Spain

Introduction: Joint damage occasioned by repetitive bleedings in people with haemophilia (PWH) origins structural, proprioceptive and control motor impairment that cause imbalance. The aim of this study is to analyze if a multimodal exercises program improve balance, functionality, risk of falls and strength in this population.

Methods: A clinical trial was carried out at the facilities of the University of Valencia. At the moment, sixteen participants with severe haemophilia A with prophylaxis have been divided into two groups: intervention group (IG) $n = 11$, mean age (SD) = 37.5 (12.03) and control group (CG) $n = 5$, mean age = 35.6 (7.1). The IG performed a personalized one-hour-program of multimodal balance and strength exercises twice a week for 3 months, while the CG continued with their daily physical activity. The intensity of the exercises were controlled by a scale of perceive exertion. The effectiveness of multimodal exercise program was probed by pre-post measurements: i) balance control using a Wii Balance Board® and Berg scale, ii) the risk of falls using the modified fall efficacy scale (mFES), iii) the functionality using the Timed "Up and Go" (TUG), sit-to-stand (STS) and 2-minute walk (2MW) tests and strength (dynamometer).

Results: Data showed statistically significant ($p < .0125$) changes in the IG in Berg scale: Pre mean (SD) = 51 (5.2)/Post = 55.1 (1.2); mFES: Pre = 120.5 (19.2)/Post = 129.3 (14.1) and functional tests: Pre-TUG = 7.1 (1.7)/Post-TUG = 5.5 (1.3); Pre-STS = 6.4 (1.9)/Post-STS = 5 (1.3); Pre-2MW = 162 (37.1)/Post-2MW = 190.6 (37.7) as well as in gluteus medial strength (Pre-right = 22.1 (7.5)/Post-right = 28.3 (6.4); Pre-left = 21.8 (7.6)/Post-left = 27 (5.8)). Although there were no statistically significant changes in the balance measured with the Wii Balance Board® in IG, improvements between 5% and 43% were observed in the variables evaluated (sway area, mean total velocity and standard deviation of amplitude antero-posterior and medio-lateral). The

CG did not show statistical significant changes in the pre-post outcomes. There were no bleedings episodes or dropouts associated with the exercises program.

Conclusions: Our results suggest that a personalised multimodal balance and strength exercises program improve functionality and balance in PwH. In addition, the absence of bleeding episodes during the program indicates that it is a safe program for patients with severe haemophilia A with prophylaxis.

P-025 (1035319) | Usage of a live-feedback measurement with sensor soles to monitor partial weight-bearing in physiotherapeutic treatment of haemophilic patients after endoprothetic upper ankle joint surgery: a case report

Daniel Taubenschmid; Emanuel Reiter; Jürgen Ableitner
CIM-Competence In Motion, Austria

Introduction: Advantages of pressure feedback sensor soles in physiotherapeutic treatment of haemophilic patients after total ankle arthroplasty. Due to a haemophilic disease, followed by a destruction of the upper ankle joint, an implantation of a total endoprosthesis was the chosen method. In this special case the patient has been treated with a period of 12 weeks of partial weight-bearing to optimize the osseous integration of the implant. In the post-surgical treatment of haemophilic patients, the control of the doctors given limits is very important, to avoid too high or too low impact load on the joint. Keeping the exact partial weight bearing is extremely difficult to handle without any technical feedback systems during the physical therapy and in everyday life.

Methods: The post-surgical physiotherapy after a total ankle-joint endoprosthesis with partial weight-bearing was supported with stapp one sensor soles. The insoles are completely wireless and are inserted into the patient's shoe. The measured data of the partial weight bearing was transmitted via bluetooth to a handheld tablet and was presented in real-time to the patient. The software application enables visual, acoustic or haptic live-feedback.

Results: The application of bio pressure sensor soles has proven as an uncomplicated and simple therapy tool. It was quite simple to keep the doctors range of load in the partial-weight bearing. With this live-feedback-system, the patient as well as the physiotherapist had a constant and exact control of load on the operated leg during all the time of therapy. Afterwards the patient was able to control the weight bearing without a visual feedback.

Conclusions: The independent usage of this sensor soles also allow to monitor pressure feedback during stairs climbing or walking on bumpy ground outdoors. So the patient got well prepared for different loads without leaving the range of partial weight bearing. Furthermore, it has to be mentioned, that the potential of these sensor soles is much higher than only measuring a partial weight-bearing. For instance, the load distribution or physiological gait parameters like cadence or speed can be better analysed. This innovative tool has shown added value for Hemophilia patients after endoprosthesis in the rehabilitation process.

P-021 (1036010) | Correlations between HJHS and FISH in the physiotherapeutic evaluation of hemophilic patients referred to radioactive synovectomy – Brazil experience

Fernando Zikan; Sylvia Thomas; Susie Medeiros
Federal University of Rio de Janeiro, Rio de Janeiro, Brazil

Introduction: Hemophilic arthropathy is the main object of the physiotherapist's performance in patients with hemophilia. These patient's joint health be constantly evaluated, assessing the severity of joint damage and musculoskeletal injury (through Hemophilia Joint Health Status - HJHS) and their functional status through Functional Independence Measure for Hemophilia (FISH). The study aimed to evaluate patients submitted to Radioactive Synovectomy, and to establish a correlation between the HJHS and FISH evaluation instruments (Pearson correlation coefficient),

Methods: It was carried out at the national referral center of this procedure, with the proper authorization of patients and caregivers and approved by the Research Ethics Committee. All patients were evaluated through HJHS and FISH one day before the Radioactive Synovectomy, by the physiotherapist group.

Results: A total of 200 evaluations were performed in 2018 until February 2020, with a patient mean age of 19.38 years old, and the knees being the most frequent target joint. The HJHS Total Score averaged 15.95 points, and the Global Gait average was 1.2 points. When we divided by age group, we found Total Score HJHS of 8.0 points, FISH 28,85 points in 34 patients from 0 to 10 yo; HJHS of 12.3 points, FISH 29,17 points in 92 patients from 11 to 20 yo; HJHS of 22.9 points, FISH 26,6 points in 48 patients from 21 to 30 yo and HJHS of 26.2 points, FISH 23,38 points in 26 patients over 30 yo. Pearson correlation coefficient to HJHS and Age is +0,48; FISH and Age is -0,42 and HJHS and FISH is -0,58, this being then a moderate negative correlation.

Conclusions: Thus, we could identify that the greater the age, the greater the HJHS score, which means greater joint health damage. Moreover, the older the patient is, the lower the FISH score also is, meaning less functional independence of these patients. Thus, these two instruments are fundamental for the evaluation, since the Scores are inversely proportional and the age was the determining variable for both.

P-020 (1036501) | Adherence to Physiotherapy and its Association with the level of Joint pain among adult patients attending Haemophilia clinics of two teaching hospitals in Sri Lanka

Hiran Karunaratne¹; Chamodi Karunasena²; Indra Karunathilake¹; Niludi Yasaratna¹

¹*Faculty of Medicine, University of Colombo, Western Province, Sri Lanka;*

²*Faculty of Medicine, University of Colombo, Southern Province, Sri Lanka*

Introduction: Although physiotherapy improves the condition of haemophilia patients by minimizing the level of joint pain, some

patients are reluctant to obtain physiotherapy treatments. This descriptive and analytical cross sectional study was carried out to assess adherence to physiotherapy, its associated factors and its relationship with level of joint pain.

Methods: An interviewer administered questionnaire was used to assess the adherence to physiotherapy according to two parameters; engagement of physiotherapy at hospital and/or domestic setting and the compliance to advices of physiotherapist. Determinants of adherence to physiotherapy were also assessed using the questionnaire. Horizontal visual analogue scale was used to assess the level of joint pain and pain was categorized into no, mild, moderate and severe pain according to the scores obtained from the scale.

Results: Of the 111 adult patients, majority were adherent to physiotherapy from both parameters; engaging in physiotherapy at home/hospital (56.8%) and being compliant to advices of the physiotherapists (52.3%). Age and occupation were significantly associated with adherence to physiotherapy according to both parameters ($p < .05$). Walking ability, family encouragement and the belief that 'joint pain is reduced by physiotherapy' were positive determinants significantly associated with adherence to physiotherapy according to both parameters ($p < .05$). There were significant negative relationships ($p < .05$) between adherence to physiotherapy and these influential factors; BMI, fatigue during physiotherapy, joint pathologies and worsening joint pain. Daily activity level and doing other weekly activities were positive determinants significantly associated with adherence to physiotherapy only on the parameter of engagement in physiotherapy in any setting ($p < .05$). Majority of the participants were having mild pain (38.7%) within a one month duration. 27% and 26.1% experienced moderate and severe pain during that period.

Conclusions: Age < 40 , Unemployed, being able to walk, high daily and weekly activity levels, belief that joint pain will be reduced and family encouragement were positive determinants for physiotherapy. BMI > 23 , existing joint pathologies, belief that joint pain will worsen and fatigue were negative determinants for physiotherapy. Adherence to physiotherapy according to both parameters were significantly associated with the low levels of joint pain ($p < .01$).

P-022 (1036829) | Manual therapy treatment on quality of life in patients with hemophilic elbow arthropathy

Rubén Cuesta-Barriuso¹; Raúl Pérez-Llanes²; José Antonio López-Pina³; Elena Donoso-Úbeda²; Javier Meroño-Gallut⁴

¹Department of Physiotherapy, University of Murcia; ²Department of Physiotherapy, Catholic University San Antonio-UCAM; ³Department of Basic Psychology and Methodology, University of Murcia; ⁴Tú Bienestar 360°, Physiotherapy and Medical Center, San Javier-Murcia, Spain

Introduction: Pain and hemophilic arthropathy have a negative impact on the quality of life in patients with hemophilia. Few studies have evaluated the effectiveness of physiotherapy in improving the quality of life for these patients. The objective of this study was to evaluate the effect

of physiotherapy on the perceived quality of life, pain intensity and joint health in people with hemophilia.

Methods: Randomized single-blind clinical trial. Sixty-nine patients were allocated to the experimental group and they were applied a manual therapy protocol for patients with elbow arthropathy. Patients in the control group received no intervention whatsoever. The variables evaluated were perceived quality of life (Short-Form Health Survey-36), pain intensity (Visual Analog Scale) and joint health (Hemophilia Joint health Score) at baseline and at 3 and 12 weeks.

Results: We found significant differences ($p < .01$) in the repeated measures analysis depending on the time of assessment in the two physical variables: joint health ($F(2, 134) = 21.92$) and pain intensity ($F(1.23, 82.64) = 18.68$). For the perceived quality of life, there were no intragroup differences in physical function ($F(2, 134) = 2.91$; $p = .04$), physical role ($F(1.7, 114.3) = 52.86$; $p < .001$), body pain ($F(2, 134) = 5.72$; $p = .004$), emotional role ($F(2, 134) = 194.69$; $p < .001$) and mental component ($F(1.83, 123.13) = 15.66$; $p < .001$). When analyzing the intergroup effect, there were significant differences ($p < .001$) in group interaction for joint health ($F = 55.31$), intensity of elbow pain ($F = 24.74$), physical role ($F = 49.22$) and emotional role ($F = 229.71$), and the mental component of perceived quality of life ($F = 9.86$). A high effect size ($n_2 p > .14$) was observed in the intergroup effect in the variables joint health ($\eta^2_p = .45$), pain intensity ($\eta^2_p = .27$), and physical ($\eta^2_p = .42$) and emotional role ($\eta^2_p = .77$).

Conclusions: A physiotherapy program for patients with hemophilic elbow arthropathy can improve their perceived quality of life, pain intensity and joint health.

P-024 (1036854) | Self-mobilization using a Foam Roller in adult patients with hemophilic knee arthropathy

Rubén Cuesta-Barriuso¹; Javier Meroño-Gallut²; Roberto Ucero-Lozano³; Elena Donoso-Úbeda⁴; Raúl Pérez-Llanes⁴

¹Department of Physiotherapy, University of Murcia; ²Tú. Bienestar 360°, Physiotherapy and Medical Center, San Javier-Murcia; ³Department of Physiotherapy, European University of Madrid, Spain; ⁴Department of Physiotherapy, Catholic University San Antonio-UCAM, Spain

Introduction: Knee arthropathy is common in patients with hemophilia. Arthropathy is characterized by limited range of motion, chronic pain, and loss of muscle strength, among other symptoms. The objective of this study was to evaluate the safety and efficacy of a protocol of self-mobilization using a Foam Roller in patients with hemophilic knee arthropathy.

Methods: Prospective cohort study. Twenty-nine patients were recruited. The patients performed the exercises of the self-mobilization protocol using a Foam Roller, 7 days a week, for 12 weeks. The duration of each session was 15 minutes. This protocol included superficial glides on the ventral and dorsal area of the leg (quadriceps, hamstrings and gluteus), and on the popliteal hollow, as well as by applying deep to the lower limb. The variables evaluated were frequency of bleeding (self-record of bleeding and a telephonic

follow-up), pain intensity (Visual Analog Scale), knee range of motion (goniometer) and muscle strength (quadriceps dynamometry) at baseline and at 12 weeks. Assessments at baseline and after the intervention were carried out by a blinded physiotherapist. The student T test for paired samples was used to compare the means in the two evaluations carried out in the study.

Results: During the treatment period, none of the patients included in the study presented hemarthrosis in the knees. Although patients could be on demand treatment and there were patients with inhibitors, no bleeding episodes in muscles or joints were reported. After the treatment period there were significant changes in knee pain intensity (means difference, MD = .87; $p = .005$; 95%CI = .28; 1.45), flexion (MD = -2.31; $p = .012$; 95%CI = -4.06; -.55) and extension (MD = 1.55; $p = .004$; 95%CI = .54; 2.55) range of motion, and quadriceps strength (MD = -40.42; $p < .001$; 95%CI = -60.27; -20.56).

Conclusions: A protocol of self-mobilization using a Foam Roller does not appear to produce muscle or joint hemorrhages in patients with hemophilic ankle arthropathy. Self-mobilization using a Foam Roller can improve pain intensity, range of motion, and quadriceps strength in patients with hemophilic ankle arthropathy.

REHABILITATION

FP-W-01.03 | Iliopsoas haemorrhage with femoral neuropathy among patients with haemophilia

Tachit Jiravichitchai¹; Monratta Panuwannakorn¹; Surapong Lertthammakiat²; Suphaneewan Jaovisidha³; Nongnuch Sirachainan²; Ampaiwan Chuansumrit²
Departments of ¹Rehabilitation Medicine; ² Pediatrics; and ³Radiology, Ramathibodi Hospital, Mahidol University, Thailand

Introduction: Iliopsoas haemorrhage is an uncommon muscular bleeding found among patients with haemophilia. However, its major complication of femoral neuropathy could lead to further serious comorbidities such as atrophy and weakness in the quadriceps muscle, hip flexion contracture and gait abnormality. The study aimed to identify the occurrence of femoral neuropathy and its recovery among patients with haemophilia exhibiting iliopsoas haemorrhage.

Methods: Medical records of 188 patients with haemophilia (A159, B 29) ≤ 20 years of age between 2007 to 2020 were retrospectively reviewed. Patients, who received a diagnosis of iliopsoas haemorrhage and confirmed by ultrasonography and/or CT-scan, were recruited. Femoral neuropathy and recovery in each episode of iliopsoas haemorrhage was evaluated.

Results: Fifty-four episodes of iliopsoas haemorrhage among 22 patients with haemophilia (A19, B3) were included. The cause of bleeding was spontaneous occurrence in the majority of patients (80%) while the remaining patients were trauma-related. Low inhibitor titre of .66 and 4.2 BU was found among 3 patients with haemophilia A while high inhibitor titre was found among 8 patients with haemophilia A ranging from 18.8 to 84 BU and one patient with haemophilia

B (113 BU). All patients without and with inhibitor ≤ 32 BU were responsive to the appropriate replacement therapy and rehabilitation. Three patients (A2, B1) with high inhibitor at 84, 76.5 and 113 BU received limited amounts of bypassing agents with unresponsiveness. However, one received additional plasma exchange, followed by high dose factor VIII administration and survived while the other two succumbed to extensive bleeding with superimposed infection. Nineteen episodes of femoral neuropathy (19/54 = 35.2%) were identified among 7 of 22 patients (31.8%). The occurrence and recovery of femoral neuropathy was found at 8.3% and 9.3% per person per episode, respectively. The median times of the occurrence of femoral neuropathy and recovery were at 3 (IQR 2–6) and 20 days (IQR 9–30) after the onset of iliopsoas haemorrhage, respectively. High titre inhibitor (≥ 5 BU) showed a significant association with femoral neuropathy.

Conclusions: Femoral neuropathy following an iliopsoas haemorrhage among patients with haemophilia was not uncommonly found especially in patients with high inhibitor titre.

P-026 (1033242) | The management of hemophilic patients in a rehabilitation ward during the COVID-19 pandemic

Marco Martinelli; Valter Emilio Passeri; Stefania Pivetti; Vincenzo Gatteri; Rossella Tosini; Luisa Cigolini; Sonia Chiar
Fondazione Teresa Camplani Domus Salutis health care Brescia Italy, Italy

Introduction: During the periods of reduced Covid impact, we have implemented a specific rehabilitation program for hemophilic patients. The overall length of stay represents an interesting outcome within the implementation of the new infection containment rules.

Methods: The period considered goes from June 2020 to the present day. Thirteen hemophilic patients were treated in the post-surgery period for major orthopedic problems. The main inpatient's path consists of 4 phases. The first phase is the selection of the patient, when a nasal mid-turbinate swab operation would be the initial screening for COVID-19 during the 48 and 24 hours prior to admission in our rehabilitation center. The negativity result is the condition for the hospital rehabilitation. The second phase consists of isolation in a single room for 5 days. During this time a physiotherapist protected (water repellent overcoat, shoe covers, mask and visor) interacts with the patient in the room. All necessary instruments being part of the therapy program such as the apparatus for the passive movement of the lower limb (for knee and ankle prosthesis), the pedal exerciser with the cycle ergometer (ankle prosthesis and hip prosthesis) and electrotherapy for muscle strengthening (all prosthesis) are used daily. The swab test is performed on the fifth day; if negative, the patient, in the third phase, gains access to a rehabilitation path outside the room but only within the confinement of the ward. Finally, for the fourth phase, after 5 additional days, in the absence of symptoms and, yet, with another negative swab, the patient is able to be admitted to the common gym in order to continue his rehabilitation program.

Results: None of the patients presented symptoms leading to covid infection and all the swabs performed in search of SARS Cov2 were negative. All treated patients passed the four phases and completed the rehabilitation treatment with an average length of stay comparable with the non-covid period (19.3 days versus 19.6).

Conclusions: The completion of the rehabilitation treatment in a reasonable time, despite the "covid" period and the unusual condition, is probably due to the immediate and extensive use of dedicated instruments, and repeated daily physiotherapy activity.

P-027 (1035955) | Telerehabilitation experiences in people with severe haemophilia during the COVID-19 pandemic: A qualitative study

Verónica Aliaga-Castillo¹; Giselle Horment-Lara¹; Felipe Contreras²; Carlos Cruz-Montecinos¹

¹Department of Physical Therapy, University of Chile, Region Metropolitana, Chile; ²Chilean Society of Haemophilia, Region Metropolitana, Chile

Introduction: The global pandemic of coronavirus disease 2019 (COVID-19) caused social distancing measures to be imposed in most countries around the world to control the spread of the virus. These measures affected access to health care for people with chronic conditions, including people with haemophilia. In order to provide rehabilitation care to people with severe haemophilia, the San José Hospital, national reference center for the treatment of haemophilia in Chile, implemented the Telerehabilitation Program in Hemophilia (TPH). The aim of this study was to describe the experiences in telerehabilitation of people with severe haemophilia during the COVID-19 pandemic in Chile.

Methods: A qualitative exploratory-descriptive study was conducted using a descriptive phenomenological approach. A criterion sampling technique was used and included all participants of the TPH. Two online focus groups were performed. Four themes were identified in advance. Three of them are elements described in the Donabedian model for evaluation of quality in health care (structure, process and results).

Results: Within the structure, the technology used and the availability of a physiotherapist expert in haemophilia were described as advantages, and the lack of adequate equipment and the reduced schedule of sessions are described as disadvantages. Within the process, the opportunity to access rehabilitation care for people from distant regions and the savings in time and money were described as advantages, and the perception that the physiotherapist does not have the possibility to perform an appropriate physical examination and the lack of direct supervision were described as disadvantages. All participants reported feeling satisfied with the program and having obtained good results, in addition to increase in well-being and to promotion of autonomy and responsibility for self-care.

Conclusions: The TPH was positively assessed by all participants, who provided proposals for improvement based on their experiences. The

program should continue its implementation to provide access to care to people who previously did not have it and to give continuity to people who had begun their face-to-face rehabilitation process. The implementation of the program depends on the availability of prophylaxis for all participants and a healthcare team with expertise in haemophilia.

REPLACEMENT SURGERY

P-028 (1036470) | Lateral approach to the total knee arthroplasty in haemophilic arthropathy: Case series

Alessandro Zorzi¹; Rodrigo Pagnano¹; Glenda Feldberg²; Janaina Bosso³; Margareth C Ozelo²

¹University of Campinas - UNICAMP; ²Hemocentro UNICAMP, University of Campinas, Campinas; Department of Internal Medicine, School of Medical Sciences UNICAMP; ³UNICAMP, Sao Paola, Brazil

Introduction: Total knee replacement (TKR) in patients with severe haemophilic arthropathy (HA) and valgus deformity remains challenging. The lateral approach, also known as Kewish's approach, has advantages, including ligament balancing, preservation of the medial retinaculum and the medial neurovascular structures for the supply of the patella. It was demonstrated that lateral approach enhances patellar tracking and improves clinical function in osteoarthritis patients. However, there is doubt about feasibility in patients with HA, because adhesions can hinder a good joint exposure. The objective of this study is to describe the results of lateral approach in haemophilic patients submitted to TKR.

Methods: Case series. Eight haemophilic patients (10 knees) were submitted to TKR between June 2017 and November 2019. Four knees had a valgus deformity corrected by a lateral approach. Primary outcome was incidence and severity of complications according to National Cancer Institute-Common Terminology Criteria for Adverse Events (NCI-CTCAE) List Version 4.0. Secondary outcomes were the radiographic alignment of the limb (FTA), HJHS scale, FISH scale, WOMAC scale, ROM and Timed-Up and Go test (TUG).

Results: Incisions healed by first intention in all cases. There were no severe adverse events and no vascular or neurological complications. A primary non-constrained implant was used in all cases of valgus deformity. No case needed tibial tuberosity osteotomy. HJHS scale before surgery ranges from 29 to 84 (58.3±23.3) and after it ranges from 25 to 47 (38±11.5). FISH scale before surgery ranges from 13 to 18 (16.3±2.2) and after from 17 to 18 (17.7±.6). WOMAC pain before surgery ranges from 0 to 13 (9.3±6.2) and after from 0 to 6 (3±3). TUG before surgery ranges from 9.5 to 19.8 (15.7±4.8) and after from 7.2 to 12.2 (9.4±2.6).

Conclusions: Kewish's lateral approach is a safe and effective way to perform total knee arthroplasty in patients with severe HA and valgus deformity.

SPORT/FITNESS/PHYSICAL ACTIVITY

FP-W-01.02 | The relationship between self-reported physical activity, treatment regimen, mental health and pain in persons with hemophilia enrolled in NHF's community voices in research

Michelle Witkop¹; Maria Santaella²; Rosaura Vidal³; Cynthia Nichols⁴
¹National Hemophilia Foundation, Traverse City, Michigan; ²National Hemophilia Foundation, Key Biscayne, Florida; ³National Hemophilia Foundation, New York, New York; ⁴National Hemophilia Foundation, Mancelona, Michigan

Introduction: Community Voices in Research (CVR) is a community-powered registry hosted by the National Hemophilia Foundation (NHF) open to persons affected with an inherited bleeding disorder and their unaffected relatives. Objectives: Assess the relationship between self-reported physical activity, treatment regimen, mental health, and pain in persons with hemophilia (PWH) enrolled in CVR.

Methods: Enrollment occurred between 3/2019-11/2020. Self-reported data was collected through electronic surveys by PWH (both genders, all severities). Specific data points included demographics, physical activity participated in, aspirational activity, limitations on activity, joint bleeds in the past 6 months, joint limitations, pain, and mental health. A non-validated weighted activity risk score was created to compare activity risk as listed in NHF's Playing It Safe booklet with Levels 1-5 being the lowest to highest risk groups.

Results: A total of 254 PWH participated (male 167; female 67; no gender response 11; hemophilia A 197) (Table 1). Persons with moderate hemophilia (PVMH) (n = 73) self-reported the lowest BMI and less: mental health concerns, chronic pain, joint bleeds in the past 6 months, joint limitations, use of prophylaxis, and engaged in less aggressive physical activity than their mild or severe counterparts. Persons with severe hemophilia (PWSH)(n = 118) reported more engagement in the highest activities risk group vs. other severities yet still would like to be more physically active. The most frequent reasons for physical activity limitations varied but the top reason was painful joints. There was a significant negative correlation between the weighted activity risk score and pain limiting rank: $Rho = -.266$, $P < .001$. This was most highly significant for PWSH: $Rho = -.351$, $P = .0001$.

Conclusions: Despite education to the contrary, PWSH continue to engage in high-risk, aggressive physical activities and would like to be even more physically active. As treatment options progress, offering more opportunity for physical activity, research is required to understand an acceptable balance between benefit and harm in PWH.

SYNOVITIS

FP-T-01.02 | The effects of intra-articular injection of TNF-alpha Antagonists in treatment of haemophilic synovitis

Mengyang Xu¹; Qin Yang²; Binglan Xia²; BAOLAI HUA¹; Junjiang Sun³
¹Departments of ¹Hematology and ²Ultrasonography, Clinical Medical College, Yangzhou University, China (People's Republic); ³Division of Molecular Pharmaceutics, Eshelman School of Pharmacy, University of North Carolina, Chapel Hill, North Carolina

Introduction: Hemophilic arthritis (HA) is a serious complication of hemophilia due to recurring joint bleeding. To observe the efficacy and adverse events (AEs) of TNF α antagonists via intra-articular injection in treatment of hemophilic synovitis.

Methods: A single dose of 25mg Etanercept was injected intra-articularly into the "target joint" of patients with HS. Clinical data (bleeding frequency, VAS, HJHS) and Ultrasound Score (HEAD-US-C) were collected on D0, D30 and D90. The synovial thickness and vascularity in the fixed layers were monitored by PDUS. TNF α level was measured by ELISA. AEs were recorded.

Results: 28 PHS with 32 "target joints" were enrolled with a median age of 24 years. 50 joints in 14 patients receiving clotting factor, either by prophylaxis or on-demand therapy, served as the controls with comparable distribution of "target joints" and age. On D30, 31 out of 32 joints had positive responses. Compared with D0, the bleeding frequency, VAS, HJHS and HEAD-US-C scores were all decreased significantly (Table 1). The maximum synovial thickness, mean synovial thickness and maximum synovial thickness change were decreased by 22.2%, 18.9%, 34.7%, respectively; the synovial vascularity was reduced, 0-graded increased from 16.5% to 31.5%, while 3-graded decreased from 23.6% to 11.0%. No significant variation of synovial thickness and vascularity in 50 control joints was seen throughout the follow-up period. On D90, the efficacy was still significant as evidenced by the observed parameters in D30 (Table 1). After treatment, TNF α of synovial fluid (n = 9, 175.6 \pm 319.8ng/ml) was decreased significantly in 6 patients (24.7 \pm 9.2ng/ml), with a fluctuant TNF- α levels in two patients and decreased significantly afterwards. No AEs were reported during the follow-up period.

Conclusions: Intra-articular administration of TNF α antagonist could be a promising treatment for HS.

FP-T-01.03 | Is PRP an effective treatment for chronic synovitis in haemophilic patients?

George Thivaivos¹; Calliope Zerzi²; Anna Kouramba²; Styliani-Despoina Christidi¹; George Veziris¹; Anna Zannou²; Olga Katsarou²

¹Orthopaedic Department and ²Blood Transfusion Centre and National Reference Centre for Congenital Bleeding Disorders, Laiko General Hospital, Athens, Greece

Introduction: Repetitive joint bleeding episodes (BE) in haemophilic patients (PWH), can lead to chronic synovitis and arthropathy, with joint deformity, chronic pain and disability. Platelet-rich plasma (PRP) contains a high concentration of growth factors, coagulation factors, adhesion molecules and cytokines that can have an anti-inflammatory effect and stimulate the healing processes. The aim of the study is to access the efficacy of intra-articular PRP injection in the management of chronic synovitis in PWH.

Methods: A total of 23 PWH, mean age 43 years (IQR 19–70), with chronic synovitis in 33 joints, underwent intra-articular PRP injections between December 2019 and December 2020 with 1 year follow up. A mean volume of 3–5 mL of PRP was injected into the joint affected. Haemophilia Joint Health Score (HJHS), Visual Analogue Scale (VAS), range of motion (ROM) and joint perimeter were assessed for each joint before treatment, 3, 6 and 12 months after treatment. The number of BE was measured during the month preceding PRP injection and between 0–3, 3–6 and 6–12 months.

Results: Seventeen (17) patients were hemophilia A (12 severe, 5 mild/moderate), 4 patients hemophilia B (3 severe, 1 moderate) and 2 patients had severe deficiency of FVII. The affected joints included 18 ankles, 8 elbows and 7 knees. VAS and HJHS scores showed a statistically significant reduction 3 months after treatment compared to pre-treatment scores [median (range) 6.00(2-9) and 8.00(3-17) vs 2.00(0-7) and 5.00(0-16) respectively, $p < .001$], as well as 3 to 6 months after [median(range) 2.00(0-7) and 5.00(0-16) vs 1.00(0-8) and 3.00 (0-11) respectively, $p < 0.05$]. Joint perimeter presented a statistically significant decrease ($p < .05$) 3 months after treatment in all joints, while 3 to 6 months after, significant reduction remained only in knees. All parameters were improved 12 months after treatment compared to pre-treatment levels. All patients noticed relief of their symptoms with statistical decrease of BE from a median (range) 2 (0-4) before to 0(0-1)3 months after($p:0.001$). No complications were described.

Conclusions: PRP is a simple, painless, inexpensive and effective treatment of chronic synovitis in PWH and could contribute to the deceleration of the disease process.

P-030 (1024945) | The role of hypoxic inducible factor-1 α and vascular endothelial growth factor pathway in hemophilic arthritis

Xiaobo Yan¹; Chunjin Xiong²; Qingqing Shi¹; Binglan Xia²; BAOLAI HUA¹; Junjiang Sun³

Departments of ¹Hematology and ²Ultrasonography, Clinical Medical College, Yangzhou University, China (People's Republic); ³Division of Molecular Pharmaceutics, Eshelman School of Pharmacy, University of North Carolina, Chapel Hill, North Carolina

Introduction: Hemophilic arthritis (HA) is a serious complication of hemophilia with hypoxia as the hallmark in joint microenvironment. Hypoxia induces hypoxia inducible factor (HIF) and then promotes the expression of Vascular Endothelial Growth Factor (VEGF), an important pro-angiogenic regulator. We hypothesize that the HIF-1 α -VEGF signaling pathway may synchronize to promote the development of synovitis and angiogenesis in HA. To investigate the role of HIF-1 α -VEGF signaling pathway in the pathogenesis of HA, by exploring the levels of protein and mRNA expression of HIF-1 α and VEGF in patients with HA:

Methods: Twenty HA patients with age-paired 20 healthy controls were enrolled. Clinical, laboratory and ultrasound data were collected. Synovial fluid was aspirated from HA patients. Protein of HIF-1 α , VEGF and their mRNAs in serum and synovial fluid, were measured.

Results: In serum, VEGF protein and VEGF mRNA were increased compared to healthy controls. No difference of HIF- α levels existed between HA and healthy controls, while HIF- α mRNA were increased. In synovial fluid, both VEGF and VEGF mRNA, both HIF-1 α protein and mRNA levels were significantly higher compared to serum. In HA patients, both VEGF protein and VEGF mRNA levels in blood correlated positively with joint fluid. The HIF-1 α mRNA was positively correlated with blood and joint fluid. The levels of VEGF mRNA and HIF-1 α mRNA were highly positively correlated in blood and also in joint fluid. VEGF mRNA and HIF-1 α in blood were significantly positively correlated with the total score and each itemized score of HEAD-US and HJHS.

Conclusions: The HIF-1 α -VEGF pathway may be involved in the development of synovitis and angiogenesis in HA, and served as an indicator for the assessment of joint lesions. Regulating HIF-1 α -VEGF signaling pathway can be a potential target for the treatment of HA.

P-029 (1036512) | Radiosynoviortesis in the treatment of bleeding and synovitis: challenges and evolution of the protocol used in a university service in Brazil

Alessandro Zorzi¹; Rodrigo Pagnano¹; Glenda Feldberg²; Janaina B S Ricciardi³; Margareth Ozelo⁴

¹University of Campinas - Unicamp, Sao Paulo, Brazil; ²Hemocentro UNICAMP, University of Campinas, Campinas, SP, Brazil; ³Department of Internal Medicine, School of Medical Sciences UNICAMP, University of Campinas, Campinas, SP, Brazil, Brazil; ⁴Hemocentro UNICAMP, University of Campinas, Campinas, SP, Brazil 2Department of Internal Medicine, School of Medical Sciences UNICAMP, University of Campinas, Campinas, SP, Brazil, Brazil; ⁴UNICAMP, Brazil

Introduction: The use of radiopharmaceuticals, drugs that emit ionizing radiation, is a well-known resource and has been used for many decades in the treatment of joint problems caused by Hemophilia. However, the high cost and lack of local production of these radiopharmaceuticals can make access to their use in poor and developing countries difficult. The purpose of this presentation is to show the challenges faced in Brazil and the solutions found in our service, which can inspire other services in a similar situation so that they do not give up this valuable tool.

Methods: A review of the publications made by our service in the last 10 years was carried out, associated with the description of our current protocol for the use of radiosynoviortesis. Three publications were found with clinical data of patients treated at this center.

Results: Three studies published in the last 10 years with clinical data of patients treated by radiosynoviortesis in our service were found. In 2010, 38 joints in 36 patients had been treated. Two radiopharmaceuticals were used: Yttrium-90 and Samarium-153. In 2016, that number increased to 98 joints in 71 patients. There was an increasing trend in the use of Sm153, due to easy access (low cost and national production). In 2020 the result of a comparative clinical study between Sm153 versus Yt90 was published, which led to the confirmation of the effectiveness of Sm153 even in larger joints.

Conclusions: The use of radiosynoviortesis is an important tool in the fight against refractory bleeding and chronic synovitis. In addition to improving symptoms, it may play a role in preventing the progression of arthropathy. The use of low-cost, locally produced radiopharmaceuticals is an alternative for performing therapy in poor or developing countries.

TELEHEALTH

FP-W-01.01 | Teleguidance enables patient self-imaging of arthropathic joints using mobile ultrasound devices

Peter Aguero; Andres Flores; Annette von Drygalski
Division of Hematology/Oncology, Department of Medicine, University of California, San Diego, California, USA

Introduction: Background: New tele-guidance technology on portable devices opens the possibility of patient self-imaging of hemophilic joints. Aims: Elucidate the feasibility of patient self-imaging with/without tele-guidance.

Methods: Adult hemophilia patients (University of California) were invited to participate in a prospective joint self-imaging study. Patients received ultrasound teaching and manuals explaining how to self-image 11 views for hemarthrosis detection in ankles, elbows and knees (Joint _{tissue} Activity and Damage Exam). Patients were asked to acquire 5 randomly selected views with the Butterfly/IQ probe (butterflynetwork.com) and an iPad without assistance 2, 6 weeks and 12 weeks later, followed by a tele-assisted session (Butterfly/IQ tele-guidance function). Image acquisition was timed, patients identified anatomic landmarks, and images were graded on a scale of 0–4 (0 = not similar to 4 = very similar; maximum score = 20/5 views). Patients and facilitator completed questionnaires describing their experience. Hemophilia Joint Health Score (HJHS) indicated arthropathy status.

Results: Of 132 hemophilia patients 10 [median age 52 years (range 29–73)] opted for study inclusion and ultrasound training. Most had severe Hemophilia A, were white/non-Hispanic, employed, and with at least a high school degree. Overall, not different from the other 120 patients. At 2 and 6 weeks after training, ~80% images were acquired correctly [image grading: median ~12 (range 4–18.5) compared to 53% at 12 weeks [image grading: median = 6 (range 0–18)]. Accuracy of landmark recognition was ~55%. With tele-guidance, all images were acquired correctly, with near-perfect image quality overall [Image grading: median 20 (range 18–20); $p \leq 0.01$ compared to 12 weeks]. Median HJHS of scanned joints was 11.5 (range = 2–20) at each time point, demonstrating a similar spectrum of arthropathic changes. Median time of image acquisition was similar with and without tele-guidance [median 01:04 (mm:ss) (range 00:17–04:56) vs. median 01:02 (range 00:16–06:01)], but differed slightly between arthropathic and non-arthropathic joints. Study participants and facilitator rated that it was easy to navigate mobile technology and acquire images with tele-guidance.

Conclusions: Mobile ultrasound with tele-guidance for joint self-imaging is feasible and warrants further exploration.